Dedicated to

Lloyd Malugani Aiello, MD
A kind, humble genius

&

The memory of Mariana Dieste Mead, RN, MD
A source of inspiration for ophthalmologists in training
Contents

1  General Medicine  1
2  Fundamentals of Ophthalmology  23
3  Optics and Refraction  55
4  Ocular Pathology  83
5  Neuro-ophthalmology  120
6  Oculoplastics  158
7  Uveitis  196
8  Glaucoma, Lens, and Anterior Segment  216
9  Cornea, External Disease, and Refractive Surgery  239
10  Pediatric Ophthalmology and Strabismus  298
11  Retina and Vitreous  361
12  International Ophthalmology  406
It is a great pleasure to write the foreword to the third edition of Massachusetts Eye and Ear Infirmary Review for Ophthalmology.

Dr. Rama D. Jager and Dr. Jeffrey C. Lamkin, the primary organizers and executors of this project, have spent an extensive amount of time creating and updating this work. Rama has served as a vitreoretinal fellow within the Department of Ophthalmology, and Jeff is a former Chief Resident. Together, they have created a work that helps physicians maintain and update their knowledgebase within the ever-growing field of ophthalmology.

Ophthalmology has indeed expanded considerably over the last several years. We expect this expansion to accelerate at an exponential rate. Keeping track of new developments within the field can often be challenging, especially for those of us who specialize in one specific field within ophthalmology or for those of us who are more interested in particular aspects within the field and less focused on other ophthalmic subspecialties. Yet, maintaining a familiarity with all the subspecialties within the field of ophthalmology is a requisite for any good ophthalmologist or ophthalmologist-in-training.

One way of keeping abreast of current developments is through self-assessment. Self-assessment and continuing medical education should not be undertaken simply because of board certification requirements—instead, they should be the cornerstones of our professional lives!

Regular self-assessment allows us to continuously update our knowledgebase, and an up-to-date knowledgebase helps us in serving our patients in a way that we ourselves would want to be served.

One of Dr. Harvey Cushing’s associates at the Peter Bent Brigham Hospital, Dr. Merrill Sosman, taught his residents, “We see only what we look for. We look for only what we know.” The questions in this book have been prepared with great care in an attempt to help the reader “know” the clinically significant areas within ophthalmology and the basic science concepts from which our care is derived.

As always, no effort has been made to reconstruct questions from any form of current or previous board examinations; rather, the questions have been created to review the essentials of the field for the ophthalmologist or ophthalmologist-in-training who wishes to be informed of recent scientific discoveries and the refashioned foundations of clinical science.

I congratulate Jeff and Rama for their contribution to the field.

Joan W. Miller, MD
Henry Willard Williams Professor of Ophthalmology
Chief and Chair, Department of Ophthalmology
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It is a great pleasure to write a brief foreword to *The Massachusetts Eye and Ear Infirmary Review Manual for Ophthalmology*. Dr. Jeffrey Lamkin, the primary organizer and executor of this project with the secondary help of the residents of the Massachusetts Eye and Ear Infirmary, initially conceived it as part of his teaching obligation to the residents while serving as Chief Resident. A highly professional and scholarly set of questions was prepared. It became apparent that many other residents and ophthalmologists seeking to identify the strengths and weaknesses in their ophthalmic knowledge could profit from the dissemination of the material.

The past decade witnessed an explosion in ophthalmic knowledge that was truly daunting and unprecedented. We can expect this expansion to accelerate in the future at an exponential rate. How does one keep abreast of new developments, and how does one form judgments as to what is clinically relevant knowledge and what is of more theoretical or marginal interest at the moment? Most of us are highly specialized in either the knowledge industry or in our clinical practice patterns. If we wish to stay in touch with other specialties of ophthalmology, we tend to need help. Self-assessment and continuing medical education should by now be cornerstones of our professional lives, but these tasks will also be progressively shared with others! We will be increasingly held to higher quality assurance standards, and recertification examinations are to be put in place. Furthermore, the ability to practice medicine in the future may become aligned with profiles of the clinical outcomes of the patients we treat, as could be mandated by federal agencies and insurance payors. Sound and constantly remodelled knowledge will thus be the best basis for protecting and advancing our clinical practices.

The questions in this publication have been prepared with great care, in order to achieve balance among the various subspecialties of our field and to highlight clinically significant subjects and basic science concepts and findings that under-gird them. No effort has been made to reconstruct questions from Board or OKAP examinations; rather, the questions have been created in a way that proceeds from what one group of people has determined to be an essential database for the practicing ophthalmologist who wishes to be informed of recent scientific discoveries and the refashioned foundations of clinical science. As they say in the movies, any resemblance to questions that appear on formal examinations is completely coincidental and reflects the fact that people who are serious students of the subject have independently identified topics that should be within the purview and common fund of knowledge of a contemporary clinician. The questions themselves are less important than the subjects they represent. A well-positioned cadre that can serve as an arbiter for this fund of knowledge is residents-in-training who are eager to equip themselves for their impending clinical professional lives, and who daily critically assess and internalize the rivulets of information that are flowing toward them from their teachers and clinical preceptors.

This textbook is the distinctive product of a unique mind and constellation of talents possessed by Jeffrey Lamkin. I can think of nobody better suited to shepherd this project than Jeffrey because he is guided by an insightful and retentive mind, a capacity for detail and global integration, and an ardent love of knowledge and its transmission to those around him. Due to the depth of his commitment to resident education and that of other colleagues, he has made a tremendous gift of his energies and time to our entire field.

Frederick A. Jakobiec, MD
More than six years have passed since the second edition of the *Massachusetts Eye and Ear Infirmary Review Manual for Ophthalmology* was released. The aim of this work is to provide medical students, residents, fellows, and general ophthalmologists with a comprehensive and updated review of ophthalmology through a series of challenging, pertinent questions.

The third edition differs from the second in several ways. The new edition has been significantly updated to reflect current knowledge in the field. Several hundred new questions have been added to this edition. Many explanations have been revised to provide the reader with additional information. Two new chapters, “General Medicine” and “International Ophthalmology,” have also been added. The “Pediatric Ophthalmology” and “Strabismus” chapters of the previous edition have been combined into a single chapter in this edition. Finally, several new color figures depicting ocular pathology and ophthalmic conditions have been added to this new edition as well.

The purpose of this book is to provide the reader with a useful method for self-assessment in his or her attempt to retain and to update knowledge in the field of ophthalmology. This book should not serve as the sole method of preparation for any specific examination and is no way intended to duplicate the American Board of Ophthalmology (ABO) certification exam, or for that matter, any other ophthalmology examination. Instead, we hope to help the reader retain the knowledge already gleaned from his or her reading, as well as provide a tool for accurate self-assessment.

We would like to thank all of the people involved in helping create this book. The physicians who helped create these editions have been very generous with their time and effort in making this edition, in our opinion, an outstanding book for self-assessment and review. We would also like to thank our fellow physician colleagues (and friends), Drs. Alfredo Gomez-Leal, Hugo Quiroz-Mercado, Abelardo Rodriguez-Reyes, Ricardo Agurto-Rivera, Dhananjay Shukla, and Ramasamy Kim for graciously providing us with and allowing us to use some of their images in our book.

Finally, we also would like to thank both Jonathan Pine, the Senior Executive Editor who supported this project from the start, and Anne Jacobs, the Managing Editor at Lippincott Williams & Wilkins who has been extraordinarily patient and helpful with our work.

Updating this edition has been a fantastic experience. We hope that you, the reader, will benefit from our efforts.

Rama D. Jager, MD, MBA

Jeffrey C. Lamkin, MD
General Medicine

Questions

1. Which of the following statements about the normal microbial flora is false?
   a. The microorganisms on the epithelial surfaces of the body remain in place primarily through adherence.
   b. Humans possess a large multitude of microorganisms on the skin, gastrointestinal (GI), vaginal, and upper respiratory tracts.
   c. There is little benefit of these microorganisms to humans.
   d. Severe infections can result from the normal microbial flora if the body’s mechanical defenses are compromised.
   e. Most Neisseria organisms are normal inhabitants of the alimentary and upper respiratory tracts.

d. Haemophilus influenzae.

2. Which of the following statements about Staphylococcus is true?
   a. S. epidermidis is found on <50% of skin cultures.
   b. Vancomycin is the drug of choice for Staphylococcus-infected prosthetic heart valves.
   c. Acute staphylococcal infections rarely require intravenous (IV) therapy.
   d. Of the tertiary care isolates of S. aureus, approximately 90% are resistant to beta-lactam antibiotics.
   e. All of the above.

3. Which of the following microorganisms is the most common cause of prosthetic heart valve infections?
   a. Staphylococcus epidermidis.
   b. Staphylococcus viridans.
   c. Staphylococcus aureus.
   d. Haemophilus influenzae.
   e. Streptococcus pneumoniae.

4. Which of the following associations about Streptococcus is incorrectly paired?
   a. S. pneumoniae: Lancet-shaped diplococci that cause alpha-hemolysis on blood agar.
   c. S. pyogenes: Highly sensitive to penicillin G.
   d. S. pneumoniae: Virulence is mediated by a polysaccharide capsule of which there are >80 different serotypes.
   e. S. pneumoniae: The organism can cause pneumococcal pneumonia, which is highly sensitive to penicillin and has a very low mortality rate in elderly patients.

5. Which of the following patients would not require endocarditis prophylaxis during invasive surgery?
   a. a patient with a prosthetic heart valve.
   b. a patient with a previous history of bacterial endocarditis.
   c. a patient with mitral valve prolapse with valvular regurgitation.
   d. a patient with a history of severe coronary artery disease.
   e. a patient with hypertrophic cardiomyopathy.
6. What type of ocular surgery would require endocarditis prophylaxis?
   a. cataract surgery.
   b. vitrectomy.
   c. orbital floor implant.
   d. tear duct reconstruction.
   e. c. and d.

7. Which microorganism is commonly associated with pseudomembranous colitis?
   a. Enterococcus.
   b. Escherichia coli.
   c. Clostridium difficile.
   d. Staphylococcus epidermidis.
   e. Pseudomonas aeruginosa.

8. Which of the following antibiotics would most likely not be effective against Haemophilus influenzae?
   a. erythromycin.
   b. augmentin.
   c. ceftazidime.
   d. trimethoprim–sulfamethoxazole.
   e. ciprofloxacin.

9. Which of the following statements is false?
   a. Gonococci are not part of the normal microbial flora.
   b. Fifty percent of women and 95% of men infected with gonococci are symptomatic.
   c. Chlamydia trachomatis is found in as many as half of all women and one third of all men infected with gonococci.
   d. Macrolides and quinolones are generally not good agents to treat gonococcal infections.
   e. Quinolone-resistant strains of gonococci have been reported.

10. Which of the following statements about syphilis is false?
    a. The treatment of choice for patients with neurosyphilis is penicillin G, 2.4 million units intramuscularly weekly for 3 weeks.
    b. Transplacental transmission from an untreated, pregnant female patient to her fetus before 16 weeks gestation can result in congenital syphilis.
    c. The classic gummas of tertiary syphilis are characterized by caseating granulomas.
    d. In neurosyphilis, the serum venereal disease research laboratory (VDRL) test result may be negative.
    e. Serum fluorescent treponemal antibody absorption (FTA-ABS) and VDRL tests are used to determine whether an individual has a history of syphilis.

11. Which of the following spirochetes causes Lyme disease?
    a. Treponema pallidum.
    b. Ixodes scapularis.
    c. Borrelia burgdorferi.
    d. Leptospira interrogans.
    e. Brachyspira pilosicoli.

12. Which of the following defines a positive purified protein derivative (PPD) reaction in an immunocompetent individual?
    a. induration of 10 mm or greater, read 24 hours later.
    b. induration of 10 mm or greater, read 48 to 72 hours later.
    c. induration of 5 mm or greater, read 24 hours later.
    d. induration of 5 mm or greater, read 48 to 72 hours later.
    e. none of the above.

13. Which of the following is not a herpesvirus?
    a. human immunodeficiency virus (HIV).
    b. cytomegalovirus (CMV).
    c. varicella zoster virus (VZV).
    d. herpes simplex virus (HSV).
    e. Epstein–Barr virus (EBV).

14. Which of the following statements about herpesviruses is false?
    a. Epstein–Barr virus (EBV) is associated with nasopharyngeal carcinoma.
    b. EBV is associated with Burkitt’s lymphoma.
    c. Intravitreal ganciclovir has been shown to be effective against systemically disseminated cytomegalovirus (CMV) in immunocompromised patients.
    d. Tricyclic antidepressants have been shown to be effective in some patients with postherpetic neuralgia from varicella zoster virus (VZV).
    e. Untreated neonatal herpes infection carries a 70% mortality rate.
Questions

15. Which of the following methods of transmission of hepatitis viruses is incorrect?
   b. hepatitis B: fecal–oral route and sexual transmission.
   c. hepatitis C: blood transfusions.
   d. hepatitis D: sexual transmission.
   e. hepatitis E: fecal–oral route.

16. Which of the following statements about acquired immunodeficiency syndrome (AIDS) is true?
   a. Approximately 500 million individuals in the world have AIDS.
   b. Most individuals infected with human immunodeficiency virus (HIV) live in the United States.
   c. The enzyme-linked immunosorbent assay (ELISA) test for HIV is 99% sensitive but only 50% specific for HIV.
   d. Infected breast milk can also be a method of transmission of HIV.
   e. Occupational exposure to HIV during ocular surgery does not require chemoprophylaxis.

17. T or F Multidrug resistance (MDR) of bacteria is often caused by combination therapy (e.g., using several different antibiotics at the same time) to treat an infection.

18. T or F Ocular complications of systemic hypertension can include retinovascular disease (e.g., central retinal vein obstruction [CRVO]).

19. Hypertension is defined as:
   a. blood pressure readings of >140/90 mm Hg on three separate occasions.
   b. a single blood pressure reading of 140/80 mm Hg.
   c. blood pressure readings of >150/80 mm Hg on three separate occasions.
   d. blood pressure readings of >160/90 mm Hg on three separate occasions.
   e. none of the above.

20. Causes of secondary hypertension include all of the following except:
   a. pheochromocytoma.
   b. pseudotumor cerebri.
   c. hyperaldosteronism.
   d. polycystic kidney disease.
   e. Cushing’s syndrome.

21. Which of the following statements about lifestyle factors affecting blood pressure is false?
   a. Processed foods account for 75% of sodium intake in the United States.
   b. Patients with mild hypertension who smoke a pack of cigarettes a day have a fivefold higher risk of coronary artery disease.
   c. Alcohol consumption of >3 mL of ethanol (300 mL of wine or 720 mL of beer) is associated with resistance to antihypertensive therapy.
   d. Regular aerobic exercise contributes to reduced mortality and morbidity from hypertension.
   e. Weight loss in obese individuals and adequate mineral intake can help them achieve blood pressure normalization.

22. Which medication is incorrectly paired with its common side effect?
   a. doxazosin: postural hypotension.
   b. atenolol: bronchospasm.
   c. spironolactone: gynecomastia.
   d. minoxidil: hirsutism.
   e. amlodipine: drug-induced lupus.

23. What types of antihypertensives should be used for initial therapy for hypertension in newly diagnosed patients?
   a. beta-blockers and diuretics.
   b. calcium channel blockers and angiotensin-converting enzyme (ACE) inhibitors.
   c. angiotensin II receptor blockers.
   d. alpha-agonists.
   e. none of the above.

24. In the order of decreasing frequency, what are the most common causes of death in the United States?
   a. 1. stroke, 2. cancer, 3. heart disease, 4. accidents, 5. diabetes.
   b. 1. cancer, 2. heart disease, 3. stroke, 4. diabetes, 5. chronic lower respiratory diseases (e.g., chronic obstructive pulmonary disease [COPD]).
   c. 1. heart disease, 2. stroke, 3. cancer, 4. COPD, 5. accidents.
   d. 1. heart disease, 2. cancer, 3. stroke, 4. COPD, 5. accidents.
   e. 1. heart disease, 2. cancer, 3. stroke, 4. accidents, 5. COPD.
25. What is the definition of a transient ischemic attack (TIA)?
   a. a loss of neurologic function caused by ischemia that lasts for <24 hours and that clears without residual signs.
   b. a loss of neurologic function that lasts for >24 hours, but for <7 days, and that leaves no lasting symptoms or signs.
   c. a loss of neurologic function that leaves a persistent minor disability but does not progress to a major stroke.
   d. an ischemic attack, but not infarcted area of the brain, which has been shown to have some plasticity with regard to recovery.
   e. none of the above.

26. Which of the following statements about stroke is false?
   a. Clopidogrel has a better side effect profile than ticlopidine with regard to bone marrow suppression.
   b. The use of tissue plasminogen activator (TPA) within 24 hours of the onset of symptoms improves outcome in patients with stroke.
   c. Carotid endarterectomy (CEA) is beneficial in reducing the incidence of stroke in symptomatic patients with carotid stenosis of >70%.
   d. Generalized seizures are common in patients with intracerebral hemorrhage but occur in <10% of cases of patients with cerebral infarction only.
   e. Intracranial hemorrhage comprises 15% of all cerebrovascular accidents.

27. T or F Magnetic resonance imaging (MRI) is better than computed tomography (CT) at detecting early cerebral infarction.

28. T or F Carotid bruits are a better predictor of arteriosclerosis than they are of stroke.

29. The annual risk rate of stroke in patients with isolated amaurosis fugax is:
   a. 2%.
   b. 4%.
   c. 6%.
   d. 8%.
   e. 10%.

30. In patients with a transient ischemic attack (TIA), the risk of stroke within the first year is:
   a. 2%.
   b. 4%.
   c. 6%.
   d. 8%.
   e. 10%.

31. After the first year, in patients with a transient ischemic attack (TIA), the annual risk of stroke is:
   a. 2%.
   b. 4%.
   c. 6%.
   d. 8%.
   e. 10%.

32. Folic acid can reduce the risk of stroke in patients by reducing the plasma levels of:
   a. high-density lipoprotein (HDL) cholesterol.
   b. homocysteine.
   c. hemoglobin.
   d. triglycerides.
   e. low-density lipoprotein (LDL) cholesterol.

33. Which of the following statements about carotid endarterectomy (CEA) is false?
   a. CEA should only be considered if the surgeon performing the operation has a perioperative morbidity rate of <3%.
   b. In the Asymptomatic Carotid Atherosclerosis Study, patients with asymptomatic carotid stenosis of >60% who underwent CEA did not show a significantly lower risk of having another major ischemic stroke, compared to patients who were solely on medication.
   c. Asymptomatic carotid bruits occur in 10% of the population older than 40 years.
   d. In the North American Symptomatic Carotid Endarterectomy Trial, CEA was shown to have increasing benefit with higher degrees of carotid stenosis.
   e. Patients with transient ischemic attack (TIA) or previous stroke in the territory of the carotid circulation are judged to be “symptomatic.”

34. All of the following can impede the supply of oxygen to the myocardium except:
   a. anemia.
   b. coronary artery stenosis.
35. All of the following will increase the demand for oxygen by the myocardium except:
   a. increased heart rate.
   b. decreased ventricular contractility.
   c. increased ventricular wall thickness.
   d. increased stroke volume.
   e. all of the above will increase the demand for oxygen supply by the myocardium.

36. What occurs when the demand by the myocardium exceeds the supply?
   a. myocardial infarction (MI).
   b. coronary artery occlusion.
   c. ischemia.
   d. penumbra.
   e. none of the above.

37. What is the most common cause of cardiac ischemia?
   a. coronary arteriosclerosis.
   b. coronary spasm.
   c. coronary arteritis.
   d. hypoxemia.
   e. anemia.

38. Which of the following is a clear risk factor for ischemic heart disease (IHD)?
   a. obesity.
   b. stress.
   c. personality type.
   d. diabetes.
   e. use of oral contraceptives.

39. T or F Variant angina is directly related to physical exertion.

40. The difference between a non-Q-wave myocardial infarction (MI) and unstable angina is that:
   a. non-Q-wave infarcts will have angina at rest, whereas unstable angina will not.
   b. non-Q-wave infarcts will have elevated cardiac enzymes, whereas unstable angina will not.
   c. non-Q-wave infarcts can have T wave inversions, whereas unstable angina will not.
   d. non-Q-wave infarcts can have ST segment depression on electrocardiogram (ECG), whereas unstable angina will not.
   e. none of the above.

41. What percentage of all myocardial infarctions (MIs) are painless?
   a. 5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

42. Half of all myocardial infarctions (MIs) involve the _________ wall.
   a. superior.
   b. lateral.
   c. posterior.
   d. inferior.
   e. anterior.

43. T or F Twenty percent of patients with subendocardial infarctions experience an acute Q-wave infarction within 3 months after initial infarct.

44. Cardiogenic shock carries a mortality rate of __________ and is seen in __________ of patients with myocardial infarction (MI).
   a. 70%; 10%.
   b. 80%; 20%.
   c. 100%; 30%.
   d. 50%; 20%.
   e. 20%; 10%.

45. Dressler’s syndrome is characterized by all of the following except:
   a. pericardial friction rub.
   b. fever.
   c. arthralgias.
   d. glossitis.
   e. pleural pain.

46. Which of the following can cause sudden cardiac death?
   a. Wolfe-Parkinson-White syndrome.
   b. long QT syndrome.
   c. torsade de pointes.
   d. atioventricular (AV) block.
   e. all of the above.
47. T or F Patients with a normal electrocardiogram (ECG) can have myocardial ischemia.

48. T or F Cardiac troponins are not usually present in the serum of healthy individuals.

49. T or F Cardiac troponin levels are usually elevated after skeletal muscle injury.

50. Troponin levels remain elevated for ____________ days after myocardial infarction (MI).
   a. 1 to 7.
   b. 1 to 14.
   c. 1 to 21.
   d. 1 to 30.
   e. 1 to 180.

51. Within 6 to 12 hours after a suspected infarct, which of the following is the most sensitive marker for infarct?
   a. troponins I and T and creatinine kinase MB.
   b. lactate dehydrogenase.
   c. erythrocyte sedimentation rate (ESR).
   d. C-reactive protein.
   e. homocysteine.

52. T or F Greater troponin I levels are associated with an increased likelihood of mortality in patients with acute coronary syndrome.

53. Coronary artery stenosis is hemodynamically substantial when the cross-sectional area is reduced by more than ____________ or when the coronary artery diameter is stenosed by more than ____________.
   a. 25%; 25%.
   b. 30%; 30%.
   c. 40%; 40%.
   d. 50%; 50%.
   e. 75%; 50%.

54. T or F Coronary artery bypass grafting (CABG) should never be considered in patients with stable angina.

55. All of the following can be used concurrently to treat acute coronary syndrome except:
   a. aspirin.
   b. low-molecular-weight heparin.
   c. nitroglycerin.
   d. atenolol.
   e. nifedipine.

56. Percutaneous transluminal coronary angioplasty (PTCA) is usually superior to thrombolytic therapy in acute coronary syndromes in all of the following situations except:
   a. initiation of therapy >/= 90 minutes after acute coronary syndrome.
   b. cardiogenic shock.
   c. patients with an increased risk of intracranial hemorrhage.
   d. patients with a prior history of coronary artery bypass grafting (CABG).
   e. patients with a history of recent extensive abdominal surgery.

57. Clinical signs of acute left-ventricular heart failure can include all of the following except:
   a. use of accessory muscles for respiration.
   b. hemoptysis.
   c. diaphoresis.
   d. mental status changes.
   e. hepatomegaly.

58. The lower limit normal for ejection fraction is:
   a. 30%.
   b. 40%.
   c. 50%.
   d. 60%.
   e. 70%.

59. What is the most common cause of congestive heart failure (CHF) in the United States?
   a. left-ventricular hypertrophy.
   b. mitral regurgitation.
   c. sarcoidosis.
   d. myocarditis.
   e. ischemic heart disease (IHD).

60. Causes of left-ventricular hypertrophy include:
   a. systemic hypertension.
   b. idiopathic hypertrophic subaortic stenosis.
   c. coarctation of the aorta.
   d. all of the above.
   e. none of the above.

61. Potential causes of high-output cardiac failure include:
   a. beriberi.
   b. Paget’s disease.
   c. hyperthyroidism.
   d. severe anemia.
   e. all of the above.
62. The afterload for the left ventricle is determined by ____________, whereas the afterload for the right ventricle is determined by ____________.
   a. left atrial pressure; right atrial pressure.
   b. mean arterial pressure; mean venous pressure.
   c. pulmonary venous pressure; inferior vena cava pressure.
   d. aortic pressure; pulmonary artery pressure.
   e. none of the above.

63. The most common cause of right-sided heart failure is:
   a. left-sided heart failure.
   b. primary pulmonary hypertension.
   c. portal hypertension.
   d. coarctation of the aorta.
   e. none of the above.

64. Usually, the most effective way of treating systolic dysfunction is to:
   a. reduce preload.
   b. increase preload.
   c. reduce afterload.
   d. increase afterload.
   e. reduce contractility.

65. The calcium channel blocker of choice in patients with congestive heart failure (CHF) with ischemic heart disease (IHD) is:
   a. captopril.
   b. amlodipine.
   c. diltiazem.
   d. doxazosin.
   e. prazosin.

66. The management of diastolic dysfunction can be accomplished by:
   a. reducing preload.
   b. reducing contractility.
   c. increasing preload.
   d. increasing afterload.
   e. none of the above.

67. T or F Cardiac transplantation is an efficacious treatment of patients with refractory congestive heart failure (CHF).

68. The primary pacemaker of the heart is the:
   a. sinoatrial (SA) node.
   b. atrioventricular (AV) junction.
   c. bundle of His.
   d. chordae tympanae.
   e. none of the above.

69. T or F Sinus bradycardia on an electrocardiogram (ECG) is almost never innocuous.

70. First-degree atrioventricular (AV) block is diagnosed by:
   a. a prolongation of the PR interval >0.2 seconds on electrocardiogram (ECG).
   b. progressive PR prolongation before a nonconducted P wave.
   c. QRS complex drops at regular intervals.
   d. complete asynchrony between the P wave and the QRS complex.
   e. irregular pulse.

71. The ideal treatment of choice for paroxysmal atrial tachycardia (PAT) is:
   a. intravenous adenosine.
   b. Valsalva maneuvers.
   c. carotid massage.
   d. beta-blockers.
   e. digoxin.

72. The preferred first-line therapy to treat arrhythmias in patients who have had a prior myocardial infarction (MI) is:
   a. automated implantable cardioverter-defibrillator (AICD).
   b. pacemaker.
   c. amiodarone.
   d. ibutilide.
   e. adenosine.

73. In most patients, first-line therapy to reduce cholesterol level should be:
   a. diet and exercise.
   b. simvastatin.
   c. atorvastatin.
   d. small amounts of alcohol.
   e. none of the above.

74. Major risk factors that modify low-density lipoprotein (LDL) goals in patients include all of the following except:
   a. an age of 45 years or greater in men and 55 years or greater in women.
   b. cigarette smoking.
   c. low level of high-density lipoprotein (HDL) cholesterol (<40 mg/dL).
75. Common adverse reactions to beta-hydroxy-beta-methylglutaryl-coenzyme A (HMG-CoA) reductase inhibitors include:
   a. incontinence and constipation.
   b. diarrhea and liver enzyme elevation.
   c. renal stones and diabetes.
   d. palpitations and anxiety.
   e. hypotension and dehydration.

76. T or F Beta-hydroxy-beta-methylglutaryl-coenzyme A (HMG-CoA) reductase inhibitors have been shown to substantially increase the risk of cataracts.

77. Characteristics of metabolic syndrome include all of the following except:
   a. abdominal girth >102 cm (40 in.).
   b. systolic blood pressure >130 mm Hg and diastolic blood pressure >85 mm Hg.
   c. fasting glucose level >110 mg/dL.
   d. triglycerides level >150 mg/dL.
   e. total cholesterol level >240 mg/dL.

78. T or F Elevated serum triglycerides concentration are an independent risk factor for coronary heart disease.

79. T or F Reduced high-density lipoprotein (HDL) cholesterol is an independent risk factor for congenital heart disease (CHD).

80. What occurs when bronchospasm narrows airways, and exhaled air is forced through these narrow airways?
   a. dyspnea.
   b. hyperventilation.
   c. wheezing.
   d. rales.
   e. none of the above.

81. Which of the following is not an irreversible obstructive disease?
   a. asthma.
   b. chronic obstructive pulmonary disease (COPD).
   c. chronic bronchitis.
   d. peripheral airway disease.
   e. none of the above.

82. An FEV₁ (forced expiratory volume at 1 second) of <80% suggests:
   a. obstructive disease.
   b. restrictive disease.
   c. fibrosis of the lung parenchyma.
   d. pneumothorax.
   e. a decreased total lung capacity.

83. T or F Nasal continuous positive airway pressure (CPAP) can increase intraocular pressure (IOP) in patients with glaucoma.

84. Which of the following is not a selective beta-2 agonist?
   a. albuterol.
   b. bitolterol.
   c. metaproterenol.
   d. all of the choices are selective beta-2 agonists.
   e. none of the above.

85. Which of the following could be used to manage an acute asthma attack?
   a. subcutaneous epinephrine.
   b. salmeterol.
   c. inhaled beclomethasone.
   d. cromolyn sodium.
   e. none of the above.

86. T or F Theophylline is a common first-line agent for asthma treatment.

87. Which of the following has been proved to increase survival in patients with severe chronic obstructive pulmonary disease (COPD)?
   a. oral corticosteroids.
   b. nitroglycerin.
   c. captopril.
   d. supplemental oxygen.
   e. hydrochlorothiazide.

88. The average lifespan of an erythrocyte is:
   a. 6 hours.
   b. 2 weeks.
   c. 6 weeks.
   d. 120 days.
   e. 1 year.
89. Erythropoiesis is stimulated by erythropoietin, which is produced mainly in the:
   a. lungs.
   b. liver.
   c. spleen.
   d. kidney.
   e. bone marrow.

90. T or F Normal hemoglobin levels are higher in men than in women.

91. Which of the following is not a cause of microcytic anemia?
   a. iron deficiency.
   b. thalassemia.
   c. sideroblastic anemia.
   d. pernicious anemia.
   e. none of the above.

92. Levels of ___________ can be used to differentiate pure iron deficiency anemias from pure anemia of chronic disease.
   a. transferrin receptor(s).
   b. hemoglobin.
   c. mean corpuscular volume (MCV).
   d. red cell distribution width.
   e. total iron-binding capacity.

93. What percentage of African Americans in the United States have sickle cell disease?
   a. 0.2% to 0.4%.
   b. 1.0% to 2.0%.
   c. 5% to 10%.
   d. 10% to 20%.
   e. 20% to 30%.

94. Which of the following substances is not used by the body to prevent widespread clotting?
   a. antithrombin III.
   b. protein C.
   c. protein S.
   d. thrombin.
   e. tissue factor pathway inhibitor.

95. What blood test is typically used to measure the effect of heparin therapy?
   a. prothrombin time (PT).
   b. partial thromboplastin time (PTT).
   c. bleeding time.
   d. international normalized ratio (INR).
   e. none of the above.

96. What blood test is typically used to measure the effect of warfarin therapy?
   a. prothrombin time (PT).
   b. partial thromboplastin time (PTT).
   c. bleeding time.
   d. international normalized ratio (INR).
   e. none of the above.

97. What blood test is typically used to measure the effect of subcutaneous low-molecular-weight heparin therapy?
   a. prothrombin time (PT).
   b. partial thromboplastin time (PTT).
   c. bleeding time.
   d. international normalized ratio (INR).
   e. none of the above.

98. Which of the following blood tests will be affected by aspirin ingestion?
   a. prothrombin time (PT).
   b. partial thromboplastin time (PTT).
   c. bleeding time.
   d. international normalized ratio (INR).
   e. all of the above.

99. T or F Connective tissue disorders are almost never associated with hemorrhage.

100. Scurvy is an uncommon disease in the United States secondary to deficiency of:
    a. ascorbic acid.
    b. niacin.
    c. folic acid.
    d. vitamin B₁₂.
    e. riboflavin.

101. Causes of immunologic destruction of platelets include all of the following except:
    a. thrombotic thrombocytopenic purpura (TTP).
    b. idiopathic thrombocytopenic purpura (ITP).
    c. posttransfusion reactions.
    d. quinidine.
    e. gold.

102. Which of the following types of medications can affect platelet function?
    a. antihistamines.
    b. tricyclic antidepressants.
    c. aspirin.
d. nonsteroidal antiinflammatory drugs (NSAIDs).
e. all of the above.

103. The most common cause of abnormal bleeding in individuals is:
a. platelet disorders.
b. hemophilia A.
c. von Willebrand’s disease.
d. vitamin K deficiency.
e. liver dysfunction.

104. Which of the following is not a fat-soluble vitamin?
a. vitamin A.
b. vitamin C.
c. vitamin D.
d. vitamin E.
e. vitamin K.

105. Which of the following statements about vitamin K is false?
a. Vitamin K is necessary for the production of factors II, VII, IX, and X in the liver.
b. Vitamin K should not be given intramuscularly because of the risk of sudden death from an anaphylactoid reaction.
c. Celiac sprue, cystic fibrosis, and biliary obstruction can be causes of vitamin K deficiency.
d. Vitamin K is routinely administered to newborns to prevent hemorrhagic disease.
e. Vitamin K deficiency leads to prolongation of both prothrombin time (PT) and partial thromboplastin time (PTT).

106. T or F Hyperhomocysteinemia is an independent risk factor for both arterial and venous thrombosis.

107. Which of the following is an ophthalmic complication of the phospholipid antibody syndrome?
a. central retinal vein obstruction (CRVO).
b. choroidal infarction.
c. anterior ischemic optic neuropathy.
d. central retinal artery obstruction (CRAO).
e. all of the above.

108. The most common rheumatic disorder is:
b. relapsing polychondritis.

109. Felty’s syndrome is a triad of:
a. aspirin sensitivity, asthma, and nasal polyps.
b. rheumatoid arthritis, splenomegaly, and leukopenia.
c. rheumatoid arthritis, hepatomegaly, and thrombocytopenia.
d. rheumatoid arthritis, portal hypertension, and polycythemia vera.
e. none of the above.

110. Ocular complications of rheumatoid arthritis include all of the following except:
a. bull’s-eye maculopathy.
b. scleritis.
c. keratitis sicca.
d. marginal corneal ulcers.
e. episcleritis.

111. Which of the following slow-acting antirheumatic drugs (SAARDs) used to treat rheumatoid arthritis is the most toxic?
a. hydroxychloroquine.
b. methotrexate.
c. chlorambucil.
d. sulfasalazine.
e. oral gold.

112. The mechanism of action of leflunomide is:
a. inhibition of pyrimidine synthesis.
b. inhibition of pyrimethamine synthesis.
c. inhibition of pyrazinamide synthesis.
d. inhibition of pyridoxine synthesis.
e. none of the above.

113. The mechanism of action of etanercept and infliximab is:
a. inhibition of messenger ribonucleic acid (mRNA) synthesis.
b. inhibition of tumor necrosis factor-alpha (TNF-alpha).
c. inhibition of the p53 suppressor gene.
d. all of the above.
e. none of the above.
114. Which of the following is not a seronegative spondylopathy?
   a. ankylosing spondylitis.
   b. acne-associated arthritis.
   c. Whipple’s disease.
   d. psoriatic arthritis.
   e. relapsing polychondritis.

115. T or F Approximately 50% of patients with nongranulomatous anterior uveitis presenting acutely are positive for human leukocyte antigen (HLA)-B27.

116. Most patients with Reiter’s syndrome present with __________ as their only symptom.
   a. conjunctivitis.
   b. vaginitis.
   c. iritis.
   d. nongonococcal urethritis.
   e. none of the above.

117. T or F Both ankylosing spondylitis and reactive arthritis are much more common in men than in women.

118. Which of the following findings would lead a clinician to suspicion of psoriatic arthritis?
   a. atrophy of the distal phalanges (“pencil-in-cup” appearance on radiographic films).
   b. fusion of the spine (bamboo spine).
   c. heel pain.
   d. interphalangeal arthritis (“sausage digits”).
   e. none of the above.

119. T or F Still’s disease commonly manifests with ophthalmic symptoms.

120. Which of the following is not part of the criteria needed to diagnose systemic lupus erythematosus (SLE)?
   a. serositis.
   b. oral ulcers.
   c. arthritis.
   d. photophobia.
   e. antinuclear antibody positivity.

121. The most common ocular complication of systemic lupus erythematosus (SLE) is:
   a. secondary Sjögren’s syndrome.
   b. retinal vascular lesions (e.g., cotton-wool spots, intraretinal hemorrhages).
   c. optic neuritis.
   d. discoid lesions of the eyelids.
   e. cortical blindness.

122. T or F Systemic lupus erythematosus (SLE) and scleroderma are much more common in women than in men.

123. More than 90% of patients with scleroderma have:
   a. calcinosis.
   b. the Raynaud phenomenon and esophageal involvement.
   c. sclerodactyly.
   d. telangiectasias.
   e. none of the above.

124. A heliotrope rash and Gottron’s papules would make one suspicious for:
   a. scleroderma.
   b. dermatomyositis.
   c. polymyositis.
   d. relapsing polychondritis.
   e. none of the above.

125. A patient who has a Schirmer-1 test of <5 mm in 5 minutes, who uses tear substitutes more than three times per day, and who is positive for rheumatoid factor probably has:
   a. scleroderma.
   b. Sjögren’s syndrome.
   c. juvenile rheumatoid arthritis.
   d. relapsing polychondritis.
   e. all of the above.

126. The most common clinical findings in relapsing polychondritis are:
   a. laryngeal collapse.
   b. conjunctivitis and iritis.
   c. aortic insufficiency and vasculitis.
   d. arthropathy, auricular, and nasal chondritis.
   e. gastritis.

127. Cogan’s syndrome is associated with __________ in 50% of cases.
   a. Churg-Strauss angiitis.
   b. Wegener’s granulomatosis.
   c. polyarteritis nodosa (PAN).
128. The gold standard for diagnosing giant cell arteritis (GCA) is:
   a. erythrocyte sedimentation rate (ESR) (Westergren method).
   b. ESR (Wintrobe methods).
   c. C-reactive protein.
   d. a. and c.
   e. none of the above.

129. The most common ophthalmic complication of giant cell arteritis (GCA) is:
   a. central retinal vein obstruction (CRVO).
   b. central retinal artery obstruction (CRAO).
   c. diplopia.
   d. amaurosis fugax.
   e. ischemic optic neuropathy.

130. The most common clinical manifestation of Behçet’s disease is:
   a. oral ulcers.
   b. genital ulcers.
   c. polyarthritis.
   d. erythema nodosum.
   e. deep vein thrombosis.

131. All of the following are potential side effects from administration of exogenous corticosteroids except:
   a. hypokalemia.
   b. avascular necrosis of the femoral head.
   c. hyperglycemia.
   d. pseudotumor cerebri.
   e. orthostatic hypotension.

132. The most common side effect of nonsteroidal antiinflammatory drugs (NSAIDs) is:
   a. myelosuppression.
   b. gastrointestinal (GI) upset.
   c. hepatic toxicity.
   d. corneal melt.
   e. punctate keratopathy.

133. T or F Diabetes mellitus can be diagnosed with a fasting blood sugar level of <126 mg/dL.

134. What percentage of Americans with diabetes have type 2 diabetes?
   a. 10%.
   b. 25%.
   c. 50%.
   d. 75%.
   e. 90%.

135. T or F Patients with impaired glucose tolerance are at risk for diabetic retinopathy and should be screened routinely.

136. T or F Most children diagnosed with cancer are alive after 5 years.

137. What is the difference between an oncogene and a tumor suppressor gene?
   a. An oncogene stimulates cell division, whereas a tumor suppressor gene slows down cell division.
   b. An oncogene slows tumor growth, whereas a tumor suppressor gene stops tumor growth.
   c. An oncogene stimulates apoptosis, whereas a tumor suppressor gene suppresses it.
   d. An oncogene is a direct inhibitor of a tumor suppressor gene.
   e. None of the above.

138. T or F In the United States, breast cancer is more common than lung cancer in women.

139. T or F In the United States, lung cancer kills more women each year than breast cancer.

140. The dose-equivalent average amount of radiation delivered to each person per year from human-made sources is approximately:
   a. 50 millirem (mrem).
   b. 100 mrem.
   c. 200 mrem.
   d. 300 mrem.
   e. 500 mrem.

141. The lethal dose 50% (LD50) of radiation from acute, whole body exposure in humans is approximately:
   a. 20 rads.
   b. 100 rads.
   c. 500 rads.
   d. 1,000 rads.
   e. 10,000 rads.
142. Which of the following statements about radiation retinopathy is false?
   a. Radiation retinopathy usually develops within 2 to 3 years after radiation exposure.
   b. Radiation retinopathy may develop earlier in patients who are undergoing chemotherapy or patients with diabetes.
   c. Radiation retinopathy is rare in individuals who have been exposed to <5,000 rads over 5 to 6 weeks.
   d. The earliest manifestation of radiation retinopathy is usually cotton-wool spots.
   e. The clinical findings of radiation retinopathy are very different from diabetic retinopathy.

143. Endostatin and angiostatin are types of ____________ that are undergoing clinical trials to treat cancer.
   a. angiogenesis inhibitors.
   b. monoclonal antibodies linked to radioisotopes.
   c. cancer vaccines.
   d. interferons.
   e. interleukins.

144. What percentage of cervical cancers are positive for human papillomavirus (HPV)?
   a. 5%.
   b. 25%.
   c. 50%.
   d. 75%.
   e. 95%.

145. The American Cancer Society recommends monthly breast self-examinations for women, starting after age:
   a. 20 years.
   b. 25 years.
   c. 30 years.
   d. 35 years.
   e. 40 years.

146. The most common cancer in men in the United States (in the order of decreasing prevalence) are:
   a. 1. lung, 2. prostate, 3. colorectal.
   b. 1. colorectal, 2. lung, 3. testicular.
   c. 1. lung, 2. testicular, 3. prostate.
   d. 1. colorectal, 2. lung, 3. prostate.
   e. 1. prostate, 2. lung, 3. colorectal.

147. The most common forms of cancer in women in the United States (in the order of decreasing prevalence) are:
   a. 1. lung, 2. breast, 3. colorectal.
   b. 1. colorectal, 2. lung, 3. breast.
   c. 1. lung, 2. breast, 3. cervical.
   d. 1. breast, 2. lung, 3. colorectal.
   e. 1. breast, 2. colorectal, 3. cervical.

148. Which of the following is not a risk factor for developing cervical cancer?
   a. low socioeconomic status.
   b. cigarette smoking.
   c. a high number of sexual partners.
   d. a large number of ovulations.
   e. a history of infection with human papillomavirus (HPV) type 16.

149. Which of the following is not a risk factor for developing breast cancer?
   a. a first-degree relative with breast cancer.
   b. a prior history of breast cancer.
   c. nulliparity or first pregnancy after the age of 30.
   d. early menarche.
   e. early menopause.

150. The incidence of prostate cancer is higher in African American males than the remainder of the US male population.

151. What percentage of patients with lung cancer in men are smokers?
   a. 5%.
   b. 10%.
   c. 30%.
   d. 70%.
   e. >70%.

152. Total and high-density lipoprotein (HDL) cholesterol screening should be performed in all patients, starting at age:
   a. 25 years.
   b. 30 years.
   c. 35 years.
   d. 40 years.
   e. 50 years.
153. Which of the following statements about colon cancer is false?
   a. A high-fiber, low-fat diet has been associated with a decreased risk in the development of colon cancer.
   b. Occult blood testing and annual sigmoidoscopy is recommended annually in individuals older than 50 years to detect colon cancer.
   c. An individual’s lifetime probability of developing colon cancer is 6%.
   d. An individual’s lifetime probability of dying from colon cancer is 3%.
   e. The mean 5-year survival rate for all colon cancer patients is <50%.

d. a. and b.
e. none of the above.

158. What preoperative tests are routinely recommended for general anesthesia in healthy men younger than age 40 years?
   a. electrocardiogram (ECG).
   b. complete blood count (CBC).
   c. creatinine.
   d. all of the above.
   e. no preoperative tests are recommended.

d. all of the above.

159. T or F Preoperative testing in patients undergoing elective cataract surgery has been shown to decrease perioperative morbidity and mortality.

d. T

e. F

160. Which of the following situations should prompt an ophthalmologist to delay elective eye surgery?
   a. a glucose level >300 mg/dL in a patient with diabetes who is fasting.
   b. a glucose level of 200 mg/dL in a fasting patient not known to have diabetes.
   c. a patient with a history of myocardial infarction (MI) who has electrocardiogram (ECG) changes on the day of surgery.
   d. a. and c.
   e. all of the above.

d. a. and c.
e. all of the above.

161. Which of the following statements about latex allergy in patients is incorrect?
   a. Patients with latex allergy having surgery should be the first case of the day in the operating room.
   b. Health care workers with a history of atopy have a higher risk of developing latex allergy.
   c. Patients with specific food allergies are more likely to have latex allergy as well.
   d. Patients who have undergone multiple urinary catheterizations and have myelodysplasia or spina bifida are more likely to have latex allergy than those without.
   e. Patients who have cystic fibrosis are more likely to have latex allergy.

d. a. and b.
e. none of the above.

162. Oral ingestion of clear liquids up to a minimum of _________ hours before surgery does not increase the risk of aspiration during surgery.
   a. 2.
   b. 3.
   c. 4.
163. Patients taking aspirin usually recover their platelet function within ____________ days after ceasing its use.
   a. 2.
   b. 4.
   c. 6.
   d. 8.
   e. 10.

164. Which of the following preoperative patients should absolutely not have their medication withheld on the day of cataract surgery despite being NPO (nothing by mouth)?
   a. a patient with hypothyroidism taking levothyroxine.
   b. a patient with bradycardia taking digoxin for congestive heart failure (CHF).
   c. a female patient with mitral valve prolapse taking an antibiotic for endocarditis prophylaxis.
   d. a patient taking insulin undergoing cataract surgery.
   e. a young anxious male patient with a history of seizures taking an anticonvulsant.

165. Echothiopate iodide eyedrops, although infrequently used, should prompt the anesthesiologist not to use ____________ during anesthesia.
   a. halothane.
   b. succinylcholine.
   c. lidocaine.
   d. benzocaine.
   e. mepivacaine.

166. Which of the following is a potential complication of Nadbath blocks?
   a. dysphagia.
   b. respiratory distress.
   c. urinary incontinence.
   d. a. and b.
   e. all of the above.

167. What is the earliest sign of malignant hyperthermia?
   a. elevated body temperature.
   b. tachycardia.
   c. metabolic acidosis.
   d. muscular rigidity.
   e. diaphoresis.

168. Treatment of malignant hyperthermia includes all of the following except:
   a. actively cooling the patient with ice and iced intravenous saline.
   b. hyperventilating the patient with 100% oxygen.
   c. stopping the administration of succinylcholine.
   d. giving dantrolene and calcium channel blockers.
   e. giving sodium bicarbonate.

169. T or F Patients with type 2 diabetes are prone to Graves’ disease, vitiligo, pernicious anemia, and Hashimoto’s thyroiditis.

170. Patients with type 1 diabetes without autoantibodies have:
   a. type 1 idiopathic diabetes.
   b. type 1 iatrogenic diabetes.
   c. type 2 diabetes.
   d. there are no patients with type 1 diabetes who do not have autoantibodies.
   e. none of the above.

171. Which of the following statements is false about gestational diabetes?
   a. A fasting glucose value of 126 mg/dL is one of the requirements needed to make the diagnosis.
   b. Four percent of all pregnancies in the United States are complicated by gestational diabetes.
   c. Within 10 years of diagnosis, 30% to 50% of women with gestational diabetes will develop type 2 diabetes.
   d. All of the above.
   e. None of the above.

172. Which of the following hormones does not increase plasma glucose concentration?
   a. cortisol.
   b. epinephrine.
   c. glucagon.
   d. thyroxine.
   e. none of the above; all of these hormones increase plasma glucose.

173. Which of the following describes the Somogyi phenomenon?
   a. high glucose concentration following episodes of hypoglycemia.
   b. hypoglycemia following episodes of very high glucose levels.
   c. normoglycemia following episodes of hypoglycemia.
d. hyperglycemia following insulin administration.
e. hypoglycemia following insulin administration.

174. __________ is the most common acute ocular complication of diabetes mellitus.
   a. Cataract.
   b. Clinically significant macular edema.
   c. Refractive error.
   d. Neovascularization of the optic disc.
   e. None of the above.

175. The glycosylated hemoglobin assay (HbA1c) target level in diabetics is:
   a. <6%.
   b. <7%.
   c. >6%.
   d. >7%.
   e. >8%.

176. The metabolically active form of thyroid hormone is:
   a. thyroxine (T4).
   b. 3,5,3'-triiodothyronine (T3).
   c. thyroglobulin.
   d. thyrotropin.
   e. thyrotropin-releasing hormone.

177. The most sensitive and specific screening test for thyroid disease is:
   a. free 3,5,3'-triiodothyronine (T3).
   b. free thyroxine (T4) and thyroid-stimulating hormone (TSH).
   c. transthyretin.
   d. radioactive iodine uptake.
   e. thyroid microsomal antibody detection.

178. 85% to 90% of patients with Graves's disease have antibodies to:
   a. thyroid stimulating hormone (TSH).
   b. TSH receptor.
   c. smooth muscle.
   d. 3,5,3'-triiodothyronine (T3).
   e. thyroxine (T4).

179. Changes in the aging eye include all of the following except:
   a. decreased corneal sensitivity.
   b. decreased visual field sensitivity.
   c. decreased contrast sensitivity.
   d. refractive error.
   e. lid retraction.
Answers

1. c. There is extensive benefit of normal microbial flora to humans through priming of the immune system, as well as excluding other pathogenic microorganisms from causing harm.

2. b. *Staphylococcus epidermidis* is found on >90% of skin cultures and is ubiquitous inhabitant of the skin. Acute *Staphylococcus* infections almost always require intravenous (IV) therapy. Only 25% of tertiary care isolates are resistant to beta-lactam antibiotics, although this number is increasing.

3. a. *Staphylococcus epidermidis* is the most common cause of prosthetic heart infections and also commonly causes cerebrospinal fluid (CSF) shunt and IV catheter infections. *Streptococcus viridans* is a common cause of prosthetic heart valve infections after dental procedures.

4. c. Pneumococcal pneumonia caused by *Streptococcus pneumoniae* can cause a severe pneumonia in elderly patients, whose mortality rate approaches 25%. Part of this may be because of the increasing resistance of *S. pneumoniae* to penicillin. In recent studies, penicillin resistance was >25%.

5. d. Patients with a history of coronary artery disease generally do not require endocarditis prophylaxis for invasive surgeries. In all of the other situations, endocarditis prophylaxis should be considered.

6. c. Endocarditis prophylaxis for ocular surgeries is usually not necessary except for cases involving the nasolacrimal drainage system or sinuses.

7. c. Pseudomembranous colitis most commonly occurs from *Clostridium difficile* after the administration of oral antibiotics. It usually occurs within 2 weeks of starting antibiotics, and patients complain of fever and diarrhea. Initial treatment involves discontinuing the antibiotic and administering metronidazole for 10 days. Vancomycin can be used in patients who are in their first trimester of pregnancy or those who cannot tolerate metronidazole.

8. a. Most strains of *Haemophilus influenzae* are resistant to macrolides. Fluoroquinolone resistance has also been observed.

9. d. Macrolides and quinolones are generally good agents to treat gonococcal infections. They have an added benefit in that they also can concomitantly treat *Chlamydia trachomatis* infection, which is often found in patients with concurrent gonococcal infections.

10. a. The treatment of choice for neurosyphilis is 2.4 million units of penicillin G Intramuscularly every 4 hours for 10 days. Early stage syphilis (e.g., within 1 year of infection) is treated with one dose of 2.4 million units of penicillin G Intramuscularly. The serum Venerale Disease Research Laboratory (VDRL) test result may be negative in patients with neurosyphilis, but the cerebrospinal fluid VDRL (CSF VDRL) test result is positive. False-positive VDRL test results can occur in patients with systemic lupus erythematosus (SLE), liver disease, pregnancy, or other treponemal infections.

11. c. *Ixodes scapularis* is the deer tick, which transmits the spirochete *Borrelia burgdorferi* to deer and humans. Lyme disease is the most common vector-borne disease in the United States.

12. b. This is the definition of a positive purified protein derivative (PPD).

13. a. Human immunodeficiency virus (HIV) is a retrovirus. All the other viruses belong to the herpesvirus family, which are double-stranded deoxyribonucleic acid (DNA) viruses.

14. c. Intravitreal and intraocular ganciclovir treatments are only effective against cytomegalovirus (CMV) retinitis and are not effective against other systemic manifestations of CMV infection.

15. b. Hepatitis B is parenterally or sexually transmitted.

16. d. By 2001, >35 million people in the world were living with human immunodeficiency virus (HIV)/acquired immunodeficiency syndrome (AIDS). Most of these individuals (>95%) live in developing countries. The enzyme-linked immunosorbent assay (ELISA) test is 99% sensitive and 99% specific for HIV, but false negatives can occur in the first few weeks after initial infection. Chemoprophylaxis is recommended when a large volume of infected blood or blood with a high HIV titer is transmitted during percutaneous occupational exposure.

17. False. Multidrug resistance is thought to be caused by inappropriate administration of antibiotics, poor compliance with antibiotic regimens, and sequential therapy of antibiotics. Combination therapy of antibiotics is felt to prevent the increasing incidence of multidrug resistance (MDR).

18. True. One of the most common causes of central retinal vein obstruction (CRVO) is hypertension.

19. a. The Seventh Report of the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure has defined stage 1 hypertension as a systolic blood pressure from 140 to 149 mm Hg and a diastolic blood pressure from 90 to 99 mm Hg. They have also added a “prehypertension” diagnosis of systolic of 120 to 139 mm Hg or diastolic of 80 to 89 mm Hg.

20. b. All of the other conditions listed are causes of secondary hypertension, which should be suspected in patients with increasing hypertension.

21. b. Cigarette smoking is associated with a 25-fold higher risk of coronary artery disease in patients with mild hypertension. In patients without hypertension, cigarette smoking increases the risk fivefold.

22. c. Hydralazine is commonly reported to have drug-induced lupus as a side effect. Doxazosin and other
alpha-blockers can result in postural hypotension. Beta-blockers often can cause bronchoconstriction and should be avoided in patients with asthma.

23. a. Beta-blockers and diuretics are the treatment of choice for initial hypertension, unless the patient has contraindications or for them. Angiotensin-converting enzyme (ACE) inhibitors or angiotensin II receptor blockers should be considered over calcium channel blockers in diabetics.

24. d. The average life expectancy according to the National Center for Health Statistics is 77.2 years as of 2001. Although heart disease is the leading cause of mortality in the United States, stroke is the leading cause of long-term disability in the United States.

25. a. A transient ischemic attack (TIA) lasts <24 hours but must be taken as a harbinger for repeat strokes. Answer b is the definition of a reversible ischemic neurologic disability and is differentiated from a TIA by the length of time that the patient experiences symptoms. Answer c defines a partial non-progressive stroke. Answer d is the definition of the penumbra, a term used to describe the ischemic area of brain tissue surrounding the main infarct. The penumbra has been shown to recover in some cases, leading to a more aggressive treatment of stroke than in the past.

26. b. Tissue plasminogen activator (TPA) is best used within 3 hours of onset of symptoms and is associated with improved outcomes in select patients. Unfortunately, TPA is often underutilized because of the lack of availability, awareness, and because patients can present to the hospital/emergency department several hours after the onset of symptoms.

27. True. Magnetic resonance imaging (MRI) is better at detecting early infarction, whereas computed tomography (CT) scan is better at detecting acute intracranial hemorrhage.

28. True.

29. a.

30. c.

31. c.

32. b. High homocysteine levels have been associated with an increased risk of stroke and vascular disease. Folic acid helps reduce homocysteine levels and can be recommended to all patients with cardiovascular or atherosclerotic disease.

33. c. Only 4% of individuals over the age of 40 have asymptomatic carotid bruits. Carotid endarterectomy (CEA) was shown to be increasingly effective in symptomatic patients (as defined by answer c) with higher levels of stenosis. CEA should be considered in symptomatic patients with carotid stenosis of >70% unless there is a contraindication to surgery, a completion of maximal neurologic deficit, or patients with acute stroke.

34. e.

35. b. Increased ventricular contractility will increase the demand for oxygen.

36. c. Prolonged ischemia will lead to myocardial infarction (MI) and then to cardiac necrosis.

37. a.

38. d. A family history of ischemic heart disease, hypertension, elevated serum cholesterol level, smoking, and diabetes are risk factors for ischemic heart disease.

39. False. Variant, or Prinzmetal, angina is not related to physical exertion, and occurs at rest. The anginal episodes are caused by coronary vasospasm, and underlying arteriosclerosis is present in 60% to 80% of cases.

40. b. Both unstable angina and non-Q-wave myocardial infarctions (MI) can have very similar presentations. The presence of elevated cardiac enzymes can serve to differentiate non-Q-wave MI from unstable angina.

41. c. Painless myocardial infarcts (MIs) are more common in patients with diabetes and elderly patients, and painless MI can present as syncope or congestive heart failure (CHF). Women can also present with atypical symptoms of an MI (e.g., stomach upset, malaise) instead of the classic “chest pain radiating down the arm” presentation.

42. d.

43. True. Consequently, patients with non-Q-wave MI should be monitored carefully and have aggressive management to prevent future infarction.

44. a. Cardiogenic shock is a severe complication of myocardial infarction (MI) and has a high mortality rate.

45. d. Dressler’s syndrome occurs after myocardial infarction (MI), typically 2 to 3 days later, and is characterized by pericarditis with fever, arthralgias, and pleural and pericardial pain. Dressler’s syndrome is typically treated with steroids, nonsteroidal antiinflammatory drugs (NSAIDs), or aspirin.

46. c. Arrhythmias, such as ventricular tachycardia or fibrillation, are usually the cause of sudden cardiac death. Other causes include torsade de pointes, Wolf-Parkinson-White syndrome, long QT’s syndrome, and pulmonary embolism.

47. True. The absence of electrocardiogram (ECG) changes does not preclude myocardial ischemia, and patients with anginal episodes can have a normal ECG in between episodes.

48. True. Cardiac troponins, unlike creatinine kinase MB, are not usually present in the serum of healthy individuals.

49. False. Cardiac troponins, unlike creatinine kinase MB, are not usually elevated after skeletal muscle injury.

50. b. Troponins can be used in patients who delay seeking medical attention when an infarct is suspected.

51. a.

52. True. A New England Journal of Medicine study demonstrated that the level of troponin I is directly associated with the likelihood of mortality at 42 days in patients with acute coronary syndrome.
53.  c.
54.  False. Patients with high-risk stable angina (e.g., those with severe angina, ejection fraction of <40%, probable three-vessel disease) may be considered for coronary artery bypass grafting (CABG) to treat stable angina.
55.  e. Short-acting rapid-release dihydropyridine calcium channel blockers such as nifedipine should not be used concomitantly with adequate beta-blockade because of the increased risk of cardiac death.
56.  a. Thrombolysis is preferable if it is anticipated that >90 minutes will elapse before percutaneous transluminal coronary angioplasty (PTCA) can be performed.
57.  e. Hepatomegaly, pedal edema, and cyanosis are signs of right-ventricular heart failure.
58.  c.
59.  e. All of the other answers listed can also be the causes of congestive heart failure (CHF), but ischemic heart disease (IHD) is the most common cause.
60.  d. All of the answers listed are causes of left-ventricular hypertrophy.
61.  e. All of the answers listed are potential causes of high-output congestive heart failure (CHF), where the heart eventually fails because it cannot sustain the prolonged excess cardiac output necessary to meet the excess demand for oxygen.
62.  d. Afterload refers to the level of pressure required by the ventricle in order to contract.
63.  a.
64.  c. Reducing afterload is usually the most effective way of treating systolic dysfunction. Angiotensin-converting enzyme (ACE) inhibitors are the medication of choice to accomplish this, although angiotensin receptor blockers, hydralazine, and other medications can be considered if the patient is unable to tolerate ACE inhibitors.
65.  b. Amlodipine has been shown to be safe in patients with congestive heart failure (CHF). Other calcium channel blockers such as diltiazem have shown to actually increase mortality in patients with CHF. Doxazosin and prazosin are alpha-adrenergic blockers. Captopril is an Angiotensin-converting enzyme (ACE) inhibitor (ACE inhibitors commonly end with the suffix “-pril”).
66.  a. Reducing preload is a common way of managing diastolic dysfunction. Diuretics are commonly used to achieve this.
67.  True. Corticosteroids and immunosuppressive agents have decreased the rate of rejection for cardiac transplants, leading to an increased survival rate.
68.  a. The sinoatrial (SA) node is the primary pacemaker of the heart and is located in the right atrium just inferior to the entrance to the superior vena cava.
69.  False. Sinus bradycardia is usually harmless and can be found in normal individuals and athletes. Treatment is usually not indicated.
70.  a. Answer b. defines second-degree Wenckebach atrioventricular (AV) block. Answer c. defines type II Mobitz AV block.
71.  a. All of the other choices are also potential ways to treat paroxysmal atrial tachycardia (PAT).
72.  a. Automated implantable cardioverter defibrillators (AICDs) are now the first-line therapy of choice to treat arrhythmias in patients with prior myocardial infarctions (MI) or prior episodes of hemodynamically unstable ventricular tachycardia.
73.  a.
74.  e. All of the answers listed are risk factors that would modify low-density lipoprotein (LDL) goals in patients. A high level of high-density lipoprotein (HDL) cholesterol is actually a “negative” risk factor and allows one to remove one risk factor from the total count. Anyone with coronary heart disease or diabetes should have an LDL of <100 mg/dL. Anyone with two or more risk factors listed in the answers should have an LDL goal of <130 mg/dL. Anyone with zero to one risk factor should have an LDL of <160 mg/dL.
75.  b. Beta-hydroxy-beta-methylglutaryl-coenzyme A (HMG-CoA) reductase inhibitors, also known as “statins” (e.g., atorvastatin), can cause diarrhea and increase liver transaminases. Baseline liver function tests as well as subsequent measurements should be taken in anyone who is starting a statin.
76.  False.
77.  c. High-density lipoprotein (HDL) cholesterol <40 mg/dL in men, and <50 mg/dL in women, is the other risk factor in the identification of metabolic syndrome, which is closely linked to the disorder of insulin resistance.
78.  True.
79.  True.
80.  c.
81.  a. Asthma is a reversible obstructive disease secondary to bronchospasm.
82.  a. The FEV₁ represents the volume of air exhaled forcefully in one second. An FEV₁ of <80% predicted suggests obstructive disease, such as asthma.
83.  True. Nasal continuous positive airway pressure (CPAP) is often used to treat obstructive sleep apnea. It has been shown in some studies to increase intraocular pressure (IOP) in patients with glaucoma.
84.  d. The selective beta-2 adrenergic agonists have a greater bronchodilatory effect with less cardiac stimulation.
85.  a. Subcutaneous epinephrine can be used to manage an acute asthma attack. Salmeterol is a long-acting beta-2 adrenergic agonist and should not be used to manage acute exacerbations. Cromolyn sodium is a mast cell stabilizer and is not used acutely; neither are inhaled steroids, such as beclomethasone.
86.  False. Theophylline is a bronchodilator with a narrow therapeutic index and is usually reserved for asthma.
cases that are not controlled with standard therapy of beta-2 agonists or ipratropium (an anticholinergic).

87. d. The Nocturnal Oxygen Therapy Trial (NOTT), a multicenter randomized trial, demonstrated that continuous low-flow oxygen therapy for patients with severe chronic obstructive pulmonary disease (COPD) resulted in improved survival.

88. d.
89. d.

90. True. Normal hemoglobin level ranges from 14 to 18 mg/dL in men and from 12 to 16 mg/dL in women.

91. d. Pernicious anemia is usually a megaloblastic anemia, where the mean corpuscular volume (MCV) is >95 femtoliters.

92. a. Transferrin receptor levels can be useful in differentiating iron deficiency anemias from anemia of chronic disease, although patients with anemia of chronic disease can also have a concomitant iron deficiency anemia.

93. a. Sickle cell disease affects 0.2% to 0.4% of all African Americans in the United States.

94. d. Thrombin is an enzyme that converts fibrinogen into fibrin and is crucial for coagulation and hemostasis.

95. b.

96. d. Although the prothrombin time (PT) does change with warfarin, the international normalized ratio (INR) is a more useful test to measure the therapeutic effect of warfarin. For deep vein thromboses and tissue replacement heart valves, the INR typically is maintained between 2.0 and 3.0; for mechanical prosthetic replacement heart valves, the INR is maintained between 2.5 and 3.5.

97. c. Blood tests are not typically used to measure the effects of low-molecular-weight heparin.

98. c. Aspirin will cause an increase in bleeding time for at least 48 hours after ingestion.

99. False. Many connective tissue disorders are associated with hemorrhage, such as osteogenesis imperfecta (easy bruising) and pseudoxanthoma elasticum (gastrointestinal [GI] bleeding).

100. a. Scurvy is characterized by vascular fragility and systemic hemorrhages secondary to deficient collagen synthesis.

101. a. Platelet destruction in thrombotic thrombocytopenic purpura (TTP) is mediated by a nonimmunologic mechanism; all of the other answers are immunologic causes.

102. c. Many different types of medications can affect platelet function, and drugs are the most common cause of platelet dysfunction.

103. a.

104. b. Malabsorption syndromes can decrease the levels of fat-soluble vitamins (A, D, E, and K). Vitamin C is water-soluble.

105. b. Vitamin K should not be given intravenously because of the risk of an anaphylactoid reaction.

106. True. All of the other primary hypercoagulable states (e.g., protein C and S deficiency, antithrombin III deficiency) are only associated with venous thrombosis.

107. e. All of the answers listed can be ophthalmic complications of the phospholipid antibody syndrome, which can manifest with recurrent spontaneous abortions, deep venous thrombosis, and cerebrovascular arterial thrombosis.

108. d.

109. b. Patients with Felty’s syndrome can have hyperpigmented skin and recurrent infections.

110. a. Bull’s-eye maculopathy is a common complication of hydroxychloroquine which is used to treat rheumatoid arthritis.

111. c. Nonsteroidal antiinflammatory drugs (NSAIDs) are usually first-line agents to treat rheumatoid arthritis, followed by slow-acting antirheumatic drugs (SAARDs). Of the SAARDs, chlorambucil and cyclophosphamide are the most toxic.

112. a. Leflunomide is used to treat rheumatoid arthritis and inhibits pyrimidine synthesis, targeting rapidly dividing cells.

113. b. Etanercept and infliximab are used to treat rheumatoid arthritis, and both are inhibitors of tumor necrosis factor-alpha.

114. e. The seronegative spondylopathies (ankylosing spondylitis, reactive arthritis, enteropathic arthritis, etc.) all share an association with human leukocyte antigen (HLA)-B27. They are termed seronegative because of the lack of IgM antibodies to rheumatoid factor in serum.

115. True. The most common ophthalmic finding in patients with seronegative spondylopathies is a nongranulomatous anterior uveitis.

116. c. Most patients with Reiter’s syndrome present with arthritis alone. The triad of Reiter’s syndrome (also known as reactive arthritis) is conjunctivitis (which is the most common ophthalmic manifestation), urethritis, and arthritis.

117. True.

118. a. Whitting of the distal phalanges is seen in psoriatic arthritis. Fusion of the spine is common with ankylosing spondylitis. Heel pain and sausage digits are found in Reiter’s syndrome.

119. False. Still’s disease is a form of juvenile rheumatoid arthritis (JRA) that usually affects young children and is typically not associated with ophthalmic complications but, rather, has systemic symptoms including fever, a maculopapular rash, and polyarthritis.

120. d. Photosensitivity, rather than photophobia, is one of the criteria listed in diagnosing systemic lupus erythematosus (SLE). Other criteria include malar rash, discoid lupus, renal disorder, neurologic disorder, hematologic disorder, immunologic disorder. Four of these criteria are required to make the diagnosis of SLE.

121. b. Retinal vascular lesions are the most common complication of systemic lupus erythematosus (SLE). All
of the other answers listed are also potential complications of SLE.

122. True.

123. b. CREST (calcinosis, the Raynaud phenomenon, esophageal involvement, sclerodactyly, and telangiectasias) is a limited form of systemic scleroderma. Renal involvement is often associated with malignant hypertension and can be a major cause of mortality in patients with scleroderma. Scleroderma can also have ophthalmic manifestations, including blepharophimosis, keratoconjunctivitis sicca, and hypertensive retinopathy secondary to scleroderma kidney.

124. b. A heliotrope rash around the eyelids (violaceous erythema) and Gottron's papules (plaques on finger knuckles) are seen in dermatomyositis.

125. b. Sjögren's syndrome is characterized by rheumatoid arthritis, dry mouth, and dry eyes. The dry mouth and dry eyes is because of an inflammatory infiltrate into the lacrimal and salivary glands.

126. d. Autoantibodies to collagen leads to relapsing polychondritis. Ocular complications are found in 50% of patients with relapsing polychondritis.

127. c. Cogan's syndrome is a constellation of hearing loss, tinnitus, vertigo, and interstitial keratitis. It is associated with polyanthritis nodosa (PAN) in up to 50% of cases.

128. e. Temporal artery biopsy is the gold standard for diagnosing giant cell arteritis (GCA).

129. e.

130. a. Oral ulcers are found in >95% of patients with Behçet's disease.

131. e. Orthostatic hypotension can be an effect of rapid exogenous steroid withdrawal, which can lead to adrenal insufficiency and can be fatal in severe cases.

132. b. Corneal melts and punctate keratopathy have been reported to be associated with topical nonsteroidal antiinflammatory drug (NSAID) administration. The incidence of gastrointestinal (GI) complications is reduced with cycloxygenase-2 selective NSAIDs.

133. True. Diabetes mellitus can also be diagnosed in patients with a fasting blood sugar of <126 mg/dL, in patients with a plasma glucose level of 200 mg/dL or greater after a 75-mg 2-hour oral glucose tolerance test.

134. e. Type 2 diabetes is by far the most common form of diabetes mellitus.

135. False.

136. True. The 5-year survival rate for all childhood cancers is approximately 70%; this represents an increase from approximately 50% in the 1970s.

137. a. Oncogenes and tumor suppressor genes normally work together to allow the body to repair damaged cells or replace dead cells.

138. True. Breast cancer is the most common cancer in women.

139. True. Although breast cancer is more prevalent, lung cancer causes more mortality in women than breast cancer.

140. b. The average amount of radiation delivered per person per year from human-made sources such as medical diagnostic equipment, global fallout from atomic testing, and nuclear power is 106 mrem per person per year. The average dose of natural radiation per person per year living at sea level is 80 mrem per person per year. Individuals that live at high altitudes receive more natural radiation than those living at sea level.

141. c. The lethal dose 50% (LD50) is the lethal dose of radiation for which 50% of a sample exposed population would die. The LD50 in humans is approximately 450 rads (4.5 Gy).

142. e. Radiation retinopathy is very similar in clinical presentation to diabetic retinopathy. Most patients exposed to >8,000 rads develop radiation retinopathy.

143. a. Angiogenesis inhibitors are a new class of drugs which are being studied to starve tumors by cutting off their blood supply. They are also of interest in proliferative vascular disorders of the eye such as diabetic retinopathy. All of the other answers listed are also being studied as forms of cancer therapy other than chemotherapy or radiation.

144. e.

145. a. Breast physical examinations should be done every 3 years starting at age 20 years, and every year, starting at age 40 years. Mammography should be done as a baseline between ages 35 to 40 years, every 1 to 2 years until age 50 years, and then annually after age 50 years.

146. a.

147. d. Breast cancer is the most common cancer in women, and the most common cause of death in women older than 40 years. Cervical cancer is the most common cancer in women aged 15 to 34 years.

148. d. Human papillomavirus (HPV) types 16, 18, 30, 31, and 33 are associated with cervical cancer. A history of other sexually transmitted diseases, especially Chlamydia, is also a risk factor for cervical cancer.

149. e. Late menopause is a risk factor for breast cancer. Approximately 10% of all women will develop breast cancer in their lifetime in the United States. More than 75% of breast cancer is cured with current therapy in the United States.

150. True. Prostate cancer is more prevalent in the African American community and is detected through digital examination of the prostate and prostate-specific antigen (PSA) measurements. Annual PSA measurements are recommended in men older than 50 years, and annual rectal examinations in men older than 40 years.

151. e. Smoking kills.

152. a. If the total cholesterol value is normal (<200 mg/dL) and the high-density lipoprotein (HDL) is normal, then patients should have their cholesterol rechecked every 5 years. Otherwise, they should obtain a complete cholesterol profile and be checked for dyslipoproteinemia.

153. b. Sigmoidoscopy is recommended every 3 years in healthy adults older than 50 years.
154. e. The incidence of melanoma is increasing more rapidly than all other cancers, and melanoma is the deadliest form of skin cancer. Asymmetric, raised large (>6 mm) lesions with irregular borders, variable color are suspicious for melanoma. Early detection and removal are crucial for survival.

155. b. The prevalence of tuberculosis is on the rise in the United States. Occupational Health and Safety Administration (OSHA) requires annual screening with purified protein derivative (PPD) testing of all health care workers annually.

156. d. Live attenuated vaccines are contraindicated in pregnant women because of the risk to the fetus. Both the rubella vaccine and the oral polio (Sabin) vaccine are live, attenuated viral vaccines. The Salk vaccine is a subcutaneous killed polio vaccine.

157. d. Diphtheria, tetanus, and pertussis (DTP), which has replaced diphtheria, tetanus, and pertussis (DTP) in the United States, should not be given to anyone 7 years of age or older because of the risk of neurologic complications from the pertussis component.

158. e. Healthy young men do not routinely require preoperative testing before general anesthesia.

159. False. This is relatively controversial, but a recent study from the New England Journal of Medicine of >19,000 patients showed that there was no significant benefit of preoperative testing for elective cataract surgery.

160. e. High glucose levels or the discovery of new-onset diabetes in patients should prompt the delay of eye surgery. Any changes in electrocardiogram (ECG) in patients with a history of myocardial infarcts (MIs) should also delay elective surgery.

161. e. There is no reported association between cystic fibrosis and latent allergy. Food allergies to specific foods including bananas and avocados have been associated with latex allergy.

162. a. In fact, preoperative children who have been kept nothing by mouth (NPO) for >12 hours may become dehydrated and hypotensive.

163. d. Because aspirin irreversibly inhibits cyclooxygenase, an 8-day waiting period is recommended before elective surgery.

164. e. Anticonvulsants should not be withheld preoperatively because the stress of surgery and general anesthesia can lower the seizure threshold. Digoxin should not be withheld if the resting heart rate is >90 bpm. Patients with mitral valve prolapse without regurgitation do not require prophylactic antibiotics for subacute bacterial endocarditis (SBE). Patients with diabetes undergoing short surgeries can withhold their insulin.

165. b. Echotoxiphate iodide is an iatrogenic cause of lowered pseudocholinesterase activity, and can lead to inadequate breakdown of succinylcholine causing prolonged paralysis during anesthesia. Patients who are taking echotoxiphate should always inform the anesthesiologist of this before surgery.

166. d. Nadjbath blocks are given into the stylomastoid foramen to provide facial akinesia. Unintentional anesthesia of cranial nerves IX, X, and XI can lead to dysphagia and respiratory distress.

167. b. Tachycardia and elevated carbon dioxide levels at end-tidal volumes are the earliest signs of malignant hyperthermia. This disorder can be fatal if diagnosis is delayed.

168. d. Calcium channel blockers should not be given with dantrolene because of the risk of myocardial depression and hyperkalemia. Dantrolene and sodium bicarbonate should be given. All of the other answers are correct.

169. False. Patients with type 1 disease, which in most cases is caused by autoantibodies leading to beta-cell destruction, are prone to other autoimmune diseases.

170. a. Most patients with type 1 idiopathic diabetes mellitus are of Asian or African descent.

171. a. Gestational diabetes is defined as any degree of glucose intolerance diagnosed during pregnancy. A glucose tolerance test can be used to make the diagnosis.

172. e. The only hormone that decreases glucose levels is insulin. Other hormones that increase plasma glucose levels include somatotropin and adrenocorticotropin.

173. a. The Somogyi phenomenon occurs when episodes of hypoglycemia (glucose levels <30 mg/dL) are followed by hyperglycemia. Growth hormone and catecholamines are thought to cause the Somogyi phenomenon.

174. c. The other answers are chronic complications of diabetes mellitus.

175. b. Glycosylated hemoglobin is a measure of the mean glucose levels over the previous 2 to 3 months. Target levels are <7% in patients with diabetes.

176. b. 3,3',5'-triiodothyronine (T3) is the metabolically active thyroid hormone.

177. b. This has a sensitivity and specificity of >95%.

178. b. Graves’ disease is more common in women (a 10:1 female-to-male ratio) and the incidence peaks in the mid-30s.

179. e. Lid laxity and ptosis are common changes in the elderly patients. All of the other answers are common changes in the aging eye. Lid retraction is often seen in Graves’ ophthalmopathy.

Suggested Readings


1. Which one of the following bones does not form part of the orbit?
   a. palatine.
   b. frontal.
   c. zygomatic.
   d. nasal.
   e. ethmoid.

2. What is the average volume of the human orbit?
   a. 30 mL.
   b. 35 mL.
   c. 40 mL.
   d. 45 mL.
   e. 50 mL.

3. How many bones compose the lacrimal sac fossa?
   a. 1.
   b. 2.
   c. 3.
   d. 4.
   e. none—a fossa is a hole.

4. In which bone does the lacrimal gland fossa lie?
   a. frontal.
   b. lacrimal.
   c. maxillary.
   d. zygomatic.
   e. ethmoid.

5. Which orbital wall is the strongest?
   a. medial.
   b. inferior.
   c. lateral.
   d. superior.
   e. all are equally strong.

6. Which one of the following structures does not insert into the lateral orbital tubercle of Whitnall?
   a. check ligament of the lateral rectus.
   b. suspensory ligament of the eyeball (Lockwood’s ligament).
   c. lateral canthal tendon.
   d. aponeurosis of the levator muscle.
   e. Whitnall’s ligament.

7. Which of the following is/are not transmitted in the optic canal?
   1. optic nerve.
   2. ophthalmic artery.
   3. sympathetic plexus.
   4. central retinal artery.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

8. How many axons compose a healthy adult optic nerve?
   a. 100,000.
   b. 300,000.
   c. 600,000.
   d. 1.2 million.
   e. 2.4 million.
9. On entry to the orbit, the ophthalmic artery turns in what relation relative to the optic nerve?
   a. inferior.
   b. lateral.
   c. superior.
   d. medial.
   e. variable.

10. The ciliary ganglion:
   a. has three roots, is located 1 cm posterior to the globe, and is medial to the optic nerve.
   b. has four roots, is located 1 cm posterior to the globe, and is lateral to the optic nerve.
   c. has four roots, is located 1 cm anterior to the orbital apex, and is medial to the optic nerve.
   d. has three roots, is located 1 cm posterior to the globe, and is lateral to the optic nerve.
   e. has three roots, is located 1 cm anterior to the orbital apex, and is lateral to the optic nerve.

11. T or F There is dual sympathetic innervation to the eye.

12. Which of the following statements about ocular glands is/are true?
   1. The glands of Wolfring and Krause are cytologically similar to the lacrimal gland.
   2. Almost all reflex tear secretion is produced by the accessory lacrimal glands.
   3. The glands of Moll are of the apocrine type.
   4. The meibomian glands are holocrine oil glands that are associated with cilia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

13. T or F The tarsal plates consist of cartilage.

14. T or F The smaller orbital lobe of the lacrimal gland is located posterior to the larger palpebral lobe.

15. T or F The canaliculi and lacrimal sac are lined by identical epithelia.

16. What are the average adult corneal diameters?
   a. 11 mm horizontally and vertically.
   b. 12 mm horizontally and vertically.
   c. 12 mm horizontally and 11 mm vertically.
   d. 11 mm horizontally and 12 mm vertically.
   e. 10 mm horizontally and vertically.

17. T or F The corneal endothelium reacts to injuries or cellular loss with hyperplasia.

18. T or F The sclera is highly vascularized.

19. The correct order of angle structures, from central to peripheral, is:
   a. scleral spur (SS), pigmented trabecular meshwork (TM), the Schwalbe line (SL), ciliary body band (CBB).
   b. SL, pigmented TM, nonpigmented TM, CBB, SS.
   c. SL, pigmented TM, nonpigmented TM, SS, CBB.
   d. SL, SS, pigmented TM, nonpigmented TM, CBB.
   e. SL, nonpigmented TM, pigmented TM, SS, CBB.

20. T or F The trabecular meshwork (TM) is lined by a bilayered endothelium.

21. The site of greatest resistance to aqueous outflow is the:
   a. uveal meshwork.
   b. corneoscleral meshwork.
   c. anterior, nonpigmented trabecular meshwork (TM).
   d. posterior, pigmented TM.
   e. juxtacanalicular TM.

22. The average anteroposterior diameter of a 40-year-old lens is:
   a. 2 to 3 mm.
   b. 3 to 4 mm.
   c. 4 to 5 mm.
   d. 5 to 6 mm.
   e. 6 to 7 mm.

23. The average equatorial diameter of a 40-year-old lens is:
   a. 6 to 7 mm.
   b. 7 to 8 mm.
   c. 8 to 9 mm.
   d. 9 to 10 mm.
   e. 10 to 11 mm.
24. T or F The anteroposterior lens diameter does not change throughout life.

25. T or F The equatorial diameter of the lens does not change throughout life.

26. T or F The posterior capsule is thicker than the anterior capsule.

27. T or F The anterior iris surface is lined by a single-layered endothelium.

28. Choose the one correct statement about the iris from the following:
   a. The iris is composed of six layers, three of which are epithelial.
   b. Clump cells are part of the anterior border layer of the iris.
   c. The iris dilator and iris sphincter are posterior to the iris pigment epithelium.
   d. The cell bodies of the anterior iris pigment epithelium give rise to the iris dilator muscle.
   e. The color of the iris is determined by the number and size of macrophages in the iris pigment epithelium.

29. Select the correct description of autonomic innervation to the eye:
   a. The iris sphincter muscle receives sympathetic innervation via the short ciliary nerves; the iris dilator muscle receives parasympathetic innervation via the short ciliary nerves.
   b. The iris sphincter muscle receives parasympathetic innervation via the short ciliary nerves; the iris dilator muscle receives sympathetic innervation via the short ciliary nerves.
   c. The iris sphincter muscle receives parasympathetic innervation via the short ciliary nerves; the iris dilator muscle receives sympathetic innervation via the long ciliary nerves.
   d. The iris sphincter muscle receives parasympathetic innervation via the long ciliary nerves; the iris dilator muscle receives sympathetic innervation via the long ciliary nerves.
   e. The iris sphincter muscle receives sympathetic innervation via the short ciliary nerves; the iris dilator muscle receives parasympathetic innervation via the long ciliary nerves.

30. T or F The choriocapillaris is a continuous vascular system with virtually limitless anastomoses between its different regions.

31. T or F The degree of pigmentation observed ophthalmoscopically in any human fundus depends on the amount of pigmentation of the retinal pigment epithelium (RPE).

32. T or F Retinal pigment epithelium (RPE) cells in the fundus periphery are taller, more closely packed, and contain more and larger melanosomes than those in the posterior fundus.

33. Which one of the following statements about retinal photoreceptors is correct?
   a. Rods contain photopigment discs that are not attached to the cell membrane and synapse with bipolar cells at a rod pedicle.
   b. Cones contain photopigment discs that are not connected to the cell membrane and synapse with bipolar cells at a cone pedicle.
   c. Rods contain photopigment discs that are attached to the cell membrane and synapse with bipolar cells at the rod spherule.
   d. Cones contain photopigment discs that are attached to the cell membrane and synapse with bipolar cells at a cone pedicle.
   e. None of the above.

34. Select the correct neuronal sequence for intraretinal processing:
   a. photoreceptor to Müller’s cell to ganglion cell.
   b. photoreceptor to bipolar cell to ganglion cell.
   c. photoreceptor to horizontal cell to amacrine cell.
   d. photoreceptor to horizontal cell to ganglion cell.
   e. photoreceptor to amacrine cell to bipolar cell.

35. In the entire retina, rods outnumber cones by a ratio of approximately:
   a. 2:1.
   b. 5:1.
   c. 20:1.
   d. 50:1.
   e. 1.25:1.
36. Which one of the following statements about Müller’s cells is correct?
   a. The external limiting membrane (ELM) and internal limiting membrane (ILM) of the retina are the basement membranes of Müller’s cells.
   b. Müller’s cells are the only nonneural (glial) cellular element found within the neural retina.
   c. Müller’s cells do not generate any detectable light-induced transretinal voltages.
   d. Müller’s cells intimately envelop virtually all retinal neurons.
   e. None of the above.

37. A cilioretinal artery contributes to some portion of the macular circulation in approximately:
   a. 5% of individuals.
   b. 15% of individuals.
   c. 25% of individuals.
   d. 40% of individuals.
   e. 50% of individuals.

38. The inner retinal circulation’s deepest level of penetration is the:
   a. ganglion cell layer.
   b. inner plexiform layer.
   c. inner nuclear layer.
   d. outer plexiform layer.
   e. outer nuclear layer.

39. Select the correct association of retinal layer and synaptic connections:
   a. outer plexiform layer: bipolar and ganglion cells.
   b. inner plexiform layer: bipolar and ganglion cells.
   c. outer plexiform layer: photoreceptor and ganglion cells.
   d. inner plexiform layer: photoreceptor and ganglion cells.
   e. inner plexiform layer: photoreceptor and bipolar cells.

40. Which of the following sites represent firm uveoscleral attachment?
   1. optic nerve (peripapillary tissue).
   2. vortex veins.
   3. scleral spur (SS).
   4. ora serrata.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

41. T or F The ora is smoother (i.e., fewer serrations or teeth) nasally than temporally.

42. T or F Myelinated nerve fibers in the retina result when Schwann’s cells migrate beyond the lamina cribrosa to form a myelin sheath around ganglion cell axons.

43. On entering the cranial cavity, the optic nerve runs:
   a. lateral to the internal carotid artery and inferior to the anterior cerebral artery.
   b. medial to the internal carotid artery and inferior to the anterior cerebral artery.
   c. medial to the internal carotid artery and superior to the anterior cerebral artery.
   d. lateral to the internal carotid artery and superior to the anterior cerebral artery.
   e. lateral to the internal carotid artery and lateral to the anterior cerebral artery.

44. Which of the following statements about ganglion axon decussation is true?
   a. More fibers cross in the chiasm than do not cross in the chiasm.
   b. Fewer numbers of ganglion cells cross than do not cross in the chiasm.
   c. A much greater proportion of macular fibers cross in the chiasm than peripheral fibers do.
   d. A much greater portion of peripheral fibers cross in the chiasm than macular fibers do.
   e. None of the above.

45. Which one of the extraocular muscles is served by a single nucleus that is shared by both the oculomotor nerve nuclei?
   a. superior rectus.
   b. medial rectus.
   c. inferior oblique.
   d. levator palpebrae superioris.
   e. inferior rectus.

46. Which is the only muscle supplied by the oculomotor nerve that receives crossed innervation?
   a. superior rectus.
   b. medial rectus.
   c. inferior oblique.
Questions

d. levator palpebrae superioris.
e. inferior rectus.

47. Which one of the following statements about the pupillomotor fibers of the third cranial nerve is true?
   a. These fibers run central in the nerve, in the superior division.
   b. These fibers run central in the nerve, in the inferior division.
   c. These fibers run peripheral in the nerve, in the superior division.
   d. These fibers run peripheral in the nerve, in the inferior division.
   e. These fibers are distributed evenly throughout the superior division.

48. T or F The fifth cranial nerve has both motor and sensory components.

49. Which of the following structures do not travel in the wall of the cavernous sinus at the level of the pituitary fossa?
   a. third and fourth cranial nerves.
   b. fourth and fifth cranial nerves.
   c. fifth and sixth cranial nerves.
   d. sixth cranial nerve and the internal carotid artery.
   e. the internal carotid artery only.

50. T or F Ocular colobomas arise from failure of fusion of the lips of the optic cup, most typically in a superotemporal location.

51. T or F The retinal pigment epithelium (RPE) cells are the first cells in the body to produce melanin.

52. T or F Myelination of optic nerve axons to the lamina cribrosa in an infant is rarely completed before 6 months of age.

53. T or F The lens of the eye develops from neuroectoderm.

54. Mesenchymal structures of the head, including the eye, are all derived from:
   a. mesoderm.
   b. neural crest cells.
   c. a combination of mesoderm and ectoderm.

   d. a combination of neural crest cells and ectoderm.
e. a combination of neural crest cells and mesoderm.

55. Neural crest cells give rise to the following structures:
   a. corneal epithelium, stroma, endothelium, and ciliary muscle.
   b. the entire sclera, optic nerve sheath, uveal melanocytes, entire choroid.
   c. orbital bones, fat, trochlear cartilage, extraocular muscles, and orbital connective tissues.
   d. ciliary body, ciliary epithelium, iris stroma, orbital bones, and orbital connective tissues.
   e. optic nerve sheath, uveal melanocytes, choroidal stroma, ciliary muscle, and iris stroma.

56. The mesoderm gives rise to:
   a. the pupillomotor muscles, ciliary muscle, and extraocular muscles.
   b. all vascular endothelia, extraocular muscles, and the trochlea.
   c. all vascular endothelia, pupillomotor muscles, and all blood vessels.
   d. all vascular endothelia, all extraocular muscles, and temporal sclera.
   e. ciliary muscle, extraocular muscles, all orbital blood vessels, and some orbital connective tissue.

57. The surface ectoderm supplies all of the following structures except:
   a. the lacrimal gland.
   b. the lens.
   c. the vitreous.
   d. the corneal epithelium.
   e. the substantia propria of the conjunctiva.

58. T or F The primary vitreous is slowly displaced peripherally as secondary vitreous develops centrally.

59. What factor distinguishes anophthalmia from microphthalmia?
   a. the size of the globe.
   b. the presence or absence of a globe.
   c. the presence or absence of lid fusion.
   d. the presence or absence of organic abnormalities of a globe.
   e. the presence or absence of nervous system disorders.
60. What factor distinguishes microphthalmia from nanophthalmia?
   a. the size of the globe.
   b. the presence or absence of a globe.
   c. the presence or absence of lid fusion.
   d. the presence or absence of organic abnormalities of a globe.
   e. the presence or absence of nervous system disorders.

61. All of the following conditions may present as a cystic bulge within the palpebral fissure except:
   a. cystic coloboma.
   b. orbital encephalocele.
   c. microphthalmos.
   d. nanophthalmos.
   e. congenital rhabdomyosarcoma.

62. Which one of the following statements about dermoids is false?
   a. Dermoids represent hamartomas of epidermal and connective tissues.
   b. Dermoids may be found in the conjunctiva and/or in the orbit.
   c. The solid variety of the dermoids is most frequently found at the limbus.
   d. Dermolipomas are usually solid, most commonly located between the lateral and superior rectus muscles, and often lack dermal adnexal structures (unlike a dermoid).
   e. Epibulbar dermoids are associated with Goldenhar’s syndrome.

63. Which one of the following statements about anterior segment dysgeneses is false?
   a. All varieties of anterior segment dysgeneses may be inherited as autosomal-dominant traits and may be either unilateral or bilateral.
   b. Posterior embryotoxon is the mildest of the peripheral varieties of anterior segment dysgeneses.
   c. Rieger’s anomaly consists of Axenfeld’s anomaly plus iris atrophy.
   d. Rieger’s syndrome is Rieger’s anomaly plus facial and musculoskeletal anomalies.
   e. In Peters’ anomaly, the central cornea is always opacified, and the lens is always densely adherent to the posterior corneal surface.

64. Which one of the following statements about aniridia is false?
   a. Generally, no iris tissue is present on histopathology.
   b. Aniridia may be either familial or sporadic.
   c. Foveal and optic nerve hypoplasia are often present in aniridia, leading to nystagmus and reduced visual acuity.
   d. There is an association of aniridia with Wilms’ tumor, genitourinary anomalies, and mental retardation.
   e. In aniridia associated with Wilms’ tumor, there is frequently a deletion on the short arm of chromosome 11.

65. Which one of the following statements about pediatric lenticular disorders is true?
   a. Microspherophakia is most often associated with Marfan’s syndrome.
   b. In Lowe’s syndrome, there are typically cataracts, glaucoma, and aminoaciduria, and girls are affected more frequently than boys.
   c. Cataracts in the congenital rubella syndrome are generally dense nuclear cataracts.
   d. Defects in the rubella syndrome are typically isolated (i.e., there are usually no other systemic abnormalities).
   e. Glaucoma and cataracts are frequently coincident in the congenital rubella syndrome.

66. Which one of the following statements about persistent hyperplastic primary vitreous (PHPV) is false?
   a. PHPV is generally unilateral.
   b. Long-term visual prognosis is usually excellent.
   c. PHPV is most easily differentiated from retinoblastoma by the presence of microphthalmos or cataract.
   d. PHPV may calcify.
   e. PHPV is almost always sporadic.

67. Which one of the following statements about tear secretion is false?
   a. Adrenocorticotropic hormone (ACTH) and androgens can stimulate tear secretion from the main lacrimal gland.
   b. The glands of Wolfring are located along the orbital margin of each tarsus, with the glands of Krause in the conjunctival fornix.
c. The accessory glands of Krause and Wolfring account for approximately 50% of total lacrimal secretory mass.

d. Both sympathetic and parasympathetic nerve stimuli are important for reflex tear secretion.

e. The cornea, conjunctiva, and meibomian glands also contribute to the tear film.

68. Which one of the following statements about corneal metabolism is false?

a. Oxygen is provided to the cornea by tears, aqueous humor, and lid vasculature.

b. Most of the metabolic fuel for the cornea is derived from aqueous humor.

c. Molecules with low lipid solubility have greater penetration through the corneal epithelium and stroma.

d. Mature corneal stromal fibers are composed of type I collagen.

e. The type and distribution of glycosaminoglycan in the cornea are crucial for corneal clarity.

69. Which one of the following statements about Descemet’s membrane and corneal endothelium is false?

a. Descemet’s membrane consists of type IV collagen.

b. Posterior keratoconus can be differentiated from Peters’ anomaly by the presence of fetal Descemet’s membrane.

c. Fetal Descemet’s membrane can be differentiated from adult Descemet’s membrane by its banding pattern.

d. The number of endothelial cells substantially increases with age.

e. The corneal endothelium actively maintains corneal deturgescence by a pump system that depends on Na⁺/K⁺-adenosine triphosphatase (ATPase) function and carbonic anhydrase.

70. Aqueous humor enters the posterior chamber from the ciliary processes by:

a. diffusion.

b. ultrafiltration.

c. secretion/active transport.

d. carbonic anhydrase II activity.

e. all of the above.

71. Which one of the following statements about prostaglandins is false?

a. The cyclooxygenase (COX) reaction culminates in the production of prostaglandins, prostacyclin, and thromboxane.

b. In general, prostaglandins cause mydriasis.

c. Corticosteroids inhibit both the COX and lipoygenase (LOX) pathways of arachidonic acid metabolism.

d. The effect of prostaglandins on intraocular pressure (IOP) is complex, with low doses decreasing IOP and high doses increasing IOP.

e. Prostacyclin is a potent vasodilator synthesized primarily in endothelial cells, whereas thromboxane is a potent vasoconstrictor synthesized primarily in platelets.

72. Which of the following enzymes is/are normally present at functioning levels in the aqueous humor?

1. carbonic anhydrase.

2. lysozyme.

3. hyaluronidase.

4. lactate dehydrogenase.

a. 1, 2, and 3.

b. 1 and 3.

c. 2 and 4.

d. 4 only.

e. 1, 2, 3, and 4.

73. The partial pressure of oxygen in aqueous humor is:

a. 40 mm Hg.

b. 55 mm Hg.

c. 75 mm Hg.

d. 85 mm Hg.

e. 100 mm Hg.

74. The most mitotically active lens epithelium cells are located:

a. at the anterior pole (AP).

b. at the posterior pole.

c. at the equator.

d. in a ring around the anterior lens.

e. in a ring around the posterior lens.
75. Which one of the following statements about the protein fractions of the lens is false?
   a. Lens fiber proteins are separated into two major groups—water-soluble and water-insoluble.
   b. The water-insoluble fraction is further divided into three types of crystallin proteins.
   c. Alpha-crystallin is the largest of the lens crystallins.
   d. Beta-crystallin is the most abundant of the lens crystallins.
   e. Most crystallins are expressed outside the lens and have other roles in addition to their function as refractile elements within the lens.

76. T or F There is an active pump mechanism in the lens that drives ionic movements across the lens, both anteroposteriorly and posteroanteriorly.

77. T or F The lens will opacify if deprived of glucose.

78. T or F The lens will opacify if deprived of oxygen.

79. The vitreous syneresis of aging is associated with:
   a. diffuse decreases in hyaluronic acid concentration.
   b. diffuse decreases in collagen concentration.
   c. focal decreases in collagen concentration.
   d. motion-induced collagen damage.
   e. light-induced collagen damage.

80. Which of the following conditions are associated with significant decreases in both collagen and hyaluronic acid concentrations in human vitreous humor?
   a. myopia.
   b. aphakia.
   c. diabetes mellitus (DM).
   d. a. and b.
   e. a. and c.

81. T or F Early resolution of a vitreous hemorrhage clot is more likely if the vitreous humor is liquefied.

82. T or F Vitamin A is delivered to the eye entirely as all-trans retinol.

83. T or F To regenerate 11-cis-retinaldehyde, the cis conformation must be regenerated at the retinal pigment epithelium (RPE).

84. Which one of the following statements about the effects of light on rod outer segment metabolism is false?
   a. In the dark, high concentrations of cyclic guanosine monophosphate (cGMP) maintain sodium channels open and rod outer segments depolarized.
   b. Light absorption leads to configurational changes in rhodopsin and activation of transducin.
   c. Transducin, through an amplification cascade, activates phosphodiesterase (PDE).
   d. PDE causes a fall in cGMP levels.
   e. Falling intracellular cGMP concentration leads to closure of sodium channels, with subsequent further depolarization of the rod outer segment.

85. T or F Like the lens, the retina primarily depends on anaerobic metabolism (glycolysis).

86. All the following methods of increasing ocular absorption of topically applied materials (without increasing systemic absorption) are effective except:
   a. adding a second eyedrop immediately after the first.
   b. waiting 5 to 10 minutes between administration of different medications.
   c. nasolacrimal sac compression.
   d. closing the eyes for 3 to 5 minutes after administration.
   e. addition of topical anesthetic immediately before administration.

87. All the following factors increase the amount of medication penetrating the cornea except:
   a. higher concentration of the drug.
   b. higher viscosity of the vehicle.
   c. higher pH of the drug.
   d. higher lipid solubility of the drug.
   e. addition of benzalkonium chloride.

88. Which of the following ocular structures have physiologically important cholinergic receptors?
   a. extraocular muscles.
   b. ciliary body muscle.
   c. lacrimal gland tissue.
   d. iris sphincter muscle.
   e. all of the above.
89. T or F In a patient with open-angle glaucoma, direct-acting cholinergic agents act to lower intraocular pressure (IOP) by decreasing relative pupillary block.

90. Unwanted side effects of direct cholinergic agonists include all of the following except:
   a. poor dark adaptation.
   b. decreased vision in older patients.
   c. headache in younger patients.
   d. possible aggravation or induction of angle-closure glaucoma.
   e. induced hyperopia.

91. T or F Cocaine (4% topical administration) is used to establish the presence of Horner’s syndrome, and hydroxyamphetamine is used to localize the lesion.

92. Following a unilateral dose of apraclonidine, the eye that received the medication can be identified by its:
   a. lid retraction.
   b. increased conjunctival injection.
   c. miosis.
   d. all of the above.
   e. none of the above.

93. Beta-adrenergic agonists generally:
   a. increase aqueous humor production, decrease outflow facility, and increase intraocular pressure (IOP).
   b. increase aqueous humor production, increase outflow facility, and increase IOP.
   c. increase aqueous humor production, increase outflow facility, and decrease IOP.
   d. decrease aqueous humor production, decrease outflow facility, and decrease IOP.
   e. decrease aqueous humor production, increase outflow facility, and decrease IOP.

94. T or F The mechanisms by which apraclonidine and timolol lower intraocular pressure (IOP) are related.

95. Dipivalyl epinephrine (dipivefrin) is converted to epinephrine in the:
   a. tears.
   b. conjunctiva.
   c. cornea.

96. Which would be the best single medication for treatment of primary open-angle glaucoma (POAG) in a 69-year-old patient with aphakia with chronic bronchitis, labile hypertension, and chronic depression treated with monoamine oxidase inhibitors (MAOIs)?
   a. epinephrine.
   b. dipivefrin.
   c. pilocarpine.
   d. timolol.
   e. acetazolamide.

97. T or F Aqueous humor secretion is exquisitely sensitive to the administration of minute amounts of carbonic anhydrase inhibitors (CAIs).

98. T or F Methazolamide-increased efficacy relative to acetazolamide is partly due to its ability to induce a greater degree of metabolic acidosis.

99. Carbonic anhydrase inhibitors (CAIs) should be used with great caution in all of the following types of patients except:
   a. patients with a remote history of nephrolithiasis.
   b. patients with chronic liver failure.
   c. patients on thiazide diuretics.
   d. patients on digoxin.
   e. patients with chronic schizophrenia.

100. Which one of the following statements about glucocorticoid effects is false?
   a. Glucocorticoids inhibit neovascularization.
   b. Glucocorticoids do not affect immunoglobulin E (IgE) titers.
   c. Glucocorticoids act through impaireing the efferent limb of the immune response.
   d. Glucocorticoids impair epithelial healing.
   e. Glucocorticoids act by blocking release of arachidonic acid from phospholipids.

101. T or F If a patient on chronic topical steroid therapy fails to show an increase in intraocular pressure (IOP) in 6 weeks, it is very unlikely to occur.

102. T or F Aspirin use may be associated with asthma attacks and hypersensitivity reactions caused by
shunting of products from the lipoxygenase (LOX) pathway to the cyclooxygenase (COX) pathway.

103. T or F Cromolyn sodium is most effective when used prophylactically; however, symptomatic relief may be obtained with intensive administration.

104. Which one of the following statements about penicillins and cephalosporins is false?
   a. Both penicillins and cephalosporins act by inhibiting bacterial cell wall synthesis.
   b. Both penicillins and cephalosporins contain beta-lactam bonds.
   c. These antibiotics have greater activity against Gram-positive organisms because of the oligopolysaccharide (OPS) coat of many Gram-negative bacteria.
   d. Probenecid counteracts resistance mechanisms by inactivating the beta-lactamase enzyme of bacteria.
   e. The most prominent mode of bacterial resistance to this group of antibiotics is the production of beta-lactamase enzymes.

105. Given a history of a hypersensitivity reaction to penicillin, the probability of a similar reaction to cephalosporin is approximately:
   a. 1%.
   b. 5%.
   c. 10%.
   d. 15%.
   e. 20%.

106. Which one of the following statements about antibiotic mechanisms is false?
   a. Sulfonamides act by inhibiting bacterial deoxyribonucleic acid (DNA) synthesis.
   b. Tetracycline is poorly water soluble but may be dissolved in eye drops containing mineral oil.
   c. Chloramphenicol use is most strongly associated with aplastic anemia when used orally.
   d. Aminoglycoside efficacy mostly depends on anaerobically supported antibiotic uptake.
   e. Erythromycin acts by inhibiting bacterial protein synthesis.

107. Which one of the aminoglycosides is most resistant to bacterial enzymatic inactivation?
   a. gentamicin.
   b. tobramycin.
   c. kanamycin.
   d. amikacin.
   e. neomycin.

108. The aminoglycoside that most frequently induces a topical hypersensitivity reaction is:
   a. gentamicin.
   b. tobramycin.
   c. kanamycin.
   d. amikacin.
   e. neomycin.

109. Which one of the following statements about vancomycin is false?
   a. Vancomycin inhibits bacterial replication by blocking cell wall synthesis.
   b. Empirical use of vancomycin in patients with neutropenia is recommended for infection prophylaxis.
   c. Vancomycin is one of the drugs of choice in filtering bleb-related endophthalmitis.
   d. Because of its poor gastrointestinal (GI) uptake, vancomycin is an excellent drug for pseudomembranous colitis.
   e. The nephrotoxicity of vancomycin is generally potentiated by concomitant use of aminoglycosides.

110. Which one of the following statements about the ocular antiviral agents is false?
   a. Vidarabine is an analog of adenine, whereas idoxuridine and trifluridine are analogs of thymidine.
   b. Trifluridine is more soluble than vidarabine or idoxuridine.
   c. Trifluridine is the most effective of the three ocular antiviral agents.
   d. Cross-resistance to different agents is commonly seen.
   e. The combination of topical trifluridine and interferon has been shown to speed epithelial healing in herpes simplex keratitis.

111. Which one of the following statements about trisomy 13 (Patau’s syndrome) is false?
   a. Less than 5% of patients with Patau’s syndrome will survive to 3 years of age.
   b. Commonly, the central nervous system (CNS) is severely affected.
   c. A clenched fist deformity is characteristic of Patau’s syndrome.
Questions

d. Cardiovascular and renal defects are very common in Patau’s syndrome.
e. Patau’s syndrome has been seen only in complete trisomy of chromosome 13.

112. Which one of the following statements about trisomy 18 (Edwards’ syndrome) is false?
   a. Edwards’ syndrome is the second most common chromosomal nondisjunction syndrome.
   b. Survival in trisomy 18 tends to be shorter than in trisomy 13.
   c. Outstanding features of Edwards’ syndrome include mental retardation and numerous musculoskeletal abnormalities.
   d. The effect of maternal age is important in trisomy 18.
   e. Glaucoma is more likely with trisomy 18 than with trisomy 13.

113. Which one of the following statements about Down’s syndrome is false?
   a. Ninety-five percent of Down’s syndrome are caused by meiotic nondisjunction, whereas the remaining 5% are translocation errors.
   b. A specific region of the long arm of chromosome 21 is responsible for the pathogenesis of Down’s syndrome.
   c. Classic findings of Down’s syndrome include the simian crease, hypoplasia of the middle phalanx of the fifth finger, and congenital heart disease.
   d. Patients with Down’s syndrome frequently have low serum purine levels.
   e. Down’s syndrome is the most common chromosomal syndrome, with an incidence of approximately 1:800 live births.

114. Which one of the following statements about Turner’s syndrome is false?
   a. Turner’s syndrome is the only disorder of sex chromosomes with characteristic eye findings.
   b. Physical findings of Turner’s syndrome include short stature, webbing of the neck, nonpitting edema, and coarctation of the aorta.
   c. The incidence of dyschromatopsia in women with Turner’s syndrome is identical to that in healthy women.
   d. Classic eye findings of Turner’s syndrome include prominent epicanthal folds, ptosis, and blue sclera.
   e. Patients with Turner’s syndrome are always sterile.

115. Increasing paternal age has been associated with all of the following disorders except:
   a. the craniosynostoses.
   b. Treacher Collins’ syndrome.
   c. neurofibromatosis.
   d. Down’s syndrome.
   e. Waardenburg’s syndrome.

116. Which one of the following statements about the genetics of aniridia is true?
   a. The autosomal-dominant form is the only form to feature aniridia with no other ocular abnormalities.
   b. The aniridia associated with Wilms’ tumor is most often inherited.
   c. Aniridia is usually sporadic.
   d. When confronted with a patient with newly diagnosed aniridia, an ophthalmologist should undertake a careful examination of the patient’s family members.
   e. Most cases of aniridia are unilateral.

117. In Marfan’s syndrome, the presence of ectopia lentis, arachnodactyly, aortic aneurysm, and tall habitus are examples of genetic:
   a. penetrance.
   b. variable expressivity.
   c. pleomorphism.
   d. phenocopying.
   e. pleiotropism.

118. From the following list, select the racial predilection that is incorrect:
   b. Glucose-6-phosphate dehydrogenase deficiency: Mediterraneans.
   c. Oguchi disease: Chinese.
   d. sickle cell hemoglobinopathy: African Americans.
   e. diabetes mellitus, type 2: Pima Indians.

119. An investigator attempts to show that a new topical medication is effective in reducing intraocular pressure (IOP). He states that patients who took his medication had significantly lower IOP than the patients who were given the placebo and says that his \( p \) value is equal to 0.01. Which one of the following statements about this situation is false?
   a. He is willing to reject the null hypothesis with a significance level \( \alpha = 0.01 \).
   b. If the difference in IOP between his treatment and placebo groups was actually
due to chance, he is falling victim to type I statistical error.
c. There is a 1% chance that the results that occurred were a result of chance alone.
d. If he chooses to accept the null hypothesis because he feels his probability level is too high, when in fact there was a true treatment difference, then he is falling victim to type II statistical error.
e. The power of his test is 0.99, or 1 minus his type I error.

120. Which of the following structures is/are transmitted in the inferior orbital fissure or infraorbital groove/canal?
   1. infraorbital nerve.
   2. inferior division of the oculomotor nerve.
   3. nerve from the pterygopalatine ganglion.
   4. superior ophthalmic vein.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

121. Which of the following bones do not form part of the orbital floor?
   1. zygomatic.
   2. sphenoid.
   3. maxillary.
   4. ethmoid.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

122. Which of the following muscles is/are supplied by the superior division of the oculomotor nerve?
   1. iris sphincter.
   2. iris dilator.
   3. inferior rectus.
   4. levator palpebrae superioris.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.

123. T or F The ptosis associated with third-nerve palsy is generally moderate to severe.

124. T or F The ptosis associated with Horner’s syndrome is generally moderate to severe.

125. With which of the following nerves does parasympathetic innervation to the iris sphincter travel?
   a. nerve to the levator palpebrae superioris.
   b. nerve to the inferior oblique.
   c. nerve to the superior rectus.
   d. nerve to the superior oblique.
   e. long ciliary nerves.

126. Which of the following are innervated by cranial nerve V (trigeminal nerve)?
   1. corneal sensation.
   2. pterygoid muscles.
   3. masseter muscles.
   4. orbicularis oculi.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

127. T or F The facial nerve does not have a branch within the orbit.

128. Which one of the following sets of nerve fibers synapses in the ciliary ganglion?
   a. sympathetic fibers to iris dilator.
   b. sympathetic fibers to choroid and ciliary body.
   c. parasympathetic fibers to iris sphincter.
   d. parasympathetic fibers to choroid and ciliary body.
   e. sensory fibers from the anterior globe (i.e., cornea, iris, ciliary body).

129. Which of the following extraocular muscles does not originate anatomically from the orbital apex?
   1. superior rectus.
   2. superior oblique.
   3. inferior rectus.
   4. inferior oblique.
Questions

130. Which of the following extraocular muscles does not originate mechanically from the orbital apex?
   a. superior rectus.
   b. superior oblique.
   c. inferior rectus.
   d. inferior oblique.
   e. 1, 2, 3, and 4.

131. Which of the following structures is not transmitted within the superior orbital fissure?
   a. superior ophthalmic vein.
   b. superior division of the third cranial nerve.
   c. ophthalmic artery.
   d. inferior division of the third cranial nerve.
   e. fourth cranial nerve.

132. Which of the following structures enter(s) the orbit outside the annulus of Zinn?
   a. nasociliary nerve.
   b. lacrimal nerve.
   c. inferior division of the third cranial nerve.
   d. fourth cranial nerve.
   e. 1, 2, 3, and 4.

133. In general, the last muscle to be rendered akinetic with a retrobulbar anesthetic block is the:
   a. superior rectus.
   b. superior oblique.
   c. inferior rectus.
   d. inferior oblique.
   e. levator palpebrae superioris.

134. Which is the only extraocular muscle typically supplied by one anterior ciliary artery?
   a. superior rectus.
   b. medial rectus.
   c. inferior rectus.
   d. lateral rectus.
   e. superior oblique.

135. T or F The anterior ciliary arteries usually terminate in rectus muscles as muscular feeder branches.

136. T or F The motor units of the extraocular muscles are considerably larger than those of other striated muscles.

137. Which upper-eyelid structure is considered to be analogous to the capsulopalpebral fascia of the lower eyelid?
   a. Müller’s muscle.
   b. levator palpebrae superioris.
   c. levator aponeurosis.
   d. Whitnall’s ligament.
   e. orbital septum.

138. The suspensory ligament of the globe is also known as:
   a. Whitnall’s ligament.
   b. Lockwood’s ligament.
   c. levator aponeurosis.
   d. capsulopalpebral fascia.
   e. lateral rectus check ligament.

139. Which one of the following statements about the arterial supply to the globe is correct?
   a. There are two long posterior ciliary arteries (LPCAs), and these arteries enter the sclera posteriorly near the optic nerve at the 3- and 9-o’clock positions.
   b. There are 10 to 12 LPCAs, and these arteries enter the sclera posteriorly in a circle around the optic nerve.
   c. There are seven LPCAs, and these arteries terminate in the rectus muscles.
   d. There are seven LPCAs, and these arteries terminate in the major arterial circle of the iris after providing feeders to the rectus muscles.
   e. There are two LPCAs, and these arteries enter the sclera posteriorly near the optic nerve at the 6- and 12-o’clock positions.
140. T or F Bowman’s membrane is the basement membrane of the corneal epithelium.

141. T or F Bowman’s layer is continuously produced throughout life.

142. T or F Descemet’s membrane is the basement membrane of the corneal endothelium.

143. T or F Descemet’s membrane is continuously produced throughout life.

144. T or F The capsule of the crystalline lens is the basement membrane of the lens epithelium.

145. The first retinal cells to differentiate a recognizable level are:
   a. photoreceptor cells.
   b. bipolar cells.
   c. ganglion cells.
   d. Müller’s cells.
   e. horizontal cells.

146. Which one of the following about photoreceptors is false?
   a. Tight junctions between photoreceptors and the retinal pigment epithelium (RPE) help maintain apposition between these layers.
   b. The major protein isolated from rod outer segments is rhodopsin.
   c. The chromophore for all the visual pigments is 11-cis-retinaldehyde.
   d. Differences in spectral absorption are due to different interactions between the chromophore and the protein (opsin) to which it is bound.
   e. The four visual pigment proteins have considerable sequence homology, implying a common ancestry.

147. Which one of the following about photoreceptor dynamics is false?
   a. Rods shed their outer segments shortly after dawn.
   b. Steady, constant dark adaptation will rapidly ablate rod outer segment shedding.
   c. Only the cis configuration of 11-retinaldehyde can initiate the light absorption cascade.
   d. The first step in regeneration of the chromophore is the formation of all-trans retinal.
   e. The chromophore is aligned parallel to the outer segment disc to enhance light capture.

148. Indirectly acting miotics (e.g., phospholine iodide) may dangerously increase systemic sensitivity to which of the following medications?
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

149. The cell most commonly used for karyotypic analysis is the:
   a. erythrocyte.
   b. platelet.
   c. neutrophil.
   d. B lymphocyte.
   e. T lymphocyte.

150. T or F The short arm of any human chromosome is labeled “p” for the French word petite.

151. What is the diameter of the anatomic human fovea?
   a. 0.5 mm (500 μm).
   b. 1.0 mm (1,000 μm).
   c. 1.5 mm (1,500 μm).
   d. 2.0 mm (2,000 μm).
   e. 2.5 mm (2,500 μm).

152. Which one of the following extraocular muscles is served by a contralateral brainstem subnucleus?
   a. superior rectus.
   b. medial rectus.
   c. inferior oblique.
   d. levator palpebrae superioris.
   e. inferior rectus.

153. A 19-year-old woman in the seventh month of her first pregnancy presents to an ophthalmologist complaining of visual loss. Potential explanations include each of the following except:
Questions

154. The risk of progression to proliferative diabetic retinopathy in a patient who is pregnant who has no or mild nonproliferative diabetic retinopathy before pregnancy is approximately:
   a. 0%.
   b. 0.2%.
   c. 1%.
   d. 5%.
   e. 10%.

155. Pregnancy may aggravate each of the following conditions except:
   a. Graves' disease.
   b. pituitary adenoma.
   c. meningioma.
   d. myasthenia gravis.
   e. diabetic retinopathy.

156. Which one of the following statements about tamoxifen is false?
   a. The antiestrogenic effects of tamoxifen are particularly useful for adjunctive therapy for estrogen receptor–positive breast cancers.
   b. Ocular toxicity has not been reported with cumulative doses of <10 g.
   c. Ocular toxicity has reportedly developed in 1% to 6% of treated patients in previous studies.
   d. Potential ocular findings following low-dose treatment (10 to 20 mg/day) include cornea verticillata, focal retinal pigment epithelium (RPE) damage, and crystalline retinopathy.
   e. Once retinal crystals have been detected, therapy should be continued with close follow-up.

157. Approximately what percentage of patients with newly diagnosed essential hypertension will remain well controlled on one antihypertensive agent?
   a. 10%.
   b. 25%.
   c. 50%.
   d. 67%.
   e. 75%.

158. Which type of organ transplantation procedure is most likely to be complicated by disseminated opportunistic fungal infection, including endophthalmitis?
   a. liver transplantation.
   b. kidney transplantation.
   c. pancreas transplantation.
   d. heart transplantation.
   e. lung transplantation.

159. Which of the following antiviral agents are commonly used to treat herpes zoster (shingles)?
   1. valacyclovir.
   2. famciclovir.
   3. acyclovir.
   4. ganciclovir.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

160. Which of the following cataract types is most often associated with steroid use?
   a. nuclear.
   b. cortical.
   c. posterior subcapsular.
   d. b. and c.
   e. a., b., and c.

161. Which of the following cataract types is most clearly associated with cigarette smoking?
   a. nuclear.
   b. cortical.
   c. posterior subcapsular.
   d. b. and c.
   e. a., b., and c.

162. Which of the following cataract types is most often associated with diabetes mellitus?
   a. nuclear.
   b. cortical.
   c. posterior subcapsular.
   d. b. and c.
   e. a., b., and c.

163. Which of the following cataract types is most often associated with ultraviolet light exposures?
   a. nuclear.
   b. cortical.
c. posterior subcapsular.
d. b. and c.
e. a., b., and c.

164. Which of the following cataract types is most often associated with lower educational levels?
   a. nuclear.
   b. cortical.
   c. posterior subcapsular.
   d. b. and c.
e. a., b., and c.

165. Each of the following statements are valid conclusions of the Diabetes Control and Complications Trial (DCCT) except:
   a. Among patients with no retinopathy at the outset of the study, the risk of progression of retinopathy was reduced to approximately 80% by intensive blood sugar control.
   b. Among patients with mild to moderate retinopathy at the outset of the study, the risk of progression of retinopathy was reduced to 65% by intensive blood sugar control.
   c. The study confirmed that there was an “early worsening” effect among patients initiated with intensive blood sugar control.
   d. Following documented progression, subsequent recovery was equally likely in the conventional versus intensive treatment groups.
   e. Patients with more severe retinopathy at baseline enjoyed the same reduction in risk of progression as patients with milder retinopathy.

166. Each of the following statements is true about valacyclovir except:
   a. Valacyclovir acts as a “prodrug” because it is converted into acyclovir in the small intestine and liver.
   b. Oral valacyclovir is substantially more bioavailable than oral acyclovir.
   c. Valacyclovir may reduce the incidence of postherpetic neuralgia, if given within 72 hours of onset of symptoms.
   d. Although a typical regimen for herpes zoster may be less expensive than acyclovir, the standard dosing of valacyclovir is more frequent than that for acyclovir.
   e. Concurrent use of cimetidine can increase plasma concentrations of the active drug.

167. Approximately what percentage of patients with diabetes will develop asymmetric retinopathy (proliferative disease with high-risk characteristics in one eye and no greater than moderate nonproliferative retinopathy in the other eye)?
   a. 1%.
   b. 5%.
   c. 10%.
   d. 15%.
   e. 20%.

168. Which of the following conditions retards the progress of diabetic retinopathy?
   1. ipsilateral glaucoma.
   2. ipsilateral severe carotid stenosis.
   3. ipsilateral chorioretinal scarring.
   4. ipsilateral aphakia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

169. When should a patient with newly diagnosed type 1 diabetes undergo an initial screening ophthalmic examination?
   a. immediately.
   b. 1 year later.
   c. 2 years later.
   d. 3 years later.
   e. 5 years later.

170. When should a patient with newly diagnosed type 2 diabetes undergo an initial screening ophthalmic examination?
   a. immediately.
   b. 1 year later.
   c. 2 years later.
   d. 3 years later.
   e. 5 years later.

171. Which one of the following agents presents the lowest risk of corneal or conjunctival toxicity as a preoperative antiseptic?
   a. chlorhexidine gluconate 4% (Hibiclens).
   b. povidone-iodine solution 5%.
   c. hydrogen peroxide 100%.
   d. benzalkonium chloride 100%.
   e. silver nitrate 1%.
172. Which of the following preoperative regimens most effectively reduces conjunctival bacterial colony counts?
   a. topical antibiotic for 3 days preoperatively.
   b. topical povidone-iodine for 3 days preoperatively.
   c. topical antibiotic for 3 days preoperatively, followed by topical povidone-iodine at the time of surgery.
   d. topical povidone-iodine at the time of surgery.
   e. topical povidone-iodine at the time of surgery, followed immediately by vigorous saline flush.

c. is more likely to cause cardiovascular complications.

d. has comparable or superior efficacy if administered once daily, in the evening.

e. causes less red eye.

173. Cigarette smoking has been most strongly associated with which manifestation of age-related macular degeneration?
   a. hard drusen.
   b. soft drusen.
   c. focal retinal pigment epithelium (RPE) hyperplasia.
   d. choroidal neovascularization.
   e. geographic atrophy.

174. Which of the following has been most strongly correlated with photic maculopathy following cataract surgery?
   a. hypoxemia during surgery.
   b. preoperative high myopia.
   c. duration of surgery.
   d. preoperative glaucoma.
   e. extracapsular cataract surgery.

175. What is the shortest duration of cataract surgery that has resulted in photic maculopathy?
   a. 5 minutes.
   b. 20 minutes.
   c. 55 minutes.
   d. 75 minutes.
   e. 90 minutes.

176. Potential side effects of topical prostaglandins or their analogs include each of the following except:
   a. bradycardia.
   b. red eye.
   c. iris hyperpigmentation.
   d. eczema.
   e. superficial punctate keratitis (SPK).

177. Compared with timolol 0.5% taken daily, latanoprost:
   a. has a lesser peak effect on intraocular pressure (IOP).
   b. is more likely to be associated with wide variations in IOP.

178. Each of the following is true about the vaccination for varicella zoster virus except:
   a. For children between the ages of 1 and 12 years, a single dose is sufficient.
   b. Two doses, 4 to 8 weeks apart, are recommended for individuals older than 12 years.
   c. The vaccine has been shown to reduce the incidence of shingles in immunocompetent adults with a history of childhood varicella infection.
   d. The vaccine is contraindicated during pregnancy.
   e. Salicylates should be avoided within the first 6 weeks following immunization.

179. “First pass” metabolism by the hepatic cytochrome P450 enzyme system of many drugs, including calcium-channel blockers and cyclosporine, may be considerably inhibited, leading to increased drug plasma concentrations, by which one of the following:
   a. herbal teas.
   b. grapefruit juice.
   c. home-distilled alcohol (moonshine).
   d. high-dose vitamin A.
   e. tanning beds.

180. Which of the following is most likely to be an extraocular focus of infection for endogenous bacterial endophthalmitis?
   a. liver abscess.
   b. endocarditis.
   c. orbital cellulitis.
   d. meningitis.
   e. septic arthritis.

181. Important differences between endogenous and exogenous (i.e., postoperative or traumatic) endophthalmitis include each of the following except:
   a. the most common responsible pathogen.
   b. diabetes mellitus as a risk factor for the endogenous form only.
   c. utility of vitreous tap and/or vitrectomy for diagnosis.
   d. long-term visual acuity.
   e. presence of pain.
182. Which of the following statements about the relation between cataract extraction and diabetes mellitus is false?
   a. Visual outcomes for cataract extraction in patients with diabetes are not as good as those among patients without diabetes.
   b. The most important predictor of good visual acuity following cataract extraction in a patient with diabetes is preoperative treatment with insulin (versus an oral agent).
   c. Women with diabetes and with normal body weight have better visual outcomes following cataract extraction than women who are overweight.
   d. Approximately one third of diabetics with symmetric nonproliferative retinopathy will progress only in the operated eye following cataract extraction.
   e. Potential acuity meter (PAM) readings correlate highly with final visual acuity in patients with nonproliferative diabetic retinopathy undergoing cataract extraction.

183. Which end-organ complication of diabetes is most likely to be aggravated by chronic alcohol intake?
   a. retinopathy.
   b. coronary artery disease.
   c. nephropathy.
   d. peripheral neuropathy.
   e. gastroparesis.

184. Nonsteroidal antiinflammatory agents (NSAIDs) and postmenopausal estrogen supplementation have both been shown to considerably reduce the incidence of which of the following disorders?
   a. essential hypertension.
   b. open-angle glaucoma.
   c. osteoporosis.
   d. Alzheimer’s disease.
   e. breast cancer.

185. Periconceptual supplementation with oral folic acid has been shown to reduce the incidence of which congenital malformation/syndrome?
   a. Down’s syndrome.
   b. neural tube defects.
   c. congenital heart disease.
   d. fragile X syndrome.
   e. neurofibromatosis.

186. The early treatment of diabetic retinopathy study (ETDRS) has established each of the following except:
   a. low-dose aspirin (650 mg orally each day) does not reduce the incidence of high-risk proliferative diabetic retinopathy.
   b. low-dose aspirin reduces the incidence of myocardial infarction (MI) in patients with diabetes.
   c. low-dose aspirin does not reduce the incidence of severe visual loss in patients with diabetes.
   d. low-dose aspirin does not reduce the risk of cataract formation in patients with diabetes.
   e. low-dose aspirin prolongs resolution of vitreous and preretinal hemorrhage in patients with proliferative diabetic retinopathy.

187. Each of the following statements about metformin is false except:
   a. Metformin has been available in other countries for many years.
   b. The prime mechanism of action of metformin is stimulation of pancreatic insulin secretion.
   c. The most common side effect is lactic acidosis.
   d. Like other medications for diabetes mellitus, metformin tends to induce weight gain.
   e. Metformin should be temporarily discontinued at least 2 days before any angiographic procedure, including fluorescein angiography.

188. Which one of the following is a topical H1 blocker with no chemical or structural relation to other antihistamines?
   a. naphazoline.
   b. lodoxamide.
   c. levocabastine.
   d. ketorolac.
   e. antazoline.

189. Which of the following medications has been shown to reduce the rate of progression of diabetic nephropathy?
   a. diltiazem.
   b. atenolol.
   c. metformin.
   d. captopril.
   e. furosemide.
190. The most important step in treating peptic ulcers is:
   a. upper gastrointestinal (GI) radiography.
   b. radioactive urea breath tests.
   c. upper endoscopy with gastric antral biopsy.
   d. serologic testing for antibodies to *Helicobacter pylori*.
   e. initiation of an H-2 antihistamine.

191. Influenza vaccine is recommended for each of the following groups of patients except:
   a. children between the ages of 3 and 6 years.
   b. patients with chronic asthma.
   c. adults older than 65 years.
   d. residents of chronic care facilities.
   e. patients with diabetes.

192. Which of the following is/are potential ophthalmic effects of hyperbaric oxygen (HBO) exposure?
   1. myopic shift.
   2. cotton-wool spots.
   3. cataract.
   4. nyctalopia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

193. Hyperbaric oxygen (HBO) has been used successfully to help treat each of the following disorders except:
   a. arterial gas embolism.
   b. chronic cystoid macular edema.
   c. central retinal artery occlusion (CRAO).
   d. rhino-cerebral-orbital mucormycosis.
   e. radiation optic neuropathy.

194. Oral acetazolamide would be expected to have the least clinical benefit in a patient with cystoid macular edema secondary to:
   a. retinitis pigmentosa.
   b. chronic uveitis.
   c. branch retinal vein occlusion.
   d. scleral buckling surgery.
   e. cataract extraction.

195. Once committed to a trial of carbonic anhydrase inhibitors for chronic cystoid macular edema, the most appropriate drug and dosage of choice is:
   a. acetazolamide 250 mg once daily.
   b. acetazolamide 500 mg once daily.
   c. methazolamide 25 mg twice daily.
   d. methazolamide 50 mg twice daily.
   e. there is no data available on the most appropriate choice.

196. A busy ophthalmology resident is asked to evaluate a 57-year-old inpatient with a complaint of “blurry vision” and with a history of diabetes, hypertension, hyperlipidemia, and tobacco use. She is currently admitted for congestive heart failure. She examines her patient and diagnoses bilateral clinically significant macular edema. The patient undergoes a fluorescein angiogram in the ophthalmology outpatient clinic and returns to the hospital. What blood level may be abnormal because of the angiogram?
   a. serum glucose.
   b. serum albumin.
   c. serum bilirubin (direct and total).
   d. serum digoxin.
   e. urine albumin.

197. Systemic therapy with interferon alpha has been clearly associated with ocular toxicity affecting which of the following structures?
   a. corneal epithelium.
   b. iris pigment epithelium.
   c. ciliary epithelium.
   d. posterior segment vasculature.
   e. photoreceptors.

198. What does the anterior lamellae of the upper eyelid consist of?
   a. the skin, orbicularis, and associated fascial and vascular components.
   b. capsulopalpebral fascia, tarsus, and orbital septum.
   c. Müller’s muscle, the conjunctiva, and the levator aponeurosis.
   d. orbital septum, orbital fat, and levator aponeurosis.
   e. none of the above.
199. What is usually the first branch of the ophthalmic artery?
   a. infratrochlear artery.
   b. lacrimal artery.
   c. supraorbital artery.
   d. central retinal artery.
   e. internal carotid artery.

200. Which of the following statements about eicosanoids is true?
   a. Cyclooxygenase-2 (COX-2) is present primarily in the stomach.
   b. Corticosteroids inhibit the conversion of lipoxigenase (LOX) into arachidonic acid.
   c. Flurbiprofen 0.03% (Ocufen) is often used during cataract surgery because of its action of direct and rapid pupillary dilation.
   d. Nordihydroguaiaretic acid (NDGA) blocks the LOX pathway.
   e. COX-2 inhibitors have never been shown to cause gastrointestinal toxicity.

201. Most of the human lens is composed of:
   a. water.
   b. beta-crystallin protein.
   c. alpha-crystallin protein.
   d. water-insoluble proteins.
   e. none of the above.

202. Which of the following is not a direct-acting muscarinic agent?
   a. carbachol.
   b. acetylcholine.
   c. pilocarpine.
   d. aceclidine.
   e. physostigmine.

203. All of the following are actions of direct-acting muscarinic agents except:
   a. contraction of the iris sphincter.
   b. relaxation of the zonular tension on the lens, allowing for accommodation.
   c. tension on the scleral spur.
   d. contraction of the longitudinal fibers of the ciliary body.
   e. posterior displacement of the lens.

204. All of the following are potential ocular side effects of sildenafil except:
   a. anterior ischemic optic neuropathy.
   b. decreased color vision or changes in color perception.
   c. conjunctival hyperemia.
   d. miosis.
   e. branch retinal artery occlusion.

205. For each of the following substances, indicate whether their aqueous humor concentrations are higher than, similar to, or lower than plasma concentrations:
   a. sodium. 1. higher.
   b. potassium. 2. lower.
   c. calcium. 3. similar.
   d. iron.
   e. lactate.
   f. ascorbate.
   g. glucose.

206. Rank the following from 1 to 5 in the order of decreasing cycloplegic duration:
   a. homatropine.
   b. cyclopentolate.
   c. atropine.
   d. tropicamide.
   e. scopolamine.

207. Match the numbered optic nerve segments listed with their appropriate length(s) and/or characteristics:
   a. longest segment. 1. intraocular.
   b. shortest segment. 2. intraorbital.
   c. segment with the most variable length. 3. intracanalicular.
   d. divided into three subsegments.
Questions

c. most vulnerable to indirect trauma.
f. most redundant segment.
g. the only segment not bathed in cerebrospinal fluid.

208. Match the lettered antibiotics with their numbered description:

<table>
<thead>
<tr>
<th>antibiotic</th>
<th>numbered description</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. penicillin G, penicillin V</td>
<td>1. highly effective against most Gram-positive and Gram-negative cocci, including anaerobes.</td>
</tr>
<tr>
<td>b. nafcillin, dicloxacillin</td>
<td>2. antimicrobial activity extended to Gram-negative species, most notably <em>Pseudomonas</em>.</td>
</tr>
<tr>
<td>c. ampicillin, cillin</td>
<td>3. less potent against amoxicillin susceptible organisms than penicillin G, but more effective against beta-lactamase-producing organisms.</td>
</tr>
<tr>
<td>d. ticarcillin, mezlocillin</td>
<td>4. moderate Gram-negative activity, including some <em>Haemophilus</em> and <em>Proteus</em> species.</td>
</tr>
<tr>
<td>e. penicillin V</td>
<td>5. storage of dietary vitamin A.</td>
</tr>
<tr>
<td>f. ticarcillin, mezlocillin</td>
<td>6. high baseline cyclic guanosine monophosphate (cGMP) levels and membrane depolarization.</td>
</tr>
<tr>
<td>g. ampicillin, cillin</td>
<td>7. amacrine and bipolar cell synapses.</td>
</tr>
<tr>
<td>h. nafcillin, dicloxacillin</td>
<td>8. flame-shaped retinal hemorrhages.</td>
</tr>
</tbody>
</table>

209. Match each lettered muscle with the numbered distance from the limbus to its tendinous insertion:

<table>
<thead>
<tr>
<th>muscle</th>
<th>distance from the limbus to its tendinous insertion</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. superior rectus</td>
<td>1. 5.5 mm.</td>
</tr>
<tr>
<td>b. medial rectus</td>
<td>2. 6.9 mm.</td>
</tr>
<tr>
<td>c. inferior rectus</td>
<td>3. 7.7 mm.</td>
</tr>
<tr>
<td>d. lateral rectus</td>
<td>4. 6.5 mm.</td>
</tr>
</tbody>
</table>

210. Match the lettered structures, functions, or characteristics with the appropriate numbered retinal layers:

<table>
<thead>
<tr>
<th>letter</th>
<th>numbered description</th>
</tr>
</thead>
<tbody>
<tr>
<td>a.</td>
<td>cell bodies whose processes project into the lateral geniculate and pretectal nuclei.</td>
</tr>
<tr>
<td>b.</td>
<td>Müller's cell bodies.</td>
</tr>
<tr>
<td>c.</td>
<td>horizontal and bipolar synapses.</td>
</tr>
<tr>
<td>d.</td>
<td>storage of dietary vitamin A.</td>
</tr>
<tr>
<td>e.</td>
<td>cell bodies whose processes form spherules and pedicles.</td>
</tr>
<tr>
<td>f.</td>
<td>high baseline cyclic guanosine monophosphate (cGMP) levels and membrane depolarization.</td>
</tr>
<tr>
<td>g.</td>
<td>amacrine and bipolar cell synapses.</td>
</tr>
<tr>
<td>h.</td>
<td>flame-shaped retinal hemorrhages.</td>
</tr>
<tr>
<td>1.</td>
<td>inner nuclear layer.</td>
</tr>
<tr>
<td>2.</td>
<td>photoreceptor layer.</td>
</tr>
<tr>
<td>3.</td>
<td>ganglion cell layer.</td>
</tr>
<tr>
<td>4.</td>
<td>nerve fiber layer.</td>
</tr>
<tr>
<td>5.</td>
<td>retinal pigment epithelium (RPE).</td>
</tr>
<tr>
<td>6.</td>
<td>outer plexiform layer.</td>
</tr>
<tr>
<td>7.</td>
<td>inner plexiform layer.</td>
</tr>
<tr>
<td>8.</td>
<td>outer nuclear layer.</td>
</tr>
</tbody>
</table>
Answers

1. d. The nasal bone lies medial to the orbit. The zygomatic, frontal, and ethmoid bones form parts of the lateral, superior, and medial bony orbit, respectively. The palatine bone forms a small portion of the posterior orbital floor.
2. a. Thirty cubic centimeters (cc, or mL for liquid measures) is approximately 2 tablespoons.
3. b. The lacrimal sac fossa is formed by the frontal process of the maxilla and the lacrimal bone. It cradles the lacrimal sac and is continuous with the nasolacrimal canal.
4. a. The lacrimal gland fossa lies in the anterolateral orbital roof within the zygomatic process of the frontal bone.
5. c. The lateral wall of the orbit is the thickest and strongest aspect of the bony orbit. It is formed by the zygoma and the greater wing of the sphenoid.
6. c. The lateral orbital tubercle of Whitnall is a small elevation in the orbital margin of the zygoma. It lies 11 mm below the frontal zygomatic suture. The tubercle is an important attachment site for all of the structures listed in the question except (ironically) Whitnall’s ligament. This structure is a condensation of fascia in the superior orbit that inserts 10 mm above Whitnall’s tubercle.
7. d. The optic canal is located in the lesser wing of the sphenoid and is approximately 8 to 10 mm in length. The central retinal artery arises from the ophthalmic artery after the latter passes through the optic canal. The central retinal artery then divides into the center of the optic nerve where it travels until it exits from the optic nerve head.
8. d. Approximately 1.2 million axons form a normal optic nerve. Each axon originates from the ganglion cell layer of the retina and extends to the lateral geniculate body. Fetal optic nerves contain a greater number of axons (approximately 3.7 million by 16 weeks), some of which regress by birth. Fewer axons may be a feature of certain optic nerve diseases (e.g., glaucoma).
9. a. The ophthalmic artery enters the orbit through the optic canal just inferior to the optic nerve. Within the orbit, it then courses lateral, superior, and finally medial to the optic nerve, until it forms the supraorbital, orbital, and uveal vasomotor nerves and sympathetic nerves to the lacrimal gland and Müller’s muscles enter the orbit with the ophthalmic artery, form the sympathetic root of the ciliary ganglion, and are distributed with the short ciliary nerves.
10. c. The ciliary ganglion receives three roots:
   1. A long sensory root that contains sensory fibers from the cornea, iris, and ciliary body—this root delivers sensation to the central nervous system (CNS) through the nasociliary nerve (V1).
   2. A short motor root that carries preganglionic parasympathetic axons to the iris sphincter—these are the only fibers that synapse here. This root arises from the lower division of CN III, the oculomotor nerve (which also supplies the inferior oblique).
   3. A sympathetic root that innervates the blood vessels of the uvea—this root arises from a plexus around the internal carotid artery and passes through the optic foramen (with the ophthalmic artery).
11. True. All of the third-order sympathetic neurons to the eye begin in the superior cervical ganglion and travel along the internal carotid artery into the cavernous sinus. The pupillomotor fibers then join the ophthalmic division of the trigeminal nerve (V1), the nasociliary nerve, and, finally, the long ciliary nerves. The orbital and uveal vasomotor nerves and sympathetic nerves to the lacrimal gland and Müller’s muscles enter the orbit with the ophthalmic artery, form the sympathetic root of the ciliary ganglion, and are distributed with the short ciliary nerves.
12. b. The accessory lacrimal glands are the glands of Krause and Wolfring, which produce primarily basal tear secretion. The meibomian glands can have aberrant cilia grow through their orifices in acquired and congenital distichiasis but normally have no associated appendages. The glands of Zeis are modified sebaceous glands associated with cilia.
13. False. The substance of the tarsus feels like cartilage, but, microscopically, it consists of densely packed type I collagen without hyalocytes.
14. False. The palpebral lobe is the smaller lobe and lies in the superolateral conjunctival fornix. (The larger orbital lobe does lie posterior to the smaller palpebral lobe.) The lobes are separated by a lateral expansion of the levator aponeurosis.
15. False. The canaliculi are lined with stratified, squamous epithelium. The lacrimal sac consists of a bilayer: a superficial columnar layer and a deep, flattened layer.
16. c. The average adult corneal diameter is approximately 12 mm (it is slightly larger horizontally) and is reached at approximately 2 years of age.
17. False. Mitosis of the endothelium rarely occurs. However, corneal endothelial cells will spread out and enlarge (polymegathism), which can be observed after surgery, inflammation, or endothelial disease (e.g., Fuch’s dystrophy).
18. False. The sclera, like the cornea, is virtually avascular except for two areas: (i) the superficial vessels of the episclera, and (ii) the intrasceral vascular plexus located immediately posterior to the limbus.
19. c. Another way of remembering this is the mnemonic (from peripheral to central) “I can (or can’t) see this stuff!” which stands for Iris, Ciliary body, Scleral spur, Trabecular meshwork, and the Schwalbe line.
20. False. The trabecular meshwork consists of thin, perforated connective tissue sheets arranged in a layered pattern. The connective tissue “beams” are lined by a...
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canalicular trabecular meshwork (TM), which is im-
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22. c. The average anteroposterior (AP) lens width of a normal
infant’s lens is 3 mm at birth and increases throughout
life to approximately 6 mm at age 80.

23. d. The equatorial diameter of the lens is approximately
6.5 mm at birth and can increase during the first three
decades of life to approximately 9 to 10 mm in diam-
et; it remains in this dimension throughout life.

24. False. The anteroposterior (AP) lens diameter gradu-
ally increases throughout life as lens epithelium cells
replicate and migrate centrally into the nucleus. An
elderly person’s lens is considerably thicker than a
young person’s lens.

25. False. The equatorial diameter of the lens does change
from infancy (6.5 mm at birth) to adulthood (9 to
10 mm); the diameter then stabilizes at this value.

26. False. The anterior capsule is almost twice as thick as
the posterior capsule and increases in thickness
throughout life.

27. False. Only the posterior surface of the iris is covered
by a continuous layer of cells. The anterior surface
consists of (unlined) stroma.

28. d. The iris is composed of five layers: the anterior
border layer, the iris stroma, the muscular layer, the
anterior pigment epithelium, and the posterior pig-
ment epithelium. The clump cells of Koganei are part
of the iris stroma. The dilator and sphincter muscles
make up the muscular layer of the iris, which is ante-
rior to the iris pigment epithelium. The color of the
iris is determined by the number and size of melain
pigment granules of the stromal melanocytes.

29. c. Parasympathetic fibers originate from the
Edinger–Westphal subnucleus in the mid brain, follow
the inferior division of the oculomotor nerve as it bi-
furcates in the cavernous sinus, continue with the
branch supplying the inferior oblique muscle, and
then synapse in the ciliary ganglion. Postganglionic
fibers are transmitted through the short ciliary nerves
to the iris sphincter. Sympathetic fibers originate in
the ipsilateral posteralateral hypothalamus and pass
through the brainstem to synapse in the intermediolat-
eral gray matter of the spinal cord between the levels
of C8–T2 (ciliospinal center of Budge). The second-
order neurons exit the spinal cord, pass over the lung
apex, and synapse in the superior cervical ganglion.
Third-order neurons travel with the internal carotid
plexus, enter the cavernous sinus, and travel with the
ophthalmic division of the trigeminal nerve (V1) to
the orbit. Thereafter, the fibers travel within the
nasociliary nerve and then the long ciliary nerves to
the iris.

30. False. Fluorescein angiography demonstrates that the
choriocapillaris is arranged in a lobular pattern, with
multiple fenestrations, especially in the posterior pole.
Functionally, the choriocapillaris is an endarteriolar
system and is densest in the macula.

31. False. It is primarily dependent on the number of pig-
mented melanocytes in the choroid. Retinal pigment
epithelium (RPE) pigmentation contributes to a lesser
degree.

32. False. Retinal pigment epithelium (RPE) cells in the
foveal area are taller, more closely packed, and have
more and larger melanosomes, contributing to the rel-
ative hypofluorescence of this area during fluorescein
angiography.

33. d. Rods and cones are characterized by three compo-
nents: the synaptic body, the inner segment, and the
outer segment. The synaptic body of a rod is called a
spherule, whereas that of the cone is called a pedicle.
Photopigment is stored in discs in the outer segments.
In rods, the discs are not attached to the cell mem-
brane, but cone discs are continuous with it.

34. b. Intraretinal processing occurs from photoreceptors
to bipolar cells to ganglion cells, with modulation by
horizontal (outer plexiform layer) and amacrine (inner
plexiform layer) cells.

35. c. Approximately 120 million rods and 6 million
cones interact with 1.2 million ganglion cells in an eye.
Therefore, the ratio of rods to cones is approximately
20:1. (Some studies cite a ratio as low as 12:1.)

36. e. The external limiting membrane (ELM) is highly
fenestrated and is composed of attachment sites of adja-
cent photoreceptors and Müller’s cells. The internal
limiting membrane (ILM) is formed by footplates of
Müller’s cells and by attachments to the basal lamina of
retinal astrocytes. The nuclei of Müller’s cells are located
in the inner nuclear layer. Although Müller’s cells are the
most prevalent glial element, two other cell types are
found—microglia are found in small numbers through-
out the retina; astrocytes are found in the proximal gan-
glion cell and nerve fiber layers of the human retina.
Note that oligodendrocytes, the myelinating fiber of the
central nervous system, are normally absent.

37. b. A cilioretinal artery contributes to the vascular sup-
ply of the retina in approximately 50% of individuals
and 30% of eyes. In 15% of individuals, it contributes
to macular circulation.

38. c. Retinal blood vessels usually do not extend beyond
the inner third of the inner nuclear layer.

39. b. The outer plexiform layer is composed of intercon-
nections between photoreceptor synaptic bodies, hori-
zontal cells, and bipolar cells. The inner plexiform
layer is composed of connections between bipolar
cells, amacrine cells, and ganglion cells.

40. a. The ora serrata is an anatomic landmark for retinal
topography but has no importance in uveoscleral
anatomy.
41. False. The ora is smooth temporally but appears serrated nasally.

42. False. Normally, axons of ganglion cells do not become myelinated until after they pass through lamina cribrosa, as part of the intraorbital optic nerve. When myelinated fibers are visible ophthalmoscopically, oligodendrocytes have migrated anteriorly.

43. b. After exiting the optic foramen, the optic nerve is bounded by the anterior cerebral artery superiorly, the internal carotid artery laterally, and the ophthalmic artery inferiorly.

44. a. Approximately 53% of optic nerve fibers are crossed and 47% uncrossed. Macular and peripheral fibers cross in roughly identical proportions.

45. d. Only the levator is served by a single subnucleus. It sits dorsal, central, and at the inferior end of the group of subnuclei that compose the two third-cranial nerve nuclei. The superior recti have two subnuclei, each controlling the contralateral nerve. In contrast, the inferior obliques and the medial recti each have individual subnuclei that control the ipsilateral nerves. The superior oblique is served by the fourth nerve. The lateral rectus is supplied by the sixth cranial nerve.

46. a. The superior recti have two subnuclei, each controlling the contralateral nerve.

47. d. The pupillomotor fibers of the third cranial nerve run in the inferior division, which carries them to the ciliary ganglion. They are among the axons in the periphery of the nerve, making them easily susceptible to compression.

48. True. The fifth cranial nerve is responsible for facial sensation and supplies motor fibers to the muscles of mastication (i.e., temporalis, masseter, and pterygoïds), as well as to the tensor veli palatini, tensor tympani, anterior belly of the digastic, and mylohyoid muscles.

49. d. The third, fourth, and fifth cranial nerves travel within the walls of the cavernous sinus. The sixth nerve and the internal carotid artery travel through the sinus itself, and therefore, they are more susceptible to injury by sinus lesions, such as meningioma and aneurysm.

50. False. Although an ocular coloboma (which is the absence of part or all of an ocular tissue) does form as the result of failure of optic cup fusion (e.g., faulty closure of the fetal fissure), a typical coloboma is found in an inferior and somewhat nasal location. More than half of colobomas are bilateral (although not necessarily symmetric).

51. True. The production of melanin in the developing retinal pigment epithelium (RPE) starts at the posterior pole and progresses anteriorly, usually reaching completion by the sixth week of embryonic development. This is the first melanin production known to take place in the body.

52. False. Myelination starts at the chiasm. Some studies have documented completion of myelination by as early as 36 weeks’ gestation. By 1 month postpartum, myelination is almost always complete (although sheaths may thicken with time).

53. False. The lens is derived from surface ectoderm.

54. e. Neural crest cells and mesoderm contribute to the mesenchymal structures of the head. (Mesenchyme is the embryonic tissue that gives rise to connective tissue.) The stroma of the iris and the muscular layer of orbital vessels are examples of neural crest contributions. The endothelium of these same vessels is of mesodermal origin.

55. e. Neural crest cells give rise to ciliary musculature, corneal stroma, and endothelium (but not corneal epithelium), most of the sclera (except for a temporal portion which is of mesodermal origin), choroidal stroma, some of the orbital bones, orbital cartilage, orbital connective tissue, nerve sheaths, and uveal melanocytes. Extraocular muscles form from paraxial mesoderm.

56. d. Blood vessel endothelia, extraocular muscles, and temporal sclera are all mesodermal in origin. The mesoderm also contributes to the formation of the vitreous. The pupillomotor muscles are neuroectodermal in origin. The trochlea originates from the neural crest.

57. e. Conjunctival epithelium is derived from surface ectoderm, but the substantia propria is derived from the neural crest.

58. False. The primary vitreous is actually displaced centrally to become Cloquet’s canal.

59. b. Anophthalmia is the absence of an identifiable eye. Microphthalmia describes the presence of a small, disorganized eye.

60. d. In nanophthalmia, the eye is smaller than normal with a disproportionately large lens, but it is otherwise unremarkable.

61. d. Nanophthalmos is, by definition, a small but otherwise normal eye. Cysts are not seen in association with it.

62. a. Dermoids are choristomas, that is, normal cells and/or tissue present in abnormal locations.

63. e. In Peters’ anomaly, the central cornea is always opacified because of the central defect in Descemet’s membrane and the absence of endothelium. The lens may be adherent to the cornea, but this is not always seen. Axenfeld’s anomaly, Axenfeld’s syndrome, Rieger’s anomaly, and Rieger’s syndrome are now considered a single entity known as Axenfeld–Rieger syndrome, which has been associated with mutations in the RIEGL1/PIXT2 gene and the forkhead gene FKHL7. Axenfeld–Rieger syndrome is most commonly inherited in an autosomal-dominant fashion and can lead to glaucoma in 50% of cases.

64. a. There is nearly always some rudimentary iris tissue present in aniridia, although it may be difficult
to visualize clinically. Aniridia is caused by a defect of the \( PAX6 \) gene on chromosome 11p13 and may be sporadic or familial. Sporadic aniridia is associated with Wilms’ tumor (usually because of deletion of the \( PAX6 \) gene and the adjacent \( WT1 \) Wilms’ tumor gene). Aniridia is not typically found in patients with autosomal-dominant Wilms’ tumor. When aniridia is associated with mental retardation, Wilms’ tumor, ambiguous genitalia, or other genitourinary anomalies, there is usually a small deletion in the short arm of chromosome 11.

65. c. Microspherophakia is observed most often with Weil–Marchesani syndrome, not Marfan’s syndrome. Patients with Marfan’s syndrome are usually tall and lean, in contrast to patients with Weil–Marchesani syndrome, who are short and stocky. Marfan’s syndrome is often associated with ectopia lentis. Lowe’s syndrome is inherited in an X-linked recessive fashion and is therefore found almost exclusively in men. Any organ in the body may be damaged by rubella, and the rubella syndrome is characterized by the triad of cataracts, deafness, and cardiac defects. Juvenile glaucoma and cataract rarely coincide with congenital rubella infection.

66. b. The visual prognosis in persistent hyperplastic primary vitreous (PHPV) (also known as PFV, or persistent fetal vasculature) is currently poor. Early cataract extraction and membrane excision may preserve some vision. An eye with leukocoria that is small and is unlikely to harbor retinoblastoma. Likewise, retinoblastoma does not typically cause cataract.

67. c. The glands of Krause and Wolfring, which produce the basal tear secretion, account for approximately 10% of total lacrimal secretory mass.

68. c. Aqueous humor provides most of the glucose needed by the endothelium, stroma, and epithelium. Hydrophobic molecules penetrate the epithelium most easily, whereas hydrophilic molecules penetrate stroma more easily.

69. d. Endothelial cells very rarely divide. As endothelial cells are damaged or die, the number of endothelial cells on a given cornea decrease, and the cells enlarge and spread out.

70. e. Diffusion and ultrafiltration are both passive, whereas secretion and carbonic anhydrase II activity are active. Most aqueous humor appears to be produced by active, energy-dependent secretion from the inner, nonpigmented ciliary epithelium involving \( \text{Na}^+/\text{K}^- \)-dependent adenosine triphosphatases (ATPases).

71. b. Topical administration of type E and type F prostaglandins, as well as arachidonic acid, causes miosis. High doses of prostaglandins will cause an increase in intraocular pressure (IOP). Low doses of some prostaglandins, in contrast, appear to lower IOP in some animal species. By blocking phospholipase, corticosteroids effectively inhibit both the lipoygenase (LOX) and cyclooxygenase (COX) pathways.

72. a. Carbonic anhydrase, although present in only trace amounts in the aqueous humor, has a high enough turnover that it is felt to be functionally significant. Hyaluronidase is present in aqueous humor and may participate in the regulation of resistance to aqueous outflow. Lysozyme is present and provides antibacterial protection. Lactate dehydrogenase, not normally detectable in aqueous humor, may be a marker for retinoblastoma.

73. b. This is approximately one third the concentration found in the earth’s atmosphere and is entirely derived from anterior chamber blood flow.

74. d. Lens epithelium cells are located anteriorly underneath the lens capsule. Epithelial cells in a ring around the anterior lens, or the germinative zone, exhibit the highest level of deoxyribonucleic acid (DNA) synthesis (the S phase in the cell cycle). Newly formed cells migrate toward the lens equator, where they differentiate into lens fiber cells.

75. b. The water-soluble proteins are divided into three types of crystallins that are fractionated into four electrophoretic groups. Alpha crystallins are the largest, with molecular weights of \( >500,000 \) daltons. Beta crystallins are the most abundant, making up approximately 55% of the water-soluble protein. Gamma crystallins are the smallest.

76. True. Na+/K+-dependent adenosine triphosphatases (ATPases) move sodium from posterior to anterior position (into the aqueous humor) and potassium from anterior to posterior position (into the lens). The posterior surface of the lens has movement (to and from the vitreous) mainly through passive diffusion.

77. True. Unlike the retina, the lens can function without oxygen but not without glucose, primarily because the lens mostly relies on anaerobic glycolysis to generate adenosine triphosphate (ATP). In a glucose-depleted environment, the lens will become hazy after several hours.

78. False. See previous answer.

79. c. Syneresis (i.e., vitreous liquefaction) is associated with a focal decrease in collagen concentration. More than half of the vitreous humor is usually liquid by the age of 80 years. Patients with Stickler’s syndrome often undergo vitreous syneresis before the age of 30 years.

80. a. Both collagen and hyaluronic acid concentration are decreased by 20% to 30% in myopic eyes (axial length >26 mm) compared to eyes with a normal axial length.

81. True. When bleeding occurs in a region of formed vitreous humor, a smaller, tighter clot that is more resistant to degradation is formed. In liquid vitreous humor, blood disperses and is cleared quickly.

82. True. Vitamin A, stored hepatically, is transported in serum as all-trans retinol.

83. True. Conversion between aldehyde and alcohol (and vice versa) occurs in the photoreceptors, whereas the
Beta-adrenergic agonists (e.g., epinephrine) are
92. a. Miosis and induced myopia are generally problem-
ease.
90. e. Dipivalyl epinephrine (dipivefrin) is a prodrug of ep-
inephrine that contains two divalyl residues. Epinephrine is released into the anterior chamber when the pivalyl groups are cleaved by corneal esterases.
96. c. Because epinephrine has been associated with a
reversible cystoid maculopathy (in approximately 25% of patients with aphakia who have been chroni-
cally treated), both epinephrine and the produg Dipivalyl epinephrine (dipivefrin) would be rela-
tively contraindicated in this case. Furthermore, alpha-agonists (epinephrine is an alpha-and beta-
agonist) are contraindicated in patients who may have an abnormally increased sensitivity to their car-
diovascular effects caused by the use of drugs such as monoamine oxidase inhibitors (MAOIs), tricyclic antidepressives, cocaine, or reserpine. In general, depression may be a side effect of beta-blockers and a relative contraindication to acetazolamide therapy. Therefore, the best single alternative drug in this case would be pilocarpine.
97. False. Clinically, carbonic anhydrase must be >99% inhibited to decrease aqueous humor secretion significantly.
98. False. Unlike acetazolamide, methazolamide is not ac-
tively secreted into the renal tubules, and metabolic acidosis is less pronounced. It is also metabolized by the liver, with consequent reduction in some of the systemic side effects (such as nephrolithiasis) of other carbonic anhydrase inhibitors. It is also less effective for reducing intraocular pressure (IOP).
99. a. A remote history of spontaneous nephrolithiasis
table 5 years earlier) is not thought to be a contraindica-
tion for starting carbonic anhydrase inhibitor (CAI)
therapy. Recent renal stones (<5 years earlier) may
be a contraindication. CAIs potentiate hepatic
encephalopathy and the potassium-wasting effects of
thiazides (hypokalemia is a side effect of CAIs).
Sensitivity to digoxin is increased by hypokalemia.
Psychiatric disturbances also may be exacerbated by
CAIs. Patients with a sulfa allergy should not be given
carbonic anhydrase inhibitors.
100. d. At the tissue level, glucocorticoids suppress early in-
flammatory responses such as local vascular congestion, edema, and hyperthermia, as well as late inflammatory
responses such as capillary and fibroblast proliferation

2: Fundamentals of Ophthalmology

trans to cis isomerization takes place in the retinal pig-
ment epithelium (RPE).
84. e. Photoreceptors are more active electrically (depolar-
ized) in the dark! With light absorption, transducin
(via phosphodiesterase [PDE]) lowers cyclic guanosine monophosphate (cGMP) concentration, which hyper-
polarizes the cell and decreases synaptic exchange with bipolar cells.
85. False. The retina is primarily oxygen dependent
(although the lens).
86. a. Adding a second drop will likely increase systemic
absorption minimally and may also simply wash out
the first drop—a normally dispensed 50-μL drop
contains far more than the 10-μL of fluid normally
found in the eye and cul-de-sac. Local anesthetic dis-
rupts corneal epithelial barrier functions, enhancing local uptake. Blinking increases drainage of topical
medications into the nasolacrimal drainage system and
subsequently into the systemic vascular system.
87. c. pH extremes trigger reflex tearing, with subsequent
dilution of the drug. Benzalkonium chloride, like local
anesthetics, disrupts the corneal epithelium.
88. e. Nicotinic cholinergic receptors mediate extraocular
muscle contraction. Muscarinic receptors mediate auto-
nomic responses (e.g., accommodation and secretions).
89. False. Although relative pupillary block may be dimin-
ished, contraction of the longitudinal fibers of the cili-
ary body “opens” the trabecular meshwork (TM) by
pulling on the scleral spur, thereby increasing outflow.
90. e. Miosis and induced myopia are generally problem-
atic in younger patients. In the elderly patients, aggra-
vation of cataractous visual loss may be important.
Because cholinergic agonists cause forward displac-
ment of the iris-lens diaphragm, angle closure can rarely result from their use.
91. True. Cocaine blocks the reuptake of norepinephrine
into presynaptic vesicles. This will cause the secondary
accumulation of norepinephrine, resulting in pupillary
dilation. In Horner’s syndrome, there is less neuro-
transmitter release, less accumulation, and therefore less
dilation. Hydroxyamphetamine causes release of norepinephrine and results in dilation. If the lesion is post-
ganglionic, there will be less norepinephrine and less
dilation. If the lesion is preganglionic, there will be nor-
mal or supranormal dilation (if there is upregulation of
receptors) because the postganglionic fibers synthesize
and store norepinephrine in a normal fashion.
92. a. Apraclonidine is a selective alpha-2 adrenergic ago-
nist. Side effects of apraclonidine that have been re-
ported include lid retraction, conjunctival blanching,
mydriasis, lethargy, xerostomia, and allergic reactions.
The first three side effects are directly attributable to its adrenergic activity.
93. c. Beta-adrenergic agonists (e.g., epinephrine) are
thought to decrease intraocular pressure (IOP),
despite their tendency to increase aqueous humor
production, by increasing uveoscleral outflow. Their
use in the medical management of glaucoma has de-
creased considerably.
94. True. Timolol maleate decreases intraocular pressure
(IOP) by reducing aqueous humor production by
cyclic adenosine monophosphate (cAMP)-mediated
mechanisms. Apraclonidine is thought to decrease IOP
(at least partially) by inhibiting aqueous humor secre-
tion through the same cAMP final common pathway.
95. c. Dipivalyl epinephrine (dipivefrin) is a prodrug of ep-
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Answers

101. False. Late steroid-induced intraocular pressure (IOP) increases are common, and IOP should be monitored during the entire course of therapy.

102. False. Aspirin causes inhibition of cyclooxygenase (COX); therefore, arachidonic acid is diverted to the lipooxygenase (LOX) pathway. This is thought to be the underlying mechanism for the asthma attacks and hypersensitivity reactions (by increased production of leukotrienes).

103. False. Cromolyn sodium is a mast cell stabilizer and has no direct antihistaminic effect (receptor blockade). It is only effective if used prophylactically.

104. d. Probenecid competitively inhibits penicillin excretion by the kidney.

105. c. Approximately 10% of patients with a history of a hypersensitivity reaction to penicillin will have cross-reactivity to the cephalosporins.

106. d. Sulfonamides indirectly inhibit bacterial deoxyribonucleic acid (DNA) synthesis by blocking the synthesis of folic acid (folic acid is a cofactor in nucleic acid synthesis). Only aerobic bacteria are susceptible to aminoglycosides. Anaerobic organisms are resistant to aminoglycosides because the mechanism by which they are taken up by microorganisms is driven by aerobic metabolism.

107. d. The most important clinical cause of acquired bacterial resistance to aminoglycosides is the production of microbial enzymes that inactivate the drug. These enzymes are genetically transmitted by bacterial plasmids. Of the aminoglycosides, amikacin is the least sensitive to inactivation by this mechanism.

108. e. Hypersensitivity reactions may occur following topical administration of any of the aminoglycosides but occur most frequently with neomycin. Such reactions occur in 6% to 8% of patients. Other important adverse reactions common to aminoglycosides include otoxicity and nephrotoxicity.

109. b. Vancomycin acts by the inhibition of cell wall synthesis. It is primarily active against Gram-positive bacteria including methicillin-resistant strains of Staphylococcus, although plasmid-mediated resistance has resulted in staphylococcal resistance. Because of increasing resistance to vancomycin, the Center for Disease Control and Prevention (CDC) has recommended avoidance of the use of empiric vancomycin in patients with neutropenia unless clear evidence for a beta-lactam–resistant Gram-positive infection can be demonstrated.

110. d. Trifluridine and idoxuridine are thymidine analogs, whereas vidarabine is an adenosine analog. All of these agents inhibit deoxyribonucleic acid (DNA) synthesis and are effective in treating herpes simplex. Trifluridine is more effective than the other two. Cross-resistance to these agents has not yet been reported.

111. c. Because the phenotypic expression of trisomy 13 (Patau’s syndrome) is caused by the presence of three copies of a portion of the long arm of chromosome 13, the syndrome can be caused by trisomy of the entire chromosome or of any portion that includes the segment from 13q14 to the q terminus. Several ocular abnormalities occur in Patau’s syndrome, including cataracts, colobomas, epicanthal folds, absent eyebrows, and hypotelorism.

112. b. There is a slightly higher mortality in trisomy 13 than in trisomy 18. Approximately 75% of infants with trisomy 13 die by 6 months of age, whereas approximately 50% of infants with trisomy 18 die by that age.

113. d. Serum purine levels in patients with Down’s syndrome are typically elevated. The enzymes required for the biosynthesis of purinies are coded by genes present on the long arm of chromosome 21. The presence of a third set of these genes presumably results in the elevation of serum purine levels.

114. c. Turner’s syndrome is caused by monosomy of the X chromosome in all cells, monosomy X mosaicism, or structural abnormalities of the X chromosome. Approximately 60% of individuals with this syndrome have monosomy X in all body cells. The absence of a second X chromosome in Turner’s syndrome can result in the full expression of X-linked recessive disorders with the same frequency as that found in healthy men (e.g., dyschromatopsia, which generally affects 5% to 8% of men).

115. d. Down’s syndrome is associated with increased maternal age. A clear correlation between advanced paternal age and Down’s syndrome has not been established.

116. d. Sporadic aniridia (i.e., nonfamilial), often associated with glaucoma or cataract, can be caused by a new deletion of an interstitial segment of chromosome 11 (11p13). This deletion also results in genital abnormalities (which may be subtle in females), mental retardation, and Wilms’ tumor. Hereditary aniridia can occur with autosomal-dominant or autosomal-recessive inheritance patterns and is associated with other ocular anomalies, including nystagmus, cataract, glaucoma, corneal abnormalities, and hypoplasia of the optic nerve. Hereditary aniridia is only rarely associated with Wilms’ tumor. Examination of family members may differentiate sporadic from hereditary aniridia. Most cases of aniridia are bilateral. Two thirds of cases are familial.

117. e. The expression of multiple discrete anomalies in various organs caused by a single gene mutation is termed genetic pleiotropism. Alterations in the connective tissues throughout the body result in the characteristic manifestations of Marfan’s syndrome, which is caused by a defect in the fibrillin gene.
118. c. Oguchi’s disease, a form of congenital stationary night blindness, is seen primarily in persons of Japanese descent.
119. e. The power of a statistical test is a measure of its ability to detect a difference between the treatment and control groups when a true difference exists. The power is mathematically defined as (1 – [type II error]). The power of a statistical test to detect a difference between the groups in a clinical trial is increased by a larger sample size and by a greater actual difference caused by the intervention.
120. b. The infraorbital nerve is a branch of the maxillary division of the trigeminal nerve, carrying sensory fibers from the cheek. It enters the orbit through the infraorbital foramen and exits through the inferior orbital fissure. The nerve from the pterygopalatine ganglion, carrying postganglionic secretory fibers for the lacrimal gland, is also transmitted through the inferior orbital fissure. The inferior division of the oculomotor nerve, as well as the superior ophthalmic vein, enters the orbit through the superior orbital fissure.
121. c. The orbital floor is formed by the zygomatic, maxillary, and palatine bones. The sphenoid contributes to the orbital roof and the medial wall, whereas the ethmoid contributes solely to the medial wall of the orbit.
122. d. The iris sphincter receives parasympathetic innervation by the inferior division of the oculomotor nerve (which also supplies the inferior oblique muscle). The inferior rectus is also supplied by the inferior division. The iris dilator is supplied by sympathetic fibers running with the nasociliary and long ciliary nerves, branches of the first division of the trigeminal nerve (V1).
123. True. The oculomotor nerve supplies the levator palpebrae superioris, and interruption of this supply results in profound ptosis.
124. False. Müller’s muscle is sympathetically innervated and provides 2 to 3 mm of eyelid “lift.” Therefore, ptosis associated with Horner’s syndrome is typically mild.
125. b. Parasympathetic fibers travel with the inferior division of the oculomotor nerve (CN III).
126. a. The orbicularis oculi muscle, responsible for reflex as well as willful eyelid closure, is supplied by the facial nerve (CN VII). The fifth cranial nerve is responsible for facial sensation and supplies motor fibers to the muscles of mastication (temporalis, masseter, pterygoids), as well as the tensor veli palatini, tensor tympani, anterior belly of the digastric, and mylohyoid muscles.
127. False. Parasympathetic secretory innervation to the lacrimal gland originates in the superior salivatory nucleus (pons) and travels within the nervus intermedius as part of the facial nerve. After branching off as the greater superficial petrosal nerve within the fallopian canal, they synapse at the pterygopalatine ganglion. Postganglionic fibers run superiorly into the orbit through the inferior orbital fissure, joining the zygomaticotemporal nerve. A branch of this nerve then runs into the lacrimal gland to stimulate reflex tearing.
128. c. These are the only fibers that synapse in the ciliary ganglion.
129. d. The inferior oblique is unique among the seven extraocular muscles in its anterior anatomic origin (from the orbital floor, just posterolateral to the lacrimal sac fossa).
130. c. Although the superior oblique has its anatomic origin at the orbital apex, it acts mechanically as if its origin were at the trochlea. Therefore, like the inferior oblique, the superior oblique generates a “pull” that is directed toward the front of the orbit.
131. c. The ophthalmic artery travels within the optic foramen.
132. c. The lacrimal, frontal, and trochlear nerves enter outside the annulus of Zinn. The nasociliary, oculomotor, and abducens nerves enter through the annulus.
133. b. Because the trochlear nerve is the only extraocular motor nerve to enter the orbit outside the muscle cone, it is usually the last to be affected by an appropriately administered retrobulbar block.
134. d. Each of the rectus muscles except the lateral, receives branches from two anterior ciliary arteries. The lateral rectus receives one. Therefore, there are a total of seven anterior ciliary arteries.
135. False. The anterior ciliary arteries supply the rectus muscles and then continue on, terminating anteriorly in the major arterial circle of the iris.
136. False. Motor units, defined as one terminal motor nerve branch and all the muscle fibers it serves, are smaller in the extraocular muscles than anywhere else in the body. This permits the finest control of force generation and muscle action possible.
137. c. The capsulopalpebral fascia is a condensation of noncontractile fascia from the sheaths of the inferior oblique and inferior rectus muscles. Like the aponeurosis of the levator muscle, it retracts the tarsus.
138. b. Lockwood’s ligament is a condensation of connective tissue in the inferior orbit and acts as a “sling” for the globe.
139. a. There are approximately 20 short posterior ciliary arteries, which enter the sclera in a circle around the optic nerve. There are two long posterior ciliary arteries, which usually enter the sclera on either side of the optic nerve at the 3- and 9-o’clock positions. The posterior ciliary arteries supply the uveal tract, the sclera, conjunctiva, and cilioretinal arteries. There are seven anterior ciliary arteries, which provide muscular feeding branches (supplying the extraocular muscles) and terminate in the major arterial circle of the iris.
140. False. Bowman’s “membrane” is an acellular condensation of corneal stroma immediately beneath the basement membrane of the corneal epithelium. It does
not regenerate when damaged. The term “Bowman’s layer” is more appropriate.

141. **False.** Once violated or destroyed by inflammatory pannus, Bowman’s layer is permanently disturbed, leaving a corneal scar.

142. **True.** Descemet’s membrane is primarily composed of type IV collagen and consists of an anterior banded portion (developed *in utero*) and a posterior non-banded portion (which increases throughout life).

143. **True.** Descemet’s membrane progressively thickens with age and with endothelial disease states.

144. **True.** The lens capsule is composed primarily of type IV collagen.

145. **c.** Ganglion cells are the first retinal cells to differentiate into a recognizable level.

146. **a.** Tight junctions do not exist between the photoreceptor layer and the retinal pigment epithelium (RPE). Spectral absorption peaks reflect different opsin structures and, subsequently, different interactions with the chromophore, which is 11-cis-retinaldehyde (11-cis-retinal) for all photoreceptors.

147. **b.** Rod outer segment shedding in animal models will persist even after several days in the dark. On the contrary, dark deprivation (i.e., constant light adaptation) will rapidly ablate normal rod disc shedding.

148. **b.** Systemic anesthetic toxicity can be seen with coadministration of indirect miotics and ester-type local anesthetics. This occurs because ester-type anesthetics are inactivated systemically by serum cholinesterase, which is blocked by the indirect miotics. Amide anesthetics are hepatically inactivated and, therefore, are not affected. Inactivation of systemic muscular depolarizing agents also depends on serum cholinesterase activity. Prolonged paralysis can result from coadministration with indirect miotics.

149. **e.**

150. **True.** The long arm of a chromosome is called the q arm.

151. **c.** Note that the diameter of the anatomic fovea and that of the optic nerve head are roughly equal.

152. **a.** Therefore, nuclear third-nerve palsy may feature contralateral upgaze palsy.

153. **a.** Pregnancy usually lowers intraocular pressure. Uveoscleral outflow is enhanced. Lowered episcleral venous pressure may contribute to enhanced trabecular outflow as well. Each of the other disorders may be precipitated by pregnancy.

154. **d.** The level of retinopathy before pregnancy is the strongest risk factor for significant progression during pregnancy. This, in turn, is usually directly proportional to the duration of diabetes.

155. **d.** The corneal and retinal pigment epithelium (RPE) damage is limited to historical reports of patients treated with much higher dosages, where the chance of toxicity is much higher.

156. **c.** Approximately half of patients with hypertension respond to monotherapy.

157. **a.** The reasons for this are not completely understood but may be secondary to impaired phagocytic functions among patients with hepatic failure.

158. **a.** Although ganciclovir has been shown to be effective against herpesviruses, it can cause myelosuppression and is only used intravenously for systemic disease (because of low oral absorption). Consequently, it is not used to treat herpes.

159. **c.**

160. **a.** There is evidence that smoking drives nuclear sclerosis progression.

161. **d.** Because of the considerable increase in diabetes over the last decade, the prevalence of cataracts secondary to diabetes will likely rise.

162. **b.** There is robust evidence that ultraviolet B light increases the risk of development of cortical cataracts.

163. **e.**

164. **d.** Once progression occurs, patients who rigorously control their blood sugars are twice as likely to recover, compared with less-controlled diabetics. Therefore, “early worsening” is a temporary effect among patients undergoing intensive treatment.

165. **d.** Valacyclovir is usually more expensive and more convenient regimen than acyclovir (t.i.d. compared to 5 times/day). Generic acyclovir can be considerably less expensive than valacyclovir.

166. **b.**

167. **b.** Carotid stenosis is somewhat controversial. Historically, it has been widely taught that reduced ocular perfusion caused by high-grade carotid stenosis reduces the vascular insult caused by diabetes. However, severe carotid stenosis may lead to an increased incidence of proliferative retinopathy as part of the ocular ischemic syndrome.

168. **e.** Patients with newly diagnosed type 1 diabetes rarely develop retinopathy within the first 5 years of diagnosis.

169. **a.** The exact date of onset of type 2 diabetes, unlike type 1 diabetes, is impossible to pinpoint. On average, patients with newly diagnosed type 2 diabetes have had the disease without being diagnosed for 7 years. Consequently, many patients with type 2 diabetes will have treatable retinopathy at the time of diagnosis of diabetes.

170. **b.** Each of the others can create considerable conjunctival hyperemia or corneal epithelial toxicity with superficial punctate keratitis (SPK).

171. **c.** Studies have shown that preoperative (72 hours) antibiotic reduces bacterial counts to a greater degree than 3 days of preoperative povidone-iodine. Adding
povidone-iodine at the time of surgery exerts a synergistic effect. Some studies have suggested that saline flushes actually increase bacterial colony counts.

173. d. In the Beaver Dam Eye Study, smoking was also correlated with retinal pigment epithelium (RPE) hyperplasia, but only in men.

174. c.

175. c.

176. a. Bradycardia is not a known side effect of the prostaglandin analogs.

177. d.

178. c. Varivax is a live attenuated vaccine. The vaccine is designed to confer immunity in those patients who are not previously exposed to varicella. Once exposure has occurred, the vaccine serves no purpose. Because of concerns about possible Reye’s syndrome, salicylates should be avoided following immunization.

179. b. A human metabolite of a bioflavonoid found in grapefruit juice, but not in orange juice, inhibits several cytochrome components of the P450 isozyme group.

180. a. A 17-year prospective series and a review of 267 cases published in 2003 found that 67% of patients had an extracocular focus of infection. Liver abscess was the most common extracocular focus for endogenous bacterial endophthalmitis, followed by endocarditis.

181. b. Diabetes mellitus (DM) is a risk factor for both types of endophthalmitis. Both types of endophthalmitis are associated with pain and decreased long-term visual acuity. Vitrectomy and/or vitreous tap with intravitreal antibiotics can be used to treat both forms of endophthalmitis. In endogenous endophthalmitis, Streptococcus and Staphylococcus aureus predominate. Intravenous antibiotics may be indicated for endogenous endophthalmitis to treat the source, whereas they usually play no role in the management of postoperative endophthalmitis (especially postoperatively in patients who have undergone cataract surgery, as per the Endophthalmitis Vitrectomy Study [EVS] study).

182. b. The most important predictor of good acuity postsurgery is the absence of preoperative retinopathy. More than 90% of individuals without diabetes will have vision of 20/50 or better following cataract extraction, compared with only 60% to 70% of patients with diabetes.

183. d.

184. d. Although no definite causative relation has been established, there is a clear association between each of these classes of medication and a reduction in the risk of subsequent Alzheimer’s disease. Nonsteroidal anti-inflammatory drugs (NSAIDs) may reduce inflammation caused by the deposition of amyloid. Estrogen is known to be a trophic factor in the central nervous system, particularly for cholinergic neurons, the most vulnerable type.

185. b. Oral folic acid supplementation starting 1 month before conception and continued during early fetal development has been shown to decrease the incidence of neural tube defects by 50%.

186. c. Low-dose aspirin therapy was shown to have virtually no effect on any ocular findings in patients with diabetes, including retinopathy and cataract. It did offer a small survival advantage and reduced the rate of myocardial infarction to a larger degree.

187. a. Concerns about lactic acidosis, an uncommon but serious adverse effect, delayed its approval in the United States. It acts by reducing hepatic gluconeogenesis and increasing peripheral glucose uptake. The most common adverse effects involve the gastrointestinal system (i.e., nausea, diarrhea, and dysgeusia). Metformin does not cause hypoglycemia or weight gain, unlike other oral hypoglycemic agents. Metformin should be discontinued before studies with iodinated contrast material.

188. c. Naphazoline and antazoline are traditional antihistamines. Levocabastine is a cyclobexylpiperdine. Lodoxamide is a mast cell stabilizer used for prophylaxis of atopic conditions, not for acute treatment. Ketorolac is a traditional nonsteroidal anti-inflammatory drug.

189. d. Angiotensin-converting enzyme (ACE) inhibitors, such as captopril, lower efferent arteriolar resistance and glomerular capillary pressure, independent of systemic blood pressure. The medication is generally started at the earliest sign of nephropathy. ACE inhibitors may also play a preventive role in diabetic retinopathy, although the evidence for this is not clear.

190. c. Most ulcers that are not temporally related to the use of nonsteroidal antiinflammatory drugs (NSAIDs) are now believed to be a complication of infection by the Gram-negative organism Helicobacter pylori. Traditional treatment of such ulcers will likely lead to its recurrence. Antibiotics (e.g., amoxicillin, bismuth, tetracycline, or metronidazole) are generally added to an antisecretory regimen (e.g., omeprazole and ranitidine).

191. a. Children between the ages of 3 and 6 years are at increased risk of Haemophilus influenzae infections. A vaccine for influenza virus will confer no protection. However, children with asthma should receive the vaccine. Children that need long-term aspirin therapy should also receive the vaccine (because of the risk of developing Reye’s syndrome).

192. a. Rod sensitivity is decreased, but this is not clinically evident.

193. c. Typical central retinal artery occlusion (CRAO) does not respond to hyperbaric oxygen (HBO). However, CRAO secondary to gas embolism may respond to HBO.

194. c. Carbonic anhydrase inhibitors are most likely to be of benefit in disorders of the retinal pigment epithelium.
Answers

(RPE) and significantly less likely to help in cases of primary retinal vascular disease. Trials have shown an unpredictably beneficial effect of acetazolamide in the other disorders listed.

195. b. Fishman and colleagues have established that 500 mg/day of acetazolamide is more effective than 250 mg/day. They have subsequently demonstrated that methazolamide 50 mg twice daily is less effective than acetazolamide.

196. d. No urine test results have been found to be affected by recent angiography. Cortisol, thyroxine, and quinidine are affected for up to 3 hours, with digoxin being affected for up to 12 hours. Serum creatinine levels may be affected, but this appears to be short-lived (<3 hours).

197. d. Typical lesions of interferon retinopathy include cotton-wool spots and retinal hemorrhages, particularly around the optic disc, which usually appear within 3 months of the onset of therapy. Ischemic optic neuropathy and proliferative retinopathy have also been reported.

198. a. The anterior lamellae consist of the skin, orbicularis, and associated fascial and vascular components. A vertical insufficiency of the anterior lamella can lead to congenital ectropion.

199. d. The central retinal artery is usually the first branch off of the opthalmic artery. The opthalmic artery is a branch of the internal carotid artery.

200. d. Cyclooxygenase-1 (COX-1) (not COX-2) is present primarily in the stomach. Therefore, selective COX-2 inhibitors have less gastrointestinal toxicity compared to nonsteroidal antiinflammatory drugs (NSAIDs), although there have been reports of gastrointestinal (GI) toxicity even with COX-2 inhibitors. Corticosteroids block the creation of arachidonic acid. Flurbiprofen is used to prevent prostaglandin-mediated miosis during surgery. Nordihydroguaiaretic acid (NDGA) is a lipoxygenase (LOX) inhibitor.

201. a. The lens is approximately 65% water. Lens proteins are divided into water-soluble and water-insoluble proteins. The water-soluble component can be divided into the alpha-crystallins and the beta-gamma crystallins (beta-crystallin proteins are the most abundant of the crystallins). The water-insoluble component can be divided into uvea-soluble and uvea-insoluble proteins.

202. c. Physostigmine is an indirect muscarinic agent that acts by reversible inhibition of acetylcholinesterase.

203. e. Direct-acting muscarinics (e.g., pilocarpine) produce forward displacement of the lens, leading to myopia.

204. d. Sildenafil (Viagra) can cause mydriasis. All of the other answers have been reported to occur.

205. a. 3, b. 3, c. 2, d. 3, e. 1, f. 1.

206. 1. c (atropine—1 to 2 weeks), 2. e (scopolamine—1 week), 3. a (homatropine—1 to 3 days), 4. b (cyclopentolate—12 to 24 hours), 5. d (tropicamide—2 to 4 hours).

207. a. 2, b. 1, c. 4, d. 1, e. 3, f. 2, g. 1.

208. a. 1, b. 3, c. 4, d. 2.

209. a. 3, b. 1, c. 4, d. 2.

210. a. 3, b. 1, c. 6, d. 5, e. 8, f. 2, g. 7, h. 4. Note that the outer plexiform layer is the region of photoreceptor—bipolar synapses as well as bipolar—horizontal interactions. The inner plexiform layer is the region of bipolar—ganglion cell synapses as well as bipolar—amacrine interactions.

Suggested Readings


Optics and Refraction

Questions

1. Which of the following statements about the wave properties of light is/are true?
   a. Light energy is proportional to its wavelength.
   b. When light enters a medium of higher index of refraction, it slows down.
   c. When light enters a medium of higher index of refraction, its frequency diminishes.
   d. The index of refraction of any material varies with the wavelength of incident light.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

2. T or F A photon of blue light has greater energy than a photon of red light.

3. The laser interferometer utilizes the principles of:
   a. constructive interference.
   b. destructive interference.
   c. high coherence.
   d. low coherence.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

4. The Haidinger brush phenomenon is caused by which special characteristic of light transmission?
   a. interference.
   b. polarization.
   c. diffraction.
   d. scattering.
   e. reflection.

5. Which of the following statements about diffraction is/are true?
   a. Diffraction is responsible for a limit on pinhole acuity of approximately 20/25.
   b. Diffraction is a limiting factor for visual acuity with pupils smaller than approximately 2 mm.
   c. Long wavelengths are diffracted more than short wavelengths.
   d. Diffraction is responsible for the blue color of the sky.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

6. The features of laser light that enhance its intensity or brightness include:
   a. directionality.
   b. coherence.
   c. polarization.
   d. polychromaticity.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
3: Optics and Refraction

7. T or F The material for which each laser is named corresponds to the substance that is actually emitting the amplified light.

8. Which of the following is/are means of increasing laser power per delivery?
   1. increasing energy per delivery.
   2. Q-switching.
   3. mode locking.
   4. increasing time of exposure at a set energy.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

9. Which of the following statements about refraction of light at interfaces is/are true?
   1. Light will bend toward the normal as it enters a medium of higher index of refraction.
   2. The index of refraction of any given substance is greater for longer wavelengths.
   3. Total internal reflection renders the anterior chamber angle invisible by a slit lamp.
   4. Light traversing a plane parallel plate at any incident angle is not refracted.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

10. T or F The minimum angle of deviation produced by a prism occurs when incident light strikes the prism perpendicular to its anterior face.

11. Which of the following statements about prisms and their units and calibration is/are true?
   1. The power in prism diopters is the number of centimeters that light is displaced perpendicularly for every centimeter that the light travels.
   2. Glass prisms are calibrated while held in the angle of minimum deviation.
   3. The Prentice position may be most closely approximated by placing a prism in the frontal plane.
   4. Real images created by prisms are deviated toward the prism base.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

12. T or F Any real image can be focused on a screen or photographic film.

13. Which of the following statements about linear magnification is/are true?
   1. Magnification is equal to the ratio of object size to image size.
   2. A magnification value <0 implies inversion of the image relative to the object.
   3. Magnification is equal to the image vergence divided by the object vergence.
   4. A magnification value >−1 and <+1 implies that the image is smaller than the object.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

14. T or F Any real image can be focused on a screen or photographic film.

15. If the average eye’s power is 60D, and its average (internal) index of refraction is 1.33, what is its approximate primary focal length in air?
   a. 17 cm.
   b. 17 mm.
   c. 22 cm.
   d. 22 mm.
   e. none of the above.

16. What is this average eye’s approximate secondary focal length?
   a. 17 cm.
   b. 17 mm.
   c. 22 cm.
   d. 22 mm.
   e. none of the above.
17. In a thick lens, refraction may be considered to take place at the:
   1. primary focal plane.
   2. primary principal plane.
   3. secondary focal plane.
   4. secondary principal plane.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

18. Which of the following statements about the cardinal planes of a thick lens is/are true?
   1. In optical systems with the same refractive medium on both sides, the nodal points and principal points coincide.
   2. If the media on either side have different indices of refraction, the nodal points are displaced toward the medium with the higher index of refraction.
   3. In the human eye, the principal planes may be considered to be superimposed.
   4. In the human eye, the nodal points may be considered to be superimposed.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

19. T or F The absolute value of the back vertex power of a meniscus spectacle lens is always greater than that of the front vertex power.

20. The angular magnification of a retinal image afforded by direct ophthalmoscopy in an emmetrope is approximately:
   a. 5×.
   b. 10×.
   c. 15×.
   d. 20×.
   e. 25×.

21. Which of the following statements about surgical loupes and telescopic low-vision aids is/are true?
   1. These devices are basically small astronomical telescopes.
   2. These devices generally feature an “add” lens.

22. Which of the following statements about optical aberrations of spherical lenses is/are true?
   1. The cornea’s steeper peripheral curvature tends to counteract spherical aberration.
   2. The typical interval of chromatic aberration in the human eye is approximately 2.5D.
   3. Tilting a lens along the horizontal axis will induce a sphere of the same sign and a cylinder of opposite sign along that axis.
   4. In an emmetrope, blue light is focused more anterior to the retina than the red light, which is focused posterior to it.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

23. T or F The image perceived when looking through a Maddox rod is virtual and perpendicular to the real image formed by the rod.

24. If a point source of light is placed a great distance to the left of a +2.00 +2.00 × 180 lens, what shape will an image located 37.5 cm to the right of the lens have?
   a. a vertical line.
   b. a vertical oval.
   c. a circle.
   d. a horizontal oval.
   e. a horizontal line.

25. Which of the following is/are cross-cylinders?
   1. +0.50 × 90, +0.50 × 180.
   2. +1.00 −0.50 × 90.
   3. +0.50 −0.50 × 180.
   4. +0.50 −1.00 × 90.
26. T or F The angular size of the field of view for an observer looking at a plane mirror may be increased by backing away from the mirror.

27. A patient with a corneal scar is carefully refracted. His best-corrected visual acuity is 20/40. With a pinhole over his correction, his acuity improves to 20/25. The best explanation for this is:
   a. spherical aberration.
   b. myopic astigmatism.
   c. cataract.
   d. irregular astigmatism.
   e. malingering.

28. Vernier acuity is important in taking measurements with which of the following ophthalmic instruments?
   1. keratometry.
   2. lensometry.
   3. applanation tonometry.
   4. automated refractors.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

29. In each of the following situations, which of the following pairs represent(s) conjugate planes?
   1. indirect ophthalmoscopy: the patient’s retina and the observer’s retina.
   2. the uncorrected nonaccommodating ametrope: the retina and the far point of the eye.
   3. neutralization in retinoscopy: the patient’s retina and the peephole of the retinoscope.
   4. indirect ophthalmoscopy: the patient’s pupil and the examiner’s pupils.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

30. T or F The binocular amplitude of accommodation is generally the same as the monocular amplitude of accommodation.

31. The accommodative amplitude of a 60-year-old healthy person is approximately:
   a. 14.0D.
   b. 10.0D.
   c. 6.0D.
   d. 1.5D.
   e. 0.5D.

32. In a group of healthy American adults, the average refractive error is approximately:
   a. −1.00D.
   b. −0.50D.
   c. plano.
   d. +0.50D.
   e. +1.00D.

33. Methods of reducing cylinder-induced distortion and asthenopia include:
   1. plus cylinder lenses.
   2. minimizing vertex distance.
   3. use of Jackson cross-cylinders of lowest possible power.
   4. rotation of the cylinder axis toward 90 or 180 degrees.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

34. Which of the following is/are problems with aphakic spectacle lenses?
   1. barrel distortion.
   2. ring scotoma.
   3. image minification.
   4. jack-in-the-box phenomenon.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

35. T or F Posterior chamber intraocular lenses (PCIOls) cause no image magnification.
36. A 57-year-old patient undergoing phacoemulsification with posterior chamber intraocular lens (Phaco/PCIOL) implantation has an average preoperative keratometry measurement of 44.5D and an axial length of 23.8 mm. The patient spends a lot of time driving and requests optimal correction for distance. Mistakenly, the surgeon enters 43.5D in his intraocular lens (IOL) power calculator (with the correct axial length), which recommends +18.00D for distance emmetropia. This lens is subsequently implanted. Which of the following statements about this situation is/are true?

1. The implant used is “too strong” for this eye.
2. The patient would probably have been happy if the surgeon had entered 24.2 mm for the axial length (instead of using the correct axial length).
3. The patient will require minus spectacle correction for optimal distance correction.
4. The miscalculation will not affect the choice of an appropriate anterior chamber intraocular lens (ACIOL).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

37. The most important factor in determining the A-constant of an intraocular lens (IOL) is the:

a. number of haptics.
b. chemical nature of the haptics.
c. configuration of the lens (e.g., biconvex, planoconvex).
d. use of surface passivation.
e. final lens position in the eye.

38. T or F Enhancement of the retinoscopic reflex is obtained by fully rotating the sleeve.

39. Which of the following statements about retinoscopic neutralization of a moderate refractive error is/are true?

1. As neutralization is approached, the streak moves faster.
2. As neutralization is approached, the streak becomes brighter.
3. As neutralization is approached, the streak becomes wider.
4. As neutralization is approached, the streak becomes narrower.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

40. An examiner sits 50 cm from a patient being refracted by retinoscopy. With the streak oriented in the horizontal meridian (sweeping vertically), a +3.00D sphere neutralizes the reflex, with the streak oriented in the vertical meridian (sweeping horizontally), a +5.00D sphere neutralizes the reflex. What is the patient’s final retinoscopic refraction?

a. +3.00 +2.00 × 090.
b. +3.00 +2.00 × 180.
c. +1.00 +2.00 × 90.
d. +1.00 +2.00 × 180.
e. +1.00 −2.00 × 180.

41. Which of the following retinoscopic reflex phenomena is/are useful for approximating the axis of small cylindrical errors?

1. intensity.
2. break.
3. skew.
4. thickness.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

42. T or F Astigmatic dial refraction is designed for use with plus cylinders.

43. T or F Cross-cylinder estimation of cylinder axis and power is best performed with the patient fogged.

44. T or F The red–green duochrome test is best introduced with the patient slightly fogged.

45. T or F The binocular balancing test is best performed with the patient fogged.
46. A 5-year-old child is noted to have a 20-prism-diopter esotropia that increases to 45 prism diopeters while reading at 20 cm. The patient’s pupillary distance is 50 mm. With the child reading through his distance correction, a +3.00D lens placed over each eye decreases the esotropia at near to 20 prism diopters. The patient’s accommodative convergence to accommodation ratio (AC/A), as determined by the gradient method, is:
   a. 3:1.
   b. 5:1.
   c. 8:1.
   d. 10:1.
   e. 15:1.

47. For the child described in the previous question, what would the accommodative convergence to accommodation ratio (AC/A) be, as determined by the heterophoria method?
   a. 3:1.
   b. 5:1.
   c. 8:1.
   d. 10:1.
   e. 15:1.

48. A 35-year-old patient with myopia presents to an ophthalmologist complaining of difficulty in reading. She recently started wearing soft contact lenses to enhance her career as a television news-caster. Potential reasons for her new difficulty is/are most likely:
   1. dry eyes.
   2. increased convergence demand.
   3. increase in relative magnification.
   4. increased accommodative demand.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

49. A patient is refracted to 20/20 visual acuity (at distance). Spheres that are +3.00D are added to this correction, and he is asked to read a 20/30 near target, which is brought progressively closer. He is able to read the target at distances between 20 and 40 cm. His comfortable working distance is approximately 40 cm. What is the correct power add for this man’s bifocals?
   a. +0.75D.
   b. +1.25D.
   c. +1.75D.
   d. +2.50D.
   e. +3.00D.

50. The age of the patient in question 49 is approximately:
   a. 36 years.
   b. 40 years.
   c. 44 years.
   d. 48 years.
   e. 52 years.

51. Assuming that the patient’s far point is actually 0.33 m, the distance refraction in question 49 must be:
   a. incorrect by +1.00D (i.e., 1.00D additional power).
   b. incorrect by +0.50D.
   c. completely appropriate.
   d. incorrect by −0.50D (i.e., 0.50D additional power).
   e. incorrect by −1.00D.

52. A 64-year-old architect presents to an ophthalmologist complaining of difficulty in reading at work but no difficulty with distance vision. His current correction is +5.00D in both eyes, with a +2.00D add. His range of clear vision with this current prescription includes:
   1. infinity to 100 cm.
   2. 20 to 15 cm.
   3. 50 to 33 cm.
   4. infinity to 15 cm.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

53. The architect in question 52 would like to be able to read his drawings at approximately 25 cm, as well as work on a drafting board at a distance of 50 to 60 cm. His corrected distance acuity is 20/20. The best prescription to address his needs might be:
   a. +5.00D with a +3.50D add.
   b. +6.00D with a +3.50D add.
   c. +5.00D with a +2.00D near add and a +1.00D intermediate add.
Questions

d. +5.00D with a +3.50D near add and a +1.50D intermediate add.

e. a referral to your least favorite partner.

54. T or F Image jump is primarily a problem for presbyopes with significant anisometropia.

55. T or F Image displacement is primarily a problem for presbyopes with significant anisometropia.

56. T or F For the anisometrope, image displacement is generally more troublesome than image jump.

57. A patient wearing new glasses comes in complaining of diplopia. The prescription is +3.00 −1.00 × 180 oculus dexter (OD) and −2.50 −1.00 × 90 oculus sinister (OS) with a +2.00D add oculus unitase (OU). You note a tropia present when the patient is reading at a comfortable distance. While the patient is reading, the visual axis is 2 cm nasal and 2 cm inferior to the distance optical center and 0.5 cm nasal and 1.2 cm above the optical center of the add in each lens. What is the induced prismatic effect for this patient?

a. 3 prism diopters base-up OD and 1 prism diopter base-out OD.

b. 3 prism diopters base-up OD and 1 prism diopter base-in OD.

c. 9 prism diopters base-down OD and 13 prism diopters base-out OD.

d. 9 prism diopters base-up OD and 13 prism diopters base-out OD.

e. 9 prism diopters base-up OD and 1 prism diopter base-out OD.

58. Alternate cover testing at near of the patient in question 57 probably revealed:

a. right hyper- and exodeviation.

b. left hyper- and exodeviation.

c. left hyper- and esodeviation.

d. right hyper- and esodeviation.

e. you cannot tell because you are thoroughly confused.

59. T or F The complaints of the patient in question 57 are due strictly to image jump.

60. Suitable methods for correcting the problem in question 58 might include:

1. press-on prisms.

2. “slab-off” grinding.

3. dissimilar segments.

4. contact lenses with reading glasses.

a. 1, 2, and 3.

b. 1 and 3.

c. 2 and 4.

d. 4 only.

e. 1, 2, 3, and 4.

61. Image jump is most troublesome for bifocals of the:

a. fused type.

b. round-top type.

c. flat-top type.

d. ribbon type.

62. T or F Ultraviolet (UV)-A light contains longer wavelengths than UV-C light.

63. A 72-year-old patient with bilateral macular degeneration has a distance acuity of 20/100. The add required for this patient to read newspaper print is:

a. +1.00D.

b. +3.00D.

c. +4.00D.

d. +5.00D.

64. The patient in question 63 should be informed that his working distance would be approximately:

a. 10 cm.

b. 20 cm.

c. 25 cm.

d. 35 cm.

e. 100 cm.

65. A 32-year-old patient with Stargardt’s disease has a distance acuity of 20/200. The add required for this patient to read newspaper print comfortably is:

a. +1.00D.

b. +3.00D.

c. +4.00D.

d. +6.00D.

e. +10.00D.

66. The patient in question 65 should be informed that his working distance would be approximately:

a. 10 cm.

b. 20 cm.
67. The advantages of hand-held magnifiers as low-vision aids include which of the following?
   1. greater working distance.
   2. greater ease of use for patients with poor manual dexterity.
   3. wider range of available magnifying powers.
   4. wider field of view.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

68. The main advantage of telescopic aids for near work is:
   a. decreased convergence requirement.
   b. wider field of view.
   c. greater depth of focus.
   d. greater working distance.
   e. greater flexibility regarding head position.

69. Which of the following is/are important in determining the oxygen flux across a contact lens?
   1. the diffusion coefficient for oxygen in the lens.
   2. the thickness of the central portion of the lens.
   3. the partial pressure gradient of oxygen across the lens.
   4. the solubility of oxygen in the lens.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

70. T or F As the diameter of a rigid contact lens increases (with a fixed radius of curvature), the lens becomes effectively steeper.

71. T or F As the radius of curvature of a rigid contact lens increases (with a constant diameter), the lens becomes steeper.

72. Which of the following patients has/have significant lenticular astigmatism?
   1. refraction: $-1.00 -1.00 \times 180$; keratometry: 43.0D at 90 degrees, 42.0D at 180 degrees.
   2. refraction: $-5.00 -3.00 \times 90$; keratometry: 44.0D at 90 degrees, 42.0D at 180 degrees.
   3. refraction: $-1.00 -2.00 \times 90$; keratometry: 42.0D at 90 degrees, 44.0D at 180 degrees.
   4. refraction: $-4.00 -1.00 \times 180$; keratometry: 42.0D at 90 degrees, 42.0D at 180 degrees.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

73. A patient requesting rigid gas-permeable (RGP) contact lenses has a best refraction oculus dexter (OD) of $-4.00 +1.25 \times 90$. Keratometry oculus dexter (OD) is 44.0D at 90 degrees and 42.5D at 180 degrees. The conversion to radius of curvature is 7.70 mm at 90 degrees, 7.95 mm at 180 degrees. The only posterior curve available is 7.80 mm. The contact lens power that should be prescribed for best vision is:
   a. $-2.00D$.
   b. $-2.75D$.
   c. $-3.00D$.
   d. $-3.50D$.
   e. $-4.00D$.

74. Which of the following are typically not causes of acquired myopia?
   a. miotics (e.g., pilocarpine).
   b. keratoconus.
   c. lenticous.
   d. peripheral lens dislocation.
   e. posterior lens dislocation.

75. T or F Diameter variations are more important in soft contact lens fitting than in rigid gas-permeable (RGP) contact lens fitting.

76. A patient requesting soft contact lenses has a spectacle correction of $-6.00 +1.00 \times 90$ (vertex distance = 15 mm) and keratometry oculus dexter (OD) of 44.0D (7.70 mm) at 90 degrees and
42.5D (7.95 mm) at 180 degrees. A lens with base curve 8.6 mm and diameter 14.5 mm is selected. The lens power for best acuity is:

- a. −5.00D.
- b. −5.50D.
- c. −6.00D.
- d. −6.50D.
- e. −7.00D.

77. Which of the following factors is/are important for increasing oxygen transmission across extended-wear contact lenses?

1. decreased lens thickness.
2. a minus carrier.
3. increased gas permeability constant (Dk).
4. ballasting.

- a. 1, 2, and 3.
- b. 1 and 3.
- c. 2 and 4.
- d. 4 only.
- e. 1, 2, 3, and 4.

78. T or F Images generated in fundus biomicroscopy using the Goldmann contact lens and +90D lenses are real inverted images.

79. Applanation tonometry measures the amount of force required to flatten an area of cornea with a diameter equal to:

- a. 3.06 mm.
- b. 6.12 mm.
- c. 1.53 mm.
- d. 0.06 mm.
- e. 3.06 cm.

80. T or F For every 4.00D of corneal astigmatism, applanation tonometry will be incorrect by 1 mm Hg.

81. Which one of the following statements about direct ophthalmoscopy is false?

- a. The linear magnification is 15 ×.
- b. The optic disc of a myopic eye appears larger than that of an emmetropic eye.
- c. The image is a virtual, upright image.
- d. The dial on a direct ophthalmoscope is intended to neutralize both the examiner’s and the patient’s refractive error.
- e. The optic disc of a hyperopic eye appears smaller than that of an emmetropic eye.

82. Which one of the following statements about indirect ophthalmoscopy of an emmetropic eye is false?

- a. The image the examiner observes is a real, inverted image in the focal plane of the condensing lens.
- b. Conjugate planes include the patient’s and the examiner’s retinas and the patient’s and examiner’s pupils.
- c. If the examiner uses a 20D condensing lens, the lateral magnification is approximately \(3 \times\) and the axial magnification approximately \(2.25 \times\).
- d. A 30D condensing lens provides greater magnification and a larger field of view than a 20D condensing lens.
- e. Aspheric condensing lenses are preferred for indirect ophthalmoscopy.

83. Which one of the following statements about keratometry is false?

- a. Corneal curvature is measured by using the cornea’s power as a convex mirror.
- b. A central image is doubled to negate the effect of eye movement.
- c. Conventional keratometry measures the curvature of the central 6 mm of the cornea.
- d. The refractive power of the average cornea equals 337.5 divided by its radius of curvature (in mm).
- e. Manual keratometry may be misleading following radial keratotomy or corneal transplantation.

84. A slide is placed 25 cm to the left of a lens. The image is in perfect focus on a screen 1 m to the right of the lens. What is the power of the lens?

- a. +0.25D.
- b. +3.00D.
- c. +4.00D.
- d. +5.00D.
- e. +3.00D.

85. How far from a +24D camera lens should an object be placed to be focused onto film 5 cm to the right of the lens?

- a. 25 cm to the left of the lens.
- b. 50 cm to the right of the lens.
- c. 50 cm to the left of the lens.
- d. 25 cm to the right of the lens.
- e. 20 cm to the left of the lens.
86. Where should a +8.00D lens be placed to form a real image 0.5 m from a real object (assuming that light travels from left to right)?
   a. 50 cm to the left of the object.
   b. 25 cm to the right of the object and 25 cm to the left of the image.
   c. 100 cm to the right of the object and 50 cm to the right of the image.
   d. 75 cm to the right of the object and 25 cm to the left of the image.
   e. 12.5 cm to the right of the object and 37.5 cm to the left of the image.

87. A patient wears a −10D spectacle lens at a vertex distance of 20 mm for distance correction. What power contact lens will be required for proper distance correction?
   a. −8.25D.
   b. −9.00D.
   c. −12.50D.
   d. −11.50D.
   e. −10.00D.

88. Where is the far point of an eye with a +4.00D “error lens”?
   a. 17 mm in front of the cornea.
   b. 25 cm in front of the cornea.
   c. 5.5 mm inside the cornea.
   d. 25 cm behind the cornea.
   e. at the retina.

89. Where is the far point of the eye corrected with a +4.00D lens?
   a. 17 mm in front of the cornea.
   b. 25 cm in front of the cornea.
   c. 5.5 mm inside the cornea.
   d. 25 cm behind the cornea.
   e. at the retina.

90. A patient with aphakia wears a spectacle correction of +10.0D at vertex distance of 20 mm. Where should a +3.0D lens be placed to correct this patient for distance?
   a. 10 cm in front of the eye.
   b. 25 cm in front of the eye.
   c. 33 cm in front of the eye.
   d. 35 cm in front of the eye.
   e. 50 cm in front of the eye.

91. A crystal ball with an opaque rear surface sits on a pedestal. Its internal radius of curvature is 50 cm. Its index of refraction is 3.00. Which of the following statements about this “lens” are correct (considering only the front surface of the crystal ball)?
   1. Its refractive power is +8.00D.
   2. Its primary focal length is 25 cm.
   3. Its secondary focal length is 25 cm.
   4. Light originating at infinity will come to focus inside the crystal ball.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

92. If the crystal ball’s radius is the same (50 cm), but its index of refraction is 1.50, which of the following statements is/are true?
   1. The refractive power of the crystal ball is +1.00D.
   2. The primary focal length of the crystal ball is 1 m.
   3. The secondary focal length of the crystal ball is 1.5 m.
   4. Light from infinity would be focused inside this crystal ball.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

93. Consider another crystal ball with an index of refraction of 1.50 and internal radius of curvature of 10 cm. Which of the following is/are true?
   1. Its primary focal length is 20 cm.
   2. Its power is +15.0D.
   3. Its secondary focal length is 30 cm.
   4. Light from infinity would come to focus inside this crystal ball.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

94. T or F If the center of curvature of a spherical refractive surface lies on the same side as object
Questions

95. T or F If the center of curvature of a spherical refractive surface lies on the same side as the medium of higher index of refraction, the net power of the surface is > 0.

96. A child is watching her goldfish swim inside a tank with internal radius of curvature of 1/3 m. If the goldfish swims up to her and returns her gaze at a distance of 1/3 m from the tank wall, where will the child believe the fish to be (assume the index of refraction of water is 4/3)?
   a. 1/5 m inside the tank.
   b. 4/15 m inside the tank.
   c. 1/2 m inside the tank.
   d. 4/9 m inside the tank.
   e. 1/3 m inside the tank.

97. A penlight is dropped into a rectangular aquarium filled with water so that the bulb burns 1/2 m from the aquarium side it faces. To an observer looking in, where will the bulb appear to be burning?
   a. 1/2 m inside the tank.
   b. 1/2 m outside the tank.
   c. at the tank surface.
   d. 3/8 m outside the tank.
   e. 3/8 m inside the tank.

98. Which of the following statements about the reduced schematic eye is/are true, if the effective power is assumed to be +60D and the internal index of refraction is assumed to be 1.33?
   1. The primary focal point is approximately 17 mm in front of the cornea.
   2. The secondary focal point is approximately 17 mm inside the cornea.
   3. The nodal point of the eye can be located with this information.
   4. The eye acts as a simple magnifier with power 4X.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

99. If a biconvex thin lens has a front surface with a radius of curvature of 10 cm, a back surface with a radius of curvature of 5 cm, and an index of refraction of 1.50, what is its total power in air?
   a. +5D.
   b. +10D.
   c. +15D.
   d. −5D.
   e. −10D.

100. An intraocular lens (IOL) implant is labeled as +18.0D. Its specifications list an index of refraction of 1.50. Which one of the following statements about the IOL power is correct?
   a. It is +18.0D in air and +54.0D in the eye.
   b. It is +18.0D in air and +6.0D in the eye.
   c. It is +18.0D in the eye and +6.0D in air.
   d. It is +18.0D in the eye and +54.0D in air.
   e. It is +18.0D in both the eye and air.

101. In the image ray diagram shown below, which of the following is/are true (assume all light travels left to right)?

   1. Image ray no. 1 must have passed through the lens’s secondary focal point.
   2. Image ray no. 2 must have been traveling parallel to the lens axis.
   3. This lens must be negative in power.
   4. Image ray no. 3 must have passed through the lens undeviated.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

102. T or F The true power of a meniscus lens is always greater than the back vertex power.

103. An afocal telescope is constructed with −10D and +4D lenses. What is the distance between the lenses?
   a. 5 cm.
   b. 15 cm.
c. 20 cm.
d. 25 cm.
e. 30 cm.

104. Which of the following statements about the image formed by this afocal telescope of an object at infinity is/are true (assuming an emmetropic user)?

1. With the −10D lens held closer to the eye, image magnification is 2.5×.
2. The image is upright.
3. The image is virtual.
4. No accommodation is required for viewing this image.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

105. An afocal telescope is constructed from +10D and +4D lenses. What is the distance between the lenses?

a. 10 cm.
b. 15 cm.
c. 20 cm.
d. 25 cm.
e. 35 cm.

106. Which of the following statements about the image formed by the afocal telescope in question 105 is/are true of an object 1 m away (assuming an emmetropic user)?

1. If the +10D lens is held near the eye, the image is magnified by a factor of 2.5×.
2. The image is upright.
3. The image is virtual.
4. No accommodation is required to view the image.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

107. Afocal surgical loupes are constructed with an eyepiece of −20D and an objective of +10D. A +5D add is placed immediately adjacent to the objective lens. Assume that the vertex distance (from eyepiece to eye) is zero. What is the accommodation required for viewing an object 25 cm from the eye?

a. 0D.
b. 1D.
c. 3.5D.
d. 10D.
e. 12D.

108. Using the loupes from question 107, approximately what accommodation is required to view an object 15 cm from the eye?

a. 0D.
b. 1D.
c. 3.5D.
d. 10D.
e. 13.5D.

109. What is the magnification of the loupes in question 107?

a. 1×.
b. 2×.
c. 2.5×.
d. 4×.
e. 5×.

110. A patient with aphakia uses a +12.5D contact lens for distance correction. How much image size change is generated with this correction?

a. 0%.
b. 7% magnification.
c. 7% minification.
d. 33% magnification.
e. 33% minification.

111. What is the appropriate spectacle to give the patient in question 110 for distance correction (assume vertex distance is 20 mm)?

a. +3.5D.
b. +8.0D.
c. +10D.
d. +12.5D.
e. +16.5D.

112. How much image size change will be generated by the spectacle correction in question 111?

a. 0%.
b. 33% magnification.
c. 33% minification.
d. 33 × magnification.
e. 33 × minification.
113. A high myope is corrected with a $-10\text{D}$ contact lens. How much image size change is generated with this correction?
   
   a. $0\%$.
   b. $5\%$ magnification.
   c. $5\%$ minification.
   d. $5 \times$ magnification.
   e. $5 \times$ minification.

114. What would be the correct spectacle correction for distance for the patient in question 113 (assume that vertex distance is 20 mm)?
   
   a. $-8.50\text{D}$.
   b. $-9.50\text{D}$.
   c. $-11.00\text{D}$.
   d. $-12.50\text{D}$.
   e. $-15.00\text{D}$.

115. What will be the image size change generated by the spectacle correction in question 114?
   
   a. $0\%$.
   b. $24\%$ magnification.
   c. $24\%$ minification.
   d. $24 \times$ magnification.
   e. $24 \times$ minification.

116. A man stands 1 m in front of a plane mirror. Which of the following statements is/are true about the image?
   
   1. The image is 50 cm in back of the mirror.
   2. The image is real.
   3. The man could see more of himself by moving farther away from the mirror.
   4. The image is upright.
   
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

117. Where is the image of a real object located 50 cm in front of a convex mirror with radius of curvature equal to 20 cm?
   
   a. The image is 8.33 cm in front of the mirror.
   b. The image is 12.5 mm in front of the mirror.
   c. The image is 8.33 cm in back of the mirror.
   d. The image is 12.5 cm in back of the mirror.
   e. The image is 12.5 mm in back of the mirror.

118. T or F The image created in question 117 is virtual.

119. T or F The image created in question 117 is magnified by a factor of $6 \times$.

120. T or F The image created in question 117 is upright.

121. Assume that the cornea has an index of refraction equal to 1.38, that its front surface has a radius of curvature 7.7 mm, and that its rear surface has a radius of curvature equal to 7.6 mm. Also assume that the index of refraction of aqueous humor is 1.33. Which one of the following is false?
   
   a. The posterior refractive surface of the cornea has negative power.
   b. The total refractive power of the cornea is $+42.8\text{D}$.
   c. The anterior reflecting surface of the cornea has negative power.
   d. The total reflecting power of the anterior corneal surface is $-42.75\text{D}$.
   e. The net refractive power of the cornea may be represented by one spherical surface with a reduced radius of curvature of 7.9 mm and an index of refraction of 1.3375.

122. A first-year ophthalmology resident decides to verify the size of an anterior chamber intraocular lens (ACIOL) implant in a new patient whose implant card records an optic diameter of 6.0 mm. Assuming that the patient’s cornea is identical to that described in question 121 and that the implant rests on the iris 3 mm posterior to the anterior corneal surface, what will the resident record as the optic diameter with his slit lamp calipers?
   
   a. 4.4 mm.
   b. 5.5 mm.
   c. 6.0 mm.
   d. 6.7 mm.
   e. 7.8 mm.

123. A patient’s pupillary reflex (reflection) is checked with a penlight held 40 cm away from her cornea. Where is the image of the penlight located (using the average corneal parameters given in question 121)?
   
   a. 3.8 mm outside the eye.
   b. 3.8 mm inside the eye.
   c. 2.5 mm outside the eye.
d. 2.5 mm inside the eye.
e. at the anterior corneal surface.

124. To facilitate indirect ophthalmoscopy through a small pupil, an examiner can:
a. move the condensing lens closer to the patient.
b. move the condensing lens away from the patient.
c. move his/her head toward the lens.
d. move his/her head away from the lens.
e. increase illumination of the patient’s fundus.

125. Novices at indirect ophthalmoscopy tend to move closer to the patient than the optimal examining distance to:
a. enhance image depth.
b. enhance image size and detail.
c. ease examination through small pupils.
d. rest their arms.
e. increase illumination of the patient’s fundus.

126. Annoying reflexes during indirect ophthalmoscopy may be moved out of the line of visualization if the examiner:
a. moves the condensing lens closer to the patient.
b. moves the condensing lens away from the patient.
c. moves his/her head toward the lens.
d. moves his/her head away from the lens.
e. tilts the lens obliquely.

127. In retinoscopy with plano-mirror technique, which of the following statements is/are true?
1. The image of the streak is between the examiner and patient.
2. “With” motion is neutralized using plus lenses.
3. With a Welch-Allyn retinoscope, the handle must be up.
4. “Against” motion implies that the patient’s far point is between the examiner and the patient.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

128. T or F By Knapp’s rule, correction of any anisometropia with a lens at the primary focal point of the eye will result in no disparity in retinal image sizes.

129. During an average night’s sleep, the human cornea:
a. becomes 10% to 12% thinner.
b. becomes 4% to 8% thinner.
c. does not change in thickness.
d. becomes 4% to 8% thicker.
e. becomes 10% to 12% thicker.

130. Which of the following statements about diurnal refractive fluctuations following incisional refractive surgery is false?
a. Diurnal refractive fluctuations are more common with incisional keratotomy than with photorefractive keratoplasty.
b. Fluctuations in the second eye closely parallel those of the first eye.
c. Diurnal fluctuations have been measured up to 11 years after refractive surgery.
d. The most likely explanation for the fluctuation is diurnal alterations in corneal thickness.
e. The most frequently seen pattern is a hyperopic shift from morning to evening.

131. Which one of the following statements about postoperative findings in photorefractive keratectomy (PRK) is false?
a. Pain is generally more severe and long-lasting in PRK than in radial keratotomy (RK).
b. Topical nonsteroidal antiinflammatory drugs (NSAIDs) are used to reduce postoperative pain.
c. Topical NSAIDs are used to reduce postoperative regression following PRK.
d. Topical NSAIDs are used to reduce the frequency of overcorrection following PRK.
e. Overcorrection and regression of the refractive effects of PRK are both more common with increasing degrees of preoperative myopia.

132. Which of the following is/are potential explanations for reduction in best-corrected visual acuity following photorefractive keratectomy for myopia?
1. corneal stromal haze.
2. residual stromal central islands.
3. irregular epithelial thickening.
4. the expected reduction in magnification.
Questions

133. Keratomileusis has become a useful treatment for higher degrees of myopia, as well as low to moderate hyperopia. Which innovation(s) has/have led to its increased prevalence?
   a. topical antimetabolites to reduce corneal scarring.
   b. immunosuppressives to prevent rejection.
   c. bandage contact lenses and topical non-steroidals to reduce pain.
   d. hydrogel corneal implants to reduce late regression of the surgical effect.
   e. automation microkeratomes and computer laser nomograms to enhance predictability.

134. Which of the following definitions is incorrect?
   a. Photocoagulation: thermally induced structural changes in tissue through the use of a laser.
   b. Photodisruption: ionization of tissue and subsequent rupture of surrounding tissue.
   c. Photoablation: removal of tissue through light- and thermal-induced damage with surrounding tissue destruction.
   d. Laser coherence: propagated energy from the laser source is in phase.
   e. Laser intensity: the power of a laser beam of a given angular size.

135. If light traveling in glass has a vergence of +1.00D and if the index of refraction of glass is 1.5, then what is its reduced vergence?
   a. +0.66D.
   b. +0.75D.
   c. −0.50D.
   d. +1.50D.
   e. none of the above.

136. Which of the following will yield artificially high intraocular pressure (IOP) measurements during applanation?
   a. applanation over corneal edema.
   b. applanation against a soft contact lens.
   c. applanation after scleral buckling surgery.
   d. applanation after laser-assisted in situ keratomileusis (LASIK) surgery.
   e. applanation over a corneal scar.

137. Which of the following substances has the highest index of refraction?
   a. air.
   b. cornea.
   c. vitreous humor.
   d. polymethylmethacrylate (PMMA).
   e. high-index refractive lenses.

138. Why does the sky appear blue?
   a. Blue light is scattered more than other light because of its longer wavelength.
   b. Blue light is scattered less than other light because of its longer wavelength.
   c. Shorter wavelengths are scattered more.
   d. Coherence.
   e. None of the above.

139. Which of the following terms is defined correctly?
   a. aniseikonia: a difference in perceived image size between the two eyes.
   b. anisocoria: a difference in pupillary size.
   c. anisometropia: a refractive error difference between the two eyes (typically >2D).
   d. coma: an effect of spherical aberration which causes light rays to be distributed in the shape of a comet.
   e. all of the above.
3: Optics and Refraction

Answers

1. c. Light energy is proportional to its frequency. When light enters a medium of higher refractive index, its velocity decreases; its frequency remains the same, but its wavelength decreases. (The velocity of any wavelength of light, \( c \), in any transparent medium is always equal to the product of its wavelength and its frequency.) The index of refraction decreases with increasing wavelength (i.e., shorter wavelengths are refracted more).

2. True. The energy of a photon of light is proportional to its frequency as given by the equation:

\[
E = h \times \text{frequency}
\]

where \( h \) = Planck's constant. Blue light is of higher frequency than red light and therefore has greater energy per photon.

3. a. The laser interferometer uses the highly coherent light of the laser to create an interference pattern on the retina. The pattern seen by the patient consists of light and dark bands created by constructive and destructive interference, respectively.

4. b. The Haidinger brush phenomenon is useful in sensory testing of the fovea. The phenomenon is created by rotating a polarizer continuously in front of a uniform blue field. The normal subject will see a rotating figure that looks like a double-ended brush. The effect is created because Henle's layer of the macula (outer plexiform layer) is oriented in such a way as to polarize incoming light.

5. a. Because of diffractive effects, pinhole vision is rarely better than 20/25 even with an optimal pinhole aperture of 1.2 mm. Pupil sizes less than approximately 2.5 mm will create diffractive effects that limit acuity. The sky is blue because light of higher frequency is scattered more than light of lower frequency. Diffraction increases with the wavelength of incident light.

6. a. A laser beam's high directionality, coherence, and linear polarization enhance its intensity. Laser light is monochromatic (one precise wavelength).

7. True. The active medium is the chemical environment (e.g., argon, krypton) that supports stimulated emission of coherent light.

8. a. Power is defined as energy per unit time. Increasing energy while keeping exposure time constant increases power. Decreasing the time over which an amount of energy is delivered also increases power. Q-switching and mode locking are means of increasing the peak power a laser is able to generate.

9. b. The index of refraction for any medium is greater for shorter wavelengths. Refraction will occur at any interface between media with different indices of refraction. According to Snell's law, light rays striking the interface perpendicular to the interface will not be bent. Light traversing a plane parallel plate is refracted twice (at the front and back surfaces).

10. False. The angle of minimum deviation is produced by a prism when light undergoes equal bending at the two faces. When light strikes the prism perpendicular to one of its surfaces (Prentice position), the angle of deviation is greater.

11. d. One prism diopter is defined as a 1-cm perpendicular displacement over a 100-cm distance. Glass prisms are usually calibrated in the Prentice position, in which one face is perpendicular to the incident light. Plastic prisms and prism bars are usually calibrated with the angle of minimum deviation. When measuring strabismus with glass prisms, they must be held in this position to be accurate; that is, glass prisms must be held with one surface perpendicular to the visual axis. Real images are deviated toward the prism base. Virtual images are deviated toward the apex.

12. False. Real images can usually be formed on a screen. One exception is the real image of the retina formed by an indirect ophthalmoscope. This real image cannot be formed on a screen because the screen would block the illumination system of the ophthalmoscope.

13. c. Magnification is equal to the ratio of image size to object size (\( i/o \)) or object vergence to image vergence (\( U/V \)), since size and vergence are reciprocally related. Magnification \(<0\) (i.e., a negative value) implies an inverted image. Magnification \(<1\) (or, if negative, \( >-1\)) implies an image smaller than the object (minification).

14. False. Reduced vergence is equal to vergence multiplied by an index of refraction of the medium the light is traveling in. The reduced vergence is greater than the vergence of an object because the refractive index of the medium the light is traveling in is \( >1\).

15. b. The primary focal length (in meters) of any lens system equals the index of refraction of the object space divided by the lens power \( (f_1 = n_1/P) \). In air, \( n_1 = 1\), so \( f_1 = 1/P \). The reciprocal of 60 D (the power of the average eye) is 0.017 m, or 17 mm.

16. d. The secondary focal length (in meters) of any lens system equals the index of refraction of the image space divided by the lens power \( (f_2 = n_2/P) \). The refractive index of the ocular media is 1.33;

\[
f_2 = \frac{1.33}{60D} = 0.0222 \text{ m} \text{, or 22.2 mm.}
\]

17. c. In a thick lens system, the primary principal plane acts as an infinitely thin refracting surface at which object light originating from the primary focal plane is converted into parallel light rays. The secondary principal plane acts as an infinitely thin refracting surface at which incoming parallel light is made to converge
Answers

toward the secondary focal point. No refraction is considered to occur at the primary or secondary focal planes in a thick lens system.

18. c. One should also remember that if the refractive media on the two sides are different, the principal points are displaced toward the medium of lower index of refraction.

19. True. Because the focal length measured from the “rear” vertex of a meniscus lens is always shorter than the corresponding “front” focal length, the absolute value of the back vertex power (P = n/f, where n = 1 in air) is always greater than that of the front vertex power. The true power is somewhere between the two.

20. c. The magnification of a simple plus lens is defined as the ratio of the angular size of the image produced by the lens to the angular size of the object viewed at 25 cm. The formula for angular magnification by a simple plus lens of power P is M = P/4. Because the average power of an emmetropic eye is +60D, angular magnification M = 60/4 = 15 X.

21. c. The devices mentioned are usually small Galilean telescopes, since Galilean telescopes are shorter and provide an upright image without additional optical elements. The total magnification is equal to the magnifying power of the “add” (P/4, magnification of a plus lens; see answer 20) multiplied by the power of the Galilean telescope—which is equal to ([power of eyepiece]/[power of objective]).

22. d. The flatter (not steeper) peripheral cornea tends to counteract spherical aberration. The total interval of chromatic aberration in the human eye is approximately 1.25D. Tilting a lens along its horizontal axis induces cylinder of the same (not opposite) sign and along the same axis, as well as sphere of the same sign. Shorter wavelengths of light are refracted more than longer wavelengths. When polychromatic light is focused on the retina (as is the case in emmetropia or proper spectacle correction), blue light is 0.87D anterior to the retina, green light is 0.37D anterior to the retina, and red light is 0.37D posterior to the retina.

23. True. A Maddox line produces two line images of a point light source: a real image that is too close to the eye to be seen, as well as a virtual image running through the light source. The real image is parallel to the cylinders’ axes, whereas the virtual image is perpendicular. This is true for all spherocylindrical lenses (even those with no power in one meridian).

24. b. The power of this lens is +4.00D at the 90-degree meridian. Therefore, a horizontal focal line will be formed 25 cm from the lens. The power of this lens is +2.00D at the 180-degree meridian. Therefore, a vertical focal line will be formed 50 cm from the lens. The circle of least confusion is located halfway, dioptrically, between the two focal lines (i.e., at the spherical equivalent). In this case, the circle of least confusion is at 3.00D, or 33 cm. The image at 37.5 cm is between the circle and the vertical line. Thus, the image will be a vertical oval, as shown in the figure below.

25. d. A cross-cylinder is a lens made of two cylinders of equal but opposite power at right angles to one another. The spherical equivalent of any cross-cylinder is zero. Notice that option 1 is a +0.50 sphere.

26. False. The angular size of the field of view for an observer looking at a plane mirror is independent of the observer’s position.

27. d. The corneal scar may be producing irregular astigmatism, which cannot be corrected by spherocylindrical spectacle lenses. The pinhole greatly minimizes nonaxial light rays, which require refraction to come into focus on the retina.

28. a. Vernier acuity refers to the ability of a normal human eye to discriminate between two line segments in the frontal plane separated by as little as 8 seconds of arc (one third the diameter of a cone). Determination of accurate end points with the keratometer, lensometer, and applanation tonometer relies on this degree of spatial discrimination.

29. c. Conjugate planes are related by being the object and image of one another.

30. False. The binocular amplitude of accommodation usually exceeds the monococular amplitude of accommodation.

31. d. Accommodative amplitude decreases with age with the following approximations:

1. Accommodative amplitude at age 40 = 6.0D, at age 44 = 4.5D, and at age 48 = 3.0D.
2. Below age 40, accommodative amplitude increases by 1.0D for every 4 years.
3. Above age 48, accommodative amplitude decreases by 0.5D for every 4 years. Here, the accommodative amplitude for a 60-year-old equals:

\[ 3.0D - 1.5D = 1.5D. \]

(Please note that many strabismus and optics experts object to the use of these accommodative tables because they are approximations and may be incorrect in
Empiric formulas have been developed that predict the magnification associated with any corrective intraocular lens. Consequently, it is possible to determine the proper intraocular lens (IOL) power for emmetropia (PCIOLs) that will yield less magnification than anterior chamber intraocular lenses (ACIOLs) to 119 (for biconvex posterior chamber intraocular lenses [PCIOLs]) to 113 (for anterior chamber intraocular lenses [ACIOLs]).

Although the development of the refractive state requires that the surgeon estimate the IOL required for emmetropia, general trends appear evident. Low-grade hyperopia in infancy and childhood usually drifts toward emmetropia in adult life. Obviously, posterior chamber intraocular lenses (PCIOLs) yield less magnification than anterior chamber intraocular lenses (ACIOLs).

Most distortions of aphakic spectacle lenses stem from their position anterior to the pupil and include image magnification, ring scotoma, pincushion distortion, and the so-called jack-in-the-box phenomenon. Aphakic spectacle lenses can also create cosmetic problems not only because of their size but also because they can make the patient’s eyes look magnified.

The A-constant depends on factors that determine the final lens position in the eye, such as anterior versus posterior chamber placement, haptic angulation, and the shape of lens. The A-constant usually ranges from 113 (for anterior chamber intraocular lenses [ACIOLs]) to 119 (for biconvex posterior chamber intraocular lenses [PCIOLs]).

If the refractive error is very large, the beam will initially become more narrow as neutralization is approached, then widen again near the appropriate correction.

The working distance is 50 cm, so 2.00D must be subtracted from the spherical portion, giving a net result of +1.00 + 2.00 × 90.

At the correct axis, the following are true:
1. The break phenomenon disappears (the intercept and reflex are parallel).
2. The width of the streak is narrowest.
3. The intensity is brightest.
4. Skew motion is no longer observed.

Intensity and skew are primarily useful for small astigmatic errors, while break and width are best judged with an enhanced streak. Astigmatic errors of <1.0D do not enhance well, so break and width are not readily appreciable.

To use an astigmatic dial, the patient is fogged to relax accommodation, and the “sharpest and clearest” line is sought, representing one of the principal meridians. The astigmatism is neutralized by adding minus cylinder perpendicular to this line until all lines are equally clear. Remember that the axis of this correcting cylinder is the mirror image of the clockline when placed over the patient. A simple rule for converting from clock hours to trial frame axis is as follows: Take the lower of the two clock hours belonging to the line 90 degrees from the clearest line on the dial. Multiply this number by 30. This represents the axis for the correcting minus cylinder. For example, if the fogged patient reports that the 2 to 8 line is sharpest, the minus cylinder must be placed with its axis parallel to the 11 to 5 line. The lower of the two numbers is 5. Multiplying by 30, the minus cylinder should be placed with its axis at 150 degrees.
accommodation, then adding minus sphere until vision is sharpest. At this point, the circle of least confusion is on the retina. Then, the cross-cylinder is introduced to find axis and power. The circle of least confusion must remain on the retina throughout cross-cylinder testing. When fogged, the circle of least confusion is anterior to the retina.

44. True. The sphere end point can be verified by the duochrome test, but this test does not relax accommodation. Therefore, the test should be introduced with the patient slightly fogged, such that the letters on the red side are clearer (the red letters will focus behind the green letters, closer to the retina). Then, minus sphere is added until letters on the green and red sides are equally clear.

45. True. Except during cycloplegic refraction, fogging is needed to remove unequal accommodative influences. If the eyes are not balanced, fogging will degrade vision less in the eye that is more overminused.

46. c. Using the gradient method, the ratio of prism diopeters of accommodative convergence to diopeters of accommodation (AC/A) is calculated by placing plus lenses in front of each eye and determining the change in the deviation at near. This change, divided by the power of the plus lens used (amount of weakened accommodation), gives the number of prism diopeters of accommodative convergence per diopeter of accommodation. In this case, 45 – 20 = 25, divided by 3 is (approximately) 8:1. Gradient method:

\[ \frac{AC}{A} = \frac{\text{deviation with lens} - \text{deviation without lens}}{\text{lens power}} \]

In these calculations, esodeviations are positive and exodeviations are negative, by convention.

47. d. The heterophoria method makes use of similar reasoning, while taking into account the effect of interpupillary distance (PD); that is, larger PDs require greater convergence for fusion. Heterophoria method:

\[ \frac{AC}{A} = \frac{\text{deviation at near} - \text{deviation at distance}}{\text{PD}} \]

where PD is interpupillary distance in cm. The denominator represents the number of diopeters of accommodation required for the near target (the reciprocal of the reading distance in meters). In this case, PD is 5 cm, the deviation at near is 45 prism diopeters, and the deviation at distance is 20D. Accommodation will be 5D at 20 cm. Thus,

\[ \frac{AC}{A} = \frac{45 - 20}{5} = 8:1 \]

48. c. There are two factors: The shorter vertex distance of contact lenses results in a greater accommodative demand for myopes and a decreased accommodative demand for hyperopes. More convergence also will be required for near work because the patient will lose the helpful base-in prismatic effect of her myopic spectacles.

49. b. This is the type of measurement that can be made using a Prince rule. After being “fogged” with the +3.00D adds, the patient is found to have an effective far point of 40 cm (2.5D) and an effective near point of 20 cm (5.0D). This patient must have a range of accommodation of 5.0 – 2.5, or 2.5D. Experience has shown that, in general, about half of this is available without causing strain (accommodative reserve). Our patient would therefore have approximately 1.25D of readily available accommodative reserve. His working distance of 40 cm calls for 2.50D of total accommodation, of which he can comfortably provide 1.25D. Therefore, an add of +1.25D (2.50 – 1.25) would be most appropriate.

50. c. See answer 31.

51. d. The patient’s distance correction must be approximately 0.5D “overminused,” since fogging with +3.00D brought his far point to 0.40 m (2.5D), rather than the expected 0.33 m for an appropriately corrected eye.

52. b. A typical 64-year-old has a 1.0D amplitude of accommodation. Assuming proper distance correction, the range of clear vision with distance correction is from infinity (relaxed accommodation) to 100 cm (using his full 1.0D of accommodation). His +2.00D add provides clear vision from 50 cm (relaxed accommodation) to 33 cm (using his full 1.0D of accommodation and his +2.00D add).

53. d. One way to fulfill all his visual needs is with a trifocal combination. An add of +1.50/ +3.50 gives an intermediate range of 40 to 67 cm (+1.50D lens coupled with +1.00D maximal accommodation) and near range from 22 to 30 cm (+3.50 plus 1.0D of accommodation). Remember, he will only be able to comfortably use 0.5D of his full 1.0D accommodative amplitude.

54. False. Image jump is a phenomenon related to encountering the sudden prismatic effect of a reading add as the gaze is moved downward into the add. The sudden addition of the prismatic effect of the add causes images to “jump,” typically upward. Image jump can occur in any correction whose add does not have its optical center at the top of the segment. Round-top segments typically produce more image jump than flat-top segments, as they have much lower optical centers.

55. True. Image displacement occurs in all corrections as gaze moves away from the optical center of the correcting lens; this is unlikely to be significantly distressing unless an imbalanced displacement occurs, as in anisometropia. The other situation where image displacement is troublesome is when an increased demand is made on an already taxed vertical fusional system ability, as in compensated vertical phoria.

56. True. On the other hand, occupational demands can often make image jump more troublesome.
57. e. Solving this problem is facilitated by preparing diagrams marked with the powers in the 90- and 180-degree meridians, the visual axes, and the optical axes for the distance corrections and adds. Image displacement in prism diopters is:

\[(\text{cm from the optical axis in the given meridian}) \times (\text{power of the lens in diopters})\]

This needs to be done for both the distance corrections and the adds separately. After this, the results are combined—first for each side, then for a net effect on both eyes, as shown in the figure below.

- **OD**: Distance
  - Vertical meridian: \(2.0 \times +2.00 = 4.0 \text{ base up}\)
  - Horizontal meridian: \(2.0 \times +3.00 = 6.0 \text{ base out}\)
- **Add**
  - Vertical meridian: \(1.2 \times +2.00 = 2.4 \text{ base down}\)
  - Horizontal meridian: \(0.5 \times +2.00 = 1.0 \text{ base out}\)

**Net OD**: 1.6 base up, 7.0 base out

- **OS**: Distance
  - Vertical meridian: \(2.0 \times -2.50 = 5.0 \text{ base down}\)
  - Horizontal meridian: \(2.0 \times -3.50 = 7.0 \text{ base in}\)
- **Add**
  - Vertical meridian: \(1.2 \times -2.00 = 2.4 \text{ base down}\)
  - Horizontal meridian: \(0.5 \times -2.00 = 1.0 \text{ base out}\)

**Net OS**: 7.4 base down, 8.0 base in

Note that when determining a net prismatic effect for two eyes, vertical prisms of different base orientation (base up plus base down) are additive, whereas horizontal prisms of same orientation (base in plus base in, or base out plus base out) are additive. For the net effect over one eye, prisms of the same orientation are additive (e.g., base up plus base up, base in plus base in).

58. a. It is easiest to visualize the patient’s deviation by considering what correcting prisms would be needed: 9 prism diopters base up, and 1 prism diopter base out OD is corrected with 9 prism diopters base down and 1 prism diopter base in OD. Since the gaze deviation is away from the base of any correcting prism, right hyper- and right exodeviations must be present.

59. False. This patient’s problem with diplopia is caused by image displacement–induced phoria/tropia, not image jump. Note that the image displacement is mostly caused by the anisometropic distance segments.

60. e. Press-on prisms are occasionally helpful. Slab-off grinding and dissimilar segments act by changing the position of the optical centers of the lenses to lessen the differential prism. Contact lenses obviate the problem entirely. This is true because the visual axis is always through the optical center of a contact lens, regardless of viewing position. Thus, no prismatic effect from anisometropic correction is created.

61. b. Image jump occurs at the segmentation line when the optical center of the add is not at its upper edge. A round-top bifocal segment has its optical center farther from its upper edge than other types and therefore causes the greatest image jump. These are commonly used only for aphakic spectacles to minimize image displacement.

62. True. UV-A: 400 to 320 nm—90% of UV light from sun. UV-B: 320 to 280 nm—10% of UV light from
sun. UV-C: <280 nm—absorbed by the ozone layer (which unfortunately appears to be diminishing over time).

63. d. For near vision aids, the reciprocal of the Snellen fraction at distance gives the approximate accommodation needed to read newspaper print. This is known as Kestenbaum’s rule. With distance vision of 20/100, accommodation equal to 100/20, or +5.0D, is required. Because a 72-year-old has <1.00D of accommodative amplitude, he will require the entire +5.00D as an add.

64. b. By Kestenbaum’s rule, the reciprocal of the distance acuity is the working distance (in diopters) required for reading newsprint. The working distance in centimeters is equal to 100 divided by this diopter value.

65. d. A 32-year-old has approximately 8.00D of accommodative amplitude, 4.00D of which can be comfortably used (accommodative reserve). Because this patient requires 10.00D of accommodation to read newsprint (20/200 distance acuity), an add of at least +6.00D is required for comfortable reading.

66. a. The working distance in diopters is given by Kestenbaum’s rule as the reciprocal of the distance acuity. The distance in centimeters is 100 divided by this diopter value. Note that the power of the add has nothing to do with the working distance in younger patients.

67. b. Hand-held magnifiers have a greater working distance (i.e., a greater eye to object distance). They are available in a range of powers from +3.00 to +68.00D. However, they do have a smaller field of view than high adds and must be held in the hand.

68. d. Afocal telescopes magnify without decreasing the working distance. Adds may be used to lessen the accommodative demand of these aids. However, they do have a small field of view and small depth of field, so the head must be positioned precisely.

69. e. \[ O_2 \text{ flux} = \frac{(D \times K \times P)}{L}, \]
where \( D \) = diffusion coefficient of oxygen in the lens, \( K \) = solubility of oxygen in the lens, \( P \) = partial pressure gradient of oxygen across the lens, and \( L \) = central thickness of the lens.

70. True. Holding the radius of curvature constant, larger diameter lenses are effectively steeper.

71. False. At a given diameter, radius of curvature and lens steepness are inversely proportional.

72. c. Lenticular astigmatism is defined as astigmatism detected on refraction in excess of the corneal astigmatism. In cases where the refractive cylinder matches the keratometric cylinder, there can be no significant lenticular astigmatism.

73. d. When fitting rigid contact lenses, if there is a >0.2 mm difference between the radii of the principal corneal meridians, then a lens of intermediate curvature, steeper than the flatter meridian by one third to one half the difference between meridians, can be used. This creates a positive tear film lens (add 0.25D per every 0.05-mm difference in radius of curvature), so minus power must be added to the original power. The starting power is the sphere of the refraction written in minus cylinder form and corrected for vertex distance if necessary (powers \( >\pm 4.00 \text{D} \)).

74. c. Posterior lens dislocation typically causes induced hyperopia (anterior lens dislocation is usually a cause of induced myopia). Peripheral lens dislocation (e.g., with ectopia lentis) usually causes acquired myopia. Some causes of acquired hyperopia include conditions which push the retina forward (e.g., central serous chorioretinopathy, orbital masses causing pressure on the posterior surface of the globe), aphakia, and conditions which weaken accommodation (e.g., Adie’s tonic pupil).

75. False. Lens diameter is important in fitting both soft and rigid gas-permeable (RGP) contact lenses. However, diameter is one of the main factors affecting RGP-lens centration. Typically, RGP-lens diameter should be approximately 3 mm smaller than the visible iris diameter.

76. a. For soft lenses, the spherical equivalent is used, after correction for vertex distance. The spherical equivalent of this correction is \(-5.00 \text{D}\). Correcting for the vertex distance of 15 mm, the appropriate contact lens has a power of \(-5.00 \text{D}\).

77. b. Extended-wear lenses are currently available as hydrogel soft lenses. Increasing the gas transmissibility (either by increasing the DK or by decreasing lens thickness) will increase oxygen transmission. Minus carriers and ballasting play no role in oxygen transmission. Extended-wear contact lenses should have a gas transmissibility of at least 90 units to avoid corneal hypoxia.

78. False. A +90D aspheric lens produces a real inverted image anterior to the lens, similar to the condensing lens of indirect ophthalmoscopy. Both the Hruby lens (\(-55 \text{D}, \text{ plano-concave}\)) and the Goldmann fundus contact lens (\(-64 \text{D}, \text{ plano-concave}\)) essentially nullify the refractive power of the cornea and create an upright, virtual image of the fundus approximately 2 cm posterior to the lens.

79. a. The Goldmann applanation tonometer uses a split-field plastic prism to indicate when a corneal area 3.06 mm in diameter has been flattened. The force in dynes required to flatten this area, multiplied by 10, is equal to the intraocular pressure (IOP) in mm Hg.

80. True. Measurement of intraocular pressure (IOP) with applanation can be inaccurate if significant
corneal astigmatism is present. An elliptical rather than a circular area will be applanated. Splitting the ellipse at a 43-degree angle to the major axis gives the best results. Alternately, taking the mean of readings at 90 degrees and 180 degrees can reduce the error. With-the-rule cylinder causes underestimation (1 mm Hg/4D) and against-the-rule cylinder causes overestimation of IOP.

81. a. The direct ophthalmoscope operates on the optical principle that light emanating from the retina of an emmetropic patient will be focused on the retina of an emmetropic observer. Lenses are used between the patient and observer to correct for nonemmetropic situations. In emmetropia, magnification obtained with the direct ophthalmoscope is approximately $15 \times$ because the patient’s eye acts as a simple plus magnifier of power (P) 60D (angular magnification (M) = P/4 or 60D/4 = $15 \times$). In conditions of ametropia, the interposition of lenses creates a Galilean telescope. Therefore, if the patient is myopic, the retina appears more magnified, and if the patient is hyperopic or aphakic, it appears less magnified. Remember that in afocal systems, magnification must be considered in angular, rather than linear, terms.

82. d. A 30D lens offers less magnification ($2 \times$) but a larger field of view than a 20D lens. Although axial magnification is the square of linear magnification, it must be reduced by a factor of 4 for indirect ophthalmoscopy because the patient’s pupil is expanded by a factor of 4 for visualization by the examiner.

83. c. Conventional kerometry measures the curvature of only the central 3 mm of the cornea.

84. d.

We can use the formula $U + P = V$, where $U =$ vergence of light from the object, $P =$ power of the lens, and $V =$ vergence of light forming the image. The power of the lens to focus the light on the screen can thus be calculated. We can calculate $U$ and $V$ respectively, because we know the distance of the object to the lens ($u = 0.25$ m) and the distance from the lens to the screen ($v = 1$ m), using the formulas: $U = -1/(0.25) = -4D$ and $V = 1/1 = +1D$.

Therefore, $P = V - U = 1 - (-4) = +5.00$.

Note that $U$ is negative because it is a real object; therefore, light rays must be diverging from it, as shown in the figure above. All real objects are composed of divergent (minus) light. All real images are composed of convergent (plus) light.

85. a.

Again, we can use the formula $U + P = V$ (see question 84). Because we know the power of the lens and the distance of the image from the lens, we can solve for $U$ and therefore $u$, the distance from object to lens.

$P = +24D$ and $V = 1/v = 1/0.05 = +20D$ (real image).

Thus, $U = V - P = 20 - 24 = -4D$.
Finally, $U = 1/u$ or $u = 1/U = 1/(-4D) = -0.25$ m or 25 cm to the left of the lens, as shown in the figure above.

86. b.

This question is more difficult. If the object and image are 50 cm apart, then $u$ (object–lens distance) + $v$ (lens–image distance) is 0.5 m. However, since the object is real, $u$ must be negative. Hence $-u + v = 0.5$ m, or:

$$-1/U + 1/V = 0.5.$$  \hspace{1cm} (3.1)

Multiplying through by $-U$ gives:

$$+1 - U/V = -U/2.$$  \hspace{1cm} (3.2)

Multiplying through by $V$ gives:

$$V - U = -UV/2.$$  \hspace{1cm} (3.3)

Given that we know the power is $+8.00D$, and $U + P = V$, we know that $U + 8 = V$.

Substituting in $U + 8$ for $V$:

$$-U + 8 = -U(U + 8)/2.$$  \hspace{1cm} (3.4)
Reorganizing, this leads to:

\[ U^2 + 8 U + 16 = 0. \]  
(3.5)

which is the equivalent to \((U + 4)^2 = 0\) or \(U = -4\), so \(u = 1/(4D) = -0.25\ m, or 25\ cm\) to the left of the lens. Because \(V = 8 + U = 4\), \(V = 1/v = 25\ cm\) to the right of the lens, as shown in the figure at the beginning of this answer.

87. a. The far point is defined as that point in space from which light will focus sharply on the retina, with accommodation fully relaxed. For the emmetropic eye, this point is at optical infinity. The near point is defined as that point in space from which light will focus sharply on the retina, with accommodation fully active. An eye with a \(+4.00\ D\) “error lens” is \(+4.00\ D\) too strong and is corrected (neutralized) with a \(-4.00\ D\) lens. Myopic eyes can be thought of as having positive error lenses. The far point of a myopic eye (with a positive error lens) is in front of the eye, at a distance in meters equal to the reciprocal of the power of the “error lens” in diopters. In this case, this is \(1/4\) of a meter, or \(25\ cm\) in front of the eye.

88. b. The far point is defined as that point in space from which light will focus sharply on the retina, with accommodation fully relaxed. For the emmetropic eye, this point is at optical infinity. The near point is defined as that point in space from which light will focus sharply on the retina, with accommodation fully active. An eye with a \(+4.00\ D\) “error lens” is \(+4.00\ D\) too strong and is corrected (neutralized) with a \(-4.00\ D\) lens. Myopic eyes can be thought of as having positive error lenses. The far point of a myopic eye (with a positive error lens) is in front of the eye, at a distance in meters equal to the reciprocal of the power of the “error lens” in diopters. In this case, this is \(1/4\) of a meter, or \(25\ cm\) in front of the eye.

89. d. Because this eye requires a \(+4.00\ D\) lens for correction, it is hyperopic. A hyperopic eye is too weak. Thus, for light to fall on the retina of a hyperopic eye, it must be converging. The far point of the hyperopic eye is where the convergent light would intersect (as a virtual object), if it were not bent onto the retina by the refractive power of the hyperopic eye. The far point of any ametropic eye is simply the secondary focal point of the corrective lens or \(f_2 = 1/4D\), 25 cm behind the cornea.

90. b. Again, the secondary focal point of a corrective lens must coincide with the eye’s far point. This eye’s far point is 10 cm \((100/10D)\) behind the spectacle plane. Given a \(2\-cm\) vertex distance, the far point is 8 cm behind the cornea. \(+3.00\ D\) lens must be placed with its secondary focal point at this same point. Because its secondary focal length is 33 cm, it must be placed 25 cm in front of the cornea, as shown in the figure above.

91. c. The refracting power of a spherical interface separating two media of different indices of refraction is given by:

\[ P = (n_2/n_1)/r, \]

where \(P\) = power of the surface, \(n_1\) = refractive index of the first medium that the light travels in, \(n_2\) = refractive index of the second medium that the light travels in, and \(r\) = radius of curvature of surface in meters. \(n_2 = 3.00\), \(n_1 = 1.00\) \((air)\), and \(r = +0.5\ m\). The sign of “\(r\)” is assigned by convention: \(r\) is positive if the center of curvature of the surface is on the opposite side of the origin of the incident light. If the center of curvature is on the same side of the interface as the origin of light, then \(r\) is negative.

Thus,

\[ P = 3 - 1/0.5 = +4D. \]

Primary focal length = \(n_2/P = 25\ cm\).
Secondary focal length = \(n_2/4 = 75\ cm\).

Because the diameter of the ball is 100 cm (since its radius is 50 cm), the secondary focal point \((75\ cm\ inside)\) lies inside the ball. The secondary focal point is where light originating at infinity will come to focus.

92. a. Because \(P = (n_2/n_1)/r, P = (1.50 - 1.00)/+0.5 = +4D.\)
Primary focal length, \(f_1 = n_2/P = 1\ m.\)
Secondary focal length, \(f_2 = n_2/4 = 1.5\ m.\)
Because \(f_2 = 1.5\ m\) and the diameter of the crystal ball is \(1.0\ m\), light coming from infinity could not come to focus inside this crystal ball.
93. b. Because \( P = (n_2 - n_1)/r \), \( P = (1.50 - 1.00)/0.10 \text{ m} = +5 \text{D} \).

Primary focal length, \( f_1 = n_1/P = 1/5 \text{ m}, \) or 20 cm.

Secondary focal length, \( f_2 = 1.5/5 \text{ m}, \) or 30 cm.

Light from infinity would not come to focus inside this crystal ball because its diameter is 20 cm.

94. **False.** The radius of curvature of a refractive surface is defined as positive if the center of curvature of the surface is on the side of the surface opposite to the origin of incident (object) light.

95. **True.** The following is a simple method to determine the sign of the power of a spherical refractive surface:

If the center of curvature of the refracting surface is on the same side as the medium of higher index of refraction, the surface is positive, regardless of direction of light propagation.

96. c. Because \( P = (n_2 - n_1)/r \), \( P = (1.33 - 1.00)/0.33 \text{ m} = +1 \text{D} \).

We know that \( U + P = V \), but in this case we must use “reduced vergences” to account for the index of refraction of the object space (1.33) and the image space (1.00). So:

\[
U = n_i/u, \text{ and } V = n_2/v.
\]

\[
U = -(4/3)/(1/3) = -4 \text{D}.
\]

\[
V = U + P = -4 + 1 = -3 = n_2/v.
\]

\( n_2 = 1.00 \) (air), so \( v = 33 \text{ cm} \) inside the tank. The image is virtual (composed of divergent light) and upright (since magnification = \( U/V > 0 \)).

97. e.

The power of a plane sheet of glass is zero, because \( r \) is infinite. Because \( U + P = V \) and \( P = 0 \), \( U = V \). Because the index of refraction of the object space (\( n_{1 \text{D}} = 1.33 \)) is different from that of the image space (\( n_{\text{air}} = 1.00 \)), \( u \) does not equal \( v \). Thus,

\[
U = -n_i/u = (4/3)/(1-2/1) = -(8/3) \text{D}.
\]

\[
V = n_2/v = U = -(8/3) \text{D}.
\]

Because \( n_2 = 1.00 \), \( v = -3/8 \text{ m} = 37.5 \text{ cm} \). Because the light rays are diverging (\( V < 0 \)), the image must be virtual and inside the tank, as shown in the figure above.

98. b. In the schematic eye, the primary focal point is located approximately 17 mm anterior to the eye (\( f_1 = n_1/P = 1/60 \text{D} = 0.017 \text{ m}, \) or 17 mm).

The distance between the secondary focal point and the secondary nodal point is always equal to the primary focal length. The secondary focal point is located approximately 22.5 mm inside the eye (\( f_2 = n_2/P = 1.33/60 \text{D} = 0.0222 \text{ m}, \) or 22.2 mm). Thus, the nodal point must be approximately 5 mm inside the eye. Because the indices of refraction are different on either side of the cornea, the primary, and secondary focal lengths are not equal.

99. c. The total power of the lens is equal to the sum of the powers of the two refractive surfaces. For the front surface,

\[
P = (n_2 - n_1)/r_1 = (1.5 - 1.0)/0.10 = +5 \text{D}
\]

The power is positive because the center of curvature is on the same side as the medium with higher index of refraction. For the rear surface,

\[
P = (n_1 - n_2)/r_2 = (1.0 - 1.5)/(0.05) = +10 \text{D}
\]

The total power is the sum of these two powers, or +15D. An alternative method for this problem utilizes the formula for total power of two combined spherical refractive surfaces in medium \( n_1 \), with medium \( n_2 \) between them:

\[
P = (n_2 - n_1) \times (1/r_1 + 1/r_2).
\]

100. d. Intraocular lens (IOL) powers are labeled according to their power immersed in aqueous humor, which has an index of refraction of approximately 1.33. In this case, the power of the IOL would be +18D in aqueous. The power of the lens in air can be calculated by using a ratio of the difference in the indices of refraction:

\[
P_{\text{air}}/P_{\text{eye}} = (n_{\text{air}} - n_{\text{IOL}})/(n_{\text{IOL}} - n_{\text{eye}})
\]

Here, \( P_{\text{air}}/P_{\text{eye}} = (1.30 - 1.00)/(1.30 - 1.33) = 3 \).

Thus, \( P_{\text{air}} = 3 \times P_{\text{eye}}, \) so \( 3 \times 18 \text{D} = 54 \text{D} \).

Note that the lens’s effective power is greater when the difference in index of refraction between the lens and the surrounding medium is greater.

101. c. Image ray no. 1 leaves the lens parallel to the lens axis and therefore must have passed through the primary focal point (not the secondary focal point). Image ray no. 2 passes through the secondary focal point. Therefore, its object ray must have been traveling parallel to the lens axis. The lens shown is a converging lens with its secondary focal point to the right of the lens and, therefore, it must be positive in power. Image ray no. 3 is traveling through the nodal point, and it therefore travels through the lens undeviated.

102. **False.** See answer 19. For both plus and minus lenses, the absolute value of the powers may be described as follows:

\[
|P(\text{back})| > |P(\text{true})| > |P(\text{front})|.
\]
In an afocal telescope, the lenses must be placed such that the secondary focal point of the objective lens coincides with the primary focal point of the eyepiece. If both lenses are positive, as in an astronomical telescope, the distance between the lenses is the sum of the focal lengths. If the eyepiece is negative, as in a Galilean telescope, the distance between the lenses is the difference of the focal lengths. In this Galilean telescope, \( f_{\text{objective}} = 25 \, \text{cm} \), and \( f_{\text{eyepiece}} = 10 \, \text{cm} \). The difference between the two, 15 cm, is the length of the device, as shown in the figure above.

### Answers

**103. b.**

\[
\text{Loupe length} = f_2 (\text{objective}) - f_1 (\text{eyepiece})
\]

\[
\begin{align*}
&| & 10 \, \text{cm} \\
&| & f_1 (\text{eyepiece}) \\
&| & 25 \, \text{cm} \\
&| & f_2 (\text{objective})
\end{align*}
\]

In an afocal telescope, the lenses must be placed such that the secondary focal point of the objective lens coincides with the primary focal point of the eyepiece. If both lenses are positive, as in an astronomical telescope, the distance between the lenses is the sum of the focal lengths. If the eyepiece is negative, as in a Galilean telescope, the distance between the lenses is the difference of the focal lengths. In this Galilean telescope, \( f_{\text{objective}} = 25 \, \text{cm} \), and \( f_{\text{eyepiece}} = 10 \, \text{cm} \). The difference between the two, 15 cm, is the length of the device, as shown in the figure above.

**104. e.** The magnification of an afocal telescope (M) equals \(- (\text{eyepiece power})/\text{objective power}\).

In this case,
\[
M = -(-10 \, \text{D}) / 4 = +2.5 \times
\]

The image formed by a Galilean telescope is upright (M > 0). No accommodation is required for viewing this image because the object is at infinity and would require no accommodation to image without the telescope. The image is virtual because it is composed of parallel light.

**105. e.**

\[
\text{Loupe length} = f_2 (\text{objective}) + f_1 (\text{eyepiece})
\]

\[
\begin{align*}
&| & 25 \, \text{cm} \\
&| & f_1 (\text{eyepiece}) \\
&| & 10 \, \text{cm} \\
&| & f_2 (\text{objective})
\end{align*}
\]

In an astronomical telescope, both the eyepiece and objective are plus lenses. The distance between the lenses is the sum of the two focal lengths, or 35 cm, as shown in the figure above.

(Compare the size of this telescope with the telescope in question 103.)

**106. b.** The magnification (M) of this astronomical telescope also equals \(- (\text{eyepiece power})/\text{objective power}\).

In this case,
\[
M = -(-10) / 4 = -2.5 \times
\]

The image formed by any astronomical telescope is inverted (M < 0). Accommodation is needed unless the objective is at infinity or an add is constructed into the telescope. The image is virtual because it is composed of divergent light (because the object is made of divergent rays).

**107. a.** Surgical loupes are generally Galilean telescopes.

This afocal Galilean telescopic system must be 5 cm long (the difference of the focal lengths of the two lenses, 10 cm and 5 cm). The add of +5.00D is used to decrease the accommodative demand. An object 25 cm from the eye (20 cm from the loupes) will have an object vergence of -5D. The +5D add converts new object vergence for the objective to 0D (\( U + P = V \), or \( -5 + 5 = 0 \)). Because the objective lens power is +10D, the image vergence \( V \) from the objective is +10D (\( U + P = V \), or \( 0 + 10 = 10 \)). The light will come to focus 10 cm from the objective lens. Because the eyepiece is 5 cm from the objective, this objective image is 5 cm from the eyepiece. Because the light is convergent, the “object” for the eyepiece has a vergence of +20D (1/0.05 m). The light leaves the telescope with zero vergence. Therefore, no accommodation is required for this viewing distance (20 cm from the add).

**108. e.** For an object at 10 cm from the loupes (15 cm from the eye).

Initial \( U = -10 \, \text{D} \).

For the add:
\[
U + P = V = -10 \, \text{D} + (+5 \, \text{D}) = -5 \, \text{D}.
\]

Since the add and objective are superimposed:
\( U \) for the objective = \( V \) from the add = -5D.

So for the objective,
\[
U + P = V = -5 \, \text{D} + 10 \, \text{D} = +5 \, \text{D}.
\]

The objective will create an image approximately 20 cm (100/5D) from itself. The eye piece is 5 cm from the objective, so the objective image (the eyepiece object) is 15 cm from the eye. The objective image (the eyepiece object) is virtual because it is composed of divergent light (because the object is made of divergent rays). Because the eyepiece is at a negligible distance from the eye, this is the accommodation needed (+13.3D).
3. Optics and Refraction

109. c. The angular magnification (M) of loupes with no add equals \((-\text{eyepiece power})/\text{objective power}\).

If there is an add to relieve accommodation, this contributes angular magnification as a simple magnifier. The total angular magnification is the product of these two magnification factors, or:

\[
\left(\frac{P_{\text{add}}}{4}\right) \times \left(-\frac{P_{\text{eyepiece}}}{P_{\text{objective}}}\right)
\]

In this case,

\[
\left(\frac{+5}{4}\right) \times \left(-\frac{-20}{10}\right) = \left(1 + 1.25\right) \times (-2) = 2.5 \times .
\]

110. b. Image size change can be calculated by considering the eye to have an “error lens” located at the nodal point 5 mm behind the corneal surface. The corrective lens is considered the objective of a Galilean telescope, and the error lens is the eyepiece. Proper distance correction requires that the secondary focal point of the corrective lens be located at the eye’s far point, which is the primary focal point of the “error lens.”

As the figure above shows, the secondary focal point of the +12.5D contact lens is located at

\[
f = 1/12.5D = 0.08 \text{ m}
\]

or 8 cm behind the corneal surface. Because the primary focal point of the “error lens” is in the same location, while the error lens is located 0.005 m behind the corneal surface, the power is given by:

\[
P_{\text{error}} = -1/(0.08 \text{ m} \times 0.005 \text{ m}) = -13.3D.
\]

(The power of the corrective lens and the error lens must be opposite.) If the error lens is thought of as the eyepiece and the corrective lens as the objective, then magnification of this Galilean telescope is given by:

\[
M = -P_{\text{eyepiece}}/P_{\text{objective}}
\]

\[
= -(-13.3D)/12.5D = 1.07 \times .
\]

That is, the image is 1.07 times the size of the object, giving 7% magnification. 

111. c. With a vertex distance of 20 mm, the corrective lens is located 25 mm in front of the “error lens.” The focal length of the corrective lens must be:

\[
f = f_{\text{error lens}} + 0.025 \text{ m} = (1/13.3D)
\]

\[
+ 0.025 \text{ m} = 0.10 \text{ m}
\]

Therefore, \(P = 1/f = 10.0D\).

112. b. \(M = -(-13.3D)/10.0D = 1.33 = 33\%\) magnification.

(See questions 108 and 109.)

113. c.

In a myope, the far point is in front of the eye, and the positions of the secondary focal point of the corrective lens and the primary focal point of the error lens must be located in the same place. Because a \(-10.0D\) contact lens located 5 mm in front of the nodal point gives proper correction, the focal length of the error lens is given by:

\[
f_{\text{error}} = -f_{\text{correction}} + 0.005 \text{ m} = -1/f_{\text{correction}} + 0.005 \text{ m}
\]

\[
f_{\text{error}} = -(1/10.0D) + 0.005 \text{ m} = 0.105 \text{ m}.
\]

The power is:

\[
P_{\text{error}} = 1/0.105 \text{ m} = 9.5D.
\]

The magnification is:

\[
M = -P_{\text{eyepiece}}/P_{\text{objective}} = -9.5D/10.0D = 0.95.
\]

This represents 5% minification, as shown in the figure above. Because the power of the eyepiece is positive and the objective negative, this is like looking through a Galilean telescope backward, resulting in minification.

114. d. With a vertex distance of 20 mm, the distance between the spectacle lens and the “error lens” is 25 mm. The focal length of the corrective lens is therefore:

\[
f_{\text{correction}} = f_{\text{error}} - 0.025 \text{ m} = 0.105 \text{ m} - 0.025 \text{ m} = 0.08 \text{ m}.
\]

The power is:

\[
P = 1/0.08 \text{ m} = -12.5D.
\]

115. c. \(M = -P_{\text{eyepiece}}/P_{\text{objective}} = -9.5D/12.5D = +0.76 = 24\%\) minification

116. d. The formula for the power of a mirror is

\[
P = -(2/r), \text{ where } r \text{ equals the radius of curvature of the mirror in meters. The vergence formula for mirrors is the same as that for lenses:}
\]

\[
U + V = P
\]
A plane mirror has an infinite radius of curvature and, therefore, no power. It does not change the vergence of light but changes only its direction. Because the object is 1 m in front of the mirror, the object vergence at the mirror is:

\[ U = -1/1 \text{ m} = -1 \text{D} \]
\[ V = U + P = -1D + 0D = -1D \]
\[ v = 1/V = -1/1D = -1 \text{ m} . \]

The image therefore is located 1 m behind the mirror and is virtual. The magnification for a mirror is the same as for a lens, \( M = U/V \):

\[ M = U/V = (-1D)/(-1D) = 1 \times . \]

The image and object are the same size, and because the magnification is positive, the image is upright. Because the angle of incidence and the angle of reflection are equal, a mirror half the height of the man will allow him to see his toes, regardless of where he stands.

117. c. As shown in the figure below, the reflective power of the mirror is:

\[ P = -2/r = -2/0.20 \text{ m} = -10 \text{D}. \]

(Convex mirror—adds divergence 
\( r > 0 \)

Radius of curvature 
\( r \)

(By definition, a mirror convex toward light has a positive radius of curvature, and a mirror concave toward light has a negative radius of curvature. Therefore, any convex mirror has negative power; any concave mirror has positive power.)

Concave mirror—adds convergence 
\( r < 0 \)

As shown in the figure above, the object is located 50 cm in front of the mirror, so:

\[ U = -1/0.5 \text{ m} = -2 \text{D} \]
\[ V = U + P = -2D - 10D = -12D \]

and the image is located:
\[ v = 1/V = 1/(-12D) = -0.083 \text{ m}, \text{ or } 8.3 \text{ cm behind the mirror.} \]

118. True. See answer 117. A convex mirror always adds divergence to light. The image light in this question is composed of divergent light. Real images, by definition, are made up of convergent light. Think of convex mirrors as producing “virmin” – virtual, minified images.

119. False. Magnification by a mirror is given by the same formula as for a lens, \( M = U/V \). Here, \( M = -2/-12 = 1/6 \). The image is 1/6 the size of the object (minified by a factor of 6).

120. True. If the magnification value > 0, the image is upright (this is true for lenses, as well). Convex mirrors produce minified images.

121. d. The refractive power of the anterior surface of the cornea is given by:
\[ P_{\text{front}} = (n_2 - n_1)/r = (1.38 - 1)/(0.0077 \text{ m}) = +49.4 \text{D}. \]

The refractive power of the posterior surface of the cornea is given by:
\[ P_{\text{back}} = (1.33 - 1.38)/0.0076 \text{ m} = -6.6 \text{D}. \]

The total refractive power of the cornea, therefore, is:
\[ P_{\text{front}} + P_{\text{back}} = 49.4 \text{D} - 6.6 \text{D} = 42.8 \text{D}. \]

The reflecting power of the front surface of the cornea is given by:
\[ P = -2/r = -2/0.0077 \text{ mm} = -260 \text{D}. \]

A single refractive surface at an interface between air and a medium with an index of refraction of 1.3375, having a radius of curvature of 7.9 mm, has refractive power of:
\[ P = (1.3375 - 1)/0.0079 \text{ m} = +42.7 \text{D}. \]

which is the same refractive power as the cornea. This is where the formula for conversion from radius of curvature to power is derived:
\[ P = 0.3375/r, \text{ or } r = 0.3375/P. \]

122. d. Consider the implant as the object. The optic diameter measured is a measurement of what we see, namely the image of the optic, not the optic itself. The optic is in aqueous (\( n = 1.33 \)), so its object vergence (\( U \)) is:
\[ U = -1.33/0.003 \text{ m} = -443.3 \text{D} \]
\[ P = 42.8 \text{D} \]
\[ U + P = V = -443.3D + 42.8D = -400.5 \text{D} \]
\[ M = v/u = U/V = -443.3D/-400.5D = 1.11 \times . \]

The image of the optic is, therefore, 1.11 times larger than the iris, so if \( o \) is the object height and \( i \) is the image height:
\[ M = i/o = 1.11 \times \]
\[ i = 1.11 \times o = 1.11 \times 6.0 \text{ mm} \]

Rounded, this equals 6.7 mm.
3: Optics and Refraction

123. b. The reflecting power of the corneal surface is given by:

\[ P = -\frac{2}{r} = -2/0.0077 \text{ m} = -260 \text{D}. \]

For a penlight held 40 cm (0.40 m) in front of this mirror:

\[ U = 1/0.4 \text{ m} = -2.5 \text{D} \]
\[ U + P = V = -262.5 \text{D} \]
\[ v = 1/V = 1/(-262.5 \text{D}) = -0.0038 \text{ m} \]

or 3.8 mm inside the eye. Light rays reflected off the cornea are divergent, so the virtual image appears to be inside the eye.

124. d. By moving his/her head away from the condensing lens, a smaller area of the patient’s pupil requires illumination, leaving more available for image transmission.

125. b. Image brightness, clarity, and detail are enhanced by moving closer. This requires a larger pupil, however (see question 124).

126. e. Higher-power condensing lenses have reflexes that require greater tilting of the lens than lower-power lenses to shift them out of the viewing axis.

127. c. With the retinoscope in the plano-mirror position (handle up on the Copeland model, handle down on the Welch-Allyn), the streak image is behind the patient or at infinity. “With” motion indicates the patient’s eye is a minus lens system, neutralized with plus lenses. “Against” motion implies that the patient’s eye is a plus system of sufficient power to cause the new streak image (acted on by the eye) to be located between the patient and the observer.

128. False. Knapp’s rule holds only for axial anisometropia. Most anisometropia is mixed, that is, both refractive and axial.

129. d. Corneal thickness increases during sleep, but there is also considerable variation in the cornea during waking hours.

130. c. Gradual steepening of the cornea throughout the waking hours leads to progressive myopia. Some patients require myopic correction as the day progress.

131. d. Nonsteroidal antiinflammatory drugs (NSAIDs) have been shown to increase the incidence of overcorrection.

132. a. Surgical reduction of myopia would be expected to reduce the minimification caused by myopic refractive correction.

133. e. Older methods were fraught with complications, including inadvertent perforation caused by the manual keratomes. Automated laser keratomileusis (ALK) and laser-assisted in situ keratomileusis (LASIK) have almost eliminated these complications and enhanced safety and predictability. The use of anterior stromal flaps rather than removable discs has also moved the science forward.

134. c. Photoablation specifically does not cause surrounding tissue destruction—hence corneal photoablation does not cause surrounding corneal opacification.

135. d. Reduced vergence is vergence multiplied by the refractive index of the medium the light is traveling in. Therefore,

\[ \text{vergence} \times n_{\text{glass}} = +1.00 \text{D} \times 1.5 = +1.50 \text{D}. \]

136. c. Applanation over a corneal scar will yield artificially high intraocular pressure (IOP) measurements. All of the other answers listed will yield artificially low IOP measurements. Increased corneal thickness may also yield an artificially high IOP measurement during applanation.

137. c. High-index lenses, have, by definition, a high index of refraction (1.6 to 1.8), which is greater than that of polymethylmethacrylate (PMMA) (n = 1.49). Air has the lowest index of refraction (1.00). In general, it is felt that the cornea (n = 1.37) has a slightly higher index of refraction than vitreous (n = 1.34).

138. c. Blue light has shorter wavelengths that are scattered more toward your eyes.

139. e. All of the above definitions are correct.

Suggested Readings


1. A Russell body is:
   a. an accumulation of granules within eosinophils.
   b. proteinaceous debris within giant cells.
   c. accumulated lysosomal degradation products within polymorphonuclear leukocytes (PMNs).
   d. proteinaceous antigen debris within macrophages.
   e. concentrated immunoglobulin within plasma cells.

2. T or F The hallmark of granulomatous inflammation is the presence of giant cells.

3. Which one of the following statements about fungal keratitis is false?
   a. Generally, these infections are initiated by trauma involving plant or animal matter.
   b. Topical corticosteroids generally enhance organism replication and penetration.
   c. The actual etiologic agent is dependent on geographic locale.
   d. Most of the important corneal pathogens are molds.
   e. The cornea is an effective barrier to intraocular fungal penetration.

4. Which one of the following statements about chlamydial ocular infections is true?
   a. The cornea is typically spared in chlamydial conjunctivitis.
   b. Disease manifestations are dictated by organism serotype.
   c. The only difference between inclusion conjunctivitis and trachoma is the presence of subconjunctival scarring.
   d. Wright stain is the preferred method of demonstrating the pathognomonic findings.
   e. Typically, the inclusion bodies of chlamydial disease are intranuclear.

5. Which one of the following statements about viral ocular disease is false?
   a. The intranuclear inclusions of molluscum contagiosum may cause cellular rupture, leading to an adjacent inflammatory reaction.
   b. The host response to herpetic infection is non-granulomatous in primary cases and may be granulomatous in reactivation.
   c. Subacute sclerosing panencephalitis, a chronic central nervous system measles infection, may be associated with retinitis.
   d. Cytomegalovirus (CMV) causes a retinochoroiditis that is associated with both intracytoplasmic and intranuclear inclusion bodies.
   e. Interferon may be an important host defense mechanism in many viral ocular infections.

6. Which one of the following statements about intraocular infections is false?
   a. The pork tapeworm *Taenia solium* may cause an intense granulomatous reaction when an intraocular cyst dies.
   b. *Toxoplasma gondii* induces a retinochoroiditis characterized by the eosinophilic abscess.
c. The cat is the definitive host for *Toxoplasma gondii*.
d. *Toxoplasma* cyst forms are extracellular, and reactivation requires cyst conversion to a trophozoite.
e. *Cryptococcus* may cause a meningoencephalitis with optic neuritis and retinitis.

7. T or F Herpes simplex keratitis is characterized by multinucleated giant cell infiltration adjacent to Descemet’s membrane.

8. Which of the following may present as ocular inflammation (or pseudoinflammation)?
   1. uveal melanoma.
   2. non-Hodgkin’s lymphoma.
   3. retinoblastoma.
   4. melanocytoma.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

9. Which one of the following statements about lens-induced uveitis and glaucoma is false?
   a. The term phacoantigenic is preferable to phacoanaphylactic.
   b. Phacoantigenic glaucoma is probably on the same spectrum as phacolytic glaucoma, simply being more severe.
   c. *Propionibacterium acnes* may be an important contributor to lens-induced uveitis, particularly in pseudophakic eyes.
   d. The classic pattern of inflammation in phacoantigenic glaucoma is the zonal granuloma centered about a site of injury to the lens.
   e. The stimulus for phacoantigenic uveitis appears to be lens cortical protein.

10. All of the following are conditions that may be associated with granulomatous ocular inflammation except:
   a. sarcoidosis.
   b. rheumatoid arthritis.
   c. Behçet’s disease.
   e. syphilis.

11. All of the following are conditions that may be associated with nongranulomatous ocular inflammation except:
   a. choroidal melanoma.
   b. juvenile rheumatoid arthritis.
   c. trauma.
   d. Reiter’s syndrome.
   e. cryptococcal endophthalmitis.

12. T or F The distinction between Vogt-Koyanagi-Harada syndrome (VKH) and sympathetic ophthalmia (SO) must be made clinically because there are no distinguishing histologic features.

13. Which one of the following statements about healing in specific ocular tissues is true?
   a. Epithelial migration and ingrowth generally leads to complete replacement of structures lost in full-thickness skin wounds by the sixth week after injury.
   b. Central corneal healing of a perforated wound is accomplished only after granulation tissue is extruded from the wound, approximately 4 to 6 weeks after injury.
   c. The most important components in limbal-wound healing are surrounding scleral vessels and resident cells.
   d. The crucial step in healing central and peripheral corneal wounds is metaplasia or migration of endothelial cells to seal the internal defect.
   e. Because of its marked vascularity and rich innervation, the iris generally shows exuberant and rapid wound-healing processes.

14. Which one of the following statements about retinal response to injury is true?
   a. Mechanical injury leads to a typical fibrovascular healing response.
   b. Cellular proliferation in retinal healing takes place mainly within the photoreceptor and inner plexiform layers.
   c. Retinal pigment epithelium (RPE) hyperplasia rarely plays an important role in retinal healing.
   d. Retinal fibrosis is responsible for the healing strength of a laser or cryotherapy mark.
   e. A rupture of Bruch’s membrane during injury will lead to choroidal participation in the human response with subsequent fibrosis.
15. T or F The histopathologic features of epithelial downgrowth are most consistent with abnormally invasive corneal epithelium.

c. inner plexiform layer.
d. nerve fiber layer.
e. subinternal limiting membrane space.

16. Postoperative sympathetic ophthalmia is most likely to develop following:
   a. uncomplicated intracapsular cataract surgery.
   b. uncomplicated extracapsular cataract surgery.
   c. penetrating keratoplasty in a patient with herpes simplex keratitis and a history of uveitis.
   d. scleral buckling procedure using scleral implants.
   e. filtration procedures on end-stage glaucomatous eyes.

17. Which one of the following statements about the pathology of penetrating keratoplasty is false?
   a. Graft failure implies an immunologic rejection response to the donor cornea.
   b. A graft rejection may be centered on the epithelium, stroma, or endothelium.
   c. Epithelial rejection is generally self-limited and resembles epidemic keratoconjunctivitis (EKC).
   d. Stromal rejection most typically presents 2 to 4 weeks postoperatively.
   e. The pathognomonic sign of active corneal graft rejection is the Khoudadoust line, which consists of inflammatory precipitates in linear form on the endothelium.

18. Select the correct pairing of eponym and pathologic findings:
   a. Elschng’s pearls—retention of lens fibers in the equatorial area of the capsular fornix.
   b. Elschng’s pearls—clumps of iris pigment epithelial cells on the anterior lens capsule following trauma.
   c. Vossius’ ring—retention of lens fibers in the equatorial area of the capsular fornix.
   d. Vossius’ ring—clumps of iris pigment epithelial cells on the anterior lens capsule following trauma.
   e. Soemmerring’s ring—clumps of iris pigment epithelial cells on the anterior lens capsule following blunt trauma.

19. The earliest accumulations of cystoid macular edema (CME) fluid occur in the:
   a. subretinal space.
   b. outer plexiform layer.

20. Which one of the following statements about intraocular hemorrhage is true?
   a. Corneal blood staining is generally the result of pressure-induced displacement of intact red blood cells (RBCs) through a damaged Descemet’s membrane.
   b. In hemosiderosis bulbi, hemoglobin accumulation leads to epithelial damage (anterior subcapsular cataract, glaucoma, and retinal degeneration).
   c. Ghost cells may be identified by phase contrast microscopy or by stained preparations documenting the presence of Heinz’s bodies.
   d. Synchysis scintillans is a condition in which hemoglobin breakdown products crystallize in the dependent portion of the vitreous.
   e. An ochre membrane results from vitreous hemorrhage coating the posterior lens capsule.

21. T or F A secluded pupil (seclusio pupillae) implies a complete membranous covering of the pupil caused by inflammation.

22. Which of the following features is/are considered necessary for the diagnosis of phthisis bulbi?
   1. abnormally small (shrinkage).
   2. disorganization.
   3. atrophy.
   4. calcification.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

23. Because of its variation in thickness, scleral rupture after blunt ocular trauma is prone to occur at all of the following locations except:
   a. at the equator of the globe.
   b. at the sclera immediately posterior to the lateral rectus insertion.
   c. in an arc at the limbus opposite the site of impact.
   d. at the optic nerve head.
   e. at the sclera immediately posterior to superior rectus insertion.
24. Which of the following statements about the phakomatous choristoma is/are true?
   1. Phakomatous choristoma may be associated with ocular colobomata.
   2. Acquired forms of phakomatous choristoma have been reported.
   3. There is frequently associated hyperpigmentation in phakomatous choristoma.
   4. The most frequent site of involvement is the eyelid.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

25. All of the following are histopathologic manifestations of angle recession except:
   a. a cleavage plane between longitudinal ciliary body fibers and sclera.
   b. atrophy of the ciliary muscle fibers.
   c. a change in the ciliary body shape from triangular to fusiform.
   d. a posterolateral displacement of ciliary processes.
   e. an epitheliallike membrane covering the trabecular meshwork and angle structures (in some cases).

26. Which one of the following statements about retinal dialysis is false?
   a. Unlike typical retinal tears, the posterior vitreous is frequently attached in the setting of retinal dialysis.
   b. Most nontraumatic dialyses are superotemporal.
   c. Trauma is the most common cause of suprachoroidal dialyses.
   d. Retinal detachment caused by retinal dialysis is typically rapidly progressive.
   e. Many patients with nontraumatic retinal dialysis will have a family history of retinal detachment.

27. A 44-year-old man is hammering metal on metal and suddenly experiences pain and loss of vision in his right eye. He is examined and is noted to have an intravitreal foreign body, which has passed through the cornea and central lens. A correct description of his injury would be:
   1. double perforating injury.
   2. corneal and lenticular perforating injury.
   3. double corneal and lenticular penetrating injury.
   4. ocular penetrating injury.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

28. Which one of the following statements about intraocular contamination is true?
   a. Siderosis bulbi is generally caused by damage to ocular epithelial tissues by ferric (Fe$^{3+}$) iron.
   b. Copper foreign bodies with >85% elemental copper generally lead to chalcosis, characterized by copper deposits in basement membranes.
   c. Glass, platinum, and porcelain are similar in their relative inert status as intraocular foreign bodies.
   d. Sterile vegetable matter will generally elicit no intraocular inflammatory response.
   e. Lead is an inert intraocular foreign body.

29. Which one of the following alkaline solutions constitutes the greatest threat for intraocular chemical injury?
   a. hydrochloric acid.
   b. ammonium hydroxide (ammonia).
   c. calcium hydroxide (lime).
   d. boric acid.
   e. petroleum.

30. Which of the following associations regarding radiation and ocular side effects is incorrect?
   a. microwave radiation—cataract.
   b. infrared radiation—true lens exfoliation (glassblower’s cataract).
   c. ultraviolet (UV) radiation—corneal epithelial damage.
   d. ionizing radiation—retinopathy similar to diabetic retinopathy.
   e. ionizing radiation—conjunctival ulceration and atrophy.

31. T or F A hamartoma consists of normal tissue in an abnormal location, whereas a choristoma consists of abnormal tissue in a normal location.

32. T or F An in utero insult at the beginning of the first trimester is more likely to lead to
Questions

33. Which of the following is/are variations of the hyaloidolenticular vascular system?

- Bergmeister’s papilla.
- persistent hyaloid artery.
- Mittendorf’s dot.
- persistent pupillary membrane.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

34. Which one of the following statements about common congenital adnexal conditions is true?

a. Epidermoid and dermoid cysts frequently arise in continuity with bony suture lines and underlying orbital periosteum.
b. Eyelid hemangiomas are usually of the capillary type and have only cosmetic implications.
c. Conjunctival dermoids are hamartomatous lesions that may arise as part of Goldenhar’s syndrome.
d. The primary difference between conjunctival dermoid tumors and dermoid cysts of the eyelids and orbit is the presence of pilosebaceous units in the latter.
e. Dermolipomas of the conjunctiva are related to dermoids and are generally easily surgically removed.

35. T or F The basic feature of the spectrum constituting Peters’ anomaly is central absence of Descemet’s membrane with a cloudy cornea (leukoma).

36. Which of the following conditions may be associated with congenital ectopia lentis?

- Marfan’s syndrome.
- Ehlers-Danlos syndrome.
- congenital glaucoma.
- Peters’ anomaly.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.

37. Which of the following phakomatoses has/have an autosomal recessive inheritance pattern?

- neurofibromatosis.
- tuberous sclerosis.
- Sturge-Weber syndrome.
- ataxia-telangiectasia.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

38. Which of the following disorders is/are commonly associated with congenital or juvenile glaucoma?

- von Hippel-Lindau syndrome.
- Sturge-Weber syndrome.
- ataxia-telangiectasia.
- neurofibromatosis.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

39. Which of the following disorders commonly has/have posterior segment findings?

- von Hippel-Lindau syndrome.
- Sturge-Weber syndrome.
- ataxia-telangiectasia.
- neurofibromatosis.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

40. Which one of the following statements about persistent fetal vasculature (PFV) is/are true?

a. The size of the eye is generally normal.
b. The occurrence of angle-closure glaucoma early in life is common.
c. The retina generally remains attached despite central fibrovascular proliferation.
d. Visual prognosis for the eye is often excellent.
e. Cataract is rarely seen in this condition.
41. Retinal dysplasia may be seen in association with:
   1. microphthalmos.
   2. trisomy 13.
   3. uveal colobomata.
   4. trisomy 18.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

42. Foveal hypoplasia has been associated with:
   1. aniridia.
   2. albinism.
   3. PAX6 gene mutations.
   4. mutations on chromosome 11.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

43. Congenital cataracts may be associated with which of the following chromosomal disorders?
   1. trisomy 21 (Down’s syndrome).
   2. trisomy 13 (Patau’s syndrome).
   3. Lowe’s syndrome.
   4. Turner’s syndrome.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

44. Fetal alcohol syndrome may be associated with ocular conditions including:
   1. retinal vascular tortuosity.
   2. optic nerve hypoplasia.
   3. esotropia.
   4. ptosis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

45. T or F Preseptal cellulitis may lead to orbital cellulitis in a normal eye.

46. T or F Dermal amyloid infiltration is generally a localized phenomenon, whereas conjunctival amyloid infiltration typically suggests systemic disease.

47. Approximately what percentage of patients with xanthelasma will have a systemic dyslipidemia?
   a. 20%.
   b. 25%.
   c. 33%.
   d. 50%.
   e. 75%.

48. Which one of the following statements about juvenile xanthogranuloma (JXG) is false?
   a. Lesions appear as yellow-orange skin nodules.
   b. Touton’s giant cells are a prominent feature.
   c. The color of these lesions arises from intercellular lipofuscin deposition.
   d. Tan or orange nodules on the iris are a common manifestation.
   e. Intraocular JXG may present as a spontaneous hyphema.

49. Which one of the following statements about cystic structures of the eyelids is true?
   a. An epidermoid cyst may be separated from a dermoid cyst at the slit lamp by transillumination characteristics.
   b. The prime feature differentiating an epidermoid cyst from a ductal cyst is the characteristic of the fluid filling the cyst.
   c. Histologically, the lining of a ductal cyst is always a cuboidal eccrine epithelium.
   d. The key histopathologic feature differentiating epidermoid from dermoid cysts is the nature of the cyst wall.
   e. Dermoid cysts are frequently associated with Goldenhar’s syndrome.

50. Which of the following definitions of epidermal pathologic findings is/are correct?
   1. hyperkeratosis–thickening of the prickle cell layer.
   2. parakeratosis–thickening of the keratin layer.
   3. acantholysis–loss of the normal keratin layer.
   4. dyskeratosis–intraepidermal keratinization.
Questions

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

51. **T or F** Collagen with a bluish hue on hematoxylin-eosin (H & E) staining is frequently a marker for previous ultraviolet radiation damage.

52. **T or F** The main difference between seborrheic and actinic keratoses is the presence of pigmentation in the former.

53. The lesion least likely to be clinically or pathologically mistaken for squamous cell carcinoma of the eyelid is:
   a. actinic keratosis.
   b. keratoacanthoma.
   c. seborrheic keratosis.
   d. pseudoepitheliomatous hyperplasia.
   e. basal cell carcinoma.

54. Which one of the following statements about basal cell carcinoma of the eyelid is false?
   a. It is the second most common malignant tumor of the eyelid.
   b. The lower lid is more frequently involved than the upper lid.
   c. Medial canthal tumors generally carry a poorer prognosis.
   d. Important histologic clues to the basal cell origin of this tumor include nests and cords of epidermal basilar cells with peripheral palisading and cracking artifact.
   e. Morpheaform (fibrosing) basal cell carcinoma is almost always undifferentiated.

55. Which one of the following statements about sebaceous neoplasms is false?
   a. Sebaceous neoplasms may arise from meibomian, Zeis, or caruncular sebaceous glands.
   b. A history of exposure to radiation is a risk factor.
   c. Benign sebaceous tumors may be associated with gastrointestinal malignancy.
   d. Histopathologic clues to the diagnosis include pagetoid spread of tumor cells, foamy tumor cells, and cystic necrosis of cell nests.
   e. Typically, hematoxylin-eosin (H & E) staining is sufficient to make the diagnosis.

56. Which one of the following statements about pigmented eye lesions is false?
   a. Unlike congenital nevi, acquired nevi rarely become malignant.
   b. Most eyelid nevi are acquired.
   c. Acquired nevi typically present at approximately 10 years of age.
   d. Nevi are best described by the location of cellular proliferation.
   e. The degree of elevation of a nevus is directly proportional to the extent of epithelial or junctional involvement.

57. Which ocular manifestation is most typical for untreated primary central nervous system lymphomas (PCNSLs)?
   a. nodular conjunctival infiltration.
   b. nodular iris infiltration.
   c. endogenous *Candida endophthalmitis*.
   d. vitreous infiltration.
   e. nodular retinal infiltration.

58. Which one of the following statements about ocular amyloidosis is false?
   a. Conjunctival foci most frequently herald primary localized disease.
   b. The deposits, regardless of location, are prone to spontaneous hemorrhage.
   c. The stain of choice is Congo red, which will bring out the deposits’ dichroism and birefringence.
   d. Dermal infiltration of amyloid frequently arises in cases of primary localized amyloidosis.
   e. Chronic conjunctival inflammation or irritation may lead to secondary amyloidosis.

59. Which one of the following statements about lymphoid lesions of the conjunctiva is false?
   a. They most frequently occur on the bulbar conjunctiva.
   b. They are typically salmon pink and soft.
   c. Patients with conjunctival lymphoma have a lower risk of developing systemic involvement than those with eyelid lesions.
   d. Russell’s bodies and Dutcher’s bodies frequently mark a lymphoid lesion as “reactive” (nonneoplastic).
   e. Approximately 10% of patients with systemic lymphoma will develop conjunctival infiltration.
60. Which one of the following statements about squamous cell lesions of the conjunctiva is false? 
   a. They are typically preceded in time by foci of carcinoma in situ (severe epithelial dysplasia).
   b. They typically arise in the interpalpebral fissure near the limbus.
   c. Specific variants of squamous cell carcinoma will infiltrate the globe.
   d. Hematogenous metastasis is not unusual with squamous cell carcinoma.
   e. Squamous cell lesions may simulate pagetoid sebaceous cell carcinoma.

61. T or F Conjunctival melanoma occurs more frequently in patients with ocular melanocytosis and oculodermal melanocytosis.

62. T or F The most frequent histopathologic diagnosis for a white patient in his thirties with conjunctival melanosis is junctional nevus.

63. Which of the following statements is/are true about primary acquired melanosis (PAM) of the conjunctiva? 
   1. The incidence is highest for middle-aged persons.
   2. Unremitting rapid growth is characteristic.
   3. The risk of progression to conjunctival melanoma is approximately 50% if the melanocytic proliferation appears atypical on histopathologic examination.
   4. PAM without atypia has been noted to progress to conjunctival melanoma.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

64. Which of the following statements is/are true about conjunctival melanoma? 
   1. Three fourths of cases arise de novo and one fourth arise from primary acquired melanosis (PAM) with atypia.
   2. The overall mortality rate is 25%.
   3. Like uveal melanoma, metastases frequently are first found in the liver.
   4. Prognosis is best assessed by melanocytic atypia and growth pattern.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

65. The malignant tumor whose histopathology most closely resembles a neoplastic or “transformed” pyogenic granuloma is: 
   a. squamous cell carcinoma.
   b. basal cell carcinoma.
   c. Kaposi’s sarcoma.
   d. capillary hemangioma.
   e. rhabdomyosarcoma.

66. The differential diagnosis for interstitial keratitis (IK) includes: 
   1. sarcoidosis.
   2. onchocerciasis.
   3. congenital syphilis.
   4. Acanthamoeba keratitis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

67. Which one of the following statements about “combined hamartomas” of the retina and retinal pigment epithelium (RPE) is false? 
   a. Combined hamartomas are frequently associated with other developmental ocular anomalies.
   b. Combined hamartomas are usually clinically silent until adulthood.
   c. One clinical hallmark is retinal vascular tortuosity.
   d. The peripapillary region is most frequently affected.
   e. Vitrectomy with membrane peeling is generally not effective for restoring vision.

68. T or F The diagnosis of Acanthamoeba keratitis is enhanced by special staining with calcofluor white and culturing on blood agar layered with Staphylococcus aureus.
69. Which of the following statements about corneal edema and its common etiologies is/are true?

1. The presence of guttae and thickened Descemet’s membrane is sufficient for the diagnosis of Fuchs’ dystrophy.
2. The differentiating feature between true Fuchs’ dystrophy and pseudophakic bullous keratopathy (PBK) is the thickness of Descemet’s membrane.
3. Acute increases in intraocular pressure (IOP) generally cause stromal and epithelial edema.
4. End-stage histopathology frequently reveals changes identical to map-dot-fingerprint dystrophy.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

70. Which of the following is/are anterior corneal dystrophies?

1. map-dot-fingerprint dystrophy.
2. Reis-Bückler dystrophy.
3. Meesmann’s dystrophy.
4. granular dystrophy.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

71. T or F The microcystic form of map-dot-fingerprint dystrophy may be difficult to distinguish clinically from Meesmann’s dystrophy.

72. Which one of the following corneal dystrophies is most likely to be associated with a recurrent erosion syndrome?

a. granular dystrophy.
b. Reis-Bückler dystrophy.
c. macular dystrophy.
d. lattice dystrophy.
e. central cloudy dystrophy.

73. T or F Granular dystrophy may be distinguished from macular dystrophy by the clarity of corneal stroma between its deposits.

74. T or F Granular dystrophy may be distinguished from macular dystrophy on the basis of a careful family history.

75. Posterior polymorphous dystrophy (PPMD) clinically or pathologically may resemble:

1. Fuchs’ dystrophy.
2. epithelial downgrowth.
3. congenital hereditary endothelial dystrophy.
4. the iridocorneal endothelial (ICE) syndromes.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

76. Which of the following statements about keratoconus is false?

a. It is associated with atopy, Down’s syndrome, and Marfan’s syndrome.
b. The earliest histologic change seen is thinning of the epithelium.
c. Anterior stromal scarring is typically a late finding.
d. Iron deposition in epithelial basal cells is not uncommon.
e. There may be a hereditary or familial predisposition.

77. T or F The most sensitive method of detecting an early Kayser-Fleischer ring is with thin-beam cross-sectional analysis at the slit lamp.

78. Which of the following statements about the lens capsule is/are true?

1. Anterior capsular guttae may be seen in aniridia and Lowe’s syndrome.
2. In posterior lenticous, the primary defect is an abnormally thin posterior capsule.
3. In any case of hypercupremia, copper may deposit in the lens capsule in a sunflower pattern.
4. The most common abnormality encountered clinically is the true exfoliation syndrome.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

79. Which of the following insults may induce lens epithelial replication, degeneration, or metaplasia?
   1. elevated intraocular pressure (IOP).
   2. chronic inflammation.
   3. trauma.
   4. atopic dermatitis.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

80. T or F Intralenticular fissures filled with eosinophilic globules are common artifacts caused by tissue-processing techniques.

81. Which of the following is/are true about the cataract associated with congenital rubella infection?
   1. Ocular findings are frequently coincident with abnormalities in the cardiovascular and auditory systems.
   2. A distinctive feature of rubella cataracts is the retention of nuclei within central lens fibers.
   3. Surgical removal of these cataracts frequently induces aggressive intraocular inflammation.
   4. Live virus is present within the lenses of patients with congenital rubella syndrome.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

82. The retinal circulation supplies which of the following layers?
   1. inner plexiform layer.
   2. inner nuclear layer.
   3. ganglion cell layer.
   4. outer nuclear layer.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

83. T or F The clinical macula is identical to the histologic macula.

84. Which anatomic feature(s) of the fovea contribute(s) to its dark appearance on angiography?
   1. local differences in retinal pigment epithelium (RPE) melanin.
   2. local differences in retinal pigmentation.
   3. local differences in the retinal vascular system.
   4. local differences in the choroidal melanin.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

85. T or F Lange’s fold is a sensitive indicator of ocular trauma in postmortem ocular examination.

86. Classic macular edema and exudates accumulate in the:
   a. nerve fiber layer.
   b. ganglion cell layer.
   c. inner plexiform layer.
   d. inner nuclear layer.
   e. outer plexiform layer.

87. Microaneurysms are located in the:
   a. nerve fiber layer.
   b. ganglion cell layer.
   c. inner plexiform layer.
   d. inner nuclear layer.
   e. outer plexiform layer.

88. Cotton-wool spots are located in the:
   a. nerve fiber layer.
   b. ganglion cell layer.
   c. inner plexiform layer.
   d. inner nuclear layer.
   e. outer plexiform layer.

89. The preservative/processing technique of choice for biopsy handling in presumed corneal cystinosis involves:
   a. fixation in 10% formalin.
   b. fixation in glutaraldehyde.
Questions

c. frozen sections.
d. fixation in absolute alcohol.
e. immediate sectioning after transport in saline-soaked gauze.

90. Which of the following statements about the histopathology of diabetic eye disease is/are true?

1. There is relative loss of pericytes.
2. Lacy vacuolization of the iris pigment epithelium is common.
3. Thickening of the basement membrane of the ciliary epithelium is universally present.
4. Microaneurysms generally arise within the ganglion cell layer.

  a. 1, 2, and 3.
  b. 1 and 3.
  c. 2 and 4.
  d. 4 only.
  e. 1, 2, 3, and 4.

91. T or F Pigmentary glaucoma is more common in myopic men aged between 20 and 50 years.

92. Which of the following statements about venous occlusions of the retina is/are true?

1. The inferotemporal quadrant is most frequently involved by branch retinal vein occlusion (BRVO).
2. Between 20% and 25% of patients with BRVO will develop retinal neovascularization.
3. There is no association of BRVO or central retinal vein occlusion (CRVO) with glaucoma.
4. Approximately 50% of patients with untreated BRVO will maintain vision better than 20/40.

  a. 1, 2, and 3.
  b. 1 and 3.
  c. 2 and 4.
  d. 4 only.
  e. 1, 2, 3, and 4.

93. Which of the following statements about lattice degeneration is/are true?

1. Strong vitreous adhesions are found directly over the degenerated retina.
2. Lattice degeneration may be found in up to 10% of the general population.
3. Lattice degeneration is the most common finding associated with retinal detachment.
4. The branching, white lines seen clinically correlate with histopathologically sclerotic retinal vessels.

  a. 1, 2, and 3.
  b. 1 and 3.
  c. 2 and 4.
  d. 4 only.
  e. 1, 2, 3, and 4.

94. The leading cause of new blindness in the United States is:

  a. glaucoma.
  b. cataract.
  c. age-related macular degeneration.
  d. diabetes.
  e. retinal detachment (RD).

95. Which of the following statements about drusen of the retinal pigment epithelium (RPE) is/are true?

1. Calcific drusen are associated with atrophy of surrounding RPE.
2. Hard drusen are the most common type seen in age-related macular degeneration.
3. Confluent drusen represent diffuse thickening of the inner layer of Bruch’s membrane.
4. Soft drusen represent microscopic RPE detachments.

  a. 1, 2, and 3.
  b. 1 and 3.
  c. 2 and 4.
  d. 4 only.
  e. 1, 2, 3, and 4.

96. T or F Pavingstone’s degeneration represents a disturbance of the inner retinal circulation.

97. T or F Typical peripheral cystoid degeneration (TPCD) is a common risk factor for retinal detachment.

98. Which of the following statements about the genetics of retinoblastoma is/are true?

1. The pertinent gene is located on the long arm of chromosome 13 (13q).
2. Retinoblastoma will be expressed in patients who have two abnormal retinoblastoma genes.
3. Clinically, familial retinoblastoma is transmitted in an autosomal dominant pattern.

4. New germ cell mutations of 13q do not increase the risk of nonocular malignancy.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

99. Which of the following statements about the histopathology of retinoblastoma is/are true?

1. The tumors are mitotically active with characteristic zones of perivascular proliferation separated by zones of necrosis.

2. These tumors appear particularly blue microscopically, because of liberated nucleic acid and calcification.

3. Flexner-Wintersteiner rosettes are extremely specific for retinoblastoma, with rare exceptions.

4. Homer-Wright rosettes are extremely specific for retinoblastoma, with rare exceptions.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

100. Which of the following statements about the prognosis in retinoblastoma is/are true?

1. The most common route for tumor spread is lymphatic metastasis.

2. Ninety-five percent of individuals with localized intraocular disease survive.

3. The most important prognostic factor for survival is the extent of choroidal invasion.

4. Bilaterality may increase mortality.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

101. Which of the following statements about medulloepithelioma is/are true?

1. Also known as diktyoma, it usually occurs from the ciliary body but also has been reported in the retina and optic nerve.

2. The cellular characteristics are strikingly similar to retinoblastoma.

3. Typically, the cells align themselves in a stratified ribbon distribution.

4. Heteroplastic tissue such as cartilage or smooth muscle may be found within their substance.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

102. In malignant melanoma of the choroid, the most important prognostic factor(s) is/are:

1. sex.

2. dimensions at the base of the tumor.

3. degree of pigmentation.

4. cell type.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

103. The most common site for primary nonocular tumors to develop metastasis in or around the eye in adults is the:

a. orbit.
b. choroid.
c. retina.
d. optic nerve.
e. iris.

104. T or F The myositis of idiopathic orbital inflammation (pseudotumor) is characterized by its cellular homogeneity.

105. T or F The orbit contains no resident lymph nodes.

106. T or F Orbital lymphomas are nearly always B-cell proliferations.
107. Systemic lymphoma is most likely to be associated with ocular adnexal disease characterized by:
   a. large tumor size.
   b. monoclonality.
   c. high degree of cytologic atypia.
   d. follicular pattern of growth.
   e. eyelid involvement.

108. T or F Primary orbital tumors are much more common than secondary orbital tumors.

109. The most common intraconal orbital tumor is the:
   a. meningioma.
   b. rhabdomyosarcoma.
   c. cavernous hemangioma.
   d. fibrous histiocytoma.
   e. neurofibroma.

110. Which of the following statements about rhabdomyosarcoma is false?
   a. It is one of the two most common primary solid malignancies in children.
   b. The clinical presentation is typically acute.
   c. Spontaneous lid ecchymosis is a very specific sign.
   d. The embryonal histologic pattern is the most common.
   e. The alveolar pattern has the worst prognosis.

111. Which of the following statements about neurofibromas and schwannomas is/are true?
   1. Unlike schwannomas, neurofibromas are encapsulated.
   2. Both tumors are found in increased frequency in patients with neurofibromatosis type 1.
   3. Nodular neurofibroma is classic in neurofibromatosis.
   4. Verocay bodies are seen in Antoni type A schwannomas.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

112. Which one of the following statements about lacrimal gland lesions is false?
   a. Roughly 80% are inflammatory or lymphoid lesions, and 20% are epithelial cell tumors.
   b. Among epithelial cell tumors, 50% are benign.
   c. The most common malignant epithelial tumor of the lacrimal gland is the adenoid cystic carcinoma.
   d. Pain as a presenting finding indicates either an inflammatory or malignant lesion.
   e. The most common epithelial tumor, the pleomorphic adenoma, is encapsulated and may be shelled out easily.

113. T or F The melanocytoma of the optic nerve head has not been associated with malignant melanoma of the choroid.

114. Which of the following statements about optic nerve glioma is/are true?
   1. Glioma may involve any part of the visual pathway.
   2. The typical age of presentation is between 10 and 20 years.
   3. Rosenthal fibers are not unique to optic nerve glioma.
   4. The prognosis is directly related to histopathologic cell type.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

115. Which of the following statements about orbital meningioma is/are true?
   1. Primary orbital meningioma is more common than secondary orbital meningioma.
   2. The mean age of patients with primary orbital meningioma is lower than in patients with secondary orbital meningioma.
   3. The prognosis is most closely related to cell type.
   4. Patients with meningioma in the pediatric population have a poorer prognosis.
   a. 1, 2, and 3.
   b. 1 and 3.
116. Which of the following statements about the histopathology of glaucoma is/are true?

1. Cavernous atrophy of Schnabel is only seen in primary open-angle glaucoma (POAG).
2. Disc pallor noted clinically reflects decreased vascularity of the optic nerve head.
3. With special stains and careful review of optic nerve specimens, the specific etiology of several types of glaucoma may be determined.
4. The retina shows ganglion cell dropout and nerve fiber layer atrophy in cases of long-standing glaucoma.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

117. Which of the following statements about the particulate glaucomas is/are true?

1. Excessive pigment in the trabecular meshwork (TM) is specific for pigment dispersion syndrome.
2. Pseudoexfoliation is characterized by its bilateral symmetry.
3. Pseudoexfoliative material is found only within the anterior chamber.
4. After lens extraction, the synthesis of pseudoexfoliative material typically continues.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

118. T or F The lifetime risk of glaucoma in an eye with >180 degrees of angle recession is 5% to 10%.

119. T or F A universal feature of the iridocorneal endothelial (ICE) syndromes is abnormal corneal endothelial proliferation.

120. Clinical findings suggestive of prior attacks of acute angle-closure glaucoma include:

1. white anterior lenticular opacities.
2. segmental iris heterochromia with atrophy.
3. peripheral anterior synechiae (PAS).
4. narrow angles in the contralateral eye.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

121. Which of the following eyelid tumors may be associated with extraocular malignancy?

1. pilomatrixoma.
2. trichilemmoma.
3. trichoepithelioma.
4. sebaceous adenoma.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

122. Which one of the following statements about medulloepithelioma is false?

a. Medulloepithelioma primarily affects the pediatric age group.
b. Medulloepithelioma is typically cystic on its surface.
c. Medulloepithelioma may arise at the optic nerve.
d. Medulloepithelioma generally arises from non-pigmented ciliary epithelium.
e. Hematogenous metastasis is common.

123. A pigmented lesion of the choroid is likely to be benign if:

1. its diameter is <10 mm.
2. there are drusen and associated lipofuscin deposits.
3. it is <1 mm in thickness.
4. there is an associated dependent exudative retinal detachment (RD).

a. 1, 2, and 3.
b. 1 and 3.
Questions

c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

124. Fluorescein angiography (FA) is helpful in distinguishing choroidal melanoma from:
1. metastatic tumors.
2. choroidal hemangioma.
3. large nevi.
4. subretinal hemorrhage.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

125. T or F The internal reflectivity of melanoma on A-scan ultrasonography is typically lower than that of other choroidal masses.

126. Risk factors for mortality in patients with choroidal melanoma include all of the following except:
a. anterior tumor location.
b. clinically documented tumor growth.
c. extraocular extension of the tumor.
d. large-sized tumor.
e. the histologic observation of melanin granules within the vitreous cavity after enucleation.

127. The most sensitive method for detecting hepatic metastasis in uveal melanoma is:
a. liver enzyme testing.
b. abdominal computed tomography (CT) scanning.
c. abdominal ultrasonography.
d. radionuclide liver scanning.
e. physical examination.

128. The “tomato ketchup” fundus is a classic finding in which of the phakomatoses?
a. neurofibromatosis.
b. Sturge-Weber syndrome.
c. tuberous sclerosis.
d. ataxia-telangiectasia.
e. von Hippel-Lindau disease.

129. Which of the following statements about choroidal osteoma is/are true?
1. Choroidal osteoma is more common in women.
2. Most cases of choroidal osteoma are bilateral.
3. Choroidal osteoma most frequently arises in the peripapillary region.
4. Ultrasonography is of no value in distinguishing choroidal osteoma from amelanotic melanoma.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

130. Which of the following statements about the genetics of retinoblastoma is/are true?
1. Retinoblastoma is transmitted in an autosomal-dominant manner, with approximately 80% penetrance.
2. Approximately 6% of patients with retinoblastoma will have a family history of retinoblastoma.
3. Fifteen percent of sporadic cases (i.e., no family history) represent a new germinal mutation.
4. The parent of a patient with unilateral retinoblastoma who has no family history of retinoblastoma has a 1% chance of having another child with retinoblastoma.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

131. T or F Patients with inherited retinoblastoma are at increased risk for having other tumors.

132. Common presenting symptoms or signs in retinoblastoma include:
1. leukocoria.
2. buphthalmos.
3. strabismus.
4. proptosis.
4: Ocular Pathology

133. The common sites of metastasis of retinoblastoma include:

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

134. T or F The Reese-Ellsworth classification of retinoblastoma gives prognostic information about long-term survival.

135. To be correctly termed von Hippel-Lindau syndrome, retinal capillary hemangioma must be associated with:

a. pancreatic cysts.
b. renal cell carcinoma.
c. pheochromocytomas.
d. cerebellar hemangioblastomas.

136. Which of the following statements about metastatic carcinoma involving the eye and adnexa is/are true?

a. Up to 10% of patients with known metastases will have ocular or orbital involvement.
b. Ocular involvement is more than five times more likely than orbital involvement.
c. Lung cancer is the most frequently encountered primary tumor in men, and breast cancer is the most frequently encountered primary tumor in women.
d. Gastrointestinal tumors metastasize to the eye or orbit infrequently.

137. Which of the following statements about ocular and adnexal metastasis is/are true?

a. Isolated retinal metastases are common in gastrointestinal cancers.
b. Up to 70% of patients with ocular metastasis from the lung have no history of a pulmonary lesion.
c. At least 25% of all patients with breast cancer present with uveal metastasis on diagnosis.
d. Metastasis represents the most common intraocular malignancy in adults.

138. Which one of the following clinical findings is not significantly associated with higher risk of metastasis for choroidal melanocytic lesions <3.0 mm in thickness?

a. posterior margin touching the optic disc.
b. subretinal fluid.
c. orange pigment on its surface.
d. visual disturbances.
e. associated intraretinal retinal pigment epithelium (RPE) hyperplasia.

139. Which one of the following clinical findings is not significantly associated with higher risk of growth of a choroidal melanocytic lesion <3.0 mm in thickness?

a. posterior margin touching the optic disc.
b. subretinal fluid.
c. orange pigment on its surface.
d. visual disturbances.
e. associated intraretinal retinal pigment epithelium (RPE) hyperplasia.

140. Which of the following structures is the most sensitive to radiation?

a. the cornea.
b. the retina.
c. the lens.
d. the optic nerve.
e. the iris.
141. Which of the following viruses would be most likely associated in an adult patient with the lesion pictured in the figures below?
   a. human immunodeficiency virus (HIV).
   b. herpes simplex virus (HSV).
   c. varicella-zoster virus (VZV).
   d. human papillomavirus (HPV) subtype 6.
   e. HPV subtype 16.

142. After excision of the lesion shown in the figure below (left) and examination of a histologic section shown in the figure below (right), the most likely diagnosis is:
   a. malignant melanoma.
   b. seborrheic keratosis.
   c. squamous cell carcinoma.
   d. eyelid lymphoma.
   e. keratoacanthoma.

143. In what layer of skin is the pathology of the lesions shown in the figure below found?
   a. epidermis.
   b. dermis.
   c. hair shafts/follicles.
   d. muscle (e.g., orbicularis).
   e. orbital fat.
144. The lesion shown in the figure below (left) is excised. Histopathology is demonstrated in the figure below (right). The lesion is most likely a:
   a. pilomatrixoma.
   b. basal cell carcinoma.
   c. sebaceous cell carcinoma.
   d. trichoepithelioma.
   e. cylindroma.

145. Which of the following statements about the lesion pictured in the figure below is true?
   a. It is one of the least common benign tumors of childhood.
   b. It usually involves the lower eyelid.
   c. Simple surgical removal is the treatment of choice.
   d. Strabismus rarely develops in infants with this condition.
   e. Eyelid necrosis can be a side effect of treatment.

146. Which of the following histologic sections most likely represent the lesion in question 145?
   a. the figure below on the left.
   b. the figure below in the middle.
   c. the figure below on the right.
   d. all of the above.
   e. none of the above.
147. An adult patient notes the development of the lesions shown in the figure below (left). One of them is excised and its histopathology is shown in the figure below (right). Which of the following statements is false?
   a. The lesions are likely benign.
   b. The patient’s sexual history should be obtained.
   c. Eosinophilic intracytoplasmic inclusion bodies are often observed on histopathologic examination.
   d. The lesions are probably caused by a ribonucleic acid (RNA) virus.
   e. Treatment with either cryotherapy or complete surgical excision can be effective.

148. A 1-year-old child undergoes surgical excision of the lesion shown in the figure below (left). Histopathology is shown in the figure below (right). Which of the following statements is false?
   a. The infant may be at risk for the development of visual loss because of secondary glaucoma.
   b. Histopathology, as shown in the figure below (right), demonstrates the presence of Touton’s giant cells.
   c. The lesion usually spontaneously regresses in the child by the age of 5 years.
   d. The lesion shown in the figure below (left) can undergo malignant transformation.
   e. Topical corticosteroids can help control secondary glaucoma if it develops in these patients.

149. The figure on the right shows a Gram stain of the canalicular reflux of a patient. What would you use to treat this patient?
   a. surgical excision.
   b. oral metronidazole.
   c. oral amoxicillin/clavulanic acid.
   d. intravenous ceftazidime.
   e. oral valacyclovir.
150. Over the last year, a 50-year-old woman has noticed the development of the conjunctival lesion shown in the figure below (left). The lesion is excised, and its histopathology is shown in the figure below (right). What is the chance that this lesion could progress to a conjunctival melanoma?

a. <1%.
b. 5%.
c. 10%.
d. 25%.
e. 50%.

151. Over the last year, a 45-year-old man has noticed the development of the conjunctival lesion shown in the figure below (left). The lesion is excised, and its histopathology is shown in the figure below (right). What is the chance that this lesion could progress to a conjunctival melanoma?

a. <1%.
b. 5%.
c. 10%.
d. 25%.
e. 50%.

152. The patient in the following figure on the left is concerned that his eyes “are getting larger.” A biopsy of the lesion performed at an outside hospital, shown in the following figure on the right, demonstrates that the patient likely has:

a. thyroid orbitopathy.
b. a benign condition.
c. sarcoidosis.
d. bilateral pleomorphic adenomas.
e. none of the above.
Questions

153. A patient complains of long-standing decreased vision. His cornea is pictured in the figure below (left). A biopsy is performed, shown in the figure below (right). The patient likely has:
   a. keratoconus.
   b. squamous cell carcinoma of the cornea.
   c. pellucid marginal degeneration.
   d. a history of topical anesthetic abuse.
   e. spheroidal degeneration.

154. A patient complains of photophobia, pain, and decreased vision. His cornea is pictured in the figure below (left). The patient eventually required penetrating keratoplasty, and histopathologic section of the excised cornea is shown in the figure below (right). Which of the following statements is true?
   a. The stain used in histopathologic section is Masson's trichrome.
   b. This is an autosomal-recessive disorder.
   c. Hyaline granules are the cause of pathology in the disorder.
   d. Recurrence of the condition in the corneal graft is rare.
   e. The condition is caused by a mutation in the gene responsible for keratoepithelin formation.
155. Which of the figures shown below represent granular dystrophy?
   a. figures a. and f.
   b. figures c. and d.
   c. figures b. and e.
   d. figures a. and e.
   e. figures b. and d.

156. Which of the following statements about the dystrophy pictured in the figure below (left) is false?
   a. The figure below (right) depicts the likely histopathologic correlate.
   b. Type III of this corneal dystrophy is associated with cranial nerve palsies.
   c. The stain often used in histopathologic examination is Congo red.
   d. Amyloid is the material deposited.
   e. The dystrophy is inherited in an autosomal-dominant fashion.
Questions

157. Which figure most likely represents a histopathologic section of a cornea with macular dystrophy?
   a. the figure below on the left.
   b. the figure below in the middle.
   c. the figure below on the right.
   d. none of the above.
   e. Macular dystrophy is a retinal condition (not a corneal one).

158. What stain is used in the figure below?
   a. oil red O.
   b. Masson’s trichrome.
   c. periodic acid-Schiff (PAS).
   d. Alcian blue.
   e. Congo red.

159. Which of the following descriptions would likely not be associated with the ocular histopathology shown in the figure below?
   a. a tall, long-fingered patient with a history of aortic aneurysm.
   b. a tall patient with a history of thromboembolism.
   c. a patient with sulfite oxidase deficiency.
   d. a patient with hypolysinemia.
   e. none of the above.

160. Which of the following lenticular diagnoses is demonstrated in the figure below?
   a. Cogan-Reese syndrome.
   b. Mittendorf’s dot.
   c. Ectopia lentis.
   d. Soemmerring’s ring.
   e. Morgagnian’s cataract.
161. A patient with the lesion shown in the figure below on the left (and corresponding pathology in the figure below on the right) is at risk for which of the following?
   a. malignant transformation of the iris nevus.
   b. spontaneous hyphema.
   c. ectopia lentis.
   d. endophthalmitis.
   e. none of the above.

162. T or F The figure below would be expected to demonstrate Touton’s giant cells.

163. A patient with granulomatous uveitis undergoes sputum culturing, which is shown in the figure below. Which of the following diagnoses is consistent with the figure?
   a. psittacosis.
   b. sarcoidosis.
   c. emphysema.
   d. tuberculosis.
   e. coccidiomycosis.

164. What stain is likely being used in the figure in question 163?
   a. Loewenstein-Jensen.
   b. Giemsa.
   c. periodic acid-Schiff (PAS).
   d. Masson’s trichrome.
   e. Ziehl-Neelsen.

165. A patient with the pathology shown in the figure below is at risk for:
   a. neovascular glaucoma.
   b. malignant melanoma of the iris.
   c. essential iris atrophy.
   d. Cogan-Reese syndrome.
   e. Axenfeld’s anomaly.

166. Which of the following statements about the histopathology shown in the following figure is false?
   a. The eye has atrophied and has probably shrunk from its original size.
   b. The sclera is thinner than normal.
   c. The pressure in the eye can be variable.
167. Which of the following statements about a patient with the pathology shown in the figure below is false?
   a. The patient may have genitourinary abnormalities.
   b. Most cases of this condition are bilateral.
   c. The patient may have had a history of nystagmus.
   d. This is usually an autosomal-recessive disorder.
   e. Sporadic cases can be associated with mental retardation.

168. A patient presents with a fundus shown in the figure below (left). The condition’s histopathology is shown in the figure below (right). What is your most likely diagnosis?
   a. Coats’ disease.
   b. retinal toxoplasmosis.
   c. intraocular lymphoma.
   d. hemangioblastoma.
   e. sarcoidosis.
169. What type of testing should the patient with the lesion in the left-hand figure in question 168 undergo?
   a. neuroimaging.
   b. renal ultrasonography.
   c. 24-hour urine collection for vanillylmandelic acid measurement.
   d. all of the above.
   e. none of the above.

170. What are potential treatment options for the patient with the lesion shown in the left-hand figure in question 168?
   a. cryotherapy.
   b. laser photocoagulation.
   c. antiangiogenic therapy.
   d. external beam radiation.
   e. all of the above.

171. In what layer of the retina does splitting occur in the figure below?
   a. inner nuclear layer.
   b. photoreceptor layer.
   c. outer plexiform layer.
   d. inner plexiform layer.
   e. nerve fiber layer.

172. In what layer of the retina does splitting occur in the figure below?
   a. inner nuclear layer.
   b. photoreceptor layer.
   c. outer plexiform layer.
   d. inner plexiform layer.
   e. nerve fiber layer.

173. What is the most likely cause of the idiopathic condition shown in the figure below?
   a. subretinal fluid creating breaks in Bruch’s membrane.
   b. subfoveal choroidal neovascularization.
   c. tangential and anteroposterior vitreomacular traction.
   d. foveal schisis leading to the accumulation of intraretinal fluid accumulation.
   e. none of the above.

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**Matching**

174. Match each differentiated cell lettered below with its immediate precursor.
   a. cytotoxic cell. 1. basophils.
   b. mast cell. 2. monocyte.
   c. epithelioid cell. 3. macrophage.
   d. plasma cell. 4. B lymphocyte.
   e. giant cell. 5. T lymphocyte.
   f. macrophage.

175. Match the conditions lettered below with the giant cell type typically seen in each.
   a. lipogranuloma. 1. Touton’s.
   b. sarcoidosis. 2. Langerhans’.
   c. juvenile xanthogranuloma (JXG). 3. foreign body.
   d. tuberculosis. 4. Langhans’.
   e. necrobiotic xanthogranuloma.
   f. fungal granuloma.
   g. leprosy.
   h. Erdheim–Chester disease.
176. Match the appropriate stain technique lettered below with the correct organisms.
   c. Gomori methenamine silver. 3. spirochetes.
   d. Warthin-Starry. 4. mycobacteria.

177. Match the gonioscopic and clinical findings with the appropriate condition.
   a. prominent, anteriorly placed Schwalbe line. 1. Rieger’s anomaly.
   b. “a” plus iris processes to the Schwalbe line. 2. posterior embryotoxon.
   c. “b” plus glaucoma. 3. Rieger’s syndrome.
   d. “b” plus iris atrophy and corectopia. 4. Axenfeld’s syndrome.
   e. “d” plus dental/facial anomalies. 5. Axenfeld’s anomaly.

178. For each of the following phakomatoses, select the correct clinical manifestation. (Note that manifestations may be used more than once and that each disorder may have more than one manifestation.)
   a. von Hippel-Lindau syndrome. 1. pulsating exophthalmos.
   b. Sturge-Weber syndrome. 2. astrocytic hamartoma of the optic nerve and retina.
   c. neurofibromatosis. 3. retinal capillary hemangioma.
   d. tuberous sclerosis. 4. choroidal cavernous hemangioma.
   e. ataxia-telangiectasia. 5. irregular retinal vessels.

179. Match the pathologic condition with the potential gland of origin. (Note that each condition may be associated with more than one gland. Glands may be used more than once or not at all.)
   b. sebaceous carcinoma. 2. glands of Zeis.
   c. external hordeolum. 3. glands of Krause and Wolfing.
   d. sudoriferous cyst. 4. eccrine sweat glands.
   e. syringoma. 5. meibomian glands.

180. Match the eponyms of various iron lines with the appropriate location.
   a. Hudson-Stähli line. 1. at the base of the cone in keratoconus.
   b. Fleischer line. 2. at the base of a filtering bleb.
   c. Stocker line. 3. along the lower lid margin.
   d. Ferry line. 4. along the advancing edge of a pterygium.
Answers

1. e. A Russell body is a spherically shaped intracellular deposit that forms when antibody accumulates and displaces the nucleus of a plasma cell.
2. False. Although giant cells are often present in granulomatous inflammation, the hallmark is the presence of epithelioid cells.
3. e. Although Descemet's membrane is a barrier to many organisms, fungi can penetrate Descemet's membrane into the anterior chamber with relative ease. Candida and Cryptococcus are yeasts; most corneal pathogens are molds.
4. b. There is typically moderate to severe keratitis in trachoma, usually subepithelial. Adult inclusion conjunctivitis is typically worse inferiorly, in distinction to trachoma, which is typically worse superiorly. Giemsa stain is more effective than Wright stain at revealing intracytoplasmic inclusions. Serotypes A through C are associated with trachoma, D through K with inclusion conjunctivitis, and lymphogranuloma venereum (LGV) 1-3 with lymphogranuloma venereum.
5. a. The inclusion bodies seen in molluscum contagiosum are intracytoplasmic. They are typically quite large, occupying virtually the entire intracellular volume. Subsequent growth may cause cellular rupture with clinically evident secondary inflammation. The following is a list of organisms and their characteristic inclusion bodies:
   - Chlamydia: intracytoplasmic
   - Molluscum contagiosum virus: eosinophilic, intracytoplasmic inclusions (Henderson-Patterson bodies)
   - Cytomegalovirus (CMV): intranuclear and intracytoplasmic
6. b. Infection with Toxocara canis produces a retinochoroiditis characterized by an eosinophilic abscess. The larvae of Taenia solium, Cysticercus cellulosae, cause relatively little inflammation until the cyst ruptures (ocular cysticercosis), when an intense granulomatous reaction can result.
7. True. The host response to initial herpes simplex infection is nongranulomatous inflammation of the conjunctiva, whereas reactivation may cause granulomatous inflammation adjacent to Descemet's membrane.
8. a. Large-cell lymphoma (reticulum cell sarcoma) may present as a pseudovitritis. Retinoblastoma may present as a pseudohypopyon. Aggressive choroidal melanoma may become necrotic or cause necrosis of the overlying retina with secondary uveitis. Melanocytomas typically do not present as ocular inflammation.
9. b. Although some authors consider phacoanaphylaxis (phacoantigenic endophthalmitis), phacoanaphylactic glaucoma is not true active inflammation. Rather, there is an increase in intraocular pressure (IOP) caused by clogging of the trabecular meshwork (TM) by lens protein and macrophages. The lens capsule is typically not ruptured in phacoanaphylactic glaucoma. Phacoantigenic glaucoma describes lens-induced inflammation as a result of immunoglobulin G (IgG) directed against lens proteins. Also, because mast cells play no role in “phacoanaphylaxis,” phacoantigenic is a better descriptor.
10. c. Behçet's disease is a nongranulomatous vasculitis that causes acute uveitis with hypopyon, aphthous stomatitis, and genital ulceration. Rheumatoid arthritis causes a zonal granuloma centered on scleral collagen. Sarcoidosis typically produces a discrete granulomatous reaction, whereas sympathetic ophthalmia and Vogt-Koyanagi-Harada syndrome (VKH) generally cause diffuse granulomatous inflammation.
11. e. Juvenile rheumatoid arthritis (JRA) and Reiter's syndrome are specifically characterized by recurrent or chronic nongranulomatous uveitis, whereas choroidal melanoma and ocular trauma may incite secondary ocular inflammation, which is nearly always nongranulomatous. (In the case of a retained intraocular foreign body after trauma, there may be a granulomatous component.) Cryptococcal endophthalmitis, like other fungal causes of ocular inflammation, features marked granulomatous inflammation.
12. False. Microscopically, granulomatous inflammation of the choriocapillaris strongly favors the diagnosis of Vogt-Koyanagi-Harada syndrome (VKH) over sympathetic ophthalmia (SO). Note that sparing of the choriocapillaris favors SO but does not rule out VKH.
13. d. Perforating wounds of the cornea do not elicit a typical granulation tissue response because there are no native blood vessels. Stromal swelling is important in the very early response phase to prevent ongoing aqueous leakage. The key response for long-term healing is the closure of the internal defect with adjacent endothelial cells, via migration or metaplasia (and, only rarely, mitosis). Note that any dermal appendages lost are replaced by fibrous (scar) tissue. Episcleral vessels and resident fibrocytes are responsible for limbal healing (the sclera itself is almost entirely avascular). The iris, despite rich vascularity and innervation, reacts minimally to mechanical injury.
14. c. The retina contains no true fibrocytes or fibroblasts. Glial (Müller's) cells are the intraretinal reactive component, so that purely retinal injury leads to gliosis and proliferation primarily within the inner nuclear layer. The retinal pigment epithelium (RPE) frequently becomes involved in retinal injury, typically with a hyperplastic response, giving rise to dense hyperpigmented clumps of fundus scars. Intraretinal fibrosis implies involvement of the choroid in the repair process.
15. False. Some ultrastructural studies reveal that this epithelium resembles conjunctiva more closely than cornea.

16. c. Sympathetic ophthalmia (SO) most often occurs after accidental trauma. Most postoperative cases follow filtering procedures on blind, glaucomatous eyes.

17. a. Graft failure is a nonspecific term referring to opacification of the donor cornea. This may occur secondary to endothelial damage, inadequate donor tissue, or postoperative infection.

18. d. Elschnig’s pearls are residual proliferating lens epithelial cells that form small globular, opaque fibers, which can migrate onto the posterior capsule following cataract extraction or surgery. Soemmerring’s ring is retention of equatorial lens cortical material following cataract extraction.

19. b. Because of the oblique orientation of the axons in this layer of the fovea, fluid accumulates more easily here than in retinal layers where the neural and glial elements are oriented radially.

20. c. Corneal blood staining results from displacement of hemoglobin (not red blood cells [RBCs]) through a traumatized endothelium but intact Descemet’s membrane. In hemosiderosis bulbi, hemosiderin (a hemoglobin breakdown product) accumulation leads to epithelial damage. Synchysis scintillans results from the accumulation of red blood cell membrane breakdown products (e.g., cholesterol) in the vitreous. An ochre membrane is the organization of posterior segment hemorrhage on the posterior surface of a detached vitreous.

21. False. An occluded pupil (occlusio pupillae) satisfies this definition. In contrast, a secluded pupil implies complete (360-degree) lens–pupil apposition, because of severe posterior synchia.

22. a. Calcification may be seen in atrophic eyes but is not necessary for the diagnosis. The other three, in combination, constitute the diagnosis of phthisis bulbi.

23. d. The phakomatous choristoma (Zimmerman’s tumor) represents a congenital developmental anomaly (choristoma) with lenslike (hence, “phakomatous”) tissue forming a mass within the eyelid, usually lower. Such a growth has been reported nowhere else in or on the body. The lesion is rare and has no reported ocular associations, with surgical excision being the usual treatment.

24. a. A cleavage plane between the longitudinal ciliary body fibers and sclera represents cyclodialysis. Iridodialysis represents iris tearing at its root or insertion. In angle recession, the plane of cleavage is between circular and longitudinal fibers of the ciliary muscle.

25. d. Retinal dialysis occurs because of mechanisms other than posteroanterior vitreous traction; thus, it is more common to discover dialyses in eyes with attached posterior hyaloid. Spontaneous dialyses, often in patients with a family history of retinal dialysis and/or detachment, occur most commonly in the inferotemporal quadrant. Subsequent detachment is typically slowly progressive and often silent clinically.

26. c. Passage into (but not through) a structure is called penetrating injury. Passage through a structure is called perforating injury. This patient perforated his cornea and lens but penetrated his eye. Had the foreign body passed through the posterior sclera, he would have suffered ocular perforation, also clumsily termed a double penetrating injury.

27. c. Siderosis bulbi is caused by epithelial damage caused by ferrous (Fe^{2+}) iron. Copper foreign bodies with >85% elemental copper lead to a suppurative noninfectious endophthalmitis. Chalcosis occurs when the copper concentration is between 70% and 85%. Vegetable matter is rarely sterile; in addition, it elicits a severe granulomatous reaction. Lead does elicit an inflammatory reaction, albeit not as severe as copper or iron.

28. b. Ammonium hydroxide has the greatest capability to penetrate into the anterior chamber. Petroleum products cause damage from detergentlike actions.

29. a. There is no evidence that supports microwave radiation–causing cataracts in humans.

30. a. There is no evidence that supports microwave radiation–causing cataracts in humans.

31. False. A hamartoma consists of aberrant tissue elements in their normal location (e.g., astrocytoma of the optic nerve head), whereas a choristoma consists of normal tissue in an abnormal location (e.g., dermoid cyst of the orbit or eyelid). A useful mnemonic device is “hammer home.” “Hammer-tomas” are tissues at home (in a normal location for their various constituents).

32. False. On the contrary, an in utero insult at the beginning of the first trimester is likely to lead to cataract because this is when the lens placode is forming.

33. a. Persistent pupillary membrane is an incomplete regression of the anterior tunica vasculosa lentis. Bergmeister’s papilla is a nonpatent hyaloid artery remnant extending from the optic disc. If this structure is patent, it is called persistent hyaloid artery. Mittendorf’s dot is an opacity on the inferonasal posterior lens capsule from connective tissue associated with the central hyaloid vasculature and posterior tunica vasculosa.

34. a. Eyelid hemangiomas can have further implications such as astigmatism and amblyopia. Conjunctival dermoids are choristomas, not hamartomas (see answer 31). Dermoid cysts of the eyelids have pilosebaceous units in their walls, whereas epidermoid cysts do not. Conjunctival dermoids are solid tumors of
the conjunctiva. Dermolipomas can have significant morbidity associated with attempts at surgical removal including resultant ptosis, dry eye, and lateral rectus paresis.

35. True. Although highly variable, the basic features of Peters' anomaly are central corneal opacification associated with thinning or absence of Descemet's membrane. Iris stroma or lens may attach to this defect in cornea. Bilateral Peters' anomaly (approximately two thirds of all cases), when associated with microphthalmia and reddish linear skin lesions, can be part of a syndrome that can be fatal secondary to cardiac arrhythmias.

36. c. Most congenital ectopia lentis is associated with homocystinuria and Marfan's syndrome. However, numerous other conditions—Peters' anomaly, Weill-Marchesani syndrome, hyperlysinemia, sulfite oxidase deficiency, oxycephaly, aniridia, coloboma, congenital glaucoma, and megalocornea—also can be associated with ectopia lentis.

37. d. Five diseases characterized by disseminated hamartomas are known as phakomatoses—von Hippel-Lindau disease (angiomatosis retinae), Sturge-Weber syndrome (encephalotrigeminal angiomatosis), neurofibromatosis (von Recklinghausen's disease), tuberous sclerosis (Bourneville's disease), and ataxia–telangiectasia (Louis-Bar syndrome). Of these, only ataxia–telangiectasia is inherited on an autosomal recessive basis. Sturge-Weber syndrome is sporadic. The others are autosomal dominant.

38. c. Juvenile glaucoma has been reported with Sturge-Weber syndrome and neurofibromatosis. (Rubeotic glaucoma may be seen as a late sequela of the von Hippel-Lindau syndrome.)

39. e. All of these may have fundus findings. von Hippel-Lindau syndrome features hemangioblastoma of the retina. Sturge-Weber syndrome is associated with diffuse and focal choroidal hemangiomas (e.g., tomato ketchup fundus), and ataxia–telangiectasia with retinal vascular telangiectasias. Neurofibromatosis (and tuberous sclerosis) patients may have astrocytic hamartomas in the retina or optic nerve head.

40. b. In persistent fetal vasculature (PFV, also known as persistent hyperplastic primary vitreous, or PHPV), the fibrovascular proliferation behind the lens is associated with a small eye, and angle-closure glaucoma is common. In addition, cataract and retinal detachment (RD) can occur, and macular and/or optic nerve hypoplasia usually portends a poor visual prognosis.

41. e. Retinal dysplasia is an abnormal proliferation of the developing retina and can occur in all of the entities listed.

42. c. Foveal hypoplasia is associated with both aniridia and albinism. Foveal hypoplasia can also be inherited (possibly because of a defect in the \( PAX6 \) gene). The \( PAX6 \) gene is a transcription factor associated with many ocular abnormalities (including aniridia) found on chromosome 11.

43. e. Turner's syndrome is associated with anterior axial embryonic cataracts. Lowe's syndrome (also known as \( oculocerebrorenal \) syndrome) is a rare X-linked recessive syndrome causing bilateral congenital cataracts, hypotonia, aminoaciduria, and chronic renal failure.

44. e. The amount of, duration of, and stage of pregnancy for teratogenic alcohol exposure have not been defined. Other ocular features of this syndrome include blepharophimosis, epicanthal skin folds, strabismus, and ptosis.

45. True. One of the causes of orbital cellulitis is extension of infection from adjacent tissue, including pre-septal cellulitis. With a normal orbital septum (i.e., no prior surgery or trauma), this should only occur with delayed or inappropriate treatment.

46. False. The location of the amyloid deposit is important because conjunctival nodular amyloid is usually a local phenomenon, whereas subcutaneous amyloid suggests a systemic amyloidosis.

47. c. These benign lesions are caused by collections of subepithelial cells containing cholesterol and other lipids (called "foam cells" because of their appearance in histologic preparations). Most patients with xanthelasma do not have a systemic hyperlipidemia.

48. c. Juvenile xanthogranulomas (JXGs) are small, benign, histiocytic tumors that are usually multiple and can be found in the iris. They are highly vascular, which, along with the lipid content, contributes to their characteristic orange tint. Their rich vascularity also accounts for a propensity for spontaneous ecchymosis (when in the skin) or hyphema (when in the iris).

49. d. Epidermoid and dermoid cysts are both filled with dense secretions, mostly keratin, and thus do not transilluminate like clear, fluid-filled eccrine or apocrine hidrocystomas. Although epidermoid and dermoid cysts both contain keratinizing squamous epithelial linings, dermoid cysts also include other (dermal) structures such as hair follicles and sebaceous glands. Goldenhar's syndrome is associated with epibulbar, solid dermoids.

50. d. Besides thickening of the keratin layer, parakeratosis also implies retention of nuclei in the keratin layer. Hyperkeratosis is a thickening of the keratin layer, not the prickle cell layer (the latter is known as \( acanthosis \)). Acantholyosis occurs in the prickle cell layer with breakdown of the desmosomal intercellular connections, which gives it its name and characteristic histologic appearance.

51. True. Hematoxylin-eosin (H & E) staining of ultraviolet light–damaged collagen may reveal a characteristic bluish tint (basophilia) instead of the customary red (eosinophilia). This reflects a chemical alteration in the
collagen structure, referred to as elastosis, because of its histologic similarity to elastin.

52. False. Seborrheic keratosis is a benign epidermal lesion featuring thickening of the prickle cell layer (acanthosis) and keratin layer (hyperkeratosis). Actinic keratosis is considered a premalignant lesion for squamous cell carcinoma. Actinic keratosis has features that may include hyperkeratosis, elastosis, and dysplasia. Although seborrheic keratosis is often pigmented, this is not a distinguishing feature.

53. c. Seborrheic keratoses have a characteristic "stuck-on" superficial look that should help prevent them from being mistaken for carcinoma. The others can all easily mimic squamous cell carcinoma in clinical or histologic appearance. The sudden onset of multiple seborrheic keratoses, known as the Leser-Trélat sign, can be associated with malignancy (usually gastrointestinal [GI] adenocarcinoma).

54. a. Basal cell carcinoma is the most common malignant tumor of the eyelid (approximately 90% of all malignant eyelid tumors). Medial canthal tumors carry a worse prognosis due to their tendency to infiltrate more deeply (especially if the lacrimal drainage system or sinuses become involved). The morpheaform variant can often infiltrate into the orbit and does not always present as a classic "rodent ulcer."

55. c. Fat staining is often needed to help conclusively identify sebaceous cell carcinoma because the spreading pattern can easily mimic epithelial dysplasia and carcinoma in situ, as well as basal cell or squamous cell carcinoma. The presence of lipid within the tumor cell cytoplasm using stains such as oil red O or Sudan black is diagnostic.

56. c. Elevation is the result of migration of nevus cells into the dermis. Junctional nevus are flat.

57. d. Most (98%) of primary central nervous system lymphomas (PCNSLs) are non-Hodgkin’s B-cell lymphomas. 25% of PCNSLs have ocular involvement (and 60% of these have ocular and central nervous system [CNS] involvement, 15% may have ocular involvement alone, and 15% have visceral involvement alone). Many of these patients have vitritis with anterior chamber spillover and are mistakenly treated with systemic corticosteroids. This may result in short-term improvement of uveitis, but the effect is not long-lasting, and the uveitis tends to recur.

58. d. Dermal amyloid infiltration is an ominous herald of systemic amyloidosis that can be associated with multiple myeloma.

59. a. Conjunctival lymphoid proliferations, both benign and malignant, occur most commonly in the fornices. Approximately 63% to 70% (two thirds) of eyelid lymphomas will be associated with systemic lymphoma. Most conjunctival lymphomas derive from monoclonal B cells.

60. d. Hematogenous metastasis of conjunctival squamous cell carcinoma is rare. Spindle cell and mucoepidermoid carcinomas are forms of squamous cell carcinoma with the potential for intraocular invasion.

61. False. Ocular melanocytosis is more common in whites; oculodermal melanocytosis (nevus of Ota) is essentially the same condition, but with abnormally increased pigmentation of the eyelid skin as well. It is more often found in darker-skinned races. There is an increased risk of uveal, not conjunctival, melanoma in whites with oculodermal melanocytosis.

62. False. The junctional nevus is felt to represent a stage in the maturation of conjunctival nevi that is not commonly seen in histopathologic specimens. It is likely that most progressed to become subepithelial nevi by the age of 30 years. The diagnosis of junctional nevi in individuals older than 30 years should raise suspicion of primary acquired melanosis (PAM) of the conjunctiva.

63. b. Primary acquired melanosis of the conjunctiva is felt to be a premalignant condition only in cases with melanocytic atypia. They do occur mostly in people of middle age and change size or grow slowly over many years. Pagetoid spread and complete replacement of the epithelium are predictors of progression to invasive melanoma.

64. c. Two thirds of conjunctival melanomas arise from preexistent primary acquired melanosis (PAM) with atypia, and the overall mortality rate is about 25%. Pagetoid spread, large depth of tumor penetration (>0.75 mm), orbital or scleral invasion, and involvement of the eyelid skin margin are poor prognostic factors. Unlike uveal melanoma, conjunctival melanoma usually metastasizes to the preauricular and submandibular nodes first.

65. c. The cellular elements present in pyogenic granuloma include fibroblasts and endothelial cells. Kaposi’s sarcoma is a neoplastic proliferation of endothelium in a background of spindle cells. Benign areas within the tumor can appear similar to a pyogenic granuloma.

66. a. Interstitial keratitis (IK) is a nonulcerating inflammation of the corneal stroma leading to vascularization and scarring. Causes include congenital syphilis, herpes simplex, herpes zoster, leprosy, Lyme disease, tuberculosis, onchocerciasis, sarcoidosis (which causes a nummular keratitis), and Cogan’s syndrome (IK and vestibuloaductal dysfunction associated with polyarteritis). Acanthamoeba typically produces an ulcerative keratitis.

67. b. These rare hamartomatous lesions typically present with visual loss, strabismus, or leukocoria in children or adolescents. Colobomata, optic nerve pits, retinoschisis, and other hamartomas have been reported in association with these lesions. They also have been associated with neurofibromatosis, incontinentia pigmenti, juvenile retinoschisis, and facial abnormalities. Three features are
4. Ocular Pathology

considered essential to the diagnosis: (i) pigmentary disturbance, (ii) glial, and (iii) retinal vascular irregularities, including leakage on angiography. The lesion is usually too extensive to be successfully addressed with membrane peeling.

68. False. Calcofluor white staining will enhance identifi-

69. d. In order to diagnose Fuchs' dystrophy, corneal edema must be present. In Fuchs' dystrophy, endothelial cells are reduced in number or absent. In pseudophakic bullous keratopathy (PBK), endothelial cell may be near normal. Descemet's membrane is thickened in both disorders. Acute increases in intraocular pressure (IOP) will typically cause only epithelial edema, provided the endothelium is otherwise healthy. Intraepithelial cysts of degenerating epithelial cells, similar to map-dot-fingerprint dystrophy, can be seen in resolved or chronic corneal edema.

70. a. Granular dystrophy is a stromal dystrophy; Meesmann's dystrophy and map-dot-fingerprint dystrophy are epithelial. Reis-Bückler dystrophy is localized to Bowman's layer. The most common anterior corneal dystrophy is epithelial basement membrane dystrophy (map-dot-fingerprint dystrophy).

71. False. Three kinds of lesions are seen within the epithe-

72. b. Erosions are rare in macular and granular dystro-

73. True. Discrete, focal, white "granular" deposits in the anterior stroma are noted in granular dystrophy. The cornea between deposits is clear. In macular dystrophy, the gray-white stromal opacities have indistinct edges, and the intervening stroma is hazy.

74. True. Transmission of granular dystrophy is autosomal dominant. Macular dystrophy is inherited in an autosomal recessive pattern.

75. c. Posterior polymorphous dystrophy has a variable clinical spectrum. Findings on the posterior cornea include vesicles, stromal edema, gray lesions, bands with scalloped edges, and iridocorneal adhesions. Although variable, it is distinct from other dystrophies. Pathologically, the abnormal endothelial proliferation can be strikingly similar to that seen in the iridocorneal endothelial syndromes, as well as epithelial downgrowth.

76. b. The earliest histologic changes in keratoconus are fibrillations/breaks in the epithelial basement membrane and Bowman's layer. Corneal topography may reveal subclinical keratoconus in family members of patients with keratoconus.

77. False. The best examination technique for detecting early Kayser-Fleisher rings is gonioscopy because the deposits are first detectable in the most peripheral Descemet's membrane.

78. a. True exfoliation refers to a characteristic “scrolling” of the anterior lens capsule following excessive infrared radiation exposure (glassblowers are typically affected). It is far less common than exfoliation syndrome (pseudoxfollation).

79. e. Acute, dramatic increases of intraocular pressure (IOP) may cause degeneration and death of lens epithelial cells, forming characteristic white patches under the anterior capsule (known as glaukomflecken). Chronic anterior uveitis may cause degeneration and necrosis of the anterior lens epithelium. Ionizing radiation, chronic inflammation, or prolonged corticosteroid use also may cause epithelial hypertrophy (bladder or Wedl's cells) leading to posterior subcapsular cataract. Posterior subcapsular cataract is also a variable feature of atopic dermatitis.

80. False. Morgagnian globules, or cosinophilic globules in slitlike spaces between lens fibers, are considered a reliable sign of cortical degeneration. Empty fissures within cortex are a common processing artifact.

81. e. The lens is typically microspherophakic with re-

82. a. The retinal circulation supplies the nerve fiber layer, the ganglion cell layer, the inner plexiform layer, and the inner one third of the inner nuclear layer. The choroidal circulation supplies the remainder of the neurosensory retina and retinal pigment epithelium (RPE).

83. False. The clinical macula is a circular region one to two disc diameters centered around the foveola. The histologic macula is the central region of the retina with a ganglion cell layer that is more than one cell thick.

84. a. There are three main features to explain the dark appearance of the fovea on fluorescein angiography (FA). First, the retinal pigment epithelium (RPE) cells are taller, narrower, and filled with greater numbers of more pigmented melanosomes. Second, yellow xantho-

85. False. During fixation of normal infant eyes, the sclera shrinks considerably. This leads to redundancy of the retina, manifested as Lange's fold.
115

Answers

86. c.
87. d.
88. a.
89. d. Absolute alcohol preserves the crystalline deposits for microscopy.
90. a. Microaneurysms are generally located in the inner nuclear layer. Diabetic retinopathy is associated with thickening of the corneal epithelial basement membrane as well.
91. True. Iris transillumination defects are characteristic in pigmentary glaucoma. Laser trabeculoplasty can help reduce intraocular pressure (IOP) in these patients.
92. c. Central retinal vein occlusion (CRVO) is clearly associated with primary open-angle glaucoma (POAG), and glaucoma is a risk factor for the development of branch retinal vein occlusion (BRVO). The superotemporal arcade (63%) is most frequently involved with BRVO.
93. c. The vitreous directly over lattice lesions is liquefied, but the vitreous at the margins of the affected retina is firmly attached. Myopia is associated with 40% of rhegmatogenous retinal detachment (RRD), whereas lattice has been found in 20% to 30% of patients with RRD.
94. c. Age-related macular degeneration is the leading cause of new blindness in the United States. Cataract is the leading cause of blindness worldwide.
95. e. Types of drusen include small (<63 μm), intermediate (63 to 125 μm), large (>125μm), hard or soft, basal laminar, and calcific. The histopathology represents periodic acid-Schiff (PAS)-positive areas between Bruch's membrane and retinal pigment epithelium (RPE).
96. False. Occlusion of the choriocapillaris (outer retinal circulation) causes focal, sharply demarcated areas of retinal pigment epithelium (RPE) and retinal atrophy recognized as "paving stones" or "cobblestones."
97. False. Typical peripheral cystoid degeneration (TPCD) is a universal finding in patients older than 20 years. It does not increase the risk of rhegmatogenous retinal detachment (RRD) and consists of cystic spaces within the outer plexiform layer. The coalescence of TPCD cystic spaces leads to typical degenerative retinoschisis. Reticular PCD is less common and consists of cystic spaces within the nerve fiber layer.
98. a. The "retinoblastoma gene" is a tumor suppressor gene; that is, the normal gene suppresses the development of retinoblastoma. If both suppressors are lost, neoplasms develop. In "familial retinoblastoma," the patient inherits one defective gene from a parent, which is present throughout all cell lines. Subsequent somatic mutations inactivate the fellow (normal) gene and lead to tumor development. When new germ cell mutations occur early in embryogenesis, the gene will be distributed throughout all cell lines. This explains bilateral retinoblastoma in patients with no family history. This situation mimics familial retinoblastoma, but there is no family history (a new mutation).
99. a. Flexner-Wintersteiner rosettes have cells surrounding a central lumen. They are a characteristic feature of retinoblastomas (but have also been observed in some pinealoblastomas). Homer-Wright rosettes are seen in neuroblastoma (primary tumors only), cerebellar medulloblastoma, and medullopithelioma, as well as retinoblastoma. The lumen of Homer-Wright rosettes is filled with eosinophilic cytoplasmic processes.
100. c. Direct extension into the central nervous system is, by far, the most common route of spread. Hematogenous dissemination also may be seen. Extraocular extension, either transscleral or intraocular, is the most important risk factor for mortality.
101. c. Medulloepitheliomas (diktyomas) probably arise from primitive neuroepithelium, generally along the line of closure of the embryonic ocular fissure. Mucinous cysts are common.
102. c. The two independent prognostic factors seem to be basal diameter and cell type. Other features, particularly tumor location, have been shown to correlate, less predictably, with survival. Spindle cell choroidal melanoma has the best prognosis. Epithelioid choroidal melanoma has the worst prognosis.
103. b. In adults, the choroids are the most common site of ocular/periocular metastasis. In children, the orbit is more commonly involved.
104. False. The pleomorphic cellular infiltrate seen in idiopathic myositis may include neutrophils, eosinophils (particularly in children), and even giant cells, in addition to plasma cells and lymphocytes.
105. True. Although there have been some studies that have demonstrated orbital lymphatic vessels in the dura mater of the optic nerve and in the lacrimal gland, lymph nodes are not normally present within the orbit. This often makes the classification of lymphomas as well as the distinction between benign proliferations and lymphomas difficult.
106. True. Most are B-cell lymphomas of the non-Hodgkin's type.
107. e. The location of the tumor is the most powerful prognostic indicator of biologic behavior. Preseptal lid infiltrates are thought to signal a higher probability of systemic lymphoma than either orbital (intermediate-risk) or conjunctival (lowest risk) lymphomas. It is now known that monoclonality is not definitely predictive of progression to systemic disease. Both benign reactive hyperplasia and malignant lymphoma may show a follicular growth pattern. Tumor size and the degree of cellular atypia are not effective for prognostication.
108. False. Secondary orbital neoplasms (local extension or metastasis) are far more common than primary neoplasms.
109. c. Cavernous hemangiomas are the most common benign neoplasm of the orbit in adults.

110. c. Rhabdomyosarcoma and Wilms' tumor are the two most common primary solid malignancies in children. Typically, there is sudden and rapidly progressive proptosis that requires emergency treatment. The three histologic variants of rhabdomyosarcoma include the embryonal, which is the most common type; the alveolar; and the rare adult-differentiated pleomorphic type (which typically has the best prognosis). The alveolar type has the least favorable prognosis. Spontaneous lid ecchymoses are seen in rhabdomyosarcoma but are not specific for this condition, as they also may occur in metastatic neuroblastoma and Ewing's sarcoma.

111. c. Neurofibromas are not encapsulated but may be circumscribed. Both neurofibromas and schwannomas (neurilemoma) are found in increased frequency in patients with neurofibromatosis type 1. Isolated neurofibromas do not necessarily indicate systemic involvement. Verocay bodies are groups of cells that resemble sensory corpuscles. The plexiform neurofibroma is the classic type seen in von Recklinghausen’s disease.

112. c. For the general ophthalmologist, approximately 20% of lacrimal gland lesions are epithelial cell tumors and 80% are lymphoid or inflammatory lesions. Benign mixed tumor (pleomorphic adenoma) constitutes approximately 50% of epithelial tumors of the lacrimal gland. Although the pleomorphic adenoma may appear encapsulated, microscopic lobules of tumor may prolapse through the capsule. Furthermore, incomplete excision is clearly associated with multiple recurrences and malignant degeneration. Extraction should be performed with a rim of normal orbital tissue instead of shelling it out.

113. False. Melanocytoma of the optic nerve head is a benign, deeply pigmented melanocytic tumor but has been associated with adjacent choroidal melanoma in rare cases.

114. b. These tumors most often present in the first decade of life. Rosenthal fibers are enlarged, eosinophilic cell processes and are not unique to optic nerve gliomas. Most, if not all, visual pathway gliomas are of the relatively indolent pilocytic variety.

115. c. Most orbital meningiomas arise from arachnoid cells and are secondary (extensions of intracranial meningiomas). The mean age of presentation of primary optic nerve sheath meningioma is lower than in the secondary type (20% are seen at younger than 10 years). The meningothelial cell type is the most common form of orbital meningioma. Pediatric meningiomas can be particularly aggressive, with rapid infiltration of the central nervous system. Patients with primary orbital meningioma tend to survive longer than patients with secondary orbital meningioma (up to 19 vs. 15 years).

116. d. Disc pallor is thought to correspond to limited reorganization and proliferation of glial cells and not to decreased vascularity. Although changes in the optic nerve head may be diagnostic of glaucoma, they often convey no information as to the etiology of the specific mechanism of glaucoma. Damage from glaucoma to the optic nerve head is manifested in the retina as atrophy of both the nerve fiber and ganglion cell layers. Cavernous (Schnabel's) optic atrophy is also seen occasionally in nonglaucomatous eyes (e.g., patients with arteriosclerosis) and consists of cystoid spaces posterior to the lamina cribrosa.

117. d. Excessive pigment in the trabecular meshwork (TM) may be seen in the pigment dispersion syndrome and the pseudoexfoliative syndrome, as well as after surgery, uveitis, trauma, or uveal melanoma. Pseudoexfoliation glaucoma is characterized by its asymmetry. Pseudoexfoliative material is thought to be closely associated with elements of the elastic system (oxytalan and small elastic fibers) and has been detected within the conjunctival substantia propria. Although pseudoexfoliative material is found adjacent to the lens epithelium and on the lens capsule, it continues to be deposited after cataract extraction.

118. True. Angle recession is a tear between the longitudinal and circular muscles of the ciliary body. Approximately 5% to 10% of patients with >180 degrees of angle recession will eventually develop glaucoma.

119. True. The iridocorneal endothelial (ICE) syndromes are all characterized by abnormal proliferation of corneal endothelium affecting the cornea, angle structures, and the iris. ICE is generally unilateral and is seen in middle-aged adults.

120. c. Acute angle-closure glaucoma can result in lens epithelial necrosis (glaukomfläcken), sectoral iris ischemia with atrophy, and peripheral anterior synechiae (PAS) after iridotomy. If the fellow eye has a narrow angle, this suggests primary angle closure, because the condition is typically symmetric.

121. c. The combination of sebaceous adenomas (or keratoacanthomas) and gastrointestinal malignancy is Muir-Torre syndrome. Trichilemmoma may be associated with breast, thyroid, or gastrointestinal (GI) neoplasms, both benign and malignant. This is known as Cowden's disease (multiple hamartoma syndrome), an autosomal-dominant condition with variable expression that is associated with a mutation in the PTEN gene on chromosome arm 10q. Multiple basal cell carcinomas of the eyelid can be a part of basal cell nevus syndrome, which is associated with fibrosarcoma and medulloblastoma.

122. c. Medulloepithelioma may show either benign or malignant histologic characteristics. Even when cytologically malignant, this tumor is rarely metastatic.
Answers

123. **b.** Pigmented choroidal lesions <10 mm in greatest diameter and <1 mm in thickness are nearly always benign. Tumors >3 mm in height are nearly always melanomas. Overlying retinal pigment epithelium (RPE) degeneration and drusen are frequently associated with choroidal nevi, although these may be seen in melanoma and are not helpful diagnostically. An associated serous retinal detachment is common with choroidal melanoma but may be seen (rarely) with nevi.

124. **d.** There are no fluorescein angiographic signs that are pathognomonic for choroidal melanoma (or for any intraocular tumor, for that matter). Subretinal hemorrhage (most frequently from choroidal neovascularization) can be differentiated from melanoma by the absence of late hyperfluorescence characteristic of melanoma.

125. **True.** A-scan ultrasonography of choroidal melanoma shows low internal reflectivity, particularly compared with choroidal hemangioma, which has strikingly high internal reflectivity.

126. **c.** Mortality from systemic metastasis of choroidal melanoma peaks during the second year after diagnosis. The 5-year mortality rate for a large choroidal melanoma is as high as 30%.

127. **a.** The liver is the most common site of metastasis of uveal melanoma.

128. **b.** Sturge-Weber syndrome is a sporadic disease associated with a distinctive, diffuse choroidal hemangioma, which causes a reddish, thickened appearance of large areas of the fundus that has been likened to tomato ketchup. Glaucoma and retinal detachment can occur in eyes with a tomato ketchup fundus.

129. **b.** Choroidal osteomas are benign bony tumors that are more common in the peripapillary region of young women. Approximately 25% are bilateral. Ultrasonography demonstrates a highly reflective surface due to the presence of a bony plate, which also results in the loss of normal orbital echoes behind the tumor.

130. **e.** Approximately 94% of retinoblastoma cases are sporadic. About 15% of these cases represent mutations involving the germ cell line, and patients are therefore capable of transmitting the gene to offspring. A patient with unilateral involvement and no family history of retinoblastoma is very likely a sporadic case, and the likelihood that the parents will have another affected child is only about 1%. The inherited form of retinoblastoma is transmitted as an autosomal-dominant trait with incomplete penetrance (about 80% to 90%) according to pedigree analysis.

131. **True.** Children with inherited retinoblastoma are at increased risk of other malignancies, most commonly osteogenic sarcoma. Other associated malignancies include rhabdomyosarcoma, brain tumors, fibrosarcoma, and other soft-tissue sarcomas.

132. **b.** The most common presenting signs in retinoblastoma are leukocoria and strabismus. Rare presenting signs include iris heterochromia, spontaneous hyphema, and fixed pupil. Proptosis generally occurs only in advanced disease.

133. **a.** Retinoblastoma can metastasize either hematogenously or, in advanced cases, via conjunctival lymphatics. The lung is not a common site of metastasis of retinoblastoma except by extension of tumor from an adjacent bony metastasis.

134. **False.** The Reese-Ellsworth classification provides prognostic information about visual outcome following therapy for retinoblastoma. It does not provide prognostic information about survival.

135. **d.** Retinal capillary hemangiomatosis associated with cerebellar hemangioblastoma is termed von Hippel-Lindau syndrome. Other lesions, including renal cell carcinoma and pheochromocytoma, are associated but are not a part of the definition of the syndrome. Multiple retinal angiosmas without cerebellar tumors are referred to as von Hippel’s disease.

136. **e.** Autopsy studies have shown that up to 10% of patients with known metastatic disease will have ocular or orbital involvement. Orbital metastases are much less common than ocular metastases at autopsy. As noted, gastrointestinal primary tumors are a relatively infrequent source of metastasis to the eye and orbit, although metastases of primary colon cancer have been reported.

137. **c.** The choroid is the typical site of involvement by metastatic tumors. The retina is rarely the first ocular tissue involved. Because of the different branching patterns of the right and left carotid system, the left eye may be slightly more susceptible to metastasis (several studies have reported a ratio of about 1.5:1 of left-sided to right-sided ocular metastasis). Breast metastases typically present months or years after treatment of the primary tumor (90% of patients with uveal metastases have a known history of breast cancer). Metastasis is more common than choroidal melanoma, the most common primary intraocular malignancy.

138. **b.** Perhaps the most provocative finding in the Shields et al. review of clinical behavior (growth and metastases) of small (<3.0 mm thick) melanocytic lesions is that thickness greater than as little as 1.0 mm does increase the risk of metastasis.

139. **e.** Note that growth is a risk factor for metastasis, whereas subretinal fluid and orange pigment are risks for growth (but not independent risks for metastasis). The absence of retinal pigment epithelium (RPE) changes is a risk factor for tumor growth.

140. **c.** The lens is the most radiosensitive component of the eye, followed by the cornea, retina, and optic nerve.
141. c. Human papillomavirus (HPV) subtype 16 has been detected in adult eyelid papillomas. HPV subtype 6 has been detected in children with eyelid papillomas.

142. b. This is a benign lesion that demonstrates hyperkeratosis, as well as pseudohorn cysts, which are invaginations of epidermis.

143. b. This is a classic picture of a syringoma, a benign tumor of hair follicle involvement. The lesion shown is a benign tumor of hair follicle origin, which is removed with simple surgical excision. One can observe numerous “basaloid islands” with hair follicle involvement.

144. d. The lesion shown is a benign tumor of hair follicle origin, which is removed with simple surgical excision. One can observe numerous “basaloid islands” with hair follicle involvement.

145. c. Capillary hemangiomas are one of the most common benign tumors of childhood, usually involving the upper eyelid. Up to 33% of patients can develop strabismus, and amblyopia is always a concern in these infants. Intralesional corticosteroids can be used to treat capillary hemangiomas but can have side effects, including eyelid necrosis and subcutaneous fat atrophy.

146. a. The leftmost figure represents a capillary hemangioma. The middle figure represents a cavernous hemangioma of the eyelid.

147. d. Molluscum contagiosum virus causes the molluscum lesions shown in the figure. It is a double-stranded deoxyribonucleic acid (DNA) virus from the poxvirus family. Adult patients with acquired immunodeficiency syndrome (AIDS) can present with multiple molluscum lesions.

148. d. The lesion shown is a xanthogranuloma. It is characterized by the presence of Touton’s giant cells (shown in the figure on the right). Patients with iris lesions secondary to juvenile xanthogranulomas are at risk for spontaneous hyphema and consequent secondary glaucoma.

149. c. The patient has canaliculitis, which is often caused by Actinomyces israelii, a filamentous Gram-positive rod (as shown in the figure). It is usually susceptible to penicillins and cephalosporins.

150. a. The lesion shown is primary acquired melanosis (PAM) without atypia. Note that atypia is not determined clinically, but rather it is determined with histologic examination (as shown in the figure on the right). PAM without atypia does not progress to conjunctival melanoma.

151. c. The lesion shown is primary acquired melanosis (PAM) with atypia. Approximately 50% of PAMs with atypia progress to conjunctival melanoma.

152. b. This patient has herniated orbital fat, a common condition that occurs with weakening of the aging orbital septum. The condition is benign.

153. c. This is a classic case of spheroidal degeneration (also known as corneal elastosis or climactic droplet keratopathy). Histopathologic examination reveals the presence of basophilic globules within the stroma, as seen in the figure on the right.

154. c. The patient has Reis-Bucklers dystrophy, which is autosomal dominant and affects Bowman’s layer. Histopathologically, one can observe disruption of Bowman’s layer in an undulating fashion. Recurrent erosions and graft recurrence are common.

155. a. Granular dystrophy is abnormal deposition of hyaline material within the stroma and is best observed with Masson’s trichrome stain. Figures b. and e. represent lattice dystrophy. Figures c. and d. represent macular dystrophy.

156. b. Lattice dystrophy can be associated with the Meretoja’s syndrome (type II lattice dystrophy), which is characterized by cranial nerve palsy and lax skin secondary to amyloid deposition.

157. a. Macular dystrophy is an autosomal-recessive condition, as pictured. It is characterized by deposition of mucopolysaccharide material in the corneal stroma.

158. b. 

159. d. The figure depicts ectopia lentis, which is associated with Marfan’s syndrome, homocystinuria, sulfite oxidase deficiency, hyperlysinemia, and other medical conditions.

160. d. A Soemmerring’s ring is shown, representing incomplete removal of cortex and/or nuclear fibers leading to a doughnut-shaped configuration of the lens.

161. b. The lesion shown is a juvenile xanthogranuloma (JXG) and puts the patient at risk for spontaneous hyphema.

162. True. Touton’s giant cells are characteristic for juvenile xanthogranuloma (JXG).

163. d. The figure demonstrates acid-fast bacilli (white arrow).

164. c. Ziehl–Neelsen staining is used to detect acid-fast bacilli such as tuberculosis.

165. a. The figure demonstrates ruberosis iridis. These patients are at risk for neovascular glaucoma.

166. b. The figure is a histopathologic section of an eye with phthisis bulbi. Phthisis bulbi is characterized by atrophy, a thickened sclera, shrinkage of the eye, and disorganization. Eyes with phthisis bulbi are not necessarily with no light perception (NLP).

167. d. The figure demonstrates aniridia. Two thirds of cases are familial and are inherited in an autosomal-dominant fashion with complete penetrance and variable expressivity of a mutation in the PAX6 gene on chromosome 11p13. Sporadic cases are associated with the WAGR complex—Wilms’ tumor, aniridia, genitourinary malformations, and mental retardation.

168. d. The patient has von Hippel’s disease, or a retinal hemangioblastoma. Note the vascular engorgement of the vessels from the lesion. The figure demonstrates the classic histopathology for a hemangioblastoma. If there is a cerebellar hemangioblastoma
present, then the patient would have von Hippel-Lindau syndrome.

169. d. von Hippel-Lindau disease is an autosomal-dominant condition that can manifest with benign and malignant tumors, including retinal and cerebellar hemangioblastomas, pheochromocytomas, and renal cell carcinomas.

170. e. Patients with retinal hemangioblastomas can undergo all of the therapies listed. Cryotherapy and laser photocoagulation put the patient at risk for exudative retinal detachments. In 2002, Aiello et al. described a case of the use of a vascular endothelial growth factor (VEGF) receptor inhibitor in the successful treatment of this condition.

171. e. The figure demonstrates reticular degenerative retinoschisis. The layer of pathology is in the nerve fiber layer.

172. c. The figure demonstrates typical degenerative retinoschisis. The layer of pathology is in the outer plexiform layer.

173. c. The figure demonstrates a full-thickness macular hole. Macular hole pathogenesis is thought to be secondary to tangential and anteroposterior vitreomacular traction.

174. a. 5, b. 1, c. 3, d. 4, e. 3, f. 2. Note that macrophages can differentiate into both epithelioid and giant cells. Also note that mast cells are tissue-based basophils (i.e., not circulating).

175. a. 3, b. 4, c. 1, d. 4, e. 1, f. 3, g. 4, h. 1. Touton’s giant cells feature vacuolated cytoplasm outside a nuclear ring with eosinophilic cytoplasm inside. Juvenile xanthogranuloma, Erdheim-Chester disease, and necrobiotic xanthogranuloma feature Touton’s giant cells. Langhans’ giant cells have a peripheral, horseshoe-shaped nuclear rim and are characteristic of sarcoidosis and mycobacterial disease. Foreign body giant cells have haphazardly distributed nuclei. Note that Langerhans’ cells are dendritic macrophages (antigen-presenting cells) within epithelia.

176. a. 4, b. 1, c. 5, d. 3, e. 2.

177. a. 2, b. 5, c. 4, d. 1, e. 3. Note that Axenfeld-Rieger syndrome is a spectrum of peripheral developmental anterior segment dysgeneses, and that there is considerable overlap between these rigid definitions, which are being used less frequently.

178. a. 3, b. 4, c. 1, 2, d. 2, e. 5. Although not as common as in tuberous sclerosis, astrocytic hamartoma of the retina and optic disc can be observed in neurofibromatosis.

179. a. 3, b. 2, 5, c. 2, d. 1, 4, e. 4.

180. a. 3, b. 1, c. 4, d. 2. Each of these iron lines represents basal epithelial uptake of iron.

Suggested Readings


1. T or F The photostress recovery test may be useful in an eye with decreased vision to differentiate between macular and optic nerve disease.

2. Which one of the following statements about the visual evoked response (VER) is false?
   a. The VER is an electrical signal that must be extracted from the simultaneously generated electroencephalogram (EEG).
   b. The stimulus may consist of either a flash of white light or a pattern, presented either transiently or continuously by pattern reversal.
   c. The two crucial parameters used for functional evaluation include the height of the first positive or upward wave (amplitude) and the time between stimulus presentation and the appearance of this wave (latency).
   d. VER is not useful for distinguishing optic neuropathy from retinal disorders.
   e. Uses of the flash VER include visual acuity assessment in preverbal children, assessment of optic nerve function in suspected multiple sclerosis (MS), and reliable establishment of factitious visual loss.

3. A patient presents with a complaint of decreased vision in the left eye. His acuities are 20/20 in the right eye and 20/100 in the left eye. The examination is entirely unremarkable, and the diagnosis of factitious visual loss is considered. Tests that would be particularly useful in establishing this diagnosis include:
   1. optokinetic nystagmus (OKN) testing.
   2. gently rocking a large mirror in front of the patient, with the good eye occluded.
   3. introducing a prism base-up in front of the left eye when the patient is reading binocularly.
   4. performing a fogging refraction.

4. A patient presents with a complaint of decreased vision in the left eye. His acuities are 20/20 in the right eye and light perception in the left eye. The examination is entirely unremarkable, and the diagnosis of factitious visual loss appears in the differential. Tests that would be particularly useful in establishing this diagnosis include:
   1. optokinetic nystagmus (OKN) testing.
   2. gently rocking a large mirror in front of the patient with the good eye occluded.
   3. introducing a prism base-up in front of the left eye when the patient is reading binocularly.
   4. performing a fogging refraction.
Questions

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

5. Lesions affecting the optic tract may be associated with:
   1. unilateral decreased visual acuity.
   2. ipsilateral afferent pupillary defect (APD).
   3. contralateral APD.
   4. homonymous hemianopia.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

6. **T** or **F** Impairment of oculovestibular caloric reflexes suggests supranuclear disturbance of eye movements.

7. **T** or **F** In the setting of an upgaze paresis, up-turning of the eyes on forceful opening of closed eyelids implies a supranuclear lesion.

8. Which of the following statements about magnetic resonance imaging (MRI) is/are true?
   1. Short repetition times (TR) and short echo times (TE) are used to generate T1-weighted images.
   2. On a T1-weighted image, vitreous humor is dark, and on a T2-weighted image, vitreous humor is bright.
   3. Air and cortical bone give a dark (hypointense) signal on MRI.
   4. T1-weighted images tend to show the anatomy well, whereas T2-weighted images tend to show the pathology well.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

9. Which one of the following statements about the intracranial portion of the optic nerve is false?
   a. The intracranial portion of the optic nerve is typically 10 to 17 mm in length.
   b. It enters the intracranial cavity inferior to the frontal lobe and anterior cerebral artery.
   c. It enters the intracranial cavity medial to the internal carotid artery.
   d. Once it enters the intracranial cavity, the optic nerve no longer has a sheath.
   e. There is generally some redundancy within the intracranial optic nerve.

10. Which of the following statements about the optic chiasm is/are true?
    1. As many fibers cross as do not cross.
    2. The posterior portion of the chiasm has a high density of macular fibers.
    3. The chiasm typically lies 1 mm above the anterior pituitary gland.
    4. The inferior fibers are the first to cross.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

11. **T** or **F** Segregation of retinal ganglion cell terminals into the six layers of the lateral geniculate body (LGB) is a function of the cells’ receptive fields.

12. **T** or **F** Superior fibers of the optic radiations, carrying information from the inferior visual field, run in close relation to the internal capsule.

13. Which of the retrochiasmal location(s) shown below can induce a monocular visual field defect with a single lesion alone?
    1. lateral geniculate body (LGB).
    2. parietal lobe.
    3. temporal lobe.
    4. occipital lobe.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

14. Which of the following features is/are consistent with a temporal lobe lesion?
    1. seizures.
    2. optokinetic nystagmus (OKN) abnormalities.
    3. formed visual hallucinations.
    4. high congruity of visual field deficits.
a. 1, 2, and 3.
b. 1 and 3.
15. Which of the following features is/are consistent with a parietal lobe lesion?
   a. agnosia.
   b. right-left confusion.
   c. optokinetic nystagmus (OKN) abnormalities.
   d. homonymous hemianopia denser inferiorly.
   e. 1, 2, 3, and 4.

16. Which of the following features is/are consistent with an occipital lobe lesion?
   a. unformed hallucinations.
   b. macular sparing.
   c. sparing of the temporal crescent.
   d. optokinetic nystagmus (OKN) abnormalities.
   e. 1, 2, 3, and 4.

17. Causative events in the pathophysiology of optic disc edema include:
   a. swollen axons.
   b. extracellular fluid accumulation.
   c. interruption of axonal transport.
   d. breakdown of the blood-retinal barrier.
   e. 1, 2, 3, and 4.

18. Which of the following statements about papilledema is/are true?
   a. Loss of venous pulsations is a particularly specific finding.
   b. Symptoms accompanying papilledema may include visual loss and diplopia.
   c. The most typical visual field finding in chronic papilledema is an enlarged blind spot.
   d. Papilledema may be unilateral.

19. Which of the following is/are universal findings in patients with pseudotumor cerebri?
   a. papilledema.
   b. increased intracranial pressure.
   c. normal neurologic examination.
   d. normal neuroimaging studies.
   e. 1, 2, 3, and 4.

20. The indications for treatment of pseudotumor cerebri include:
   a. papilledema.
   b. severe headache.
   c. obesity.
   d. visual field loss.
   e. 1, 2, 3, and 4.

21. The most common cause of permanent visual loss in patients with cavernous sinus–dural fistulae is:
   a. neovascular glaucoma.
   b. corneal exposure.
   c. open-angle glaucoma.
   d. choroidal effusions.
   e. strabismus.

22. Which of following statements about giant cell arteritis (GCA) is/are true?
   a. It is exceedingly rare in patients younger than 50 years and is more common in women.
   b. Forty percent of untreated patients will develop some form of permanent visual loss.
3. Sixty-five percent of untreated patients will have contralateral involvement after permanent visual loss in one eye.

4. In a patient with anterior ischemic optic neuropathy (AION) but no other localizing signs or symptoms, a normal erythrocyte sedimentation rate (ESR) rules out the diagnosis of GCA.

23. Which one of the following statements about nonarteritic anterior ischemic optic neuropathy (NAION) is false?
   a. Visual loss is generally less severe than in the arteritic variety.
   b. The condition most frequently associated with NAION is hypertension.
   c. Fluorescein angiography (FA) is usually not of benefit in differentiating optic neuritis from NAION.
   d. There is approximately a 15% chance of subsequent occurrence in the contralateral eye within 5 years.
   e. Aspirin has no clear effect on prophylaxis of the other eye from NAION.

24. Approximately what percentage of patients with an initial attack of optic neuritis (and no lesions observed on magnetic resonance imaging [MRI] at the time of the attack) will develop multiple sclerosis (MS) at 15-year follow-up?
   a. 1%.
   b. 5%.
   c. 8%.
   d. 15%.
   e. 100%.

25. What percentage of patients with severe multiple sclerosis (MS) will, on autopsy examination, have demyelinated lesions of the optic nerves?
   a. 10%.
   b. 25%.
   c. 50%.
   d. 75%.
   e. >90%.

26. Which of the following statements about optic nerve glioma is/are true?
   1. Optic nerve glioma is more frequently seen in children than in adults.
   2. Most patients will have associated neurofibromatosis.
   3. Presenting findings include visual loss, strabismus, proptosis, or hypothalamic abnormalities.
   4. These tumors are more likely to be aggressively malignant in children than in adults.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

27. T or F Collateral vessels at the optic disc are highly specific for optic nerve meningioma.

28. Which of the following statements about optic nerve meningiomas is/are true?
   1. Optic nerve meningiomas primarily affect children.
   2. Most patients will have associated neurofibromatosis.
   3. On computed tomography (CT) scanning, affected optic nerves have a “kinked” appearance.
   4. These tumors are more likely to be aggressively malignant in children than in adults.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

29. T or F Dominant optic atrophy (DOA) is usually slowly progressive throughout life.

30. Which of the following statements about Leber’s hereditary optic neuropathy (LHON) is/are true?
   1. All offspring of a female carrier are either affected or carriers.
   2. Ten percent of female carriers will be affected.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
3. There is generally sequential asymmetric bilateral involvement.
4. A small percentage of patients will enjoy partial or complete recovery late in their course.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

35. T or F The secreting variety of pituitary tumors is more likely to present with visual loss than the nonsecreting variety.

36. T or F Bitemporal macular hemianopia (bitemporal central scotomas) may be a false-localizing sign.

37. The most common location for a cerebral aneurysm associated with acute third nerve palsy is:
   a. the junction of posterior communicating and posterior cerebral arteries.
   b. the junction of the vertebral and superior cerebellar arteries.
   c. the junction of the posterior cerebral artery and the internal carotid artery.
   d. the junction of posterior communicating artery and the internal carotid artery.
   e. the junction of the internal carotid artery and the anterior communicating artery.

38. T or F A third nerve palsy associated with a cerebral aneurysm is always painful.

39. A 42-year-old man with diabetes presents with a painful partial third nerve palsy. On his first follow-up visit, you notice that, when looking down, his upper eyelid appears to retract or lag. This finding essentially rules out the possibility of:
   a. diabetic third nerve palsy.
   b. aneurysm.
   c. meningioma.
   d. syphilitic gumma.
   e. trauma.

40. The most common cause of acquired fourth nerve palsy in adults is:
   a. tumor.
   b. trauma.
   c. vascular disorder.
   d. idiopathic palsy.
   e. aneurysm.

41. Of the following choices, the best method for detecting a decompensated congenital fourth nerve palsy is:
   a. the three-step (the Bielschowsky head tilt) test.
   b. distance and near prismatic measurements.
Questions

c. double Maddox rod testing.
d. vertical fusional amplitude measurements.
e. the Lancaster red-green test.

42. A 35-year-old Chinese man presents with right ptosis and an abduction deficit, facial hypesthesia, and keratoconjunctivitis sicca in the right eye. This picture is highly suggestive of:
   a. meningioma.
   b. chordoma.
   c. diabetic vascular neuropathies.
   d. nasopharyngeal carcinoma.
   e. aneurysm.

43. T or F Sixth nerve palsy caused by an intracavernous lesion may present as abduction palsy plus miosis.

44. The finding that all three types of Duane’s syndrome share is:
   a. a deficit of abduction.
   b. a deficit of adduction.
   c. globe retraction with adduction.
   d. esotropia.
   e. exotropia.

45. T or F The sixth nerve is more resistant to damage within the cavernous sinus because it runs within its substance rather than within its lateral wall.

46. T or F Damage to the right frontal eye field will always result in permanent, complete inability to generate a leftward saccade.

47. Which one of the following statements about oculomotor apraxia is false?
   a. Both congenital and acquired forms of oculomotor apraxia may be observed.
   b. Pursuits are generally affected more than saccades.
   c. Horizontal movements are generally affected much more than vertical movements.
   d. In the acquired form, blinks are frequently used to break fixation.
   e. In the congenital form, children frequently use compensatory, exaggerated head turns to refixe.

48. Which of the following ocular motor disorders is most associated with malignancy?
   a. square-wave jerks.
   b. ocular flutter.
   c. opsinclonus.
   d. dysmetria.
   e. ocular bobbing.

49. The saccadic movement that is often affected in progressive supranuclear palsy (PSP) is:
   a. upward.
   b. downward.
   c. leftward.
   d. rightward.
   e. saccades are equally affected in all directions.

50. T or F The latency in generating a pursuit movement is shorter than that for a saccade movement.

51. Hypertropia associated with ipsilateral adduction weakness is most suggestive of:
   a. decompensated congenital fourth nerve palsy.
   b. dorsal midbrain syndrome.
   c. “one-and-a-half” syndrome.
   d. skew deviation with internuclear ophthalmoplegia (INO).
   e. progressive supranuclear palsy (PSP).

52. The dorsal midbrain syndrome is associated with all of the following except:
   a. upward gaze paresis.
   b. accommodative abnormalities.
   c. light-near dissociation.
   d. lid retraction.
   e. paradoxic optokinetic nystagmus (OKN).

53. Which of the following would not normally be found in congenital nystagmus?
   a. oscillopsia.
   b. normal visual acuity.
   c. paradoxic optokinetic nystagmus (OKN).
   d. amplitude dampened by convergence.
   e. amplitude increased by fixation.

54. Monocular nystagmus in a toddler raises the specter of:
   a. optic nerve meningioma.
   b. craniopharyngioma.
   c. rhabdomyosarcoma.
5. Neuro-ophthalmology

d. chiasmal glioma.
e. metastatic neuroblastoma.

55. According to Alexander’s law, in which position should upbeat nystagmus be most prominent?
a. up gaze.
b. down gaze.
c. left gaze.
d. right gaze.
e. convergence.

56. Which pattern of nystagmus is most localizing?
a. upbeat nystagmus.
b. periodic alternating nystagmus.
c. vestibular nystagmus.
d. downbeat nystagmus.
e. seesaw nystagmus.

57. Which visual field defect is most likely to be associated with seesaw nystagmus?
a. central scotoma.
b. bitemporal hemianopia.
c. incongruous hemianopia.
d. congruous hemianopia.
e. a visual field defect should not be associated with seesaw nystagmus.

58. T or F Saccades, pursuits, and vergence eye movements utilize the same pathways for eye movement generation.

59. T or F Upper motor neuron facial nerve paralysis usually leaves voluntary eyelid closure intact.

60. T or F In Parkinson’s disease, volitional eyelid movements are impaired, whereas reflex and emotional movements are intact.

61. Which one of the following statements about the facial nerve is false?
a. The sensory innervation of the anterior two thirds of the tongue terminates in the nucleus solitarius, and the motor innervation to the lacrimal gland arises, in part, from the superrior salivatory nucleus.
b. The first branch of the facial nerve is the greater superficial petrosal nerve, which synapses in the geniculate ganglion.
c. Within the fallopian canal, the facial nerve gives off a motor branch to the stapedius muscle and sensory branches for the skin behind the ear.
d. Nerves from chorda tympani synapse in the geniculate ganglion, carrying sensory innervation to the tongue and motor innervation to the salivary glands.
e. The motor nerves to the intrinsic facial muscles branch within the parotid gland.

62. T or F Marcus Gunn’s “jaw-wink” reflex is an example of aberrant regeneration.

63. Which of the following therapies has proven to be effective for nonarteritic anterior ischemic optic neuropathy (NAION)?
a. levodopa.
b. hyperbaric oxygen.
c. intravenous steroids.
d. optic nerve decompression surgery.
e. none of the above.

64. What percentage of patients with Bell’s palsy will experience complete spontaneous recovery?
a. 5%.
b. 25%.
c. <50%.
d. <75%.
e. >75%.

65. T or F The facial nerve is the most frequently involved cranial nerve in neurosarcoidosis.

66. T or F Hemifacial spasm is probably a disorder of the basal ganglia.

67. Facial myokymia in a child is frequently associated with:
a. chiasmal glioma.
b. nasopharyngeal carcinoma.
c. cerebellar hemangioblastoma.
d. pontine glioma.
e. spasmus nutans.

68. Which of the following conditions may be confused with essential blepharospasm?

1. severe dry eye.
2. retained conjunctival foreign body.
3. tardive dyskinesia.
4. hemifacial spasm.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
Questions

69. Which of the following statements about the pupillary reflex is/are true?

1. The pupil pathway terminates in pretectal nuclei after passing, without synapsing, through the lateral geniculate body (LGB).
2. The decussation at the chiasm is responsible for a normal consensual pupillary response.
3. Sympathetic pupillary fibers originate in the superior cervical ganglion, travel in the cranial vault with the internal carotid artery, and enter the orbit with the ophthalmic artery through the optic foramen.
4. The pathway for accommodative miosis enters the Edinger-Westphal nucleus anterior to the pathway for light-induced miosis.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

70. Which of the following statements about light-near dissociation is/are true?

1. A key finding in the diagnosis of Argyll Robertson’s pupils is the presence of miosis.
2. Argyll Robertson’s pupils react to light, but do not have a near response.
3. The most common etiology of the dorsal midbrain syndrome in a child younger than 10 years is a pineal gland tumor.
4. The most common etiology for the dorsal midbrain syndrome in a patient older than 60 years is multiple sclerosis (MS).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

71. Pupillary dilation may be the only sign of oculomotor nerve palsy in which of the following disorders?

1. uncal herniation.
2. diabetic microvascular disease.
3. basilar meningitis.
4. cerebral aneurysm.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

72. Which one of the following statements about Adie’s tonic pupil is false?

a. Most patients will manifest Adie’s syndrome.
b. Most patients with the syndrome will have unilateral involvement.
c. Pupillary size generally diminishes after accommodative symptoms abate.
d. Pupillary constriction in response to 0.25% pilocarpine is conclusive evidence of denervation hypersensitivity.
e. The differential diagnosis of a tonic pupil includes herpes zoster, syphilis, and giant cell arteritis (GCA).

73. Which of the following statements about Horner’s syndrome is/are true?

1. The distribution of anhidrosis may be helpful in locating the lesion.
2. Four percent cocaine will dilate the pupil of the patient with Horner’s syndrome, whereas it will leave a normal pupil unchanged.
3. The evaluation of a patient whose miotic pupil does not dilate with topical cocaine but does dilate with topical hydroxyamphetamine consists, in part, of chest x-ray and careful neurologic examination.
4. Horner’s syndrome with coincident ipsilateral headache is indicative of spontaneous carotid dissection, even in the setting of a normal carotid angiogram.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

74. After determination of which pupil is abnormal, the next step in evaluating a patient with anisocoria is:

a. the swinging flashlight test.
b. testing of extraocular movements.
c. optokinetic nystagmus (OKN) testing in the vertical direction.
d. slit-lamp examination.
e. pharmacologic testing.

75. T or F The incidence of multiple sclerosis (MS) is more common in relatives of patients with the disease because there is clearly a genetic component to the disease.

76. T or F The most common cranial mononeuropathy seen in multiple sclerosis (MS) is an isolated oculomotor palsy.

77. T or F Five percent to 10% of patients with multiple sclerosis (MS) will have findings of posterior uveitis, including pars planitis or retinal periphlebitis.

78. Pheochromocytoma may be seen as part of:
   1. neurofibromatosis.
   2. tuberous sclerosis.
   3. von Hippel–Lindau disease.
   4. ataxia-telangiectasia.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

79. Astrocytic hamartomas of the retina or optic nerve head may be seen in:
   2. neurofibromatosis.
   3. ataxia-telangiectasia.
   4. tuberous sclerosis.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

80. The triad of adenoma sebaceum, mental retardation, and seizures is considered pathognomonic for:
   a. neurofibromatosis.
b. Sturge-Weber syndrome.
c. ataxia-telangiectasia.
d. tuberous sclerosis.
e. angiomatosis retinae.

81. Seizures are often seen in patients with:
   1. neurofibromatosis.
   2. tuberous sclerosis.
   4. ataxia-telangiectasia.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

82. Infantile or juvenile glaucoma may be seen in:
   1. neurofibromatosis.
   2. tuberous sclerosis.
   4. ataxia-telangiectasia.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

83. T or F The histopathology of the retinal tumor of von Hippel’s disease is most consistent with retinal cavernous hemangioma.

84. Chronic sinopulmonary infections may be seen as part of:
   1. neurofibromatosis.
   2. tuberous sclerosis.
   3. angiomatosis retinae.
   4. ataxia-telangiectasia.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

85. Which one of the following statements about Kearns-Sayre syndrome is false?
   a. The complete syndrome has an onset of symptoms or signs before the age of 20 years.
b. The pigmentary retinopathy is generally associated with good visual function throughout life.
c. The progressive external ophthalmoplegia (PEO) that is associated with the syndrome typically presents as diplopia.
Questions

- Heart block develops late in the course of the syndrome.
- Elevated cerebrospinal fluid (CSF) protein level may predict patients at risk of developing heart block.

86. T or F The presence of polychromatic cataract (“Christmas tree” cataract) in association with progressive external ophthalmoplegia (PEO) suggests the diagnosis of Wilson’s disease.

87. The most common sign of Graves’ ophthalmopathy is:
   - lid retraction.
   - conjunctival injection over the horizontal rectus muscles.
   - superficial punctate keratitis (SPK).
   - proptosis.
   - diplopia in upgaze.

88. The second most frequently involved extraocular muscle in Graves’ ophthalmopathy is the:
   - inferior rectus.
   - lateral rectus.
   - superior rectus.
   - medial rectus.
   - inferior oblique.

89. The least frequently involved muscle in Graves’ ophthalmopathy is the:
   - inferior rectus.
   - lateral rectus.
   - superior rectus.
   - medial rectus.
   - inferior oblique.

90. What percentage of patients with myasthenia gravis (MG) present with ocular findings only?
   - 5%.
   - 10%.
   - 25%.
   - 50%.
   - 75%.

91. What percentage of patients with myasthenia gravis (MG) will develop Graves’ disease?
   - 5%.
   - 10%.
   - 15%.

92. What percentage of patients with myasthenia gravis (MG) have thymomas visible on computed tomography (CT) scan?
   - 1%.
   - 10%.
   - 25%.
   - 50%.
   - 75%.

93. For a patient to be reassured that systemic disease is unlikely, ocular myasthenia should remain localized for what length of time?
   - 3 months.
   - 6 months.
   - 1 year.
   - 2 years.
   - 5 years.

94. Animal studies show that irreversible ischemic retinal damage occurs after what duration of retinal vascular occlusion?
   - 30 minutes.
   - 50 to 60 minutes.
   - 90 to 100 minutes.
   - 3 hours.
   - 12 hours.

95. T or F The major cause of mortality in patients with embolic central retinal artery occlusion (CRAO) is massive cerebral infarction.

96. Clues to the presence of high-grade carotid stenosis include:
   1. low-grade anterior segment inflammation.
   2. cataract.
   3. hypotony.
   4. contralateral corneal arcus.
   - 1, 2, and 3.
   - 1 and 3.
   - 2 and 4.
   - 4 only.
   - 1, 2, 3, and 4.

97. Typical symptoms in vertebrobasilar insufficiency include:
   1. ataxia.
   2. hemiparesis.
3. vertigo.
4. monocular blurring or loss of vision with phosphenes.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

98. A 29-year-old woman presents to an ophthalmologist with a complaint of pain during eye movements and blurry vision in her right eye. Review of systems documents a 3-week history of paresthesias in the right lower leg approximately 6 months before the onset of her visual disturbance. The patient reports that her visual disturbance developed over a period of 2 or 3 days before presentation. Examination discloses a visual acuity of 20/60 in the right eye and 20/20 in the left eye. She is able to interpret correctly four of 11 Ishihara plates with her right eye and 10 of 11 plates with her left eye. Visual fields disclose a central scotoma in the right eye and are normal for the left eye. There is no afferent pupillary defect (APD) noted. Which one of the following is true?
   a. The patient probably has an acute maculopathy.
   b. The patient probably has factitious visual loss.
   c. The patient probably had a similar episode affect her left eye sometime in the past.
   d. The patient probably has a hereditary optic neuropathy.
   e. Oral steroid therapy is indicated.

99. Which of the following diseases has been observed in association with Coats’ disease?
   a. facioscapulohumeral muscular dystrophy.
   b. myotonic dystrophy.
   c. cerebral angiitis.
   d. cerebrovascular congophilic angiopathy.
   e. progressive external ophthalmoplegia (PEO).

100. A 14-year-old girl is brought to the ophthalmologist by her parents after complaining that “the page swims when I try to read.” The examination is normal with the exception of pronounced downbeat nystagmus. A careful review of systems documents the presence of intermittent headaches in the occipital region, which are intensified with anger or sudden head movement. The patient denies any use of prescription or illicit drugs, including alcohol. Which of the computed tomography (CT) scans shown below most likely depicts her condition?
Questions

101. Modalities likely to successfully relieve the reading difficulties of the patient described in question 100 include:
   1. clonazepam (Klonopin).
   2. carbamazepine (Tegretol).
   3. basilar craniectomy.
   4. psychotherapy.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

102. A 9-year-old boy presents to the ophthalmologist with the complaint that he has lost his position on the school basketball team because he cannot see the basket. He notes occasional morning headaches but denies any nausea or vomiting. Examination reveals visual acuity of 20/40 in the right eye and 20/25 in the left eye. The patient has marked symmetric weakness of upgaze bilaterally. His pupils are 7 mm and are poorly reactive to light, with better reaction to a near target. There is approximately 2 mm of superior scleral show bilaterally. Fundus examination suggests optic atrophy in both eyes. A review of the systems reports increased consumption of water, with frequent urination at night. The most likely diagnosis is:
   a. pinealoma.
   b. pontine glioma.
   c. cerebellar astrocytoma.
   d. chiasmal glioma.
   e. hereditary optic atrophy.

103. After neuroimaging, an important step in the diagnostic evaluation of the patient in question 102 would be:
   a. visual evoked responses (VERs).
   b. electroencephalogram (EEG).
   c. Farnsworth D-15 color vision testing.
   d. lumbar puncture.
   e. electromyograms.

104. Which of the following statements about optic neuritis in childhood is/are true?
   1. Optic neuritis is more commonly bilateral than unilateral.
   2. The visual prognosis is almost always poor.
   3. The pathophysiology is believed to be related to autoimmune demyelination.
   4. Enlargement of optic nerves on neuroimaging implies another, more ominous, diagnosis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

105. The differential diagnosis for presumed posterior ischemic optic neuropathy (PION) includes:
   1. radiation optic neuropathy.
   2. post-coronary artery bypass graft surgery.
   3. giant cell arteritis (GCA).
   4. essential hypertension.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

106. In a patient complaining of headache and transient visual obscuration whose examination reveals bilateral disc edema, the first diagnostic intervention to be undertaken is:
   a. lumbar puncture.
   b. automated perimetry.
   c. computed tomography (CT) scanning.
   d. measurement of blood pressure.
   e. measurement of sedimentation rate.

107. Which of the following disorders is clearly associated with optic nerve drusen?
   a. migraines.
   b. pseudotumor cerebri.
   c. giant cell arteritis (GCA).
   d. retinitis pigmentosa.
   e. normal tension glaucoma.

108. Which one of the following statements about anisocoria is false?
   a. Approximately 15% to 20% of the healthy adult population will have simple (physiologic) anisocoria.
   b. Anisocoria caused by Horner’s syndrome is more apparent in the dark.
5: Neuro-ophthalmology

c. Simple anisocoria, as defined by a difference in pupillary diameters, is less apparent in bright light.
d. Anisocoria caused by Adie’s tonic pupil is more apparent in bright light.
e. Simple anisocoria, as defined by a difference in pupillary areas, is less apparent in bright light.

109. Which histopathologic variety of meningioma is most commonly seen within the orbit?
   a. angioblastic.
   b. transitional.
   c. meningothelial.
   d. fibroblastic.
   e. pilocytic.

110. A meningioma arising in which location is most likely to lead to optic disc edema?
   a. sphenoid wing.
   b. olfactory groove.
   c. planum sphenoidale.
   d. optic nerve sheath.
   e. cavernous sinus.

111. T or F Heavy consumption of alcohol and cigarettes together is sufficient for the development of tobacco-alcohol amblyopia.

112. Blood tests that are important in the evaluation of a patient with bilateral optic atrophy and cecocentral scotomas include:
   1. serum B12 level.
   2. fluorescent treponemal antibody-absorptive test (FTA-ABS).
   3. serum folate level.
   4. serum cyanide level.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

114. Which of the following statements about diabetic papillopathy is/are true?
   1. Diabetic papillopathy is typically painful.
   2. Visual loss is generally severe.
   3. The papillopathy is generally followed by the development of florid neovascularization.
   4. The development of the disorder seems to be independent of the extent of blood sugar control.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

115. Modalities useful in the treatment of optic neuropathy secondary to Graves' disease include:
   1. subtotal thyroidectomy.
   2. orbital radiation.
   3. chronic (>4 weeks) oral prednisone therapy.
   4. two- or three-wall orbital decompression.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

116. Bromocriptine treatment may be indicated in the management of:
   a. a pituitary tumor that is secreting prolactin.
   b. a pituitary tumor that is secreting growth hormone.
   c. a pituitary tumor that is secreting thyrotropin.
   d. a pituitary tumor that is nonsecreting.
   e. all pituitary tumors.

117. The development of sudden severe headache with accompanying acute visual loss is a well-recognized complication of:
   1. meningioma.
   2. intracavernous carotid artery aneurysm.
   3. hypothalamic glioma.
   4. pituitary adenoma.
   a. 1, 2, and 3.
   b. 1 and 3.
118. Which of the following features favor(s) the diagnosis of cerebral arteriovenous malformation (AVM) rather than migraine?
   - the absence of fortification spectra before visual loss.
   - family history of headaches.
   - nausea associated with visual disturbances.
   - fixed visual field defect.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

119. A 34-year-old woman presents with a complaint of double vision when reading. The examination is normal with the exception of a small-angle left hypertropia in downgaze, as well as poor elevation of the left eye. Review of systems is normal, and she denies any history of antecedent trauma. The next step in diagnosis should be:
   a. a Tensilon test.
   b. computed tomography (CT) scanning.
   c. magnetic resonance imaging (MRI).
   d. orbital ultrasonography.
   e. forcedduction testing.

120. A 40-year-old man presents to an ophthalmologist with a complaint of diagonal binocular diplopia following a motor vehicle injury. Findings consistent with a bilateral fourth nerve palsy include:
   - a right hypertropia with right head tilt.
   - a left hypertropia with left head tilt.
   - excyclodeviation >10 degrees on double Maddox rod testing.
   - a V pattern esotropia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

121. Which features are necessary to conclude a motility disturbance is a skew deviation?
   - a vertical component.
   - comitance in all gaze directions.
   - pattern of motility inconsistent with a single muscle or nerve dysfunction.
   - other obvious brainstem abnormalities.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

122. Brainstem nuclei crucial for the generation of normal vertical eye movements include:
   - paramedian pontine reticular formation.
   - the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF).
   - the abducens nucleus.
   - the interstitial nucleus of Cajal (INC).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

123. T or F The most common etiology for bilateral sixth nerve palsy is small vessel disease causing infarction of the nerve trunks.

124. T or F Most cases of unilateral abducens palsy in children are intracranial lesions.

125. What finding in a child with isolated abduction deficit most strongly argues for the diagnosis of Duane’s syndrome rather than a congenital sixth nerve palsy?
   - inability to fully abduct the eye volitionally.
   - normal abduction on oculocephalic rotational testing.
   - orthotopia in primary gaze.
   - normal adduction.
   - involvement of the left eye only.

126. Features found in all cases of internuclear ophthalmoplegia (INO) include:
   - ipsilateral adduction slowing or weakness.
   - exotropia.
131. A variant of the Guillain-Barré syndrome that involves only the brainstem and cranial nerves is known as:
   a. internuclear ophthalmoplegia (INO) skew.
   b. Foville’s syndrome.
   c. wall-eyed bilateral INO.
   d. Miller-Fisher syndrome/variant.
   e. “one-and-a-half” syndrome.

132. T or F The clinical distinction between a cavernous sinus syndrome and an orbital apex syndrome is best made by the presence of proptosis in the latter.

133. A healthy patient presents complaining of a painful double vision. Examination reveals normal visual acuity, diminished corneal sensation, and global impairment of ocular motility in the left eye, with no proptosis. There is pain with eye movements, and the globe and orbit are slightly tender. Computed tomography (CT) scanning and magnetic resonance imaging (MRI) are both normal, and the patient rapidly and completely responds to 60 mg of oral prednisone a day. The most likely diagnosis is:
   a. intracavernous carotid artery aneurysm.
   b. sphenoid wing meningioma.
   c. orbital pseudotumor.
   d. Tolosa-Hunt syndrome.
   e. cavernous sinus thrombosis.

134. Potential complications of carotid-cavernous fistulae include:
   1. retinal neovascularization.
   2. cataract.
   3. glaucomatous optic nerve damage.
   4. corneal ulceration.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

135. T or F Both high-flow and low-flow carotid-cavernous fistulae may be associated with a history of head trauma.
136. A 59-year-old man presents to the emergency room with a complaint of sudden-onset oscillopsia and diplopia. Examination reveals an alcohol odor to his breath, normal acuity, bilateral abduction deficits, and coarse binocular nystagmus. Appropriate intervention should include:
   a. intravenous glucose.
   b. intravenous naloxone.
   c. intravenous chlordiazepoxide.
   d. intravenous thiamine.
   e. coffee.

137. Which of the following optic disc lesions are distinguished by autofluorescence?
   1. myelinated nerve fibers.
   2. astrocytic hamartomas.
   3. optic nerve pits.
   4. optic nerve drusen.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

138. The approximate prevalence of giant cell arteritis (GCA) in Scandinavians is 1 in:
   a. 10.
   b. 100.
   c. 1,000.
   d. 10,000.
   e. 100,000.

139. The approximate prevalence of polymyalgia rheumatica (PMR) in individuals older than 50 years is 1 in:
   a. 20.
   b. 200.
   c. 2,000.
   d. 10,000.
   e. 20,000.

140. Clinical characteristics that may be seen with chiasmal compression include:
   1. field abnormalities more notable centrally with fainter test objects.
   2. postfixation blindness.
   3. temporal color desaturation.
   4. diplopia.

141. Diagnostic modalities useful in confirming the presence of suspected optic disc drusen include:
   1. computed tomography (CT) scanning.
   2. fundus photos with standard fluorescein angiography (FA) filters.
   3. ophthalmic ultrasonography.
   4. magnetic resonance imaging (MRI).
   a. 1, 2, and 3.
   b. 2 and 4.
   c. 1 and 3.
   d. 4 only.
   e. 1, 2, 3, and 4.

142. A woman with known multiple sclerosis (MS) presents to an ophthalmologist complaining of “a tiny blind spot in my right eye.” Examination discloses a right afferent pupillary defect (APD) and slight ocular tenderness in the right eye. The examiner attempts to confirm diagnostic suspicions by eliciting the Pulfrich phenomenon. To do this, the examiner:
   a. asks the patient to glance quickly back and forth horizontally and report any photopsias.
   b. asks the patient to climb several flights of stairs briskly and report any visual loss.
   c. asks the patient to watch the pendulum on the grandfather clock across the room and report any three-dimensional movement.
   d. spins the examining chair and watches for any nystagmus while the patient fixates on her outstretched thumb.
   e. carefully observes optic disc vasculature and color during digital pressure on the globe.

143. The test that best correlates with the pathophysiology underlying the Pulfrich phenomenon is:
   a. electrooculography (EOG).
   b. visual evoked response (VER).
   c. electoretinography (ERG).
   d. calorics and electronystagmography.
   e. fluorescein angiography (FA).
144. A 29-year-old woman presents to her ophthalmologist with a complaint that colors do not seem as bright as they used to appear. Her acuities are 20/20 in the right eye and 20/200 in the left eye. She is able to name 10 of 12 color plates correctly with the right eye but only 1 of 12 with the left. Visual fields show a cecocentral scotoma in the left eye only. Her right fundus is shown in the figure below (left). A fluorescein angiogram (FA) of her left eye is shown in the figure below (right). Each of the following statements about her situation is true except:
a. A headache may accompany the onset of visual loss.
b. The right eye is likely to be similarly affected within the next several months.
c. Diagnostic evaluation must include electrocardiography.
d. She should be counseled to avoid tobacco and heavy consumption of alcohol.
e. She should be counseled to expect no improvement in the left eye over time.

145. Each of the following statements about the disorder in question 144 is true except:
a. The patient’s brothers are more likely to be similarly affected than the sisters.
b. Other than siblings, similarly affected family members are far more likely to be maternally related than paternally related.
c. The disease is frequently associated with abnormalities of the X chromosome.
d. Unfortunately, no specific test exists to confirm the suspected diagnosis.
e. Any of the patient’s children may ultimately be affected by the disorder, to some degree.

146. A 42-year-old woman presents to her ophthalmologist with complaint of double vision. Her eye movements are shown in the figures below: Figure a. shows her in primary position; Figure b. shows her immediately after being requested to look to her right; Figure c. shows her immediately after being asked to look to her left; Figure d. shows her approximately 10 to 15 seconds after being requested to look to her left. Each of the following statements is true about her case except:
a. She may have intermittent ptosis.
b. She may have intermittent nystagmus.
c. The responsible lesion is on the left side of her brainstem.
d. There may be a history of recent closed head trauma.
e. The right eye may adduct normally with convergence.
147. The patient in question 146 may demonstrate nystagmus when looking in which field of gaze, as depicted by the figure shown there?
   a. Figure a. above.
   b. Figure b. above.
   c. Figure c. above.
   d. the patient will have nystagmus in all of the above fields.
   e. there is no reason to expect the patient to develop nystagmus.

148. A 63-year-old man presents with the pupillary findings shown in Figure a. Figure b. shows his pupils after bilateral installation of 4% cocaine. Figure c. shows his pupils after bilateral installation of 1% hydroxyamphetamine. Each of the following statements about his situation is true except:
   a. His anisocoria is worse in low ambient illumination.
   b. The right pupil is the abnormal pupil.
   c. Associated findings might include unilateral hypotony or a mild anterior chamber reaction.
   d. One potential etiology might be occult lung cancer.
   e. No workup is necessary if the findings can be shown to be longstanding.

149. If the patient in question 148 presents with acute headache, the workup must include urgent:
   a. cranial magnetic resonance imaging (MRI).
   b. carotid angiography.
   c. serum Venereal Disease Research Laboratory (VDRL) and fluorescent treponemal antibody-absorption (FTA-ABS) testing.
   d. lumbar puncture.
   e. chest computed tomography (CT) scan.
150. A 23-year-old obese woman is seen for routine ophthalmologic examination. The examination is entirely normal with the exception of bilaterally elevated discs with indistinct margins. The right optic nerve is shown in color in the figure below (left). The figure below (right) is a preinjection photo from fluorescein angiography (FA). Which one of the following statements about this patient is true?

a. The condition depicted is typically associated with mild to moderate visual loss.
b. The filters on the fluorescein camera are of poor quality.
c. The clinical and histologic findings reflect axoplasmic stasis and congestion.
d. There may be an associated arcuate field defect.
e. The findings in the picture below (right) are pathognomonic for her condition.

151. Select the computed tomography (CT) scan that is most likely to create the visual field disturbance shown in the figure below.
152. A 43-year-old man presents with a complaint of persistent red left eye, (seen in the first figure below) present ever since a car accident 6 weeks earlier. In addition to redness, he has noted intermittent horizontal diplopia without pain. The results of his examination are normal except for 4 mm of proptosis, a mild deficit in abduction, and prominent conjunctival vessels on the left. His right eye is entirely normal. Which one of the radiographic studies below is most likely to belong to this patient?

153. Which one of the following abnormalities might be expected in a young child with bilaterally poor vision and with the computed tomography (CT) scan shown below ?

a. precocious puberty.
b. mental retardation and lacunar peripheral retinopathy.
c. ash-leaf macules.
d. panhypopituitarism.
e. pheochromocytoma.
154. A unilateral brainstem lesion at the level of the central nervous system in the figure shown below is most likely to produce which one of the following neurologic deficits?
   a. alexia without agraphia.
   b. combined abducens and facial palsies.
   c. “one-and-a-half” syndrome.
   d. corneal hypesthesia.
   e. third nerve palsy and contralateral hemiplegia.

155. Which of the following would be the best choice for treatment of acute, severe migraine headache?
   a. aspirin.
   b. acetaminophen.
   c. methysergide.
   d. propranolol.
   e. sumatriptan.

156. Which of the following would be the best initial choice for prophylaxis of acute, severe migraine headache?
   a. aspirin.
   b. acetaminophen.
   c. methysergide.
   d. propranolol.
   e. sumatriptan.

157. Which one of the following statements about the Ischemic Optic Neuropathy Decompression Trial (IONDT) is false?
   a. Spontaneous improvement of vision was seen in >40% of control (nonsurgical) cases.
   b. Patients in both the surgical and the nonsurgical groups were equally likely to lose three or more lines of visual acuity.
   c. Patients in both the surgical and the nonsurgical groups were equally likely to lose three or more lines of visual acuity.
   d. The study evaluated patients with both progressive and nonprogressive forms of anterior ischemic optic neuropathy (AION).
   e. The study represented the first randomized controlled prospective trial of optic nerve sheath decompression (ONSD) for AION.

158. Which one of the following statements about pseudotumor cerebri is true?
   a. The associated headache is typically produced by certain environmental stimuli.
   b. Headache is the *sine qua non* of the disease (i.e., it is a universal symptom among patients with the disorder).
   c. Neurologic abnormalities, including abducens palsy, are common.
   d. Opening pressure and cerebrospinal fluid (CSF) protein levels are typically elevated.
   e. Many cases are probably based on decreased reabsorption of CSF.

159. Which one of the following autoantibodies are most commonly found in patients with generalized myasthenia gravis (MG)?
   a. blocking antibodies to acetylcholine.
   b. blocking antibodies to acetylcholinesterase.
   c. blocking antibodies to acetylcholine receptors.
   d. binding antibodies to acetylcholine receptors.
   e. none of the above.

160. Relative to generalized myasthenia, ocular myasthenia is:
   a. more commonly associated with Graves’ disease.
   b. more responsive to anticholinesterase drugs.
   c. more responsive to steroids.
   d. completely localized to ocular or bulbar involvement.
   e. more commonly associated with thymoma.

161. Which of the following medications is most clearly associated with drug-induced myasthenia?
   a. warfarin.
   b. methotrexate.
   c. diltiazem.
Questions

d. ranitidine.
e. D-penicillamine.

d. All of the above.

e. pharnmacologic anisocoria.

e. all of the above.

162. Afferent pupillary fibers from the optic tract exit at:
a. the lateral geniculate body (LGB).
b. the pretectal olivary nuclei.
c. the occipital cortex.
d. the frontal lobe.
e. the medial longitudinal fasciculus.

163. Which one of the following nerves supplies the cornea?
a. frontal nerve.
b. lacrimal nerve.
c. nasociliary nerve.
d. supraorbital nerve.
e. none of the above.

164. A patient with a history of bilateral occipital lobe infarcts adamantly states that he can see quite well and confabulates visual images. He most likely has:
a. Anton’s syndrome.
b. palinopsia.
c. Charles Bonnet’s syndrome.
d. blindsight.
e. none of the above.

165. While undergoing retinal examination, a patient claims to see his retinal vessels. This is an example of:
a. formed hallucinations.
b. Charles Bonnet’s syndrome.
c. blindsight.
d. Purkinje effect.
e. the Pulfrich phenomenon.

166. Which of the following statements about monocular diplopia is false?
a. Monocular diplopia is usually relieved by covering either eye.
b. It can be caused by keratoconus.
c. It can be caused by lenticous.
d. It can be caused by high astigmatism.
e. It can be caused by retinal pathologies.

167. Which of the following results in anisocoria in dim light?
b. posterior synechiæ.

c. physiologic anisocoria.

e. all of the above.

d. pharmacologic anisocoria.
173. Which of the following statements about the condition shown in the figure below is false?
   a. The condition is unilateral.
   b. Retinal vessels originating from the periphery of the disc is a classic finding.
   c. Patients with this condition are at risk for retinal detachments.
   d. The patient may have a relative afferent pupillary defect (RAPD).
   e. Visual acuity is often normal.

174. A patient with the lesion shown in the figure below most likely has:
   a. choroidal melanoma.
   b. medulloepithelioma.
   c. congenital hypertrophy of the retinal pigment epithelium (RPE).
   d. melanocytoma.
   e. none of the above.

175. Which of the following statements about the condition shown in the figure below is false?
   a. The successful use of photodynamic therapy (PDT) to treat this condition has been recently reported.
   b. Vitreous hemorrhage can occur spontaneously in this condition.
   c. Patients with no other medical history presenting with a similar fundus should undergo neuroimaging.
   d. An orbital bruit can be heard in patients with this condition.
   e. Facial nevi and intracranial calcification can be observed in patients with this condition.
Matching

176. Match each of the various disorders numbered below with its most likely visual field defect(s) shown in the lettered figures below:

1. ischemic optic neuropathy.
2. optic nerve pit with serous retinal detachment.
3. toxic/nutritional optic neuropathy.
4. glaucoma.
5. hereditary optic neuropathy.
6. optic neuritis.

a. Out to I4e stimulus
b. Out to I4e stimulus
c. Out to I4e stimulus
d. Out to I4e stimulus
e. Out to I4e stimulus
f. Out to I4e stimulus
177. Match each visual field shown with its corresponding lesion in the following numbered visual pathway locations:
1. posterior chiasm.
2. anterior chiasm.
3. optic nerve.
4. thalamus.
5. mid-chiasm.

178. Listed below are various retrochiasmal visual disorders. Match each lettered defect with its correct anatomic locus numbered below.
- a. alexia with agraphia.
- b. alexia without agraphia.
- c. cerebral dyschromatopsia.
- d. prosopagnosia.
- e. unformed hallucinations.

179. Listed below are neurologic deficits that may be associated with fascicular third nerve palsy. Match each lettered defect with its correct anatomic locus.
- a. contralateral coarse tremor.
- b. contralateral hemiparesis.
- c. contralateral ataxia.

180. Match each lettered symptom complex listed below with its appropriate name.
- a. severe throbbing headache with associated nausea and photophobia.
- b. severe throbbing headache.
181. Match the numbered ganglia with the appropriate characteristics described below (note that each ganglion may be assigned to more than one characteristic).

a. cell bodies with processes providing facial sensation.

b. origin of the postganglionic fibers to the iris dilator muscle.

c. origin of the greater superficial petrosal nerve.

d. located in Meckel's cavity.

e. origin of postganglionic fibers to iris sphincter.

f. located in fallopian canal.

g. origin of postganglionic fibers to the lacrimal gland.

h. located near the angle of the mandible.

i. located lateral to the intraorbital optic nerve.

1. pterygopalatine ganglion.

2. geniculate ganglion.

3. gasserian ganglion.

4. ciliary ganglion.
Answers

1. True. Patients who have a maculopathy will take longer (90 to 180 seconds) to recover pretest vision after looking into a bright light for 10 seconds. Patients with optic nerve disease will have a normal recovery time (<60 seconds). The test is probably only valid for patients with vision better than 20/80.

2. c. A pattern visual evoked response (VER) (as opposed to a flash VER) is required for visual acuity assessment in preverbal children. Although the VER is useful in establishing factitious visual loss, its reliability is limited by the fact that patients can produce false readings by using accommodation to fog their vision. Abnormalities in VER latency and amplitude have been reported in various maculopathies and retinopathies, and, therefore, these features cannot distinguish optic neuropathy from retinal disorders with complete reliability.

3. d. Although optokinetic nystagmus (OKN), the rocking mirror test, and the base-up prism test can help discover factitious monocular blindness, these tests are not sensitive enough to diagnose factitious visual deficit at the 20/100 level. For such mild visual deficits, a fogging refraction, stereo acuity, and red-green glasses may be useful in diagnosing factitious visual loss.

4. e. See the answer to question 3. A fogging refraction is probably the most useful tool to master this problem because it can be applied to any level of factitious visual loss.

5. e. An optic tract lesion may cause unilateral decreased visual acuity if the lesion is not complete or if the optic nerve or chiasm is also involved. If the optic nerve or chiasm is involved, the afferent pupillary defect (APD) is typically ipsilateral. Pure optic tract lesions can produce a contralateral afferent pupillary defect (because more than half of afferent fibers cross), as well as a complete homonymous hemianopia.

6. False. To the contrary, intact oculovestibular reflexes in a patient with a gaze palsy implies a supranuclear disturbance. If the vestibular system can drive the eyes in the direction of the paretic field of gaze, the gaze paresis must be supranuclear in nature. Impaired oculocephalic responses indicate that the infranuclear, end-organ function is responsible for the motor disturbance.

7. True. Upturning of the eyes upon forceful opening of closed eyelids is known as the Bell phenomenon. If forcefully opening closed eyelids stimulates upgaze, the final common pathway (infranuclear) for upgaze must be intact.

8. c. Magnetic resonance imaging (MRI) is based on applying a radio frequency pulse to a tissue within a strong magnetic field and measuring the change in the tissue’s nuclear spin and magnetic vector. The longitudinal relaxation time is termed T1. Fat is bright and water is dark on T1-weighted images. In contrast, fat is dark and water is bright on T2-weighted images. Some tissues such as cortical bone, rapidly flowing fluid (blood), and air give no signal at all on MRI.

9. c. Although there is some redundancy of the optic nerve within the orbit, the intracranial optic nerve has little “slack.” The dimensional characteristics of the optic nerve can be remembered with the mnemonic phone number 125-1017, which stands for the lengths of the intraocular (1), intraorbital (25), intracanalicular (10), and intracranial (17) portions of the optic nerve.

10. c. Fifty-three percent of the retinal ganglion cells cross in the chiasm (this difference is occasionally important clinically). The macular fibers constitute a large portion of the optic chiasm and most decussate in the posterior chiasm. The chiasm lies approximately 1 cm (not 1 mm) above the anterior pituitary gland. The inferior nasal retinal fibers cross in the anterior chiasm and were thought to loop anteriorly in the contralateral optic nerve before traveling posteriorly, leading to the term Wilbrand’s knee. It is now thought that Wilbrand’s knee may be an artifact.

11. False. The retinal ganglion cell terminals are segregated by eye (ipsilateral versus contralateral) rather than on the basis of which receptive field the ganglion cells serve. The ipsilateral ganglion cells synapse in layers 2, 3, and 5, whereas the contralateral ganglion cells synapse in layers 1, 4, and 6.

12. False. In fact, the inferior fibers of the optic radiations pass extremely close to the internal capsule. The combination of a superior hemianopic contralateral visual field defect and a contralateral hemiparesis can be produced by a small infarct in this region.

13. d. The temporal 30 degrees of a binocular visual field is perceived by the nasalmost retina of the ipsilateral eye only. These “temporal crescents” are represented in the anterior-most occipital lobe. Therefore, a lesion in this area will produce a monocular visual field defect in the far temporal periphery of the contralateral eye, the so-called temporal crescent syndrome. For example, a right anterior occipital lobe lesion would produce a far temporal field defect in the left eye. Similarly, a right posterior occipital lobe lesion may spare the far temporal field in the left eye.

14. b. Optokinetic nystagmus (OKN) abnormalities indicate lesions of the parietooccipital (slow-phase pursuit abnormalities) or the frontal lobe (fast-phase recovery abnormalities). High congruity of visual field deficits indicates a lesion in the occipital lobe. Both partial complex seizures and formed visual hallucinations may be seen with temporal lobe lesions. Inferior nerve fibers
Answers

from the superior retina course anteriorly in Meyer’s loop; therefore, lesions affecting Meyer’s loop will result in pie-in-the-sky defects contralateral to the lesion.

15. c. Parietal lobe lesions are associated with agnosia and right-left confusion. A parietal lobe lesion will also affect slow-phase pursuit movements toward the ipsilateral side. Unlike temporal lobe lesions, which produce hemianopsias that are denser superiorly, parietal lobe lesions produce hemianopsias that are denser inferiorly.

16. a. Formed hallucinations occur with temporal lobe pathology. Depending on the location of the occipital lobe lesion, the temporal crescent, the representation of which is located in the most anterior visual cortex, may be either spared (more common) or affected. Although optokinetic nystagmus (OKN) asymmetry may rarely occur with occipital lesions, this finding is generally indicative of a parietal locus of disease.

17. b. Key events in the development of true disc edema include cessation of axonal transport with swelling of axons. The increase in disc volume is because of enlargement of axons, rather than increased extracellular fluid, as seen in edema of other tissues. Breakdown of the blood-retinal barrier does occur and is detected as leakage on fluorescein angiography (FA), but this is not important in the causation of disc edema.

18. c. Although loss of spontaneous venous pulsations is an early sign of papilledema, approximately 20% of normal patients lack venous pulsations. Transient obstructions of vision (TOV) often accompany papilledema and are episodes of unilateral or bilateral visual loss, lasting only a few seconds. The most typical visual field finding in acute papilledema is an enlarged blind spot. Although rare, unilateral papilledema may occur. For example, if contralateral optic atrophy exists, papilledema may be detectable only in the viable disc.

19. c. Pseudotumor cerebri is characterized by (i) increased intracranial pressure on lumbar puncture, (ii) normal neuroimaging studies (although the ventricles may be small), and (iii) normal cerebrospinal fluid (CSF). Papilledema need not be present for the diagnosis. Although the neurologic examination is usually normal, sixth nerve palsy may occur with increased intracranial pressure of any etiology.

20. c. Obesity is not an indication for treatment, although weight loss (even as little as 6% of total body weight) often improves the condition.

21. c. Studies have documented that up to 80% of patients with cavernous sinus-dural fistulae will develop ocular hypertension. Twenty-five percent of patients will develop optic disc cupping and 20% visual field defects. Any entity that raises episcleral venous pressure can cause secondary open-angle glaucoma.

22. a. “Occult” giant cell arteritis (GCA) is a recognized entity in which atypical clinical findings are present (e.g., malaise, anorexia). Despite a normal erythrocyte sedimentation rate (ESR), biopsy results are positive and steroids are effective.

23. c. Nonarteritic anterior ischemic optic neuropathy (NAION) is far more common than arteritic anterior ischemic optic neuropathy (AAION) (approximately 95% vs. 5%) and patients have a lower mean age at diagnosis than patients with AAION (60 years vs. 70 years). It usually occurs in a younger age group and may resemble optic neuritis. The various ways to differentiate the NAION from optic neuritis include (i) the absence of pain with eye movement, (ii) the age group affected, and (iii) delayed optic disc filling present in 75% of NAION cases (whereas filling should be normal in optic neuritis). Although aspirin may prevent further strokes in patients with NAION, its effect on the contralateral eye is unclear.

24. d. 16% of patients with an initial attack of optic neuritis with a negative magnetic resonance imaging (MRI) will develop multiple sclerosis (MS) at 15-year follow-up. More than 50% of patients with three or more lesions that are visible on MRI will develop MS at 15-year follow-up.

25. c. In a retrospective review of autopsy findings, nearly 100% of patients dying of multiple sclerosis (MS) had some degree of optic nerve demyelination.

26. b. Ninety percent of optic gliomas occur in the first two decades of life. The percentage of patients with optic nerve glioma that have associated neurofibromatosis (NF-1) ranges from 14% to 60%. Malignant gliomas of the visual pathways, although rare, occur more frequently in middle-aged adults than in children. Survival averages 6 to 12 months after diagnosis.

27. False. Collateral vessels (optociliary shunt vessels) at the disc connecting the retinal and choroidal vascular systems also may be seen in association with central retinal vein occlusion (CRVO), sphenoid wing meningioma, long-standing primary open-angle glaucoma (POAG), optic gliomas, or chronic papilledema.

28. d. In contrast to optic nerve gliomas, meningiomas occur primarily in adults and are three times more common in women. Although persons with neurofibromatosis (NF-1) have a higher incidence of meningiomas than the general population, only a few patients with meningiomas have NF-1. With contrast computed tomography (CT) scanning, the peripheral part of the involved optic nerve may show enhancement, resulting in the “railroad track” sign (“kinking” is specific for optic nerve glioma).

29. False. Dominant (Kjer) optic neuropathy (DOA) manifests between 5 and 10 years of age. Visual loss may progress until the mid-teens, at which point it usually stabilizes. Color defects are almost universally present, and tritanopia (which can be detected with the Farnsworth-100 hue testing) is suggestive of DOA. Inheritance is naturally autosomal dominant, and DOA is linked to the OPA1 gene on chromosome 3.
30. c. The incidence of spontaneous recovery has been reported to be as high as 10%. Leber’s hereditary optic neuropathy (LHON) exhibits mitochondrial inheritance (inheritance from the mother).

31. c. Patients with “papillophlebitis” have normal or near normal visual acuity. It may be a form of incomplete central retinal vein occlusion (CRVO) and usually resolves spontaneously within 12 months.

32. c. Optic nerve drusen occur almost exclusively in white individuals.

33. True. Morning glory disc syndrome is characterized by a mass of glial tissue with radiating blood vessels, peripapillary atrophy, and pigmentation of chorioretinal tissue. It is usually unilateral and is more common in women than men (2:1). Visual acuity is usually poor.

34. c. Optic nerve hypoplasia is seen with greater incidence in children of diabetic mothers and following fetal exposure to antiepileptic medications, quinine, or lysergic acid diethylamide (LSD). Optic nerve hypoplasia, particularly bilateral involvement, has been associated with other midline developmental anomalies—that is, absence of the septum pellucidum and hypothalamic-pituitary abnormalities. Endocrine dysfunction is manifested as hypoglycemic seizures and growth retardation. This combination of clinical findings is known as DeMorsier’s syndrome.

35. False. Nonsecreting tumors often present with visual field loss, whereas secreting tumors present with endocrine dysfunction. An exception is prolactin-secreting tumor in male patients because the decreased libido and impotence are often not reported early in their course.

36. True. When the causative lesion is not located at the site responsible for a clinical sign, it is described as false localizing. Bitemporal macular hemianopia can only arise from compression of the posterior chiasm. Although most lesions responsible for this compression are parasellar, a lesion distant from this site (e.g., a tumor at the base of the brainstem) also may lead to chiasmatic compression. This occurs if the lesion causes obstructive hydrocephalus, which enlarges the third ventricle, compressing the chiasm.

37. d. The most common location for a cerebral aneurysm with third nerve palsy is the junction of posterior communicating artery and internal carotid artery.

38. False. Precipitous aneurysmal distention caused by hemorrhage of the communicating artery at its junction with the internal carotid artery may occur, creating a third nerve palsy. Ninety-five percent include pupillary involvement, and pain is nearly always present, although not universally. Furthermore, many third nerve palsies resulting from diabetic vasculopathy also can be intensely painful.

39. a. Third nerve aberrant regeneration never occurs with diabetic oculomotor neuropathy. Aberrant regeneration of the third nerve implies another etiology, such as aneurysm, tumor, inflammation, or trauma. Other classic findings of aberrant regeneration include persistent vertical gaze limitation secondary to simultaneous contraction of superior and inferior recti, and pupillary miosis with elevation, adduction, or depression.

40. b. The long intracranial course of the trochlear nerve leaves it especially susceptible to damage from closed head trauma. This occurs because of contrecoup injury from the free tentorial edge. Ischemic damage, usually caused by diabetes mellitus, is second, and idiopathic palsies are third. Hydrocephalus, vascular loops, or tumor can compress the trochlear nerve as well.

41. d. The three-step test is useful for diagnosis but does not differentiate between congenital and acquired trochlear nerve palsy. Large vertical fusional amplitudes (>5 prism diptors) and facial asymmetry from childhood head-tilting suggest a decompensated congenital lesion.

42. d. Nasopharyngeal carcinoma can involve numerous cranial nerves because of its proximity to the preoptic basal cistern. Most frequently, the trigeminal nerve is involved, causing facial hypesthesia or facial pain. The abducens nerve is the second most common. The hallmark of nasopharyngeal carcinoma is its propensity to involve multiple cranial nerves noncontiguously. Nasopharyngeal carcinoma is common in Chinese men. The least differentiated forms are also known as Schmincke’s and Regaud’s tumors.

43. True. Within the cavernous sinus, sympathetic branches of the paracarotid plexus join the sixth nerve briefly. Occasionally, an intracavernous lesion can produce sixth nerve palsy with postganglionic Horner’s syndrome, producing pupillary miosis.

44. c. Duane’s syndrome may be secondary to hypoplasia or aplasia of the abducens nucleus, with lateral rectus innervation by the oculomotor nerve. It has various presentations, but retraction of the adducted globe appears most consistently. The three types of Duane’s syndrome are distinguished by the relative ability to adduct or abduct: type 1, limited abduction but full adduction; type 2, normal abduction but limited adduction; and type 3, both abduction and adduction are limited.

45. False. In the cavernous sinus, cranial nerves III, IV, and V are relatively protected within the walls of the sinus, but cranial nerve VI runs in the middle of the sinus and is more prone to injury.

46. False. Horizontal saccades originate in the contralateral frontal lobe, but either hemisphere can produce ipsilateral saccades if the other hemisphere is damaged. Appropriate stimulation of the parietal or occipital cortex can also produce contralateral saccades.

47. b. Oculomotor apraxia is an inability to initiate voluntary horizontal saccades. Congenital oculomotor apraxia is characterized by striking compensatory head movements, and acquired lesions (e.g., resulting from bilateral frontal lobe strokes) usually produce defects in
Answers

initiation of bidirectional saccades—patients may blink to break fixation and then turn their head to fixate on something else. Pursuits remain relatively unaffected.

48. c. Opsoclonus consists of nonstop, random, directionally unpredictable saccades. Ocular flutter consists of spontaneous groups of back-and-forth horizontal saccades that may occur during fixation or at the end of a normal horizontal saccade. Both ocular flutter and opsoclonus may be associated with malignancy, such as metastatic neuroblastoma in children or small cell cancer of the lung in adults. These eye movements may be the first presenting sign of cancer. Multiple sclerosis (MS) also can cause ocular flutter. Square-wave jerks are a microsaccadic fixation disturbance associated with cerebellar diseases of various kinds. Ocular motor dysmetria is a back-and-forth saccadic motion about the point of fixation that occurs following an otherwise normal saccade. It is felt to represent “overshooting” of the intended fixation point. Ocular bobbing is most commonly seen in comatose or quadriplegic patients with large infarcts or brainstem hemorrhages.

49. b. In progressive supranuclear palsy (PSP), downward gaze is generally affected, becoming smaller and slower. Saccades are affected more than pursuits. This is in distinction to the dorsal midbrain syndrome, in which upward saccades are generally affected.

50. True. The period between detection of movement and the beginning of smooth pursuit (for tracking) is between 125 and 135 milliseconds. Initiation of saccadic movement requires from 150 to 200 milliseconds from the first perception of a target.

51. d. Skew deviation is, by definition, a vertical misalignment of gaze that cannot be assigned to a single nerve or muscle weakness. In the presence of an internuclear ophthalmoplegia (INO), the hypertropic eye is often on the same side as the addiction deficit (medial rectus dysfunction). Fourth nerve palsy does not produce difficulty with addition. The “one-and-a-half” syndrome results in, among other things, complete horizontal paralysis of one eye and limitation of the other to abduction only. Progressive supranuclear palsy (PSP), as discussed in answer 49, is a disorder of movement that does not produce a resting vertical gaze deviation. Dorsal midbrain syndrome (Parinaud’s syndrome) does not feature an adduction deficit (see answer 52).

52. c. Upward gaze paresis, light-near dissociation, lid retraction, accommodative abnormalities, and convergence-retraction nystagmus are all features of the dorsal midbrain (Parinaud’s) syndrome. Furthermore, skew deviation and papilledema may be seen, depending on the etiology. Lid retraction—the Collier sign—may worsen with attempted upgaze. Convergence-retraction nystagmus is also a response to an effort at upgaze that triggers medial rectus contractions. Hence, this form of “nystagmus” is worsened by upward optokinetic nystagmus (OKN) testing. Paradox OKN is not a feature (see answer 53).

53. a. Congenital motor nystagmus has several features, including normal visual acuity, paradox optokinetic nystagmus (OKN) (caused by the abnormal movements of nystagmus appearing “slow” and against the expected direction of target tracking), dampening by convergence, and aggravation by fixation. Fortunately, oscillopsia (the sensation of the world moving) is not normally a problem.

54. d. Monocular nystagmus has been associated with chiasmal and hypothalamic gliomas. It is also seen in blind eyes, multiple sclerosis (MS), and spasmus nutans—a transient, fast-beating, low-amplitude nystagmus found during the first 5 years of life.

55. a. Alexander’s law describes the increased frequency and amplitude of nystagmus movements accompanying gaze in the direction of the fast-beating component. Of the various forms of nystagmus, downbeat nystagmus most frequently violates Alexander’s law.

56. c. Upbeat nystagmus can be caused by lesions of the anterior vermis and lower brainstem, as well as drugs and Wernicke’s encephalopathy. Downbeat nystagmus may be localized to anatomic structures at the cranio cervical junction (e.g., Arnold-Chiari malformation) and may be seen in certain intoxications (alcohol, lithium). Vestibular nystagmus has its etiology within the vestibular system. Periodic alternating nystagmus also can localize to the cranio cervical junction but may be seen in other forms of posterior fossa disease. Seesaw nystagmus results from third ventricle tumors or diencephalic lesions involving the connections to the interstitial nucleus of Cajal (INC) and is therefore the most localizing of those mentioned.

57. b. Bitemporal hemianopia may be seen in acquired seesaw nystagmus because the posterior chiasm is most vulnerable to diencephalic lesions, which may provoke it.

58. False. Vergence eye movements are felt to have a pathway of their own for stimulating the brainstem motor nuclei. Saccades originate in the contralateral frontal lobe, whereas pursuits arise in the ipsilateral parietal lobe.

59. True. Because of bilateral supranuclear frontal lobe, whereas pursuits arise in the ipsilateral parietal lobe.

60. False. Spontaneous or reflexive blinking requires normal function of the basal ganglia, as do other nonvolitional facial expressions. Therefore, these are commonly affected in Parkinson’s disease. Volitional movements, in distinction, are generally not adversely influenced.

61. b. The greater superficial petrosal nerve synapses in the pterygopalatine ganglion, not in the geniculate ganglion.
62. False. Marcus Gunn’s “jaw-wink” reflex is an example of synkinesis (abnormal innervation connecting two groups of normally unrelated muscles, such that they contract together), but it is not synkinesis because of aberrant regeneration, which usually follows a peripheral neuropathy or trauma to the nerve. The “jaw-wink” is a congenital/neurogenic phenomenon.

63. c. There is currently no proven treatment for nonarteritic anterior ischemic optic neuropathy (NAION). All of the answers have been tried with limited success.

64. e. Approximately 85% of patients with Bell’s palsy spontaneously recover, although some will have some degree of aberrant regeneration. Recovery usually begins within 3 weeks of diagnosis and usually becomes complete by 2 to 3 months.

65. True. The parotid gland can become infiltrated with granulomas, and the facial nerve is involved at this site. Facial nerve involvement in sarcoidosis is frequently bilateral but asymmetric. The second most commonly involved cranial nerve is the optic nerve.

66. False. Of the three most common causes of facial nerve overactivity, only essential blepharospasm is believed to be related to basal ganglia dysfunction. Compression of the facial nerve in the cerebellopontine angle by anomalous vessels has been demonstrated in 90% of cases of hemifacial spasm. Tumors in the cerebellopontine angle also can cause hemifacial spasm. Facial myokymia is caused by disease in the pons involving the facial nucleus or fascicle. The most common cause of synkinesis (abnormal innervation connecting two groups of normally unrelated muscles, such that they contract together), but it is not synkinesis because of aberrant regeneration, which usually follows a peripheral neuropathy or trauma to the nerve. The “jaw-wink” is a congenital/neurogenic phenomenon.

67. d. See answer 66. Optic nerve gliomas are associated with compressive optic neuropathy; chiasmal gliomas may cause hypothalamic dysfunction. Spasmus nutans is a monocular nystagmus that usually occurs in the first year of life.

68. a. Tardive dyskinesia secondary to neuroleptic medication can produce facial grimacing and blepharospasm, similar to Meige’s syndrome. Hemifacial spasm is distinct from essential blepharospasm in that it is unilateral, and multiple facial muscles are involved. Reflex blepharospasm can be caused by a number of disorders, including severe dry eye, intraocular inflammation, and foreign bodies.

69. d. The afferent pupillomotor fibers exit the optic tracts just before the lateral geniculate body (LGB); they do not pass through the LGB. Although postganglionic pupillomotor fibers in the sympathetic pathway do arise from the superior cervical ganglion, the sympathetic pathway leading to these fibers is thought to originate in the posterior hypothalamus. In addition, postganglionic sympathetic fibers enter the orbit with the ophthalmic division of the trigeminal nerve through the superior orbital fissure. The consensual pupillary response is seen because of decussation at the pretectal nuclei. Were the chiasm split in half, consensual responses would be preserved.

70. b. Argyll Robertson’s pupils are miotic and irregular and do not react to light, but they have a normal near response. This is a rare finding in some patients with tertiary syphilis. In the light-near dissociation of Parinaud’s dorsal midbrain syndrome, the pupils are larger. In young children, the most common cause of this syndrome is a tumor in the region of the pineal gland. In young adults, head trauma and multiple sclerosis (MS) are frequently seen. In patients older than 60 years, stroke is most commonly to blame.

71. b. Oculomotor nerve palsy usually includes ptosis, limitation of oculomotor function, and pupillary abnormalities. In uncal herniation and basal meningitis, pupil dilation may be the only sign of third nerve palsy. In the case of a cerebral aneurysm, the pupil is usually involved, along with other functions of the third nerve. If the pupil is not involved, it is less likely that an aneurysm is the cause of the palsy. Total oculomotor palsy with a spared pupil in elderly patients usually suggests a microvascular etiology.

72. d. Although 0.25% pilocarpine is very dilute, patients with normal pupils may respond to this dose. Therefore, a weaker preparation (0.10%) is recommended. Most patients affected will have unilateral pupillary involvement and depressed tendon reflexes. After many months or years, an Adie’s pupil will become miotic.

73. b. Horner’s syndrome is defined as ipsilateral ptosis and miosis. Ptosis is secondary to lack of Müller’s muscle function. Anhidrosis may or may not be present. Localization of the lesion in Horner’s syndrome is part of the clinical workup and sometimes guided by the extent of anhidrosis. First-order lesions (central nervous system [CNS]) cause ipsilateral anhidrosis of the entire body. Second-order lesions (Pancoast’s tumor, neck trauma) cause ipsilateral facial anhidrosis. Third-order lesions cause anhidrosis only around the affected eye or none at all. In response to 4% cocaine, a normal pupil will dilate, but a Horner’s pupil will dilate poorly. Hydroxyamphetamine (Paredrine) will cause a similar response in preganglionic Horner’s syndrome only. Painful Horner’s syndrome may be caused by many disorders (neck trauma, migraine, cluster headaches), but spontaneous dissection of the common carotid artery must be ruled out with angiography or magnetic resonance imaging (MRI)/magnetic resonance angiography (MRA).

74. d. Most patients with nonphysiologic anisocoria have sphincter muscle dysfunction visible at the slit lamp. Trauma is most common, followed by Adie’s pupil.

75. True. The risk of developing multiple sclerosis (MS) for a relative of a patient with MS is nearly 20-fold higher than the general population. MS is a multifactorial disease with both genetic and environmental risk factors.
76. False. The most common cranial mononeuropathy in multiple sclerosis (MS) is optic neuritis, and it is the one of the presenting features of MS in approximately 25% of patients. The most frequently affected motor nerve is the sixth nerve.

77. True. Retinal vascular infiltration (lymphocytic) and vitreous cells may be seen in a small percentage of active multiple sclerosis (MS) cases.

78. b. Pheochromocytomas produce, secrete, and store catecholamines. They are most often derived from the adrenal medulla but may arise in any of the sympathetic ganglia. Two phakomatoses, neurofibromatosis and von Hippel-Lindau disease, are associated with the tumor.

79. c. Astrocytic hamartomas can be seen in tuberous sclerosis or neurofibromatosis, or they can be sporadic. They are located in the nerve fiber layer of the retina and sometimes can be associated with the optic disc.

80. d. This is known as Vogt’s triad and is present in 30% of patients with tuberous sclerosis. Facial angiofibromas (adenoma sebaceum) are present in at least 75% of adults with tuberous sclerosis.

81. a. Seizures are part of the classic triad in tuberous sclerosis (80% of patients have seizures). Patients with Sturge-Weber syndrome and meningeal hemangioma may have seizure disorders. Patients with neurofibromatosis also have an increased risk of seizures.

82. b. Unilateral congenital glaucoma is seen in 25% of cases of Sturge-Weber syndrome, often associated with an angioma on the upper lid. Likewise, a plexiform neurofibroma of the upper lid is closely associated with juvenile glaucoma in von Recklinghausen’s disease. Glaucoma can also be seen in von Hippel-Lindau disease.

83. False. The characteristic ocular lesion of angiomatosis retinae, or von Hippel's disease, is a retinal angiomatoma. This is a retinal capillary hemangioma (hemangioblastoma). The cavernous hemangioma of the retina is a rare lesion that can be associated with similar skin and central nervous system (CNS) lesions. Diffuse choroidal hemangiomas (tomato ketchup fundus) are associated with Sturge-Weber syndrome (encephalo-facial angiomatosis).

84. d. Patients with ataxia-telangiectasia may have associated thymic hypoplasia, defective T-cell function, and immunoglobulin A (IgA) (secretory immunoglobulin) deficiency, with severe respiratory infections.

85. c. Four signs constitute complete Kearns-Sayre syndrome: (i) progressive external ophthalmoplegia (PEO), (ii) mild pigmentary retinal degeneration, (iii) onset before 20 years of age, and (iv) heart block, potentially lethal and among the last signs to develop. Some studies indicate that elevated cerebrospinal fluid (CSF) protein levels correlates with the presence of heart block.

86. False. Many systemic syndromes include progressive external ophthalmoplegia (PEO), which is a non-descriptive term for chronically progressive loss of eye movements. One such syndrome is myotonic dystrophy. A diagnostic feature of myotonic dystrophy is the presence of polychromatic (“Christmas tree”) cataracts, which may present even in the “amytotic” forms. Wilson’s disease is associated with the sunflower cataract.

87. a. Conjunctival injection and superficial punctate keratitis are common, as is proptosis. Diplopia in upgaze is also seen in orbital floor fractures. The lid retraction and lag is the most common sign.

88. d. The most frequently involved muscle in dysthyroid orbitopathy is the inferior rectus. The medial rectus is the second most frequently affected muscle and may simulate a sixth nerve palsy.

89. b. One mnemonic for this is “I M Stuart Little” (inferior, medial, superior, lateral, in order of their involvement).

90. d. Although 75% of all myasthenics will have eye findings at presentation, only 33% to 50% will have ocular myasthenia only. A higher percentage (90%) of patients with myasthenia gravis (MG) will develop ocular symptoms during the course of the disease. Proptosis is the most common.

91. a. A tiny fraction of patients with Graves’ disease will develop myasthenia gravis (MG) as well.

92. b. Radiologic investigation is mandatory for all myasthenics in order to discover thymic enlargement. Thymectomy may be curative in this setting.

93. d. Studies of central retinal artery ligation in rhesus monkeys established this value.

95. False. Patients with embolic central retinal artery occlusion (CRAO) are at great risk of myocardial infarction (MI). Stroke is also more likely after CRAO, but the most common cause of death is heart disease.

96. e. Intraocular pressure (IOP) may be low (caused by ciliary hypoperfusion), normal, or elevated (caused by neovascularization of the iris and angle). Unilateral arcus is highly suggestive of contralateral carotid disease. Carotid stenosis protects the ipsilateral cornea from serum lipid deposition.

97. a. Transient monocular blindness is the hallmark of carotid (anterior) disease. Vertebralbasilar insufficiency typically causes binocular visual blurring or oculomotor symptoms. Other symptoms of vertebralbasilar insufficiency include transient dysarthria, drop attacks, photopsias, and hemisensory defects.

98. c. The patient’s ocular signs and symptoms clinically suggest the diagnosis of demyelinating optic neuritis. With a history of paresthesias in her right leg, the diagnosis of multiple sclerosis (MS) should be entertained. An afferent pupillary defect (APD) almost invariably occurs in the acute phase. This patient most likely had a subclinical contralateral episode of optic neuritis in the past and therefore no detectable...
5: Neuro-ophthalmology

APD. APDs are always relative—that is, comparing one optic nerve to the contralateral nerve. In the Optic Neuritis Treatment Trial, oral steroid therapy showed to offer no improvement in long-term prognosis and had a higher rate of subsequent optic neuritis recurrence.

99. a. The etiology of Coats’ disease is unknown, and there does not appear to be any genetic, familial, racial, or ethnic predisposition. However, Coats’-type retinal vascular changes have been noted in patients with facioscapulohumeral muscular dystrophy, Turner’s syndrome, Senior-Loken syndrome, and one variant of the epidermal nevus syndrome. In addition, Coats’-like retinopathy has been noted in up to 3.6% of patients with retinitis pigmentosa.

100. b. Downbeat nystagmus in primary position is localized to the cranioverical junction (or certain intoxications). This patient's clinical symptoms of intermittent occipital headaches with sudden head movements or anger suggest the diagnosis of Arnold-Chiari malformation. Arnold-Chiari malformation is one of the most common causes of downbeat nystagmus.

101. b. Clonazepam and suboccipital craniotomy have been used in the treatment of downbeat nystagmus in patients with Arnold-Chiari malformation. By contrast, carbamazepine toxicity has been associated with downbeat nystagmus.

102. a. This patient appears to have Parinaud’s dorsal midbrain syndrome, which may include the following findings: pupillary light-near dissociation, lid retraction (the Collier sign), upgaze paresis, convergence-retraction nystagmus, fixation instability, small-amplitude skew deviation, and papilledema (if ventricular outflow has been compromised). The most common cause in this age group would be a pinealoma. Other causes include stroke, hydrocephalus, and multiple sclerosis (MS).

103. d. After neuroimaging has been obtained, a lumbar puncture would be an important step in the diagnostic evaluation of this patient because pinealoma classically sheds cells into the cerebrospinal fluid (CSF).

104. b. Optic neuritis in childhood is more commonly bilateral. Visual loss can be severe, although intravenous corticosteroids can improve visual function. Diffuse enlargement of the optic nerve on computed tomography (CT) scan may be seen in this condition, mimicking a neoplasm of the optic nerve sheath. The demyelination typically follows a viral illness or vaccination by 10 to 14 days.

105. a. The differential diagnosis for posterior ischemic optic neuropathy (PION) should include radiation optic neuropathy, status post-coronary artery bypass graft, anemia, acute systemic hypotension, giant cell arteritis (GCA), and syphilis. Well-controlled essential hypertension is associated with anterior ischemic optic neuropathy (AION) but not PION.

106. d. Bilateral disc edema and headache may be caused by several reasons, but hypertension should be first on the list to be excluded because it is quite easy to do so. After checking blood pressure, a neuroimaging should be obtained immediately.

107. d. Optic disc drusen may be associated with retinitis pigmentosa and with pseudoaxanthoma elasticum.

108. e. This may sound incredibly picky, but there is clinical relevance here. Rigorous pupillary measurements typically involve measurement of pupillary diameter. With this as the parameter, simple anisocoria is less apparent in bright light, as is the case for pathologic mydriasis, such as Adie’s and third nerve palsy. If, by contrast, pupillary area is measured, then simple anisocoria can be distinguished from pathologic anisocoria because the pupillary areas will remain different by the same amount in simple anisocoria regardless of lighting conditions. With pathologic anisocoria, the difference in pupillary areas will change drastically as ambient lighting is changed. Who would measure pupillary areas? Well, the most common method of testing pupillary reactions, simple observation of pupil size without any numerical measurements, is most closely related to the perception of pupillary areas.

109. c. Meningothelial (syncytiial) meningioma is the most common histopathologic type of meningioma seen within the orbit. “Pilocytic” describes the cell type of gliomas of the visual pathways.

110. d. In general, optic nerve compression >1 cm posterior to the globe does not cause disc edema.

111. False. So-called tobacco-alcohol amblyopia seems to be seen only in heavy smokers/drinkers with poor nutrition. This has led many to believe that a combination of toxic plus nutritional insults must be necessary for the development of the disorder.

112. a. The most common etiologies of bilateral central or cecocentral scotomas include hereditary optic neuropathy and nutritional optic neuropathy (vitamin B12 and folate deficiency), drug toxicity, tobacco-alcohol amblyopia, and infiltrative disorders such as syphilis and tuberculosis. Cyanide levels are not helpful in suspected tobacco-alcohol amblyopia.

113. c. Although disc edema, disc hemorrhages, and choroidal rupture may be seen in acute traumatic optic neuropathy, the most common finding is a normal fundus. Disc pallor would be unusual in the acute setting but present in all cases after several weeks.

114. d. The development of diabetic papillopathy appears to be independent of serum glucose levels. Diabetic papillopathy is classically seen in young adults with type 1 diabetes with moderate to severe retinopathy (although it can be seen in patients with type 2 diabetes as well). It is painless, and associated visual loss is generally mild. The disorder generally resolves spontaneously.
Fortification spectra that precede visual loss suggest
117. d. The symptoms presented in this question are indicative of pituitary apoplexy, in which there is a hemorrhage into a pituitary tumor. A sudden severe headache is a common presenting symptom; other manifestations depend on the direction of the expansion of pituitary gland. Acute painful visual loss is uncommon with the other disorders.

118. d. Fortification spectra that precede visual loss suggest a diagnosis of migraine headaches. A fixed visual defect suggests a cerebral arteriovenous malformation (AVM). A family history of headaches and nausea associated with visual disturbances are more suggestive of a migraine. Field defects may be found during acute migraine headaches, but those present interictally suggest another diagnosis.

119. e. The patient described has limitation of upgaze and downgaze in the affected eye. Making the distinction between a restrictive and a paralytic process by doing forced duction testing would be the best way to further narrow the differential diagnosis.

120. e. These findings, as well as right hypertropia in left gaze and left hypertropia in right gaze, are typical features of bilateral fourth nerve palsy.

121. b. A skew deviation is a motility disturbance with a vertical component that does not have a pattern consistent with a discrete muscle underaction or nerve palsy. They are generally due to supranuclear or vestibulocochlear dysfunction and generally reflect brainstem disease. They are typically comitant but not always.

122. c. Although transient changes in vertical eye movements and slowing of vertical saccades can result from lesions of the paramedian pontine reticular formation, nuclei crucial for the initiation of vertical eye movements are the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) and the interstitial nucleus of Cajal (INC).

123. False. Small vessel disease is the most common cause of unilateral sixth nerve palsy. More common causes of bilateral sixth nerve palsy include increased intracranial pressure, head trauma, and tumors of the ventral brainstem. Congenital bilateral sixth and seventh nerve palsies are characteristic of Möbius' syndrome.

124. False. An isolated sixth nerve palsy in children is most commonly attributable to postviral inflammation occurring 1 to 3 weeks following a nonspecific viral illness of the upper respiratory tract. Recovery is generally complete and occurs within 10 to 12 weeks.

125. c. Medial rectus contracture is distinctly uncommon in Duane's syndrome. In congenital sixth nerve palsy, it is quite common and results in esotropia in primary position. Although the left eye is more commonly involved in Duane's syndrome, this does not help in distinguishing this syndrome from a congenital sixth nerve palsy.

126. b. Disruption of the medial longitudinal fasciculus (MLF), which carries projections of interneurons from the contralateral sixth nerve nucleus to the ipsilateral medial rectus subnucleus, results in ipsilateral absence of adduction and contralateral abduction nystagmus. This combination of findings is termed internuclear ophthalmoplegia (INO). Vertical nystagmus and skew deviations are frequently found in association with INO but are not universal.

127. c. Bilateral internuclear ophthalmoplegia (INO) is often associated with upbeat nystagmus.

128. False. Bilateral internuclear ophthalmoplegia (INO) is more frequent in demyelinating disease than in cerebrovascular disease. This is because the brainstem blood supply is lateralized—right and left circulations are usually discrete and end at the midline. Demyelination does not respect the midline.

129. e. A lesion of the abducens nucleus results in an ipsilateral gaze paresis caused by disruption of the motor neurons and internuclei to the contralateral medial rectus mediating gaze conjugacy by the contralateral medial longitudinal fasciculus (MLF). A lesion that disrupts both the abducens nucleus and the ipsilateral MLF will result in the combination of an ipsilateral gaze palsy and internuclear ophthalmoplegia (INO). This combination has been termed the “one-and-a-half” syndrome. The only horizontal eye movement that remains is contralateral abduction.

130. c. Exotropia in primary position can occasionally occur in association with a bilateral internuclear ophthalmoplegia (INO), resulting in a syndrome called wall-eyed bilateral INO.

131. d. Miller-Fisher syndrome is generally considered a variant of Guillain-Barré syndrome that results in ophthalmoplegia, ataxia, and areflexia. Serum immunoglobulin G (IgG) autoantibodies and elevated cerebrospinal fluid (CSF) protein may be present. Complete recovery is common.
132. False. The presence of optic nerve dysfunction, manifested by decreased vision, an afferent pupillary defect, and/or dyschromatopsia, distinguishes an orbital apex syndrome from a cavernous sinus syndrome because the optic nerve passes through the optic canal and does not enter the cavernous sinus.

133. Tolosa-Hunt syndrome is thought to be caused by a nonspecific inflammatory process of the cavernous sinus. Findings include painful ophthalmoplegia, sensory deficits of the trigeminal nerve (most commonly the ophthalmic division), and a dramatic response to systemic corticosteroid therapy. Remissions may be spontaneous, with partial or complete reversal of deficits. Episodic recurrence also can occur. Aneurysms of the circle of Willis can produce a similar picture, including steroid responsiveness. A normal magnetic resonance imaging (MRI) makes this unlikely but not impossible. Tolosa-Hunt syndrome is a diagnosis of exclusion.

134. Iris and posterior segment neovascularization, as well as rapidly progressive cataract, may all be seen as complications of the ischemic oculopathy that these fistulae generate. Corneal exposure caused by proptosis can produce a similar picture, including steroid responsiveness. A normal magnetic resonance imaging (MRI) does not image calcium, this modality is not useful. As described in answer 137, fundus photography through standard fluorescein filters will detect autofluorescence if the drusen are sufficiently near the surface of the nerve head.

135. True. In the “low-flow” fistulae, the head trauma may be only minor. More significant head trauma is the most frequent cause of high-flow carotid-cavernous fistulae and is present in up to 75% of cases. Spontaneous rupture caused by hypertension or atherosclerosis is the cause in most of the remaining cases.

136. Acute thiamine deficiency (Wernicke’s encephalopathy) can result in central scotomas as well as ophthalmoplegia, primarily affecting cranial nerves III and VI. It can be precipitated in nutritionally depleted alcoholics given intravenous glucose alone because of sudden consumption of systemic thiamine stores.

137. Autofluorescence is produced when certain tissues/material are stimulated with monochromatic blue light and emit in the yellow-green range, as fluorescein does. The two optic nerve lesions that may autofluoresce are astrocytic hamartomas and drusen. Large accumulations of lipofuscin also may autofluoresce. To demonstrate this, fundus photographs should be obtained through the standard fluorescein setup, but without fluorescein injection. Lesions will appear bright, as if they had absorbed fluorescein, although none was injected. Note that this is not the same as a “red-free” photograph (which is produced with a green filter that does not provide sufficient blue light to stimulate autofluorescence).

138. Giant cell arteritis (GCA) is common in northern European climates. On the basis of autopsy studies, the prevalence has been estimated to be 1.1% of the Scandinavian population.

139. b. Polymyalgia rheumatica (PMR) is a surprisingly common disorder. Patients with PMR are at higher risk for developing giant cell arteritis (GCA).

140. c. Chiasmal field defects are characterized by greater loss centrally. Postfixation blindness is a necessary concomitant of bitemporal hemianopia. Objects behind the point of fixation are in the temporal hemifield of each eye. With loss of these fields, nothing beyond the point of fixation is visible. Reds and greens often appear “washed out” in the temporal hemifield of affected patients. “Hemifield slip” refers to the diplopia these patients may also notice. By mechanisms that are not entirely understood, binocular input at the vertical midline seems necessary for motor fusion. When a substantial portion of the vertical midline is not overlapped by both visual fields, there may be loss of motor fusion with resultant diplopia.

141. a. The calcium that is contained in optic nerve drusen is visible on computed tomography (CT) scanning and ultrasonography. Because magnetic resonance imaging (MRI) does not image calcium, this modality is not useful. As described in answer 137, fundus photography through standard fluorescein filters will detect autofluorescence if the drusen are sufficiently near the surface of the nerve head.

142. c. The Pulfrich phenomenon probably reflects delayed conduction in the demyelinated nerve. Oscillating objects perceived by the affected eye appear to be behind the image seen with the healthy eye, simulating three-dimensional movement where there is only movement within one plane.

143. b. The delayed implicit time is the electrophysiologic correlate of the bizarre perception known as the Pulfrich phenomenon (see answer 142).

144. c. The history is suggestive of a subacute optic neuropathy. The examination confirms this and reveals pseudodisc edema: blurred, indistinct disc margins with dilated, telangiectatic peripapillary capillaries but no dye leakage on angiography. This constellation is pathognomonic for Leber’s hereditary optic neuropathy (LHON). The disease is typically asynchronous and bilateral. Many patients have asymptomatic but important electrocardiographic abnormalities, most commonly a preexcitation syndrome. It is theorized that the added metabolic burden of tobacco and alcohol use may exacerbate the defect in oxidative metabolism caused by the defects in mitochondrial and nuclear deoxyribonucleic acid (DNA) (see question 145).

145. d. As is now well understood, the primary genetic defect underlying Leber’s hereditary optic neuropathy (LHON) resides in the mitochondrial deoxyribonucleic acid (DNA) of every somatic cell. This defect is inherited strictly from the cytoplasm of the maternal egg. As paternal sperm carry no mitochondria, the disease is maternally inherited. There is clearly a gender predilection,
with 80% to 90% of affected individuals being men. Fifty percent of affected individuals will have a point mutation at position 11778 of the mitochondrial DNA, resulting in impaired adenosine triphosphate (ATP) production. No treatment has been proven effective.

146. c. The right eye’s adduction is clearly slowed, indicating a right internuclear ophthalmoplegia (INO). The lesion is always on the same side of the brainstem as the eye with impaired adduction. Therefore, her lesion is on the right side of her brainstem. Some patients with myasthenia gravis (MG) will present with a pseudo-INO virtually indistinguishable from a true INO. Any other associated motility disorders, particularly ptosis or fatigability of adduction, should prompt a Tensilon test. Traumatic INO has a good prognosis for recovery. If convergent adduction is preserved, the term “posterior” INO has been used historically. Anterior INO implies a more rostral lesion, more likely to impair convergence as well as saccadic adduction.

147. c. Patients with internuclear ophthalmoplegia (INO) typically show “abduction nystagmus” of the contralateral eye. Therefore, a patient with a right INO may have nystagmus when looking to the left.

148. d. Anisocoria aggravated by cocaine implies Horner’s syndrome. Cocaine blocks reuptake of catecholamines at the synapse. In Horner’s syndrome, the catecholamine level at sympathetic synapses (iris dilator) is lower, so the mydriasis normally caused by cocaine is lessened. Hydroxyamphetamine stimulates release of catecholamines from the presynaptic neuron at the effector junction. If hydroxyamphetamine fails to dilate the abnormal, miotic pupil (in this case, the right pupil), the lesion must be postganglionic. In Horner’s syndrome, the miotic pupil is the abnormal one, caused by a relative lack of sympathetic stimulation. In the acute phase, the affected eye may be injected (lack of sympathetic vasodilation), hypotonomous (reduced aqueous secretion), and mildly inflamed (breakdown of the blood–ocular barrier). The so-called Pancoast’s tumor, a primary pulmonary carcinoma of either upper lobe, may compress the second order sympathetic neurons, causing a preganglionic Horner’s syndrome.

149. b. Painful Horner’s is an acute dissection of the intracarotid artery until proven otherwise. The diagnosis of exclusion then becomes migraine if the angiogram is normal.

150. d. Bilaterally elevated discs with indistinct margins raise the possibility of papilledema, particularly in an obese young woman (pseudotumor cerebri). The photo demonstrates two findings arguing against this diagnosis. First, the vessels are clearly visible, with no obscuration of their borders. This is a sensitive finding in true papilledema. Second, two or three tiny excrescences with a crystalline appearance within the neural rim are visible. The right-hand figure reveals either auto- or pseudofluorescence (hyperfluorescence despite no injection of fluorescein). The clinical findings are consistent with a lesion known to be associated with autofluorescence, optic nerve drusen. Typically, visual acuity is normal, although there may be associated field defects. Axoplastic stasis is a feature of true papilledema, rather than pseudopapilledema, which is present here. Autofluorescence is well described for both optic nerve drusen and astrocytic hamartomas of the optic nerve head. Both lesions may appear quite similar clinically. A distinguishing feature is the hypervascularity of astrocytomas, best revealed with fluorescein angiography (FA).

151. d. The visual fields reflect a bitemporal hemianopia. This indicates chiasmal compression. Although this may occur with severe hydrocephalus of any cause, the primary lesion is usually parasellar. In this case, there is a huge pituitary tumor, probably nonsecreting; the chiasm can be seen riding on top of this impressive mass. Figure a. is a large occipital infarct, b. is a parietal lobe metastasis, c. is a meningioma of the falx cerebri, and e. is a left optic nerve glioma.

152. a. The corkscrew conjunctival vessels, all the way to the limbus, are highly suggestive of arteriolarization of the orbital venous system. With a history of recent trauma, carotid–cavernous fistula is likely. All his clinical findings are consistent with this, as well. Figure b. represents a large intracavernous right carotid artery aneurysm. c. shows severe bilateral Graves’ disease, d. is a “berry” aneurysm of the posterior communicating artery, and e. shows bilateral optic disc drusen.

153. d. The computed tomography (CT) scan reveals absence of the septum pellucidum, a thin layer of serous connective tissue separating the two lateral ventricles. This is clearly associated with bilateral optic nerve hypoplasia and hypothalamic–pituitary disturbances, typically underfunctioning. Aicardi’s syndrome features mental retardation, lacunar retinopathy, and/or congenital absence of the corpus callosum. Precocious puberty may be a part of polyostotic fibrous dysplasia (Albright’s syndrome). Ash-leaf macules and pheochromocytoma may be seen in various phakomatoses (tuberous sclerosis, neurofibromatosis, and von Hippel-Lindau disease).

154. e. This magnetic resonance imaging (MRI) demonstrates central nervous system anatomy at the level of the midbrain. A unilateral brainstem lesion here could result in numerous defects. Weber’s syndrome refers to an intramedullary cranial nerve (CN) III lesion within the substance of the cerebral peduncle where descending fibers of the corticospinal tract run. This results in third nerve palsy and contralateral hemiplegia (the motor fibers within the corticospinal tract decussate at the caudal medulla, distal to the lesion of Weber’s syndrome).

155. e. Sumatriptan is a serotonin antagonist available orally, by injection, or as a nasal spray. The “-triptan”
antimigraine drugs are used for symptomatic relief of migraines but are contraindicated in patients with basilar artery migraine. They can produce myocardial infarction (MI) and should be used with caution in patients with severe hypertension or coronary artery disease.

156. d. Methysergide is an ergot alkaloid with numerous potential side effects. The beta-blockers are effective and safer.

157. c. Patients in the surgical group were significantly more likely to lose at least three lines of acuity.

158. e. Pseudotumor cerebri typically (not universally) presents with visual loss and headaches. Neurologic abnormalities are usually absent except for occasional abducent paralysis. Although the opening pressure is elevated, cerebrospinal fluid (CSF) protein levels are either normal or low.

159. d. Binding antibodies to acetylcholine receptors are found in 90% of patients with generalized myasthenia gravis (MG).

160. d.

161. c. The disease is indistinguishable from myasthenia gravis (MG), with an onset averaging 6 months after initiation of therapy. Approximately 80% of patients will remit completely within 6 to 8 months of cessation.

162. b. Pupillary fibers from the optic tract exit before reaching the lateral geniculate body (LGB) and exit into the pretectal olivary nuclei.

163. c. The nasociliary nerve is a division of cranial nerve (CN) V1 and supplies the cornea.

164. a. This is a classic description of Anton’s syndrome.

165. d. The Purkinje effect is a benign condition. It is a real, subjective test and can rarely be used to determine whether a patient’s retina is attached.

166. a. Binocular diplopia is relieved by covering one eye. Monocular diplopia is relieved by covering the affected eye. All of the other answers are potential causes for monocular diplopia.

167. c. All of the above conditions (a. to d.) can result in anisocoria greater in dim light.

168. False. There are a host of systemic conditions that can become exacerbated during pregnancy. Pseudotumor cerebri is not one of them.

169. e. All of the other conditions (a. to d.) can be associated with central retinal artery occlusion (CRAO).

170. c. Craniopharyngioma is the most common cause of seesaw nystagmus. Other parasellar tumors can cause the condition as well.

171. False. Spasmus nutans usually does resolve by 6 years of age, but it is difficult to differentiate clinically from monocular nystagmus of childhood (which can be caused by a chiasmal glioma). Hence, any patient with suspected spasmus nutans should undergo neuroimaging.

172. b. Confrontation is usually not to the patient’s benefit. Symptoms may often clear on follow-up examination. None of the other answers are appropriate.

173. e. The fundus photograph demonstrates a morning glory disc anomaly, a unilateral condition usually accompanied with severe visual loss. It can also cause relative afferent pupillary defect (RAPD). The other answers are correct.

174. d. Optic nerve head melanocytomas are dark brown lesions on the optic nerve head that are typically benign but can rarely undergo malignant transformation. They can produce relative allergic photodermatitis (APDs) and visual field defects and should be followed up to document growth.

175. a. The fundus photograph demonstrates a retinal racemose angioma. Wyburn-Mason syndrome is a sporadic condition, characterized by a retinal racemose angioma with an intracranial arteriovenous malformation (AVM). Patients with Wyburn-Mason syndrome can have orbital AVMs as well (and consequent ocular bruises). As of 2004, there have been no reports of photodynamic therapy (PDT) to treat Wyburn-Mason syndrome (and it is unlikely that PDT would be effective in patients with this condition).

176. d. Optic nerve head melanocytomas are dark brown lesions on the optic nerve head that are typically benign but can rarely undergo malignant transformation. They can produce relative allergic photodermatitis (APDs) and visual field defects and should be followed up to document growth.

177. d. Optic nerve head melanocytomas are dark brown lesions on the optic nerve head that are typically benign but can rarely undergo malignant transformation. They can produce relative allergic photodermatitis (APDs) and visual field defects and should be followed up to document growth.

178. a. 3, b. 4, c. 5, d. 1, e. 2. The pathways for visual processing are not yet entirely characterized but appear to depend heavily on the dominant parietal lobe, usually the left. Right homonymous hemifields transmit information to the left occipital lobe, then to the angular gyrus of the left parietal lobe. Left homonymous hemifields are perceived by the right occipital lobe. Higher functions require processing through the splenium of the corpus callosum to the angular gyrus. This pathway may explain some of the following clinical observations. A left parietal lesion involving the angular gyrus results in the inability to process visual information: alexia with agraphia. A lesion of the corpus callosum results in the inability to process information from the left homonymous hemifield
(right occipital cortex). A left occipital lesion prevents processing of the right homonymous hemifields. However, if the left parietal lobe remains intact, reading and writing ability are not affected. The inferior occipitotemporal junction subserves color sensation for the entire contralateral hemifield. For complete (both right and left hemifields) cerebral dyschromatopsia, bilateral lesions are necessary.

179. a. 2, b. 3, c. 1. Oculomotor neurons leave their nuclear complex, then pass through the red nucleus and the medial portion of the cerebral peduncle. Therefore, involvement of adjacent structures will have additional clinical characteristics. Involvement of the red nucleus produces a contralateral “rubral” tremor, and cerebral peduncle involvement produces contralateral hemiparesis. Finally, superior cerebellar peduncle involvement may produce ataxia.

180. a. 2, b. 3, c. 1. A migraine equivalent is a scintillating scotoma that may be followed by nausea and photophobia but is not associated with a headache. A common migraine is a severe throbbing headache associated with nausea and photophobia but with no other neurologic or visual disturbance. A classic migraine is a severe throbbing headache preceded by scintillating scotoma or other auras and followed by nausea and photophobia.

181. a. 3, b. 5, c. 2, d. 3, e. 4, f. 2, g. 1, h. 5, i. 4. The geniculate ganglion is a condensation of nerve fibers without any true synapses. At this “ganglion,” the greater superficial petrosal nerve branches off the facial nerve carrying preganglionic secretory fibers for the lacrimal gland. These fibers synapse in the pterygopalatine ganglion. The gasserian ganglion is synonymous with the trigeminal ganglion. Note that the pterygopalatine and sphenopalatine ganglia are also one and the same.

■ Suggested Readings

1. T or F The medial walls of each orbit are parallel.
2. T or F The lateral walls of each orbit are parallel.
3. T or F The widest dimension of the orbit is at the anterior orbital rim.
4. T or F The volume of the average human orbit is approximately 2 tablespoons (30 mL).
5. The globe is least protected by the orbit and, therefore, most vulnerable to trauma:
   a. superiorly.
   b. laterally.
   c. inferiorly.
   d. medially.
   e. the globe is equally protected on all four sides.
6. The strongest orbital wall is the:
   a. roof.
   b. medial wall.
   c. floor.
   d. lateral wall.
   e. all four walls are equally strong.
7. The weakest orbital wall is the:
   a. roof.
   b. medial wall.
   c. floor.
   d. lateral wall.
   e. all four walls are equally strong.
8. Within which orbital bone are the optic foramen and canal contained?
   a. frontal.
   b. ethmoid.
   c. lesser wing of sphenoid.
   d. greater wing of sphenoid.
   e. palatine.
9. Which of the following terminal branches of the ophthalmic artery anastomose with branches of the external carotid system?
   1. the supraorbital artery.
   2. the lacrimal artery.
   3. the dorsonasal artery.
   4. the angular artery.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
10. Which sinus system aerates first?
    a. frontal.
    b. ethmoid.
    c. sphenoid.
    d. maxillary.
    e. pyriform.
Questions

11. Which of the following orbital disorders is/are marked by prominent pain?
   1. orbital pseudotumor.
   2. thyroid ophthalmopathy.
   3. malignant mixed tumor of the lacrimal gland.
   4. optic nerve sheath meningioma.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

12. Which one of the following disorders typically produces downward and lateral displacement of the globe?
   a. thyroid ophthalmopathy.
   b. malignant mixed tumor of the lacrimal gland.
   c. frontal sinus mucocele.
   d. squamous cell carcinoma of the maxillary sinus.
   e. optic nerve sheath meningioma.

13. All of the following disorders typically present with a rapid onset except:
   a. orbital cellulitis.
   b. rhabdomyosarcoma.
   c. benign mixed tumor of the lacrimal gland.
   d. bacterial dacryoadenitis.
   e. ruptured dermoid cyst.

14. T or F Hypertelorism and exorbitism are synonymous.

15. The upper limit of normal for exophthalmometry in white men is:
   a. 16 mm.
   b. 20 mm.
   c. 22 mm.
   d. 25 mm.
   e. 28 mm.

16. The upper limit of normal for exophthalmometry in African American men is:
   a. 16 mm.
   b. 20 mm.
   c. 22 mm.
   d. 25 mm.
   e. 28 mm.

17. The most frequent cause of bilateral proptosis in adults is:
   a. lymphoma.
   b. cavernous hemangioma.
   c. carotid-cavernous fistula.
   d. thyroid ophthalmopathy.
   e. sphenoid wing meningioma.

18. The most common cause of unilateral proptosis in adults is:
   a. lymphoma.
   b. cavernous hemangioma.
   c. carotid-cavernous fistula.
   d. thyroid ophthalmopathy.
   e. sphenoid wing meningioma.

19. The most common cause of unilateral proptosis in children is:
   a. acute leukemia.
   b. orbital cellulitis.
   c. orbital pseudotumor.
   d. thyroid ophthalmopathy.
   e. rhabdomyosarcoma.

20. T or F Unlike in children, enlargement of orbital dimensions in an adult implies a chronic process.

21. Each of the following findings is at least as common in pediatric Graves’ disease as in the adult form of the disorder except:
   a. emotional lability.
   b. goiter.
   c. polyphagia.
   d. exposure keratopathy.
   e. cardiovascular hyperactivity.

22. T or F One disadvantage of computed tomography (CT) scan compared with conventional tomography is its higher radiation dosage.

23. Most ocular and orbital ultrasound is performed in the range of:
   a. 10 Hz.
   b. 10 kHz.
   c. 10 MHz.
   d. 20 kHz.
   e. 20 MHz.

24. T or F Resolution of ultrasound images increases with stimulus frequency.
25. T or F Ultrasound depth penetration increases with stimulus frequency.

26. Which method of echography provides the greatest amount of dynamic information, particularly for vascular structures?
   a. standardized echography.
   b. A-scan echography.
   c. B-scan echography.
   d. Doppler echography.
   e. immersion echography.

27. Orbital echography reveals a rounded extraconal lesion with well-defined, smooth borders and very low internal reflectivity. Which one of the following is the most likely tissue diagnosis?
   a. orbital neurofibroma.
   b. sinus mucocele with orbital extension.
   c. orbital lymphangioma.
   d. carcinoma metastatic to the orbit.
   e. orbital pseudotumor.

28. A 34-year-old woman presents with explosive proptosis, chemosis, photophobia, and pain with exquisite tenderness. There is global limitation of ocular motility and enlargement of the lacrimal gland clinically. There is no discharge, fever, or leukocytosis. Orbital ultrasonography may have characteristics including:
   1. regular, smooth, rounded borders.
   2. poor internal sound transmission (high reflectivity).
   3. prominent vascular pulsation.
   4. enlargement of extraocular muscles (EOMs).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

29. State-of-the-art computed tomography (CT) scans permit a spatial resolution of:
   a. 1 cubic mm.
   b. 5 cubic mm.
   c. 1 cubic cm.
   d. 5 cubic cm.
   e. 10 cubic cm.

30. Which of the following features should be requested to maximize information gained from orbital computed tomography (CT) scanning?
   1. thin slices (1- to 2-mm cuts).
   2. intrathecal metrizamide injection.
   3. direct (nonreconstructed) coronal sections.
   4. 1.5-tesla.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

31. Which of the following features is/are necessary for generation of magnetic resonance images (MRI)?
   1. high-power magnetic field.
   2. atomic nuclei with electrical dipoles.
   3. radio frequency energy pulses.
   4. presence of water or calcium.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

32. T or F The magnetic field strength used to generate magnetic resonance images (MRI) approaches that of the Earth’s magnetic field.

33. Which one of the following statements regarding the physics of magnetic resonance imaging (MRI) is false?
   a. Nuclear dipoles are aligned with each other by a strong external magnetic field.
   b. The direction of nuclear dipole alignment is acutely altered by application of a radio frequency pulse, either 90 degrees or 180 degrees away from original alignment.
   c. The times required for the return of dipole alignment with the external magnetic field—relaxation times—determine the scan’s appearance.
   d. Substances with long T1 times (longitudinal relaxation time) appear bright on T1-weighted images.
   e. Substances with long T2 times (transverse relaxation time) appear bright on T2-weighted scans.
Questions

34. T or F Vitreous humor appears bright on T1-weighted images.

35. T or F Orbital fat appears bright on T1-weighted images.

36. Compared with magnetic resonance imaging (MRI), which of the following may be considered advantages of computed tomography (CT) scanning?
   1. better definition of bony tissues.
   2. better definition of soft tissues.
   3. ability to detect metallic foreign bodies.
   4. better definition of chiasmal.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

37. Relative to computed tomography (CT) scanning, magnetic resonance imaging (MRI) has which of the following advantages?
   1. greater patient comfort.
   2. lower cost and greater accessibility.
   3. less motion artifact.
   4. wider selection of scanning planes.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

38. T or F Anophthalmos is more common than microphthalmos.

39. T or F Normal orbital development is dependent on the presence of a grossly normal globe during childhood.

40. Craniofacial cleft syndromes that affect the orbit, eyelids, or eye include:
   1. Treacher Collins’ syndrome.
   2. Apert’s syndrome.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

41. The most common location for an orbital meningocele is the:
   a. medial canthus.
   b. infraorbital notch.
   c. lateral canthus.
   d. supraorbital notch.
   e. lacrimal gland fossa.

42. The most common orbital or eyelid finding in the craniosynostosis syndromes is:
   a. blepharophimosis and ptosis.
   b. V-pattern exotropia.
   c. hypertelorism and proptosis.
   d. ankyloblepharon.
   e. large orbit with apparent enophthalmos.

43. Features serving to differentiate preseptal cellulitis from orbital cellulitis include:
   1. pain with eye movement.
   2. diplopia with limitation in ductions.
   3. afferent pupillary defect (APD).
   4. marked warmth and tenderness of periorbital tissues.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

44. T or F In adults, preseptal cellulitis usually spreads rapidly to involve the orbits with bacteremia and meningitis.

45. The species most frequently implicated in preseptal cellulitis in adults is:
   a. Staphylococcus species.
   b. Streptococcus species.
   c. Haemophilus species.
   d. Pseudomonas species.
   e. Bacillus species.

46. The most common risk factor for the development of preseptal cellulitis is:
   a. recent dental surgery.
   b. recent upper respiratory infection.
   c. recent orbital fracture.
   d. recent skin trauma.
   e. sinusitis.
47. Drugs of choice in the treatment of typical preseptal cellulitis include:
   1. Trimethoprim-sulfamethoxazole.
   2. Dicloxacillin.
   3. Tetracycline.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

48. The agent most likely to cause a severe preseptal cellulitis leading to secondary orbital cellulitis and central nervous system (CNS) infection in infants and toddlers is:
   a. *Staphylococcus aureus*.
   b. *Streptococcus pneumoniae*.
   c. *Haemophilus influenzae*.
   d. *Streptococcus pyogenes*.
   e. *Neisseria meningitidis*.

49. The most common risk factor for the development of orbital cellulitis is:
   a. recent upper respiratory infection.
   b. ethmoid sinusitis.
   c. recent dental extraction.
   d. preseptal cellulitis.
   e. recent orbital fracture.

50. Decreased visual acuity in the setting of suspected orbital cellulitis must be evaluated with which of the following tests?
   1. magnetic resonance imaging (MRI).
   2. swinging flashlight test.
   3. fluorescein angiography (FA).
   4. computed tomography (CT) scanning.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

52. Tests that are important for distinguishing infectious orbital cellulitis from inflammatory orbital pseudotumor include:
   1. oral temperature.
   2. orbital computed tomography (CT) scan.
   3. complete blood count (CBC) with differential.
   4. erythrocyte sedimentation rate (ESR).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

53. A 19-year-old patient presents to the ophthalmologist with redness of his left eye. The skin of the upper and lower left eyelids is red and inflamed. There is a small eschar on the left upper lid medially. Visual acuity is normal. The patient is orthotropic in primary gaze, and ductions appear full bilaterally. On far right gaze, the patient develops a small exotropia. The deviation is slightly more noticeable when he fixes with his left eye. Hertel exophthalmometry reads 16 mm in the right eye and 18 mm in the left eye. There is no orbital tenderness, but he describes a slight discomfort with right gaze. Oral temperature is 99.2°F. White blood count is 9,100 with a normal differential. The examination finding that most strongly argues against the diagnosis of simple preseptal cellulitis is:
   a. exophthalmometry.
   b. ocular motility findings.
   c. external examination findings (erythema, warmth).
   d. oral temperature.
   e. white blood cell count.

54. The patient in question 53 undergoes computed tomography (CT) scanning, which reveals a dense left ethmoid sinusitis with irregular densities of the orbital fat on the left. The patient is hospitalized and started on intravenous antibiotics. Appropriate antibiotic choices for these findings include:
   1. nafcillin.
   2. cefuroxime.
   3. chloramphenicol.
   4. cefazolin.
Questions

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

55. Following admission, the clinical status of the patient in questions 53 and 54 slightly deteriorates over the initial 12 hours. This is followed by a 24- to 36-hour period of modest improvement. After 2 days, however, 3 mm of proptosis persists, and there is still pain with attempted adduction in the left eye. These findings suggest the possibility of:
a. cavernous sinus thrombosis.
b. spread of infection to adjacent sinuses.
c. inappropriate choice of antibiotic.
d. subperiosteal abscess.
e. meningitis.

56. What test must be performed to evaluate the findings described in question 55?
a. orbital ultrasonography.
b. ocular ultrasonography.
c. cerebral angiography.
d. lumbar puncture.
e. orbital computed tomography (CT) scanning.

57. The diagnostic test of choice in question 56 is performed and confirms suspicions. What therapeutic step must now be undertaken?
a. surgical drainage.
b. intrathecal antibiotics.
c. pars plana vitrectomy with intraocular antibiotics.
d. use of broader spectrum intravenous antibiotic agents.
e. orbital decompression.

58. T or F The phakomatoses are generalized disorders featuring multiple hamartomas—disordered growths containing tissue elements not normally found at the site of involvement.

59. Which one of the following regarding dermoid and epidermoid cysts is false?
a. They share a common pathophysiology.
b. The key distinguishing feature between the two is the nature of the wall of the cystic cavity.
c. Superficial cysts present more often during childhood.
d. In adults, nearly all of these lesions are anterior to the orbital septum.
e. Dermoid cysts may induce bony erosion on radiography.

60. Features shared by dermoid cysts and lipoder-moids include:
1. typical location.
2. general surgical strategies.
3. gross pathology.
4. cellular constituents on histopathology.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

61. T or F The definition of an orbital teratoma is a lesion containing all three germinal cell layers.

62. T or F Orbital teratomas may simulate malignancy but rarely, if ever, metastasize.

63. More than 90% of periocular capillary hemangio-mas manifest by:
a. 4 to 8 weeks of life.
b. 6 to 8 months of age.
c. 12 to 18 months of age.
d. 24 to 30 months of age.
e. 3 to 4 years of age.

64. Most capillary hemangiomas reach their peak size at approximately what age?
a. 2 to 3 months.
b. 6 to 12 months.
c. 12 to 24 months.
d. 3 to 5 years.
e. 5 to 7 years.

65. Potentially important complications of capillary hemangiomas in childhood are:
1. occlusion amblyopia.
2. proptosis with exposure keratopathy.
3. significant astigmatism.
4. ocular invasion.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
66. The nevus flammeus, which may be mistaken for a capillary hemangioma, is seen as a part of what systemic disorder?
   a. neurofibromatosis.
   b. Treacher Collins’ syndrome.
   c. von Hippel’s disease.
   d. Sturge-Weber syndrome.
   e. Goldenhar’s syndrome.

c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

72. T or F Orbital lymphangiomas are believed to develop from abnormal sequestrations of lymphatic channels from the remainder of the orbital lymphatic system.

73. Which of the following regarding the histopathology of orbital lymphangioma is/are true?
   1. unlike capillary hemangioma, the cystic spaces are large.
   2. lymphoid tissue, even follicles, may be found within the substance of the tumor.
   3. loculated areas of old hemorrhage, “chocolate cysts,” may be found.
   4. there is generally a well-defined fibrous capsule surrounding the tumor.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

74. Which of the following regarding the surgical therapy of orbital lymphangioma is/are true?
   1. careful dissection will generally lead to complete tumor excision.
   2. significant cosmetic deformity and recurrent orbital hemorrhage are the most common indications for surgical therapy.
   3. recurrence is rare.
   4. carbon dioxide laser, contact Nd:YAG laser, or orbital decompression techniques may be helpful in the surgical therapy of this tumor.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

75. Which of the following orbital tumors is least likely to present as a “masquerade syndrome”?
   a. optic nerve glioma.
   b. orbital dermoid cyst.
   c. rhabdomyosarcoma.
   d. acute leukemic orbital infiltration.
   e. metastatic neuroblastoma.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
76. Which one of the following clinical findings at presentation is inconsistent with the diagnosis of optic nerve glioma?
   a. insidious onset.
   b. afferent pupillary defect (APD).
   c. pain.
   d. axial proptosis.
   e. unilaterality.

77. Which one of the following radiographic features is considered pathognomonic for optic nerve glioma?
   a. adjacent bony erosion.
   b. “kinking” of the optic nerve.
   c. multiple cystic cavities within the optic nerve.
   d. “tram-track” enlargement of the optic nerve.
   e. traction on posterior globe with “tenting.”

78. The study of choice for defining the presence or extent of intracranial involvement with optic nerve glioma is:
   a. polytomography.
   b. ultrasonography.
   c. computed tomography (CT) scanning.
   d. magnetic resonance imaging (MRI).
   e. carotid angiography.

79. T or F Incisional biopsy of suspected optic nerve glioma may be easily misinterpreted as optic nerve meningioma.

80. Modalities that may be of use in the treatment of optic nerve glioma include:
   1. observation.
   2. radiotherapy.
   3. surgical resection.
   4. chemotherapy.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

81. Indications for therapeutic intervention for optic nerve glioma include:
   1. presence of afferent pupillary defect (APD).
   2. rapidly progressive tumor growth.
   3. proptosis.
   4. involvement of intracranial optic nerve with field loss in the contralateral eye.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

82. A patient with known neurofibromatosis presents with pulsating proptosis of long duration. Computed tomography (CT) scan of the orbit will most likely reveal:
   a. orbital neurofibroma.
   b. cavernous hemangioma.
   c. abnormality of the sphenoid bone.
   d. optic nerve glioma.
   e. carotid-cavernous fistula.

83. A 7-year-old boy presents with a 3-day history of progressive proptosis, injection, and pain of the left eye. He is systemically well with normal temperature. White blood cell count is normal, and emergent orbital computed tomography (CT) scanning reveals superonasal orbital infiltration with bony erosion. The diagnosis that must be excluded at this point is:
   a. bacterial orbital cellulitis.
   b. optic nerve glioma.
   c. frontal sinus mucocele.
   d. rhabdomyosarcoma.
   e. orbital neurofibroma.

84. Which one of the following regarding a biopsy of orbital rhabdomyosarcoma is false?
   a. Care must be taken to prevent local seeding or distant dissemination with tumor cells.
   b. Electron microscopic studies are frequently necessary to secure the diagnosis.
   c. In cases of high suspicion, lumbar puncture and bone marrow biopsy should be performed at the time of biopsy.
   d. The embryonal pattern is the most common pathologic variant, accounting for >80% of total cases.
   e. The most malignant variant, alveolar rhabdomyosarcoma, occurs most frequently in the superior orbit.

85. T or F The cell of origin for most rhabdomyosarcomas is the myocyte of extraocular muscle (EOM).

86. T or F Like in adults, metastases of nonocular primary tumors in children more frequently involve the eye (uvea) than the orbit.
87. Fungal species likely to lead to a necrotizing orbital cellulitis include:
   1. *Candida*.
   2. *Aspergillus*.
   3. *Fusarium*.
   4. *Mucor*.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

88. T or F Both mucormycosis and aspergillosis of the orbit occur only in patients with immune dysfunction (e.g., patients with diabetes).

89. A 33-year-old patient with type 1 diabetes mellitus presents with a 2-week history of gradually progressive proptosis, redness, and irritation in the left eye. Visual acuities are 20/20 in the right eye and 20/100 in the left eye. There is an afferent pupillary defect (APD) on the left, along with 4 mm of proptosis and moderate conjunctival infection. Ductions are normal on the right and globally reduced on the left. Computed tomography (CT) scanning reveals left ethmoid and maxillary sinusitis with evidence of orbital involvement. The next step in the proper evaluation of this patient is:
   a. complete blood count (CBC).
   b. blood glucose level.
   c. careful ear, nose, and throat evaluation.
   d. oral temperature.
   e. orbital magnetic resonance imaging (MRI).

90. Which of the following orbital or adnexal tumors affect women significantly more frequently than men?
   1. cavernous hemangioma.
   2. sebaceous cell carcinoma.
   3. meningioma.
   4. fibrous histiocytoma.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

91. A patient presents to the ophthalmologist with a history of gradually progressive prominence of the left eye. There are no visual complaints, although the patient notes a mild throbbing pain intermittently on the left side of his head. Visual acuities are normal and symmetric, and there is 4 mm of axial proptosis on the left. There is no lid retraction evident in primary gaze. There is questionable lid lag on downgaze. The eye examination is otherwise normal, including no evidence of inflammation. The historical feature most convincingly arguing against the diagnosis of Graves’ ophthalmopathy is:
   a. male sex.
   b. unilateral involvement.
   c. throbbing sensation.
   d. full-eye movements.
   e. no lid retraction.

92. Surveillance of the patient with Graves’ ophthalmopathy must include:
   1. visual acuity testing.
   2. visual field testing.
   3. color vision testing.
   4. fluorescein angiography (FA).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

93. T or F If laboratory evidence of a dysthyroid state fails to develop within 10 years of the diagnosis of Graves’ ophthalmopathy, the ophthalmic diagnosis should be questioned seriously.

94. Surgical procedures that may be indicated during the active, inflammatory phase of Graves’ ophthalmopathy include:
   1. strabismus surgery.
   2. lateral tarsorrhaphy.
   3. recession of the levator aponeurosis.
   4. orbital decompression.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

95. A 45-year-old woman presents with mild bilateral proptosis and lid retraction. There is no previous
history of thyroid disease, and the patient denies any periocular pain. The one blood test that is most likely to be of value in this circumstance is:

- a. total thyroxine (T4) levels.
- b. free T4 levels.
- c. total 3,5,3'-triiodothyronine (T3) levels.
- d. thyroid hormone index (total T4 and thyroid-binding globulin levels).
- e. sensitive thyroid-stimulating hormone (TSH) levels—immunometric assay.

96. T or F The pathologic changes seen in extraocular muscle (EOM) involvement with Graves’ ophthalmopathy reflect primary inflammatory destruction of muscle cells.

97. T or F The histopathologic changes evident in extraocular muscle (EOM) specimen from patients with Graves' ophthalmopathy also may be seen in the lacrimal glands.

98. Which one of the following signs is considered classic for computed tomography (CT) scanning in Graves’ ophthalmopathy?

- a. nodular muscle enlargement.
- b. solitary muscle enlargement.
- c. “kinking” of extraocular muscles (EOMs).
- d. fusiform muscle enlargement with sparing of tendons.
- e. enhancement of posterior sclera with intravenous contrast injection.

99. A patient with brittle type 1 diabetes and labile hypertension presents with obvious Graves’ ophthalmopathy. There is an afferent pupillary defect (APD), decreased visual acuity, and an abnormal visual field, all on the right side. The patient is markedly hyperthyroid, and general anesthesia is felt to be risky as the patient is on the brink of thyroid storm. The patient’s blood sugar level is in the mid-200 mg/dL range, and renal function studies are normal. Which of the following interventions might be appropriate for control of vision-threatening ophthalmopathy?

- 1. lateral tarsorrhaphy.
- 2. intravenous corticosteroids.
- 3. oral corticosteroids.
- 4. orbital radiation.

100. Before undertaking surgical correction of strabismus in Graves’ ophthalmopathy, the angle of deviation should be stable for what period of time?

- a. 1 month.
- b. 3 months.
- c. 6 months.
- d. 1 year.
- e. 2 years.

101. Typical manifestations of idiopathic orbital inflammation (pseudotumor) include all of the following except:

- a. dacryoadenitis.
- b. peripheral ulcerative keratitis.
- c. extraocular myositis.
- d. periscleritis.
- e. optic perineuritis.

102. Clinical findings that are more likely in pediatric orbital pseudotumor than in the adult variety of the disease include all of the following except:

- a. bilateral involvement.
- b. systemic symptoms and signs (malaise, fever, vomiting).
- c. minimal periocular pain.
- d. peripheral eosinophilia.
- e. uveitis.

103. Which of the following are features of sclerosing orbital pseudotumor?

- 1. insidious onset.
- 2. more frequently steroid-resistant than other variants.
- 3. may be misdiagnosed as an orbital neoplasm.
- 4. pain is a less prominent complaint.

104. A patient with presumed inflammatory orbital pseudotumor is treated with oral prednisone, 60 mg, daily for 2 weeks. No improvement in clinical findings is seen. The next therapeutic step should be:

- a. doubling of oral prednisone dosage.
- b. induction with intravenous methylprednisolone.
- c. orbital irradiation.

105. A patient with presumed inflammatory orbital pseudotumor is treated with oral prednisone, 60 mg, daily for 2 weeks. No improvement in clinical findings is seen. The next therapeutic step should be:

- a. doubling of oral prednisone dosage.
- b. induction with intravenous methylprednisolone.
- c. orbital irradiation.
d. oral cyclophosphamide.
e. orbital biopsy.

105. Which of the following regarding the biopsy findings of orbital pseudotumor is/are true?
1. Compared with orbital lymphoma, the lesions are relatively hypocellular.
2. A microscopic specimen of involved extraocular muscle (EOM) could be indistinguishable from that of Graves’ ophthalmopathy.
3. The presence of many eosinophils in an adult’s biopsy may suggest the presence of an underlying systemic vasculitis.
4. The presence of neutrophils makes the diagnosis of pseudotumor unlikely.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

106. Computed tomography (CT) scan findings that differentiate orbital pseudotumor from Graves’ ophthalmopathy include:
1. enlargement of multiple extraocular muscles (EOMs).
2. enlargement of EOM tendons.
3. unilateral involvement.
4. enhancement of posterior sclera with intravenous contrast injection.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

107. All of the following disorders may be associated with a clinical presentation indistinguishable from typical inflammatory orbital pseudotumor except:
a. systemic lupus erythematosus (SLE).
b. polyarteritis nodosa.
c. Wegener’s granulomatosis.
d. sarcoidosis.
e. Churg-Strauss syndrome.

108. For which of the following disorders is bilateral, painless enlargement of the lacrimal glands considered a typical presentation?
1. Sjögren’s syndrome.
2. sarcoidosis.
3. inflammatory orbital pseudotumor.
4. benign lymphoepithelial lesions.

109. Which of the following regarding cavernous hemangioma of the orbit is/are true?
1. Women are more frequently affected than men.
2. It is the most common etiology of neoplastic unilateral proptosis in adults.
3. A-scan ultrasonography reveals high internal reflectivity.
4. There is generally an associated ocular bruit.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

110. T or F Hemangiopericytomas are more likely to limit ocular motility than cavernous hemangiomas.

111. T or F The most important prognostic indicator for systemic mortality from orbital hemangiopericytoma is cellular morphology on biopsy.

112. A 28-year-old man presents to an ophthalmologist 1 week after being in a motor vehicle accident with resultant blunt head trauma. He complains of irritation and redness of the left eye. Visual acuity is normal bilaterally. There is marked conjunctival injection of the left eye, with prominence of the superficial and deep vessels all the way to the limbus. There is 5 mm of proptosis on the involved side. Slit-lamp examination is normal. Intraocular pressures (IOPs) are 16 mm Hg in the right eye and 25 mm Hg in the left eye. Funduscopic examination is normal on the right but reveals dilated, tortuous retinal veins on the left. On careful questioning, the patient reports hearing a rushing noise intermittently. Auscultation of the left orbit reveals a faint bruit. “Arterialization” of his orbit is most likely caused by a disturbance in the:
a. intraorbital central retinal artery.
b. intracranial ophthalmic artery.
c. intracavernous internal carotid artery.
d. cervical common carotid artery.
e. branches of the middle meningeal artery.
113. A 73-year-old woman presents to the ophthalmologist complaining of mild redness and irritation of her left eye for approximately 2 months. She denies any head trauma. Visual acuity is normal bilaterally. There is 3 mm of proptosis on the left with mild to tortuosity of retinal vasculature oculus sinister (OS). Intraocular pressure is (IOP) 12 mm Hg oculus dexter (OD) and 30 mm Hg OS. “Arterialization” of her orbit is most likely caused by a disturbance in the:
   a. intraorbital central retinal artery.
   b. intracranial ophthalmic artery.
   c. intracavernous internal carotid artery.
   d. cervical common carotid artery.
   e. branches of the middle meningeal artery.

114. T or F The clinical finding considered classic for orbital varices is an audible bruit with corkscrewing of conjunctival vessels.

115. T or F Most orbital meningiomas arise outside the orbit and invade secondarily.

116. T or F The growth patterns of meningiomas associated with neurofibromatosis are indistinguishable from isolated tumors.

117. Which of the following regarding the radiographic evaluation of meningioma is/are true?
   1. Associated bony changes may be either osteolytic or osteoblastic.
   2. Angiography generally reveals a highly vascularized tumor (“tumor blush”).
   3. Computed tomography (CT) scanning is the modality of choice in the initial evaluation of a patient with optic nerve meningioma.
   4. Magnetic resonance imaging (MRI) is particularly useful for evaluating suspected intracranial extension.

118. T or F Excisional biopsy of optic nerve meningioma generally results in visual loss.

119. Indications for the removal of orbital meningioma include:
   1. severe loss of vision.
   2. progressive tumor growth in a child.
   3. evidence of intracranial extension.
   4. afferent pupillary defect (APD).

120. T or F Anterior visual pathway gliomas in adults are particularly malignant, but only when associated with neurofibromatosis.

121. In the general ophthalmologist’s practice, what percentage of lacrimal gland lesions will be inflammatory or lymphoid?
   a. 10%.
   b. 20%.
   c. 40%.
   d. 50%.
   e. >75%.

122. The most common neoplasm of the lacrimal gland is the:
   a. adenoid cystic carcinoma.
   b. malignant mixed tumor.
   c. benign mixed tumor.
   d. adenocarcinoma.
   e. mucoepidermoid carcinoma.

123. Which of the following historical features are considered essential in the clinical evaluation of lacrimal gland lesions?
   1. presence or absence of double vision.
   2. presence or absence of pain.
   3. presence or absence of dry eye symptoms.
   4. duration of symptoms.

124. Which radiographic features of computed tomography (CT) scanning are considered essential in the evaluation of lacrimal gland lesions?
   1. degree of enhancement with intravenous contrast injection.
   2. soft-tissue outlines of glandular enlargement.
3. presence or absence of glandular calcification.
4. presence or absence of adjacent bony change.
  a. 1, 2, and 3.
  b. 1 and 3.
  c. 2 and 4.
  d. 4 only.
  e. 1, 2, 3, and 4.

125. A 30-year-old man presents with a 2-year history of gradually progressive proptosis of the right eye. There is no associated pain or inflammation. Computed tomography (CT) scanning reveals globular enlargement of the lacrimal gland with no extension anterior to the orbital rim. Which of the following statements about this condition is true?
  a. The most likely diagnosis is more frequently encountered in men.
  b. Initial approach to the patient should include an incisional biopsy.
  c. Any surgical intervention must be undertaken with very careful dissection.
  d. Definitive treatment will necessitate lateral orbitotomy.
  e. Histopathology may reveal areas of cartilage.

126. The most common malignant neoplasm of the lacrimal gland is the:
  a. pleomorphic adenoma.
  b. malignant mixed tumor.
  c. adenoid cystic carcinoma.
  d. mucoepidermoid carcinoma.
  e. adenocarcinoma.

127. T or F Adenoid cystic carcinoma of the lacrimal gland is frequently painful because of extreme reactive inflammation and orbital swelling.

128. Which of the following regarding fibrous dysplasia is/are true?
  1. Orbital involvement in a girl younger than 10 years is associated with a high probability of precocious puberty.
  2. The hallmark of the histopathology is aggregation of lamellar bone.
  3. The treatment of choice is high-dose radiotherapy.
  4. The most frequent visual sequelae are related to optic nerve compression.
  a. 1, 2, and 3.
  b. 1 and 3.
  c. 2 and 4.
  d. 4 only.
  e. 1, 2, 3, and 4.

129. Which one of the following histiocytic disorders is most likely to involve orbital bone?
  a. Letterer-Siwe disease.
  c. eosinophilic granuloma.
  d. juvenile xanthogranuloma (JXG).
  e. sinus histiocytosis.

130. T or F The therapy for localized eosinophilic granuloma is identical to that for the disseminated histiocytoses.

131. Which of the following is one of the most common mesenchymal tumor of the orbit?
  a. ossifying fibroma.
  b. fibrous dysplasia.
  c. hemangiopericytoma.
  d. fibrous histiocytoma.
  e. osteogenic sarcoma.

132. T or F Fibrous histiocytoma may behave in a locally aggressive fashion but rarely if ever metastasizes.

133. T or F Clinical or radiographic features frequently allow differentiation between reactive lymphoid hyperplasia and orbital lymphoma.

134. The systemic evaluation of a patient with biopsy-proven orbital lymphoma generally includes all of the following except:
  a. bone scan.
  b. bone marrow biopsy.
  c. liver and spleen scan.
  d. lumbar puncture.
  e. serum immunoelectrophoresis.

135. Key features supporting the diagnosis of lymphoproliferative lesions over idiopathic inflammatory orbital syndrome (pseudotumor) include:
  1. absence of significant fibrous stroma.
  2. polyclonal lymphocyte expansion.
  3. hypercellularity.
  4. cellular heterogeneity (lymphocytes plus plasma cells, eosinophils, neutrophils).
  a. 1, 2, and 3.
  b. 1 and 3.
Questions

c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

136. Exceptions to the rule of thumb that all orbital tumors are dark on T1-weighted magnetic resonance images (MRI) include:
   1. retrobulbar hemorrhage of at least 24 hours duration.
   2. melanoma.
   3. mucocele.
   4. meningioma.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

137. T or F The most common site of origin for tumors that secondarily invade the orbit is the cranial cavity.

138. The most common sinus lesion that invades the orbit is the:
   a. osteoma.
   b. mucocele.
   c. mucoepidermoid carcinoma.
   d. squamous cell carcinoma.
   e. inverted papilloma.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

139. The most common site of a primary tumor metastatic to the orbit in women is:
   a. breast.
   b. lung.
   c. ovary.
   d. colon.
   e. uterus.
140. The most common site/type of a primary tumor metastatic to the orbit in men is:
   a. colon.
   b. lung.
   c. carcinoid.
   d. prostatic.
   e. cutaneous melanoma.

141. Classic features of a “tripod” fracture include all of the following except:
   a. downward displacement of the lateral canthus.
b. infraorbital hypesthesia.
c. trismus.
d. deficit of upgaze.
e. temporal subconjunctival hemorrhage.

142. A sensitive finding in direct naso-orbital-ethmoid fracture is:
   a. subconjunctival hemorrhage.
   b. epistaxis.
   c. telecanthus.
   d. hypoglobus.
   e. infraorbital hypesthesia.

143. T or F Indirect orbital floor fractures are frequently associated with inferior orbital rim fractures.

144. Which of the following findings in orbital floor fractures is/are exacerbated by the presence of a coincident medial orbital wall fracture?
   1. infraorbital hypesthesia.
   2. enophthalmos.
   3. vertical diplopia.
   4. subcutaneous emphysema.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

145. All of the following findings are consistent with an inferior orbital wall fracture and entrapment except:
   a. infraorbital hypesthesia.
   b. enophthalmos.
   c. subcutaneous emphysema.
   d. limitation in ocular motility in all fields of gaze.
   e. hypoglobus.

146. T or F The diagnostic study of choice in probable orbital floor fracture is the Waters view (conventional radiograms).

147. T or F Forced duction testing is generally helpful in the acute setting to determine if ocular motility limitations are caused by entrapment or muscle dysfunction.

148. T or F If exophthalmometry performed at the time of orbital floor fracture does not reveal enophthalmos, it is unlikely to develop subsequently.
149. T or F Most diplopia associated with orbital contusion disappears within 7 to 14 days following injury.

150. Which of the following are considered indications for surgical repair of orbital floor fractures?
   1. disabling diplopia present 7 to 10 days after the original injury.
   2. large fracture on acute computed tomography (CT) scan.
   3. enophthalmos >2 mm.
   4. inferior rectus entrapment in a pediatric patient present 1 day after injury.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

151. T or F Larger, complex, comminuted floor fractures are more likely to lead to entrapment than small, posterior floor fractures.

152. The optimal time for surgical repair of orbital floor fractures is generally considered to be:
   a. within 24 hours of injury.
   b. 1 to 3 days following injury.
   c. 3 to 7 days following injury.
   d. 7 to 14 days following injury.
   e. 4 to 6 weeks following injury.

153. Which of the following are considered indications for removal of an intraorbital foreign body?
   1. vegetable foreign body.
   2. any lead foreign body.
   3. anterior, easily approachable foreign bodies.
   4. orbital apex location with good vision.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

154. A patient presents to an emergency room after being struck over the left eye with a baseball bat. Examination reveals a 24-year-old man in acute distress. Visual acuity is 20/20 in the right eye and barely detectable in the left eye. There is hemorrhagic chemosis 360 degrees in the left eye and a microhyphema. Funduscopic examination is normal with the exception of obvious arterial pulsations. Intracocular pressure (IOP) is 75 mm Hg. The next step to be taken is emergent:
   a. computed tomography (CT) scanning.
   b. intravenous methylprednisolone.
   c. exploration of the globe.
   d. lateral canthotomy and cantholysis.
   e. intravenous acetazolamide and mannitol.

155. A patient presents to an emergency room after tumbling off a bicycle and striking his head on the pavement. Examination reveals a 24-year-old man in mild discomfort holding his head. There is a 5-mm abrasion on the left temple. Visual acuity is 20/20 in the right eye and light perception in the left eye. There is an obvious left afferent pupillary defect (APD). There is no periorbital edema, and extraocular movements are full. The slit-lamp examination, fundus examination, and intraocular pressures (IOPs) are normal. Confrontation visual fields are full in the right eye. The next step to be taken is emergent:
   a. computed tomography (CT) scanning.
   b. oral prednisone.
   c. exploration of the globe.
   d. lateral canthotomy and cantholysis.
   e. intravenous acetazolamide and mannitol.

156. T or F The distinction between direct and indirect optic nerve trauma is the involvement of a foreign body in the former.

157. A computed tomography (CT) scan performed on the bicyclist described in question 155 is entirely normal. The next appropriate step to be taken in this situation is emergent:
   a. intravenous methylprednisolone.
   b. exploration of the globe.
   c. lateral canthotomy and cantholysis.
   d. intravenous acetazolamide and mannitol.
   e. optic nerve sheath decompression.

158. For which of the following clinical situations is enucleation, rather than evisceration, preferred?
   1. suspected intraocular malignancy.
   2. painful blind eye.
3. blind eye following significant penetrating trauma.
4. cosmetically unacceptable phthisical eye.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

159. The most common postoperative complication of enucleation is:
   a. superior sulcus deformity.
   b. socket contracture.
   c. enophthalmos.
   d. exophthalmos.
   e. extrusion of implant.

160. Which of the following features increase the risk of extrusion of orbital implants?
   1. implant too large for the orbit.
   2. conformer too large for the fornix.
   3. exposed synthetic integrated implants.
   4. orbital tissue infection.

   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

161. T or F The primary advantage of evisceration over enucleation is the lower probability of sympathetic ophthalmia (SO).

162. T or F The procedure of choice in severe socket contracture following enucleation involves the use of buccal mucosa grafts.

163. T or F The difference between subtotal and total exenteration is preservation of periorbita in the former.

164. T or F The difference between total and extended exenterations is preservation of bony orbital walls and sinuses in the former.

165. Which one of the following statements regarding the orbicularis oculi muscle is false?
   a. The orbital portion is primarily involved in forceful eyelid closure (winking and spasm).
   b. The palpebral portion is primarily involved in involuntary closure (blinking).
   c. The superficial medial head of the pretarsal portion of the muscle (Horner’s tensor muscle) is critical to adequate lacrimal drainage.
   d. The lateral terminations of the pretarsal portion of the muscle constitute the lateral canthal tendon.
   e. The lateral terminations of the preseptal portion of the muscle constitute the lateral palpebral raphe.

166. Which of the following regarding the orbital septum is/are true?
   1. The septum arises both superiorly and inferiorly from the periostium of the orbital rim.
   2. An intact, healthy orbital septum provides a relative barrier to intraorbital spread of subcutaneous infection.
   3. When fat is encountered intraoperatively, it implies retroseptal penetration.
   4. The orbital septum normally fuses with the levator aponeurosis in the upper lid in the same position as it does with the capsulopalpebral fascia in the lower lid.

   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

167. Which of the following regarding Whitnall’s ligament (superior suspensory ligament of the globe) is/are true?
   1. It is a condensation or extension of the sheath of the levator muscle.
   2. It acts as an important fulcrum for appropriate function of the levator muscle.
   3. It runs from the trochlea medially to the lateral orbital wall laterally.
   4. Standard ptosis surgery includes transection of Whitnall’s ligament.

   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
168. Which one of the following regarding the levator palpebrae superioris is false?
   a. The muscular portion is longer than the aponeurotic portion.
   b. It originates in close proximity to the superior rectus origin, just above the annulus of Zinn.
   c. Its superficial portion inserts into the orbicularis muscle and subcutaneous tissues.
   d. Its deeper portion inserts into the superior border of the tarsus.
   e. It runs from the posterior lacrimal crest medi ally to the lateral orbital tubercle laterally.

169. T or F Müller’s muscle (superior tarsal muscle) generally accounts for approximately 4 mm of palpebral fissure height.

170. T or F The superior and inferior tarsal muscles both insert into the peripheral border of their respective tarsi.

171. Which of the following regarding the lower eyelid retractors is/are true?
   1. The lower eyelid analog of the levator aponeurosis is the capsulopalpebral fascia.
   2. The capsulopalpebral fascia arises from the common sheaths of the inferior oblique and inferior rectus muscles.
   3. Both the capsulopalpebral fascia and the inferior tarsal muscle insert into the inferior conjunctival fornix.
   4. The orbital septum inserts into the capsulopalpebral fascia 3 to 4 mm inferior to the inferior tarsal margin.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

172. T or F The superior peripheral palpebral arterial arcade lies in a plane between the levator aponeurosis and Müller’s muscle.

173. T or F The accessory lacrimal glands of Krause are found in the conjunctival fornix, whereas the glands of Wolfring are found at the tarsal border in the subconjunctival space.

174. T or F The anterior insertion of the medial canthal tendon (MCT) is more important than the posterior insertion for maintenance of normal medial canthal appearance and function.

175. T or F “Antimongoloid” slant of the palpebral fissure occurs when the lateral canthus is >3 mm higher than the medial canthus.

176. The structure(s) that give(s) rise to the gray line of the eyelid margin is/are:
   a. meibomian glands.
   b. Moll’s glands.
   c. tarsal border.
   d. the mucocutaneous junction.
   e. the marginal strip of pretarsal orbicularis muscle.

177. The normal horizontal extent of the human palpebral fissure is approximately:
   a. 20 mm.
   b. 25 mm.
   c. 30 mm.
   d. 35 mm.
   e. 40 mm.

178. T or F The palpebral arterial arcades provide numerous anastomotic connections between internal and external carotid circulations.

179. Which of the following is/are considered defining features of the blepharophimosis syndrome?
   1. ptosis.
   2. epicanthus tarsalis.
   3. telecanthus.
   4. lower lid ectropion.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

180. T or F The blepharophimosis syndrome is generally inherited on an autosomal dominant basis.

181. The systemic disorder most commonly associated with the blepharophimosis syndrome is:
   a. hypospadias.
   b. dry, redundant skin.
   c. primary amenorrhea.
   d. diabetes mellitus.
   e. coarctation of the aorta.
182. T or F Congenital coloboma of the upper eyelid is generally isolated, whereas that of the lower lid is more commonly associated with other facial abnormalities.

183. T or F In cryptophthalmos, the eye underlying the lid defect is generally normal.

184. Disadvantages of intralesional steroid treatment of chalazia relative to incision and curettage include:
   1. lower success rate.
   2. potential depigmentation of overlying skin.
   3. steroid-induced glaucoma.
   4. higher incidence of infection.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

185. The most likely outcome following inadvertent suturing of the orbital septum into subcutaneous tissues when repairing a partial-thickness eyelid laceration is:
   a. ectropion.
   b. entropion.
   c. kink of the lid margin.
   d. lid retraction in downgaze.
   e. ptosis.

186. The most reliable sign that the orbital septum has been involved in a partial-thickness lid laceration is:
   a. ptosis.
   b. orbital fat herniation.
   c. entropion.
   d. ectropion.
   e. lid retraction in downgaze.

187. T or F In medial canthal tendon (MCT) avulsion, the critical maneuver in reestablishing cosmetic and anatomic integrity is reattachment of the anterior limb of the MCT.

188. T or F Transnasal wiring or titanium plate fixation of the medial canthal tendon (MCT) is indicated in the presence of posterior MCT avulsion and naso-orbital-ethmoid fracture.

189. Which of the following donor sites are considered acceptable for free skin grafts in the setting of lid lacerations with tissue loss?
   1. retroauricular skin.
   2. contralateral upper eyelid.
   3. supraclavicular space.
   4. buttock skin.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

190. To allow maximal spontaneous return of function before surgical repair, it is generally wise to observe traumatic ptosis in an adult for what period of time?
   a. 4 weeks.
   b. 2 months.
   c. 3 months.
   d. 6 months.
   e. 12 months.

191. T or F Involutional ectropion is more common in the upper eyelid than in the lower eyelid.

192. Which of the following factors are important to evaluate before planning therapy for involutional ectropion?
   1. position of the inferior punctum.
   2. stability of the lower limb of the medial canthal tendon (MCT).
   3. stability of the lateral canthal tendon.
   4. presence or absence of contracture of the orbicularis muscle.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

193. An acceptable temporizing measure for paralytic ectropion and severe corneal exposure associated with typical Bell’s palsy might be:
   a. pentagonal wedge resection of the lower eyelid.
   b. temporary lateral tarsorrhaphy.
   c. frontalis brow suspension.
d. punctal electrocautery.
e. inferior retractor recession with full-thickness skin grafting of the lower lid.

194. T or F Adequate repair of severe cicatricial ectropion can usually be accomplished solely with scar relaxation techniques combined with horizontal tightening procedures.

195. T or F Involutional entropion is more commonly seen in the upper lid.

196. T or F The “snapback” test is useful in the evaluation of a patient with presumed involitional entropion.

197. T or F Digital pressure along the inferior border of the inferior tarsus will temporarily correct involitional entropion but not cicatricial entropion.

198. Which of the following is/are true regarding cryotherapy for trichiasis?
   1. The treatment is probably more effective than electrolysis.
   2. It employs a freeze-thaw-freeze technique.
   3. Local anesthesia is required to minimize pain.
   4. The only complication associated with the treatment is failure to eliminate trichiasis completely.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

199. Critical components in the evaluation of corneal protection mechanisms prior to ptosis surgery include all of the following except:
   a. assessment of lagophthalmos.
   b. Jones (primary dye) testing.
   c. assessment of the Bell phenomenon.
   d. Schirmer testing.
   e. assessment of corneal sensation.

200. The primary abnormality seen in simple congenital ptosis is in the:
   a. levator muscle.
   b. levator aponeurosis.
   c. levator innervation.
   d. third nerve nucleus.
   e. supranuclear control of lid function.

201. T or F In acquired myogenic ptosis, the amount of ptosis generally correlates well with the degree of residual levator function.

202. The primary abnormality seen in ptosis after cataract surgery is in the:
   a. levator muscle.
   b. levator aponeurosis.
   c. levator innervation.
   d. third nerve nucleus.
   e. supranuclear control of lid function.

203. T or F In aponeurotic ptosis, the amount of ptosis generally correlates well with the degree of residual levator function.

204. T or F The ptosis of myasthenia gravis (MG) almost always responds well to systemic anticholinesterase medication.

205. The ptosis associated with Marcus Gunn’s syndrome is because of aberrant connections between the levator muscle and which cranial nerve?
   a. V.
   b. VII.
   c. IX.
   d. X.
   e. XII.

206. The procedure of choice for moderate unilateral acquired ptosis associated with good levator function and a normal upper eyelid crease should be:
   a. Müller’s muscle resection.
   b. levator muscle resection.
   c. reinsertion of levator aponeurosis.
   d. bilateral frontalis suspension.
   e. unilateral frontalis suspension.

207. The procedure of choice in a child with asymmetric but bilateral, moderate to severe congenital ptosis would be:
   a. Müller’s muscle resection.
   b. levator muscle resection.
   c. reinsertion of levator aponeurosis.
   d. bilateral frontalis suspension.
   e. unilateral frontalis suspension.
Questions

208. The procedure of choice in a patient with ptosis following cataract surgery (good levator function and a high or effaced upper eyelid crease) would be:
   a. Müller’s muscle resection.
   b. levator muscle resection.
   c. reininsertion of levator aponeurosis.
   d. bilateral frontalis suspension.
   e. unilateral frontalis.

209. T or F Unlike thyroid eye disease, the lid retraction of the Collier sign features greater retraction of the lateral lid than the medial lid.

210. T or F A subset of patients with congenital ptosis will also have impaired ipsilateral upgaze but no other signs of third nerve palsy.

211. All of the following are included in the differential diagnosis of eyelid retraction except:
   a. thyroid eye disease.
   b. progressive supranuclear palsy.
   c. a history of superior rectus resection.
   d. dorsal midbrain compression.
   e. myasthenia gravis (MG).

212. T or F In the management of lid abnormalities and thyroid eye disease, inferior lid retraction is generally more easily and successfully repaired than superior lid retraction.

213. Which one of the following papillomatous lesions of the eyelid is considered premalignant?
   a. verruca vulgaris.
   b. seborrheic keratosis.
   c. actinic keratosis.
   d. nevus verruca.
   e. acanthosis nigricans.

214. Which of the following papillomatous lesions of the eyelid may be associated with underlying systemic malignancy?
   a. verruca vulgaris.
   b. ephelis.
   c. actinic keratosis.
   d. nevus verruca.
   e. acanthosis nigricans.

215. Which of the following eyelid lesions are typically slightly elevated with a central ulcerated area or crater?
   1. molluscum lesion.
   2. basal cell carcinoma.
   3. keratoacanthoma.
   4. verruca vulgaris.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

216. Which of the following regarding basal cell carcinoma of the eyelid is/are true?
   1. It is the most common eyelid malignancy.
   2. It may be clinically indistinguishable from sebaceous cell carcinoma of the eyelid.
   3. The morpheaform pattern has the worst prognosis.
   4. It more frequently involves the lower eyelid than the upper eyelid.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

217. Which one of the following regarding the treatment of basal cell carcinoma of the eyelid is false?
   a. The 5-year mortality rate for all cases of ocular basal cell carcinoma approaches 3%.
   b. Complete surgical excision is the treatment of choice in virtually all cases.
   c. Following Mohs’ resection of basal cell carcinoma, spontaneous granulation generally leads to the optimal cosmetic outcome.
   d. Cryotherapy of adnexal lesions may be considered in patients who are extremely poor candidates for surgery.
   e. Extirpation of medial canthal lesions may require resection of the lacrimal drainage system and/or subtotal exenteration.

218. T or F The clinical course of squamous cell carcinoma of the ocular adnexa is generally more indolent than that of basal cell carcinoma.

219. Which one of the following regarding sebaceous cell carcinoma is false?
   a. The primary focus may be either eyelid or caruncle.
   b. Recognition is often delayed because of misdiagnosis as innocent eyelid inflammation.
   c. Shave biopsy techniques are inadequate.
d. Similar to basal cell carcinoma, Mohs’ resection offers optimal cosmetic outcome and long-term survival for sebaceous cell carcinoma.

e. The hallmark of the histopathology of the condition include skip areas and pagetoid intraepithelial spread of malignancy.

220. Which one of the following regarding malignant melanoma of eyelid skin is false?

a. The incidence of this neoplasm is increasing.
b. Lentigo maligna melanoma and nodular melanoma are the most common forms affecting the eyelid.
c. Nodular melanoma is the most rare eyelid melanoma and has the worst prognosis.
d. The factor of greatest prognostic significance is depth of invasion.
e. Like conjunctival melanosis, eyelid melanoma may be controlled with cryotherapy.

221. T or F In general, eyelid marginal defects of 25% or less may be repaired by direct closure, whereas larger defects require grafting or advancement techniques.

222. Which of the following may serve to distinguish blepharochalasis from dermatochalasis?

1. Age of onset.
2. History of recurrent eyelid swelling.
3. Herniation of the orbital lobe of the lacrimal gland.
4. Coincident presence of true ptosis.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

223. The most significant complication of blepharoplasty is:

a. Overcorrection.
b. Undercorrection.
c. Diplopia.
d. Orbital hemorrhage.
e. Cellulitis.

224. All of the following favor the diagnosis of benign essential blepharospasm over hemifacial spasm except:

a. No involvement of lower facial muscles along with orbicularis muscle.
b. Bilateral involvement.
c. Absence of abnormal movements during sleep.
d. Synchronous contractures of involved muscles.
e. Lack of response to neurosurgical decompression of the facial nerve.

225. Which of the following regarding the treatment of essential blepharospasm is/are true?

1. Botulinum injections offer effective but temporary relief.
2. Meticulous stripping of orbicularis muscle fibers is a surgical alternative to botulinum injections.
3. Treatment of essential blepharospasm may exacerbate coexistent dry eye syndrome.
4. Facial nerve avulsion is a surgical modality preferable to orbicularis myectomy.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

226. Which of the following regarding the secretory apparatus of the lacrimal system is/are true?

1. The lacrimal gland is split into two lobes by the check ligament of lateral rectus muscle.
2. Surgical removal of the orbital portion of the lacrimal gland with preservation of the palpebral portion will result in normal reflex tear secretion.
3. Surgical removal of the palpebral portion of the lacrimal gland with preservation of the orbital portion will result in normal reflex tear secretion.
4. Efferent fibers to the lacrimal gland originate in the superior salivatory nucleus of the mid-pons.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

227. Which one of the following ganglia serves as the home for cell bodies providing postganglionic efferent innervation to the lacrimal gland?

a. Geniculate.
b. Superior cervical.
c. Ciliary.
d. sphenopalatine.
e. gasserian.

228. **T or F** The accessory lacrimal glands of Krause and Wolfring are partially responsible for basal tear secretion.

229. The average distance from lacrimal punctum to nasolacrimal sac is:
a. 2 mm.
b. 8 mm.
c. 10 mm.
d. 30 mm.
e. highly variable.

230. The average distance from lacrimal punctum to inferior nasal meatus is:
a. 2 mm.
b. 8 mm.
c. 10 mm.
d. 30 mm.
e. 50 mm.

231. **T or F** The flap of soft tissue that prevents reflux of nasal secretions into the nasolacrimal duct system is called the valve of Rosenmüller.

232. **T or F** The sites of obstruction in congenital and acquired nasolacrimal obstruction are identical.

233. Which one of the following functional tests of lacrimal drainage is most likely to yield a false-positive result?
a. primary dye test (Jones I test).
b. secondary dye test (Jones II test).
c. Schirmer test.
d. dye disappearance test.
e. lacrimal scintigraphy.

234. Which one of the following functional tests of lacrimal drainage allows identification of a failure of the lacrimal pump mechanism?
a. primary dye test (Jones I test).
b. secondary dye test (Jones II test).
c. Schirmer test.
d. dye disappearance test.
e. lacrimal scintigraphy.

235. **T or F** As with congenital impatency of nasolacrimal system, probing of the nasolacrimal duct in acquired impatency may have long-lasting therapeutic benefit.

236. Diagnostic evaluation in the patient complaining of tearing whose functional tests are normal might include:
1. examination of tear film.
2. tear breakup-time testing (TBUT).
3. complete Schirmer testing.
4. corneal sensation testing.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

237. **T or F** Both superior and inferior canalicular systems must be patent for normal tear drainage.

238. Common causes of canalicular obstruction include all of the following except:
a. congenital.
b. trauma.
c. phospholine iodide.
d. *Actinomyces* infection.
e. idoxuridine use.

239. A 65-year-old worker presents to his ophthalmologist having noted a lump on his eyelid which has grown over the past 6 to 12 months. The lesion is shown in the figure below. Which of the following biopsy specimens most likely belongs to this patient?

   ![Image of eye with lesion]

a. Figure a. on the following page.
b. Figure b. on the following page.
c. Figure c. on the following page.
d. Figure d. on the following page.
e. Figure e. on the following page.
240. A 72-year-old woman presents for a second opinion regarding a stye. It has been drained twice, but will not resolve. Its clinical appearance is shown in the figure below. Which biopsy belongs to this patient?

a. Figure a. in question 239.
b. Figure b. in question 239.
c. Figure c. in question 239.
d. Figure d. in question 239.
e. Figure e. in question 239.

3. emergent dacryocystorhinostomy (DCR).
4. oral antibiotics.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

243. Acute, lancinating pain in the medial canthal region with minimal noninflamed enlargement of the lacrimal sac is most subjective of:
a. acute dacryocystitis.
b. chronic dacryocystitis.
c. impacted dacryolith.
d. Actinomyces canaliculitis.
e. Wegener’s granulomatosis of the lacrimal sac.

244. T or F The most common malignant tumor of the lacrimal sac is adenocarcinoma.

245. T or F Congenital swelling of the medial canthus above the medial canthal tendon (MCT), in the absence of any active inflammation, probably represents congenital dacryocele.

246. Which of the following regarding congenital obstruction of the nasolacrimal system is/are true?
1. Approximately one third are bilateral.
2. If spontaneous resolution has not occurred by age 9 to 12 months, it is unlikely to occur.
Questions

3. Successful surgical management with probing alone may become significantly less likely after the age of 1 year.
4. General anesthesia is generally necessary after the age of 6 months.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

247. T or F Following unsuccessful probings of a congenitally impatent nasolacrimal system, the next appropriate step is dacryocystorhinostomy (DCR).

248. The most common site of organic obstruction in acquired nasolacrimal obstruction is:
a. punctum.
b. canaliculus.
c. valve of Rosenmüller.
d. intraosseous nasolacrimal duct.
e. valve of Hasner.

249. Which of the following constitute indications for dacryocystorhinostomy (DCR)?
   1. recurrent acute dacryocystitis.
   2. chronic discharge or symptomatic epiphora with a positive secondary dye test (organic obstruction).
   3. persistent epiphora in a child after probing and Silastic intubation of a congenitally impatent system.
   4. chronic discharge or symptomatic epiphora with a negative secondary dye test (functional obstruction).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

250. T or F In probing the nasolacrimal system of an infant with congenital stenosis, it is better to start with the superior canaliculus.

251. The surgical approach of choice in a patient complaining of epiphora with severe canicular trauma and scarring is:
a. dacryocystorhinostomy (DCR).
b. canaliculo-DCR.
c. conjunctivo-DCR.
d. Silastic intubation of the nasolacrimal system.
e. marsupialization of the canalicular system.

252. The way(s) to minimize removal of the incorrect eye during enucleation is/are:
a. reexamining the patient immediately prior to surgery.
b. marking the eye to be enucleated before surgery.
c. checking the patient’s consent form and chart prior to surgery.
d. having the patient point to which eye to enucleate.
e. all of the above.

253. Elevation of what structure to its previous anatomic position is often used to perform midface rejuvenation?
a. suborbicularis oculi fat (SOOF).
b. preseptal orbicularis.
c. lateral canthal tendon.
d. retro-orbicularis oculi fat.
e. none of the above.

254. What is the average onset of action of botulinum toxin?
a. immediately.
b. 6 to 12 hours.
c. 48 to 72 hours.
d. 1 week.
e. 3 weeks.

255. An external photograph of a patient with a chronic history of sinusitis is shown in the following figure (left). Slit lamp examination reveals sclerokeratitis as shown in the following figure (right). What would your likely diagnosis be?
a. mucormycosis.
b. Letterer-Siwe disease.
c. eosinophilic granuloma.
d. Wegener’s granulomatosis.
e. none of the above.
256. The best treatment for the patient in question 255 is:
a. observation.
b. topical corticosteroids.
c. oral corticosteroids.
d. high-dose burst of intravenous corticosteroids.
e. prednisone and cyclophosphamide.

257. The patient (shown in the figure below left) undergoes biopsy (results as shown in the figure below right). What is his likely diagnosis?
a. benign lymphoid hyperplasia.
b. orbital cellulitis.
c. mucormycosis.
d. orbital lymphoma.
e. none of the above.

258. Which of the following orbital tumors are generally not encapsulated?
a. schwannoma (neurilemmoma).
b. lymphangioma.
c. cavernous hemangioma.
d. hemangiopericytoma.
e. all of the above tumors are encapsulated.

259. The patient shown in the following figure (left) undergoes excision. The gross surgical specimen is shown in the following figure (middle). Histopathologic sectioning is shown in the following figure (right). What is the most likely diagnosis?
a. a rodent ulcer.
b. cavernous hemangioma.
c. lymphangioma.
d. hemangiopericytoma.
e. schwannoma.
260. **T or F** The tumor shown in the previous question often undergoes malignant transformation.

c. frontal, lesser wing of sphenoid.

d. ethmoid, lacrimal, sphenoid, and maxillary.

e. orbital floor.

261. What is the most common side effect of topical cyclosporine?

- conjunctival hyperemia (red eye).
- transient blurred vision.
- ocular stinging/burning.
- pruritus.
- epiphora.

262. **T or F** The use of topical cyclosporine is contraindicated in a patient with active blepharitis.

263. **T or F** Blepharochalasis is more common in elderly patients.

264. Which of the following would be the best treatment option for a localized orbital lymphoproliferative lesion?

- surgical excision.
- surgical excision combined with chemotherapy.
- radiation.
- radiation and systemic corticosteroids.
- surgical excision combined with radiation.

265. **Matching**

266. Match the lettered orbital structures listed below with the orbital wall that contains them.

a. infraorbital groove.
- orbital roof.

b. lacrimal gland fossa, trochea.
- lateral orbital wall.

c. lacrimal sac fossa.
- medial orbital wall.

d. orbital tubercle of Whitnall.
- orbital floor.

267. Match the lettered structures below with their appropriate portals of entry or exit from the intracranial space.

a. cranial nerve VI.
- optic canal.

b. cranial nerve III.
- foramen ovale.

c. potential route for spread of infectious sinusitis.
- inferior orbital fissure.

d. ophthalmic artery.
- foramen lacerum.

e. cranial nerve V3.
- ethmoidal foramina.

f. cranial nerve IV.
- superior orbital fissure.

g. superior ophthalmic vein.
- foramen rotundum.

h. cranial nerve II.
- sympathetic nerve fibers to the iris dilator muscle.

i. sympathetic nerve fibers to ocular and orbital blood vessels.

j. infraorbital nerve.

k. cranial nerve V2.

l. sympathetic nerve fibers to ocular and orbital blood vessels.

m. internal carotid artery.
268. Match the lettered descriptions listed below with the appropriate orbital soft tissues.
   a. travel(s) within fat compartments with radiating branches.
   b. fibrous origin of the extraocular muscles (EOMs).
   c. travel(s) in orbital septa in a complex of rings.
   d. continuous with the orbital septum anteriorly and dura posteriorly.
   e. splits lacrimal gland into two lobes.
   f. acts as suspensory ligament of the globe.
   g. three layers, thinnest posteriorly and thickest anteriorly.

269. For each of the lettered sinus systems listed below select the nasal area into which it drains.
   a. sphenoid sinus.
   b. frontal sinus.
   c. anterior ethmoid air cells.
   d. posterior ethmoid air cells.
   e. maxillary sinus.
   f. nasolacrimal duct.

270. Match the lettered blood tests below with the conditions for which they are most likely to be positive or helpful.
   a. angiotensin converting enzyme (ACE).
   b. antinuclear antibody (ANA).
   c. perinuclear antineutrophil cytoplasmic antibodies (p-ANCA).
   d. cytoplasmic antineutrophil cytoplasmic antibody (c-ANCA).
   e. anti-SS A and B antibodies.

271. Match the lettered clinical characteristics listed below with the histologic patterns.
   a. best prognosis.
   b. most differentiated (most likely to see cross-striations on light microscopy).
   c. more commonly occurs in inferior orbit.
   d. most common pattern seen in children.
   e. worst prognosis.
   f. least common of the primary orbital varieties.
   g. orbital involvement reflects secondary invasion from sinuses or conjunctiva.

272. Rank the extraocular muscles (EOMs) listed below in order of decreasing frequency of involvement with Graves' ophthalmopathy (1 being the most frequently involved and 4 being the least frequently involved).
   a. superior rectus.
   b. lateral rectus.
   c. inferior rectus.
   d. medial rectus.

273. A patient with a known history of hyperthyroidism presents with bilateral proptosis, unilateral decreased vision with associated afferent pupillary defect (APD), dyschromatopsia, and visual field defects. There is associated vertical strabismus and severe lid retraction with cosmetic significance bilaterally. The corneas are normal bilaterally. Arrange the surgical procedures listed below in their proper chronologic order (1 being the first procedure to be performed and 3 being the final procedure to be performed).
Questions

274. Match the lettered clinical characteristics below with the type of lacrimal gland lesion (the numbered choices may be used more than once).

a. pain.
   1. lymphoid.
b. adjacent osteolysis.
   2. inflammatory.
c. “pancake” enlargement of lacrimal gland.
   3. benign neoplasm.
d. symptoms present for >1 year.
   4. malignant neoplasm.
e. globular enlargement of lacrimal gland.
   5. 1 and 2.
f. adjacent bony molding (pressure-induced bony change).
   6. 2 and 4.
g. symptoms present for <1 year.

h. enlargement of lacrimal gland anterior to the orbital rim on computed tomography (CT) scanning.

i. normal adjacent bone on CT scanning.

275. Match the lettered radiographic characteristics listed below with the type of facial fractures.

a. craniofacial dysjunction.
   1. Le Fort I.
b. low transverse maxillary fracture.
   2. Le Fort II.
c. orbital floor, medial wall, lateral wall fractures.
   3. Le Fort III.
d. pyramidal maxillary fracture.
e. no orbital bony involvement.
f. medial orbital floor fractures.

d. symptoms present for >1 year.

h. enlargement of lacrimal gland anterior to the orbital rim on computed tomography (CT) scanning.

i. normal adjacent bone on CT scanning.

276. Arrange in order, from superficial (1) to deep (7), the structures listed below as encountered through an eyelid incision in the upper eyelid crease.

a. levator aponeurosis.
b. skin.
c. conjunctiva.
d. Müller’s muscle.
e. orbicularis oculi.
f. orbital fat.
g. orbital septum.

277. Match the anatomic definitions lettered below with the correct terms numbered below.

a. fold of skin over the medial canthus, broader in the upper lid.
   1. euryblepharon.
b. pretarsal muscle and skin riding above the lid margin with vertical eyelashes.
   2. ankyloblepharon.
c. fold of skin over the medial canthus, broader over the lower lid.
   3. epicanthus inversus.
d. horizontal widening of palpebral fissure associated with antimongoloid slant and loss of contact between the lid and globe.
   4. epicanthus tarsalis.
e. partial or complete fusion of eyelids laterally by webs of skin.
   5. epicanthus palpebralis.
f. fold of skin over the medial canthus that is equally broad in both the upper and lower eyelids.
   6. epiblepharon.
Answers

1. True. 
2. False. Each lateral orbital wall diverges at an angle of approximately 45 degrees from the midline.
3. False. The orbit is pear-shaped. Its widest dimension approximately 1 cm posterior to the anterior rim.
4. True. The average human orbit has a volume of approximately 30 mL.
5. b. Blunt orbital trauma from the temporal side is most dangerous to the globe because it is most exposed laterally. (Of course, there is no orbital protection anteriorly.).
6. d.
7. b. The anterior portion of the medial wall is also known as the lamina papyracea, or paper-thin layer.
8. c. Blunt cranial trauma is often transmitted to the sphenoid, with secondary indirect traumatic optic neuropathy as a consequence.
9. a. These anastomoses may prove critical in the setting of high-grade carotid stenosis. (The angular artery is the terminal branch of the facial artery, a branch of the external carotid system.)
10. b.
11. b. If pain is present, Graves' disease is usually not the cause of the orbitopathy.
12. c. The proptosis of Graves' disease is variable but usually axial (as is the case with meningioma). Lacrimal gland malignancies may produce proptosis, which is typically down and inward. Maxillary sinus tumors typically push the eye up.
13. c. The history of proptosis caused by pleomorphic adenoma is characteristically >6 to 12 months.
14. False. In hypertelorism, the medial orbital walls are separated excessively. In exorbitism, the lateral walls diverge excessively (>90 degrees).
15. c.
16. d.
17. d. This is a very common question. The most common cause of bilateral proptosis in children is not definitively established, but metastatic neuroblastoma and/or leukemia should be considered.
18. d.
19. b.
20. True. Children’s orbits are more plastic, so disorders with a short duration can cause orbital enlargement. This is not the case in adults.
21. d. In pediatric autoimmune hyperthyroidism, the systemic findings are more prominent and the orbital findings less common and less severe. Furthermore, for similar degrees of exophthalmos, children seem to develop exposure problems less often.
22. False. Although computed tomography (CT) scan does use greater radiation than plain films, it involves less than plain film tomography, which requires multiple exposures. An orbital CT scan administers a dose of 1 to 2 centigrays per scan.
23. c. Ocular ultrasound is usually performed in the range of 8 to 15 MHz. Higher frequencies give better resolution. Lower frequencies give better penetration.
24. True. See answer 23.
25. False. See answer 23.
26. d.
27. b. Neurofibromas will have high reflectivity. Lymphangiomata will have irregular borders. Metastatic cancer and pseudotumor will have irregular borders and high reflectivity.
28. c. Idiopathic orbital inflammation (pseudotumor) is the likely diagnosis.
29. a. This represents considerable improvement over first-generation scanners, whose resolution was frequently no greater than 5 cubic mm.
30. b. Orbital studies are incomplete without coronal sections (preferably nonreconstructed, which requires the patient to lie prone). Thin slices enhance sensitivity. Intrathecal metrizamide is rarely used since the advent of magnetic resonance imaging (MRI), which is enhanced with powerful 1.5-tesla magnets.
31. a. Magnetic resonance images (MRI) are generated by exposing dipolar molecules to a radio frequency pulse after alignment with a powerful external magnetic field. Calcium is bright on standard x-rays but dark (hypointense) and poorly imaged with MRI.
32. False. The magnetic field used for magnetic resonance imaging (MRI) is 2,000 to 15,000 times greater than the earth’s magnetic field.
33. d. Long T1 times render a substance hypointense (dark) on magnetic resonance images (MRI).
34. False. Vitreous appears dark on T1-weighted (long T1) and bright on T2-weighted (long T2) images.
35. True. Orbital fat behaves opposite to vitreous. To determine the type of magnetic resonance imaging (MRI), look for which is brighter. If the fat is brighter, it is a T1-weighted image. If the vitreous is brighter, it is a T2-weighted image.
36. b. Computed tomography (CT) scan is better for calcified structures. Magnetic resonance imaging (MRI) is superior for soft tissues and intracranial evaluation, and also may reduce dental artifact from dental fillings seen on CT scan. MRI is contraindicated in the setting of possible metallic foreign body because it may cause motion of the foreign body which can lead to serious injury to the patient.
37. d. Magnetic resonance imaging (MRI) offers simultaneously available axial, coronal, and sagittal images. It takes longer to generate, increasing motion artifact. The “box” that patients must lie in may be claustrophobic, although “open MRIs” can obviate this to a certain degree.
38. False. Anophthalmos is rare.
39. True. Enucleation in early childhood can lead to underdevelopment of the involved orbit. This is aggravated by irradiation.
40. b. Apert’s syndrome and Crouzon’s disease are synostoses, not cleft syndromes. Treacher Collins’ syndrome (mandibulofacial dysostosis) is autosomal dominant syndrome characterized by malar and mandibular hypoplasia, microstomia, lid colobomas, and ear abnormalities. Fifty percent of patients have downward displacement of the lateral canthus (anti-mongoloid slant). Goldenhar’s syndrome (oculoauriculo-vertebral dysplasia) is a sporadic or autosomal dominant syndrome of the first branchial arch, associated with lid colobomas, limbal dermoids, preauricular skin tags, aural fistulas, and vertebral abnormalities.
41. a. A bluish bulge above the medial canthal tendon (MCT) is typically a meningocoele, whereas one below is typically a dacryocoele.
42. c. The orbits of patients with craniosynostoses are usually shallow and small, with resultant proptosis and exposure. Y-pattern exotropia is also common.
43. a. Both forms of inflammation will feature superficial erythema (rubor) and warmth (calor). Decreased visual acuity or an afferent pupillary defect (APD) suggests involvement of the orbital apex and warrants aggressive management.
44. False. An intact orbital septum is an important, but not inviolable, barrier to infectious spread. Inappropriate or delayed treatment increases the risk. Rapid central spread of preseptal infection may be seen in infants; consequently, infants with preseptal cellulitis should immediately be given intravenous (IV) antibiotics.
45. a. *Staphylococcus aureus* is the most common cause of preseptal cellulitis secondary to trauma. *Streptococcus* organisms are a close second.
46. d.
47. c. Dicloxacillin offers some advantage against beta-lactamase–producing organisms. Cephalexin is less expensive, yet still efficacious. Ampicillin–sulbactam (Unasyn) or amoxicillin–clavulanic acid (Augmentin) also can be used.
48. c. Intravenous antibiotics are indicated for *Haemophilus influenzae* cellulitis in children. The HiB vaccine has significantly reduced the incidence of preseptal and orbital cellulitis secondary to *H. influenzae*.
49. b. The other options listed are also risk factors, but ethmoid sinusitis is the most common. Ninety percent of orbital cellulitis is secondary to paranasal sinus extension.
50. c. Decreased vision suggests an acute orbital apex syndrome, requiring prompt decompression.
51. e. This is one of the most dreaded complications of orbital cellulitis, with a significant increase in systemic morbidity and mortality.
52. a. Patients with orbital cellulitis are typically febrile. For pseudotumor, this is uncommon except in children. A leukocytosis with “left-shift” is also more common in cellulitis. The sedimentation rate may be elevated in either condition, and in both, computed tomography (CT) scan may show a nonspecific retrobulbar infiltrate but may have signs specific to one of the two disorders (e.g., muscle enlargement, subperiosteal abscess).
53. b. Two millimeters of discrepancy on exophthalmometry is the upper limit of normal. The external findings are consistent with either preseptal or orbital cellulitis. The “fever” is low grade and nonspecific. Strabismus indicates possible orbital inflammation.
54. a. First-generation cephalosporins (e.g., cefazolin, cephalexin, cephalothin) are only effective against half of the isolates of *Haemophilus influenzae*, and are generally insufficient to cover anaerobes. Second-generation agents (cefuroxime) are preferred. Also note that third-generation cephalosporins (e.g., cefotaxime) have less activity than first-generation cephalosporins against *Gram*-positive organisms, especially *Staphylococcus aureus*. Beta-lactamase–resistant penicillins, combined with the appropriate cephalosporin or chloramphenicol, are routinely used to treat orbital cellulitis. Chloramphenicol use has declined because of its association with a potentially lethal aplastic anemia.
55. d. Failure to dramatically improve after 48 to 72 hours of antibiotic can indicate a subperiosteal abscess. Relapse after switching to oral antibiotics also may be a sign. Inappropriate choice of antibiotic should not be associated with modest improvement.
56. e. An orbital computed tomography (CT) scan (preferably with thin, 1-mm direct coronal cuts) would be indicated to rule out subperiosteal abscess.
57. a. Antibiotic penetration into abscesses is notoriously poor. “Pus must pass.”
58. False. The phakomatoses are hamartomatous disorders. However, hamartomas are nests of abnormal tissue growth involving normally native tissue types. Hamartomas are abnormal tissue in a normal place (think of “hammer home”); choristomas are comprised of normal tissue in an abnormal location.
59. d. Both types are ectopic epithelial rests created by aberrant “pinching off” *in utero*. The variety that is silent until adulthood is generally intraorbital (retroseptal).
60. d. Dermolipomas are more frequently on the bulbar surface and are generally solid (rather than cystic). Surgical excision is much more difficult because of unsuspected extension and periorbital infiltration.
61. False. More than one of the three germinative layers (ectoderm, mesoderm, and endoderm) must be present to label a lesion a teratoma, but all three need not be.
62. True. They are frequently quite large at birth and thus simulate malignancy.
63. b. Greater than half are obvious by 1 to 2 months of age.
64. **b.** These tumors generally spontaneously regress, starting in the second year of life. Forty percent of tumors completely regress by age 4. Eighty percent of tumors completely regress by age 8.
65. **b.** These are two of the three primary indications for intervention. The third is significant cosmetic deformity.
66. **d.** The nevus flammeus (port-wine stain) is a cavernous hemangioma of the dermis. It can be seen in Sturge-Weber syndrome, Klippel-Trénaunay-Weber syndrome, and as an idiopathic finding. Some pathology textbooks also refer to the lesion as a dermal telangiectasia.
67. **d.** Capillary hemangiomata will blanch with pressure, whereas the nevus flammeus does not. There is considerable overlap between the other features listed.
68. **False.** Many affected children have a systemic angio- genic disorder including pulmonary, soft-tissue, and/or skin lesions. Thrombocytopenia in association with visceral capillary hemangiomas is known as Kasabach-Merritt syndrome.
69. **a.** Spontaneous involution can continue over 4 to 7 years.
70. **c.** Systemic or intralesional steroids may be associated with typical side effects (glaucoma and cataract), as well as rebound growth after cessation of therapy (steroid dependence). Steroid injections typically do cause soft-tissue atrophy and hypopigmentation. Radiation, rarely used except in the most severe cases, carries the most ominous risks, including future malignancy.
71. **b.** Neither capillary nor cavernous hemangiomas have high blood flow, and neither metastasize. Cavernous hemangiomas are rarely seen in youth, whereas, as the name of each implies, the blood-filled spaces are tiny in a capillary hemangioma and large in a cavernous hemangioma.
72. **False.** The origin of orbital lymphangioma remains unclear, although vascular malformations with lymphatic and venous components may play a role. Furthermore, noncontiguous vascular malformations have been reported in one study to occur in up to 25% of patients.
73. **a.** The lack of a well-defined capsule makes complete surgical excision very difficult. The growth pattern is infiltrative.
74. **c.** Surgical strategies are frequently limited to “debulking” by any safe means possible, because complete excision is usually not possible.
75. **a.** Each of the others may present and be treated as orbital cellulitis. Metastatic neuroblastoma also may simulate trauma, presenting with spontaneous periocular ecchymosis.
76. **c.** Each of the others is considered typical. Pain is quite unusual.
77. **b.** Tram-tracking of the optic nerve is considered classic for optic nerve sheath meningioma.
78. **d.** Magnetic resonance imaging (MRI) has been a major advance in the management of patients with optic nerve tumors.
79. **True.** The tumor may induce a secondary meningotheial hyperplastic response misinterpreted as meningioma.
80. **c.** The management of optic nerve glioma in children is a subject of much discussion and honest disagreement among the experts. Any of the options may be correct in a given situation, and the ultimate choice must be individualized to the patient’s circumstances (e.g., vision, size of tumor, extent of centripetal spread, age).
81. **c.** Most experts agree that intracranial spread (or a high likelihood of such), rapid growth simulating a malignanp variety, and loss of functional vision constitute indications for intervention.
82. **c.** Dysplasia or aplasia of the sphenoid bone creates large posterior orbital defects. This leads to pulsating exophthalmos, as brain tissue herniates outward.
83. **d.** Orbital pseudotumor should not cause bony erosion. Mucocele, glioma, and neurofibromas are rarely so inflammatory. Rhabdomyosarcoma responds well to treatment and should be promptly diagnosed.
84. **c.** All the statements are true except that alveolar rhabdomyosarcoma has a predilection for the inferior orbit. The embryonal variant is more common supranasally. Metastatic workup includes lumbar puncture and bone marrow biopsy, best done under anesthesia.
85. **False.** Rhabdomyosarcoma originates from undifferentiated pluripotential mesenchymal cells in the orbit. The tumor does not arise from malignant transformation of extraocular muscle (EOM).
86. **False.** Most periorbital metastases in children are orbital, unlike in adults. The most common is neuroblastoma.
87. **c.**穆尔, Rhizopus, and Aspergillus can lead to necrotizing orbital cellulitis. Diagnosis consists of biopsy. **Muco** and **Rhizopus** will show branching, nonseptate hyphae that stain with hemotoxin and eosin (H & E). **Aspergillus** will stain with Gomori methenamine-silver and will show branching septate hyphae. IV amphotericin B can be used for treatment.
88. **False.** Mucormycosis is generally seen only in the immunocompromised (chemotherapy, posttransplant, diabetes mellitus). Aspergillosis is probably more common in this population also, but it may be seen in otherwise healthy individuals, and it also may be seen as a result of allergic aspergillosis sinusitis.
89. **c.** Orbital phycomyosiosis generally results from invasion by necrotizing fungal sinusitis. Black eschar in the nasal cavity is virtually diagnostic but is a late finding. (Its absence does not exclude the diagnosis.)
90. **a.** Fibrous histiocytoia is slightly more common in men. The others are more common in women, by a ratio of at least 3:2.
91. c. If pain is present, Graves' disease is highly unlikely.
92. a. Decreasing acuity, relative afferent pupillary defect (APD), impairment of color vision, and evidence of scotomata may all be indicative of optic nerve compression. Maculopathy is uncommon in Graves' disease.
93. False. Thyroid-related orbitopathy can occur despite a persistently euthyroid state (as indicated by clinical and laboratory findings).
94. c. Acute corneal decompensation and evidence of optic nerve compression are indications for immediate surgical intervention. Strabismus surgery should be delayed until the inflammation is quiescent and the examination has stabilized. Levator aponeurosis recession may yield unpredictable results in the setting of active inflammation.
95. c. This is the most sensitive way to detect hyperthyroidism; feedback regulation can lead to a reliably detectable decrease in thyroid-stimulating hormone (TSH), even when fluctuating levels of thyroxine (T4) and 3,5,3'-triiodothyronine (T3) may not be indicative.
96. False. The atrophy of myofibrils is felt to be secondary to compression by surrounding edema. The latter occurs with deposition of glycosaminoglycans in the presence of inflammatory cell and fibroblast infiltration. Antibodies to the thyroid-stimulating hormone (TSH)-receptor expressed on fibroblasts may make them target cells for the inflammation observed in Graves' ophthalmopathy. Primary destruction of myocytes does not seem to occur.
97. True. Fibrosis tends to be less severe, leading to partial loss of the normal glandular tissue.
98. d. The enlargement generally features smooth contours. Although sometimes only a single muscle is enlarged, this should be considered an atypical finding. Posterior scleral enhancement is more suggestive of orbital inflammatory syndrome/pseudotumor or posterior scleritis.
100. c. This allows adequate time for stability of the condition to be established, allowing for a more predictable outcome.
101. b. Changes in corneal sensation may occur, but frank ulceration—especially peripherally—is unusual.
102. c. Orbital pseudotumor is generally quite painful for both children and adults.
103. e. The lack of pain as a prominent symptom contributes to the insidious progress of this condition and the possibility of misdiagnosis as a neoplasm. Its tendency to be diagnosed at a later stage, when fibrosis has already occurred, may contribute to the diminished response to steroids (relative to other forms of the disease).
104. c. Although a dosage of 60 mg of prednisone is at the lower end of the usual beginning ranges for steroid therapy, one should consider an orbital biopsy because of the possibility of other inflammatory diseases and other orbital pathology. After the diagnosis is confirmed, additional attempts should be made to bring about a response with stronger steroid therapy, orbital radiation, methotrexate, or cyclophosphamide.
105. a. The inflammatory infiltrate is polymorphous and may include neutrophils. Graves' myositis may look identical.
106. c. Unlike Graves' ophthalmopathy, pseudotumor can and commonly does involve the muscle tendons as well as their insertions, along with posterior Tenon fascia.
107. d. Sarcoidosis is generally not associated with pain and usually spares orbital soft tissues.
108. c. Sjögren's syndrome typically would present with keratoconjunctivitis sicca and/or a dry mouth, usually along with symptoms of (or a diagnosis of) a collagen-vascular disease like rheumatoid arthritis. Furthermore, the lacrimal glands are usually small. Orbital pseudotumor is usually painful.
109. a. These lesions generally do not have the high arterial blood flow usually associated with bruits. Their composition—densely packed vascular channels—leads to a high internal reflectivity on ultrasonography. Ocular bruits can be associated with carotid-cavernous fistulas and sphenoid wing dysplasia.
110. True. Hemangiopericytomas tend to be more rapidly expansile and aggressive tumors, increasing the likelihood of adversely affecting ocular motility.
111. False. Benign-appearing tumors of this type may turn out to be invasive and may metastasize. They are encapsulated and must be completely excised because of their risk of recurrence and malignant transformation. Some that appear quite malignant, with poor differentiation and many mitotic figures, may remain relatively quiescent.
112. c. This would be a classic presentation of an internal carotid-cavernous sinus fistula, often resulting from the shearing effect of rapid acceleration/deceleration trauma on the relatively fixed/immobile artery passing through the sinus.
113. e. This is, in contrast to the previous question, a classic presentation of a cavernous sinus wall-dural shunt, featuring lower arterial flow and a more insidious onset.
114. False. Varices are venous structures that usually would not have sufficient blood flow to produce an audible bruit. The classic presentation is proptosis with the Valsalva maneuver. Spiral computed tomography (CT) scan before and during Valsalva maneuvers can demonstrate varix enlargement.
115. True. Most are of intracranial origin; however, with the widespread use of computed tomography (CT), it has become apparent that primary optic nerve sheath meningiomas are much more common than was once
6. Oculoplastics

thought—many being asymptomatic lesions found incidentally.

116. **False.** In neurofibromatosis, the growth pattern tends to be paraxial (diffusely along the nerve sheath) rather than focal.

117. **c.** Although it could be argued that magnetic resonance imaging (MRI) may better delineate the tumor initially, bony changes—particularly those that are subtle—will be better seen with computed tomography (CT) scan. Gadolinium-enhanced MRI is particularly valuable for detecting intracranial spread.

118. **True.** The fragile pial vessels supplying the optic nerve are easily disrupted by any attempt to remove the tumor. Surgery is usually undertaken if there is intracranial extension or severe proptosis and severe visual loss.

119. **a.** Because these tumors are usually histologically benign— despite being locally destructive—they often can be observed for years without risking the loss of vision that would accompany excision. When vision has become too poor to counterbalance the risk of tumor spread (if such is occurring), or if tumor growth is becoming potentially life threatening, surgical intervention is indicated. Pediatric meningiomas are more aggressive than adult tumors and are removed earlier.

120. **False.** Although optic nerve gliomas presenting in adulthood do tend to be particularly malignant, this applies to isolated tumors (not associated with neurofibromatosis). The latter generally (in adults and children) have a better prognosis.

121. **a.** In contrast, orbit specialists probably see 50% epithelial neoplasms and 50% inflammatory/lymphoid lesions.

122. **c.** Adenoid cystic carcinoma is the most common malignant neoplasm of this location.

123. **c.** Pain is more typical of acute inflammatory, infectious, or malignant lesions. Duration of symptoms is helpful in separating slowly progressive (benign) lesions, such as the benign mixed tumor, from more acute conditions, such as malignant neoplasm or inflammation. Radiologic findings are the third critical feature in the evaluation. Osteolysis strongly favors a malignant lesion.

124. **c.** Soft-tissue contour analysis can help differentiate between lymphoid tumors (elongated smooth masses—“pancakes”) from parenchymal tumors (globose masses). Adenoid cystic carcinomas usually produce bony destruction. Benign mixed cell tumors often cause concave pressure changes in the adjacent bone. Lymphoid and inflammatory lesions generally do not cause bony changes.

125. **b.** The most likely diagnosis is that of benign mixed cell tumor. The male-female ratio is 3:2. The tumor should be approached through a lateral orbitotomy with careful excision to avoid rupture of the tumor’s pseudocapsule. Incisional or incomplete biopsy techniques can lead to infiltrative tumor recurrence (in 32% of cases) and, occasionally, malignant transformation.

126. **c.** Mucopidermoid carcinoma is distinctly uncommon.

127. **False.** Although adenoid cystic carcinomas may cause severe pain, it is usually a result of perineural invasion and bony destruction.

128. **d.** Fibrous dysplasia may be monostotic (affect only one bone) or polyostotic. The polyostotic variety may present with precocious puberty and dermal hyperpigmented macules. This disorder, Albright’s syndrome, rarely involves the orbit. Orbital disease is nearly always monostotic and rarely associated with precocious puberty, regardless of age. Surgical curettage or excision is usually undertaken. Only woven bone, not cancellous, is found microscopically.

129. **c.** Eosinophilic granuloma frequently involves the orbital bones. The classic triad of Hand-Schüller-Christian disease consists of proptosis, lytic skull lesions, and diabetes insipidus. Orbital involvement is rare in Letterer-Siwe disease. Prior characterization of histiocytic disorders using these terms is being replaced by the terms diffuse soft tissue histiocytosis and unifocal/multifocal eosinophilic granuloma of bone.

130. **False.** Chemotherapy (vincristine or vinblastine) is required for patients with disseminated disease. In contrast, the lesions of eosinophilic granuloma may (i) be observed for spontaneous regression, (ii) be excised, or (iii) receive radiotherapy (if advanced), or (iv) receive systemic corticosteroid administration.

131. **d.** Most are benign (>90%), and have a storiform, or matlike, pattern on histopathology. It is usually very firm and can displace other orbital structures.

132. **False.** Ten percent have metastatic potential. Another 16% are termed “locally aggressive” but not frankly malignant.

133. **False.** It is impossible to differentiate clinically or radiologically between benign reactive lymphoid hyperplasia and orbital lymphoma. Biopsy with light microscopy, immunohistochemical staining, and electron microscopy is necessary to distinguish between them.

134. **d.** The central nervous system (CNS) is not routinely surveyed in patients with orbital lymphoma. This is in contrast to patients with intraocular lymphoma. When CNS involvement is suspected, computed tomography (CT) scan or magnetic resonance imaging (MRI) is the starting point.

135. **b.** Both orbital inflammatory syndrome and lymphoproliferation are composed of polyclonal lymphocytes (although lymphoma contains a monoclonal subpopulation). Cellular heterogeneity is a feature of both orbital inflammatory syndrome and reactive lymphoid hyperplasia. The key features separating the two are (i) prominent fibrovascular stroma and
Answers

(ii) hypocellularity, both seen in pseudotumor and not in lymphoid activation.
136. a. The three orbital deposits that are hyperintense on T1-weighted images are blood, melanin, and mucus.
137. False. The nose and paranasal sinuses are more frequently the focus of tumors that secondarily invade the orbit.
138. b. The most common sinus neoplasm to invade the orbit is squamous cell carcinoma.
139. a. Typically, the orbital metastasis occurs months to years after diagnosis and treatment of the primary tumor. A classic (but uncommon) presentation is progressive enophthalmos caused by scirrhus breast carcinoma, which contracts and scars, drawing the eye in, and causing pseudoenophthalmos.
140. b. An orbital metastasis is more likely to be the mode of presentation for bronchogenic carcinoma than for a breast primary.
141. d. The tripod complex is produced by three distinct fractures along suture lines—zygomaticofrontal, zygomaticomaxillary, and the zygomatic arch. Ocular motility may or may not be normal in pure tripod fractures, but upgaze should be spared. Repair is indicated when there is marked cosmetic deformity or potential mandibular instability.
142. c. Telecanthus is defined as an abnormally large distance between medial canthi, regardless of intraorbital distance. Telecanthus and rounding of the medial canthus are characteristic findings in direct naso-orbital-ethmoid fractures.
143. False. The qualifier “indirect” implies that forces other than direct contact with a blunt object led to orbital wall fracture (for instance, suddenly increasing intraorbital pressure). This type is rarely associated with orbital rim fracture, which is seen more.
144. c. Subcutaneous emphysema is usually produced by medial or inferior orbital wall fractures into the ethmoid or maxillary sinuses.
145. d. Orbital floor fractures usually produce vertical limitations of gaze. Global motility deficits generally indicate blunt trauma with muscle and/or nerve contusion.
146. False. Computed tomography (CT) scan of the orbit with direct coronal, axial, and sagittal views gives better definition of the soft tissue and bony structures than plain films. Magnetic resonance imaging (MRI) has a very limited role, because bone is dark.
147. False. With severe muscle contusion, edema, or hemorrhage, forced ductions may be falsely positive or impossible to interpret. They are more fruitful if performed 5 to 10 days after injury.
148. False. Enophthalmos often develops after the swelling subsides from an initially exophthalmic orbit.
149. True. Most diplopia is a result of reversible muscle contusion.
150. e. All of these are indications for orbital floor fracture repair. Pediatric patients may develop entrapment of the inferior rectus with a “trapdoor” fracture which should be repaired sooner to limit fibrosis and extraocular muscle (EOM) restriction. EOM testing may cause bradycardia in these patients because of the oculocardiac reflex.
151. False. Large, complex anterior fractures lead to marked inferior orbital herniation without entrapment. This causes hypoglobus and enophthalmos. Small, posterior fractures can cause significant entrapment as a crowded muscle belly is forced or pinched into the defect. In these cases, enophthalmos is minimal.
152. d. This time period provides a chance for orbital swelling and “contusion diplopia” to resolve and yet is early enough to avoid problems with scarring of a significant floor fracture.
153. b. Vegetable foreign bodies carry a high risk of infection. Lead can cause a granulomatous reaction, but only if far posterior should be left in place. BB pellets (often made from lead) are commonly left within the orbit without consequence. Orbital apex foreign bodies are hazardous to remove and should be pursued only if there is convincing evidence of direct optic nerve compromise (decreased vision, afferent pupillary defect [APD], dyschromatopsia).
154. d. When ocular or optic disc perfusion is severely compromised by an orbital compartment syndrome, immediate canthotomy with cantholysis should be performed to decompress the orbit.
155. a. In cases in which the ocular examination suggests optic nerve trauma (afferent pupillary defect [APD], poor vision), neuroimaging is indicated to rule out direct optic nerve injury (e.g., optic canal fracture).
156. False. In direct optic nerve trauma, something physically impinges on the optic nerve, but it need not be a foreign body (e.g., bone fragments). In indirect trauma, nothing can be found to have directly struck the nerve.
157. a. Because no obvious lesion can be identified on the computed tomography (CT) scan, the optic nerve injury is most likely indirect. Emergent high-dose intravenous methylprednisolone is indicated to reduce swelling associated with the injury. Optic canal decompression may also be considered.
158. b. Evisceration provides better cosmesis and is technically easier to perform. The advantages of enucleation are preservation of ocular pathology for microscopic review and lower risk of sympathetic ophthalmia (SO). A cosmetically unacceptable phthisical ophthalmia may be covered with a prosthetic shell. Evisceration should not be performed in eyes with intraocular malignancy or when the type of intraocular pathology is unknown.
159. a. This complication results from inadequate volume replacement when the eye is removed from the orbit.
The normal adult palpebral fissure is 27 to 30 mm horizontally and 8 to 11 mm vertically. This strip is also known as the muscle of Riolan.

Müller's muscle accounts for about 2 mm of the total lacrimal secretory mass. It is far more common in the lower eyelid.

The deep head of the pretarsal muscle (Horner’s tensor tarsi) is a localized bundle of pretarsal orbicularis that is critical to adequate tear drainage. It surrounds the lacrimal sac and, with each blink, pumps tears downward into the nasolacrimal duct.

The posterior limb of the tendon must be reattached to the posterior lacrimal crest, or the lid contour will ride away from the globe surface.

Buccal mucosal grafting is ideal for the treatment of socket contracture. Care must be taken to avoid damaging the duct from the parotid gland when harvesting the graft.

Severe ocular defects are usually hidden by the fused lids.

When the lateral tendon inserts lower than the medial tendon [MCT]), “Antimongoloid” slant occurs. The tendon must be reattached to the posterior lacrimal crest.

Artificial tears/lubricating ointments are often the only necessary treatment for temporary paralytic ectropion.

Other methods of treating trichiasis include surgery, argon laser therapy, and radio frequency epilation.
199. **b.** Jones testing will give information regarding patency of the lacrimal drainage system. Patients with positive Jones tests will have difficulties with epiphora, which can usually be managed independently.

200. **a.** Congenital myogenic ptosis, in isolation, is a muscular dystrophy (maldevelopment) typically affecting only the levator-superior rectus complex.

201. **True.** Acquired myogenic ptosis is uncommon and is usually secondary to muscular disease such as myasthenia gravis (MG), chronic progressive external ophthalmo-plegia, or ocuopharyngeal dystrophy. Levator shortening or frontalis sling procedures are usually effective.

202. **b.** Aponeurotic dehiscence has been blamed on anesthetic injections, lid specula, and bridle sutures. The exact cause is not clear.

203. **False.** The muscle is spared, so despite severe ptosis, levator function is usually good.

204. **False.** Actually, the ptosis is often resistant to anticholinesterases or steroids. Surgery is usually performed after medical therapy has been optimized.

205. **a.** Levator innervation in Marcus Gunn's syndrome is derived from the trigeminal supply (CN V) to the temporygoids and masseters.

206. **b.** Müller resections only offer 1 to 3 mm of improvement. Aponeurosis surgery is indicated for a blunted or high eyelid crease. Frontalis suspension is bilateral surgery for severe bilateral ptosis with poor levator function.

207. **d.** See answer 206.

208. **c.** See answer 206.

209. **False.** The converse is true: lateral lid retraction is greater than medial in Graves' disease.

210. **True.** A combined levator-superior rectus dystrophy is cited as the cause.

211. **c.** Recession of the superior rectus muscle may cause lid retraction. Resection will lead to ptosis.

212. **False.** Both may be difficult, but lower lid retraction is more challenging. Its repair may necessitate spacer grafts (e.g., auricular cartilage, hard palate).

213. **c.** Acanthosis nigricans is associated with internal malignancy but not transformation. Actinic keratoses is the most common premalignant skin lesion. It is related to ultraviolet (UV) exposure and may develop into squamous cell carcinoma. Although actinic keratoses resolve spontaneously within a year in up to 25% of cases, patients with multiple actinic keratoses have a 12% to 16% risk of developing squamous cell carcinoma.

214. **c.** Suddenly appearing multiple seborrhoeic keratoses may in fact be evolving acanthosis nigricans. This is known as the Leser-Trelat sign and is usually associated with a gastrointestinal (GI) malignancy.

215. **a.** Verruca are typically papilliform without central excavation.

216. **c.** The fibrosing, or morpheaform, pattern extends in thin, fingerlike projections, which can escape excision or radiation.

217. **c.** Mohs’ resection may permit healing by secondary intention (granulation), but most patients prefer early (delayed primary) closure by flaps or grafts.

218. **False.** Squamous cell carcinoma is less common but clinically more aggressive.

219. **d.** There may be multicentric foci of tumor with skip areas, and negative margins do not necessarily imply complete excision of the tumor. Hence, Mohs’ resection should never be undertaken. Wide surgical margins should be used, with map biopsies taken over the surrounding area of lesion.

220. **c.** Cryotherapy selectively destroys melanocytes but is insufficient for cutaneous melanoma and should be considered a palliative treatment.

221. **True.**

222. **a.** Blepharochalasis is a rare idiopathic disorder leading to inflammatory edema of the eyelids. It is familial, and younger patients, especially women are affected. The repeated episodes of edema may cause ptosis and herniation of the orbital lobe of the lacrimal gland. Dermatocchalasis is redundant preseptal skin caused by aging. True ptosis (involutional) may be present in either disorder.

223. **d.** Injury to orbital fat pads, vessels, or orbicularis may result in retrobulbar hemorrhage that dissect into the posterior orbit and compresses the optic nerve.

224. **d.** Hemifacial spasm is rarely bilateral and is usually caused by vascular compression of the seventh cranial nerve at the brainstem and can result in synchronous contractions of the entire side of the face. Along with partial complex seizures and myoclonic epilepsy, essential blepharospasm is effaced by sleep.

225. **a.** Any treatment for blepharospasm is designed to decrease eyelid closure. Thus, dry eye will be aggravated. Facial nerve ablation suffers from recurrence rates as high as 30% and is associated with complications such as hemifacial paralysis. Consequently, its use has significantly decreased. Orbicularis myectomy is usually preferred to facial nerve avulsion. Botulinum injection is the initial treatment of choice.

226. **d.** The lateral horn of the levator aponeurosis divides the lacrimal gland. Removal of the orbital lacrimal gland removes the efferent input and interferes with reflex tearing, whereas removal of the palpebral lobe damages the ducts from the orbital portion, which run through the palpebral lobe. This impairs reflex tearing as well.

227. **d.** The sphenopalatine ganglion receives parasympathetic fibers from the greater superficial petrosal nerve (a division of CN VII).

228. **True.** The secretory apparatus is now thought to function together as a unit, with less rigid divisions of basal and reflex tear secretion functions.

229. **c.** It is 2 mm down (ampulla), then 8 mm across (canaliculus).

230. **d.** The sac plus duct add another 20 to 25 mm, for 30 mm total from punctum to inferior meatus.
231. **False.** The valve of Hasner provides this protection. The valve of Rosenmüller prevents tear reflux from the lacrimal sac into canaliculi.

232. **False.** In congenital obstruction, the blockage is at the valve of Hasner. In acquired cases, the blockage is within the intraosseous nasolacrimal duct. Involutional stenosis is one of the most common causes of acquired nasolacrimal duct obstruction.

233. a. The Jones I test consists of placing 2% fluorescein in the conjunctival fornices and attempting to recover fluorescein with a swab at the inferior meatus. Unfortunately, one third of normal patients will have abnormal results with this test.

234. b. The Jones II test is performed after a dye disappearance test or after Jones I indicates blockage. The fornix is irrigated, and the lacrimal sac cannulated and irrigated. If dye is recovered in the inferior meatus, then incomplete blockage of the nasolacrimal duct, patent upper system, and functioning lacrimal pump is indicated. However, if only clear fluid is recovered, then a nonfunctioning lacrimal pump or blocked upper system is indicated.

235. **False.** Probing of acquired obstructions rarely yields permanent patency.

236. a. If no functional or organic obstruction is discovered, then hyperlacrimation probably accounts for tearing. The evaluation of the tear film and lacrimal secretory function should be performed. Abnormal tear film precipitates or mucus may be discovered, and tear breakup-time testing (TBUT) <10 seconds indicates poor function of the inner mucinous layer of the tear. Schirmer testing provides information on reflex and basal secretion of the lacrimal gland.

237. **False.** A single functioning punctum and canaliculus is usually sufficient.

238. a. Congenital obstructions usually occur at the valve of Hasner. Canalicular impatency is rare in infants. Other acquired causes of canalicular obstruction include Stevens-Johnson syndrome and oral contraceptive pills (OCP).

239. a. Eyelid nodules with central craters generate the following first-order differential diagnosis: (i) basal cell carcinoma (BCC), (ii) keratoacanthoma, and (iii) molluscum contagiosum. Keratoacanthoma has a more acute presentation. In a 65-year-old man with a history of significant sun exposure, the diagnosis is basal cell carcinoma until proven otherwise. Figure a. shows a biopsy of the lesion in question. Note the islands of basalioid cells with little intraepithelial spread, primarily endophytic growth, peripheral palisading, and clefting artifact (separation of the neoplastic cells from the surrounding stroma). Figure b. shows a keratoacanthoma, with its typical exophytic growth pattern. Figure c. shows a sebaceous cell carcinoma. The vacuolization of the cells is highly suggestive of the correct diagnosis. Figure d. shows an unusual tumor, the oncocytoma. The copious eosinophilic cytoplasm is characteristic of this benign tumor of the caruncle. Figure e. shows a classic spindle cell melanoma, in this case, choroidal.

240. c. See answer 239. Note the localized madarosis. This is highly suggestive of sebaceous cell carcinoma and unusual for chalazia or styes. Mortality is approximately 20% at 5-year follow-up, although various studies have published mortality rates from 3% to 41%.

241. c. Stenoses, narrow or long ducts, and nasal and sinus inflammatory disease may cause tear stasis and lead to acute dacryocystitis.

242. c. Again, less invasive maneuvers should be tried first. Probing and irrigation offer little here. Topical antibiotics are of little value in the setting of tear stasis.

243. c. This is the ocular equivalent of a kidney stone.

244. **False.** The most common malignant tumor of the lacrimal sac is squamous cell carcinoma. Treatment may consist of a dacryocystectomy and a rhinotomy.

245. **False.** Dacryocystitis is usually below the medial canthal tendon (MCT). Encephalocele is usually above the MCT.

246. c. About 90% of congenital nasolacrimal duct (NLD) obstructions spontaneously resolve by 1 year of age.

247. False. The next appropriate step is Silastic intubation, dacryocystorhinostomy (DCR) follows if this is unsuccessful.

248. d. See answer 232.

249. a. A negative secondary dye test indicates poor lacrimal pump function; dacryocystorhinostomy (DCR) would not be helpful.

250. **True.** The superior canalicus possesses more maneuverable angles and hence is easier to probe. Furthermore, there is less functional significance if a false passage is created.

251. c. Complete bypass of the canalicular system is required, and conjunctivo-dacryocystorhinostomy (DCR) with placement of a Jones tube is the only choice to provide this bypass.

252. e. Unfortunately, cases of the incorrect eye being everted have been reported. Undertaking all of these maneuvers is truly in the patient’s best interests.

253. a. Suborbicularis oculi fat (SOOF) elevation provides a youthful midface appearance. It is usually accessed through a transconjunctival or subciliary approach.

254. c. Peak botulinum effect usually occurs 2 to 3 weeks after injection.

255. d. Any patient with sclerokeratitis and coexisting sinus disease should be suspected for Wegener’s granulomatosis. Ocular disease occurs in 60% of patients with the disease. Eighty percent of patients are positive for antineutrophilic cytoplasmic antibodies, or cytoplasmic pattern (c-ANCA).
256. e. The use of immunosuppressive drugs (especially cyclophosphamide) has significantly improved the mortality rates for patients with Wegener’s granulomatosis.

257. d. The patient has orbital lymphoma. The overwhelming majority of cases are non-Hodgkin’s lymphoma, characterized by sheets of malignant lymphocytes as shown in the figure.

258. b. Lymphangiomas are not encapsulated. The other tumors listed are generally encapsulated.

259. e. The patient has a schwannoma, an encapsulated tumor displaying the classic Antoni-A spindle cells and nuclear palisading (Verocay bodies).

260. False. Schwannomas rarely undergo malignant transformation. They can recur after incomplete excision, however.

261. c. Ocular burning occurs in approximately 15% of patients using topical cyclosporine. All of the other answers are less frequent side effects.

262. True. Patients with active ocular infections should not be prescribed topical cyclosporine.

263. False. Dermatochalasis is more common in elderly patients. Blepharochalasis is an inherited variant of angioneurotic edema, which is most common in young women.

264. c. Surgical excision is not recommended, and neither are systemic corticosteroids. Radiation is the treatment of choice for these lesions.

265. a. 2, b. 4, c. 1, d. 3.

266. a. 4, b. 1, c. 3, d. 2.

267. a. 6, b. 6, c. 5, d. 1, e. 2, f. 6, g. 6, h. 1, i. 6, j. 3, k. 7, l. 1, m. 4.

268. a. 1, b. 2, c. 6, d. 5, e. 7, f. 3, g. 4.

269. a. 1, b. 3, c. 3, d. 2, e. 3, f. 4.

270. a. 2, b. 4, c. 7, d. 1, e. 3, f. 8, g. 2, h. 6, i. 5, j. 7.

271. a. 4, b. 4, c. 1, d. 3, e. 1, f. 4, g. 2.

272. a. 3, b. 4, c. 1, d. 2.

273. a. 2, b. 1, c. 3. Optic nerve decompression gets the highest priority. Strabismus surgery should be performed before lid surgery because the former can cause changes in resting lid position.

274. a. 7, b. 4, c. 5, d. 3, e. 6, f. 3, g. 8, h. 5, i. 5.

275. a. 3, b. 1, c. 3, d. 2, e. 1, f. 2.

276. a. 1, b. 2, c. 3, g. 4, f. 5, a. 6, d. 7, c.

277. a. 3, b. 6, c. 5, d. 1, e. 2, f. 4.

**Suggested Readings**


Uveitis

Questions

1. T or F Passive immunization confers immunity that is longer lasting than that offered by active immunization.

2. Which of the following lymphocytes has cytotoxic activity without a specific antigen receptor and is not antigen specific?
   a. T-lymphocytes.
   b. B-lymphocytes.
   c. plasma cells.
   d. macrophages.
   e. natural killer (NK) cells.

3. Which cellular component of the lymphoreticular system has cytotoxic activity that is antibody dependent?
   a. T-lymphocytes.
   b. B-lymphocytes.
   c. plasma cells.
   d. killer cells.
   e. natural killer (NK) cells.

4. The class of antigen(s) typically responsible in immune responses is/are:
   1. protein.
   2. lipid.
   3. polysaccharide.
   4. nucleic acid.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

5. The first cell that an antigen typically contacts in the cascade of immune response is the:
   a. natural killer (NK) cell.
   b. T-lymphocyte.
   c. B-lymphocyte.
   d. plasma cell.
   e. macrophage.

6. Typically, the first class of antibody produced against a newly encountered antigen is:
   a. IgA.
   b. IgD.
   c. IgE.
   d. IgG.
   e. IgM.

7. Typically, the antibody class produced to an antigen previously exposed to the immune system is:
   a. IgA.
   b. IgD.
   c. IgE.
   d. IgG.
   e. IgM.

8. T or F Class I cell surface antigens are typically expressed on the cell surfaces of all nucleated human cells.
9. **T or F** Class II cell surface antigens are typically expressed on the cell surfaces of all nucleated human cells.

10. The major histocompatibility antigen complex in humans (human leukocyte antigen [HLA] system) is coded for by genes located on chromosome:
   a. 6.
   b. 11.
   c. 13.
   d. 18.
   e. 21.

11. What is the second most common method of human immunodeficiency virus (HIV) transmission?
   a. sexual intercourse.
   b. blood transfusions.
   c. intravenous drug use.
   d. transplacental transmission.
   e. organ transplantation.

12. Which one of the following human leukocyte antigen (HLA) markers is associated with the presumed ocular histoplasmosis syndrome (POHS)?
   a. A29.
   b. B5.
   c. B7.
   d. B8.
   e. B27.

13. **T or F** The variable regions of the immunoglobulin molecule are the amino termini of both the light and heavy chains.

14. Which immunoglobulin class has the highest individual molecular weight?
   a. IgA.
   b. IgD.
   c. IgE.
   d. IgG.
   e. IgM.

15. Which immunoglobulin molecule has the longest serum half-life?
   a. IgA.
   b. IgD.
   c. IgE.
   d. IgG.
   e. IgM.

16. Which is the only immunoglobulin class to readily cross the human placenta?
   a. IgA.
   b. IgD.
   c. IgE.
   d. IgG.
   e. IgM.

17. Which immunoglobulin class(es) may exist in polymer form?
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

18. Which antibody class(es) fix(es) complement?
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

19. **T or F** Secretory immunoglobulin contains alpha heavy chain and is secreted by the plasma cell as a dimer.

20. Which immunoglobulin class is probably the oldest phylogenetically?
   a. IgA.
   b. IgD.
   c. IgE.
   d. IgG.
   e. IgM.

21. Which of the following immunoglobulin classes is/are important in antigen reception at the surface of lymphocytes in primary immune responses?
   1. IgD.
   2. IgG.
7: Uveitis

3. IgM.
4. IgE.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

22. Which of the following immunoglobulin classes is/are important in antigen reception at the lymphocyte cell surfaces for anamnestic immune responses?
1. IgD.
2. IgG.
3. IgM.
4. IgE.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

23. Which complement component is present in the highest serum concentrations?

a. C1q.
b. C3.
c. C4.
d. C5.
e. C9.

24. Which of the following hypersensitivity reaction types involve the participation of antibodies?
1. type I.
2. type II.
3. type III.
4. type IV.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

25. T or F The cyclooxygenase pathway leads to the production of prostaglandins, thromboxane, and prostacyclins, whereas the lipoxygenase pathway leads to the production of leukotrienes.

26. T or F In the classic wheal-and-flare reaction, edema and tissue infiltration with circulating inflammatory cells are prominent findings.

27. T or F Cytotoxicity is strictly complement-dependent.

28. T or F Neutrophil inhibitors, such as nitrogen mustard, will block the manifestations of an Arthus reaction.

29. T or F Interleukins are immune upregulators produced by white blood cells and fibroblasts.

30. Graves’ disease is a manifestation of what type of hypersensitivity?

a. type I.
b. type II.
c. type III.
d. type IV.
e. type V.

31. Which immunoglobulin has not been detected in tear samples?

a. IgA.
b. IgD.
c. IgE.
d. IgG.
e. IgM.

32. Wessely’s rings are a manifestation of what type of hypersensitivity?

a. type I.
b. type II.
c. type III.
d. type IV.
e. type V.

33. What percentage of patients with ankylosing spondylitis possesses the HLA-B27 gene?

a. 10%.
b. 25%.
c. 50%.
d. 75%.
e. >75%.

34. Dalen-Fuchs nodules may be seen in which of the following conditions?

1. sympathetic ophthalmia (SO).
2. tuberculous choroiditis.

a. 1, 2, and 3.
b. 1 and 3.
Questions

c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

35. The most common cause of anterior uveitis in the adult population is:
   a. herpes simplex keratouveitis.
   b. herpes zoster keratouveitis.
   c. syphilitic uveitis.
   d. HLA-B27 iridocyclitis.
   e. idiopathic iridocyclitis.

36. The most common cause of posterior uveitis in the adult population is:
   a. toxocariasis.
   b. sarcoidosis.
   c. toxoplasmosis.
   d. idiopathic posterior uveitis.
   e. tuberculosis.

37. A low-grade uveitis associated with nongranulomatous keratic precipitates (KPs) distributed diffusely on the corneal endothelium (i.e., both upper and lower cornea) should suggest the diagnosis of:
   a. syphilitic uveitis.
   b. HLA-B27 uveitis.
   c. Fuchs’ iridocyclitis.
   d. sarcoidosis.
   e. idiopathic iridocyclitis.

38. Which of the following conditions may be associated with iris nodules and skin findings?
   1. sarcoidosis.
   2. juvenile xanthogranuloma (JXG).
   3. neurofibromatosis.
   4. pseudoxanthoma elasticum.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

39. The most common cause of decreased vision in intermediate uveitis is:
   a. occluded pupil.
   b. posterior subcapsular cataract.
   c. vitritis.
   d. cystoid macular edema (CME).
   e. papillitis with optic atrophy.

40. The prevalence of HLA-B27 in the general population is:
   a. <5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

41. Which of the following antiretroviral agents has been shown to increase the CD8+ lymphocyte count in human immunodeficiency virus (HIV)–positive patients?
   a. zidovudine (ZDV, AZT).
   b. ritonavir.
   c. didanosine (ddI).
   d. zalcitabine (ddC).
   e. foscarnet.

42. Which of the following conditions are associated with the HLA-B27 genotype?
   1. inflammatory bowel disease.
   2. Reiter’s syndrome.
   3. ankylosing spondylitis.
   4. psoriatic arthritis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

43. What is the probability, given the HLA-B27 genotype, of sacroiliac disease?
   a. <5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

44. The most important component of long-term therapy in a young man who is HLA-B27–positive is:
   a. recurrent annual visual acuity testing.
   b. annual tonometry.
   c. systemic (oral) nonsteroidal antiinflammatory therapy.
   d. physical therapy.
   e. cardiac ultrasonography.

45. Which one of the following concerning Reiter’s syndrome is false?
   a. Ninety percent of cases occur in men.
   b. It may follow a bout of either urethritis or dysentery.
c. Most patients are HLA-B27–positive.
d. The most common ocular finding is an acute nongranulomatous anterior uveitis.
e. The characteristic skin finding is a rash similar to pustular psoriasis.

46. **T or F** Acute anterior uveitis is more likely to develop in patients with ulcerative colitis than in patients with Crohn’s disease.

47. **T or F** Acute anterior uveitis commonly develops in conjunction with both psoriasis and psoriatic arthritis.

48. Which one of the following concerning Behçet’s disease is false?
   a. The disease is more common among Japanese than Americans.
   b. The classic acute uveitis of Behçet’s disease is typically associated with a hypopyon.
   c. Anterior uveitis is more common in Behçet’s disease in men than posterior uveitis.
   d. Characteristic aphthous ulcers develop on mucous membranes, including the mouth and genital tract.
   e. A potentially lethal systemic vasculitis may be successfully treated with colchicine.

49. **T or F** The uveitis that accompanies glaucomatocyclitic crisis is typically very mild, with only a few or no keratic precipitates (KPs).

50. **T or F** The uveitis-glaucoma-hyphema syndrome is generally caused by posterior chamber intraocular lenses that are inappropriately sized or have closed loops.

51. Which one of the following concerning Kawasaki’s disease is false?
   a. Most of the affected patients are children younger than 10 years.
   b. The hallmark of the eye findings is a bilateral conjunctival congestion that spares the limbus.
   c. The hallmark of the dermatologic findings is a shedding rash affecting the extremities.
   d. Corticosteroids are agents of choice to treat the underlying disease.
   e. A mild bilateral anterior uveitis may accompany the conjunctival infection but is not present in all cases.

52. **T or F** Unlike most other uveitides, herpetic uveitis is frequently accompanied by elevated intraocular pressure.

53. **T or F** The iris atrophy of herpes simplex uveitis tends to be segmental, whereas that of herpes zoster uveitis tends to be patchy.

54. In order to be appropriately termed “chronic,” a uveitis must last at least:
   a. 2 weeks.
   b. 4 weeks.
   c. 6 weeks.
   d. 8 weeks.
   e. 12 weeks.

55. Which one of the following concerning uveitis associated with juvenile rheumatoid arthritis (JRA) is false?
   a. Most of the affected children are girls.
   b. Most of the affected children have pauciarticular arthritis.
   c. Most of the affected children will be antinuclear antibody (ANA)-positive.
   d. Most of the affected children will be rheumatoid factor (RF)-positive.
   e. The onset of anterior uveitis in the third decade of life of a patient with a remote history of pediatric arthritis does not preclude the diagnosis of JRA.

56. **T or F** The diagnosis of juvenile rheumatoid arthritis (JRA) is often made incidentally because of its insidious progress.

57. **T or F** An ocular condition identical to the uveitis associated with juvenile rheumatoid arthritis (JRA) may be seen in children who never develop arthritis.

58. All of the following are considered clinical hallmarks of Fuchs’ heterochromic iridocyclitis except:
   a. diffuse atrophy of the iris stroma.
   b. small, white, starlike keratic precipitates (KPs) diffusely scattered.
   c. mild or minimal anterior chamber reaction.
   d. anterior and posterior synechialization.
   e. cataract rapidly progressing from posterior subcapsular to complete “white-out.”

59. **T or F** In Fuchs’ heterochromic iridocyclitis, the cataractous eye typically has the lighter colored iris.
60. T or F In Fuchs’ heterochromia, the long-term vision-limiting complication is generally band keratopathy.

61. Which one of the following concerning the manifestations of sarcoidosis is false?
a. The uveitis may be granulomatous or non-granulomatous.
b. A classic orbit/adnexal finding is bilateral painless enlargement of the lacrimal gland.
c. The most commonly involved organ system is pulmonary.
d. “Candle-wax” drippings (taches de bougie) are actually irregular granulomas along retinal venules.
e. The anterior uveitis of sarcoidosis is nonscarring (i.e., few or no synechiae).

62. Laboratory tests that are helpful in the evaluation of the patient with suspected sarcoidosis include:
   1. angiotensin-converting enzyme (ACE).
   2. serum lysozyme (muramidase).
   3. chest x-ray.
   4. antineutrophil cytoplasmic antibody assay (ANCA).
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

63. T or F The key histopathologic feature separating the tubercles of sarcoidosis from those of tuberculosis is the presence of caseation in the former.

64. T or F Seventh nerve palsy in neurosarcoidosis typically is caused by primary, granulomatous neuritis.

65. Which one of the following concerning congenital syphilis is true?
a. The interstitial keratitis (IK) of congenital syphilis is generally bilateral and asymptomatic.
b. Syphilitic IK is a direct manifestation of active corneal infection.
c. The chorioretinitis of congenital syphilis is generally bilateral and nonprogressive.
d. A diagnosis of syphilitic IK mandates a full course of treatment for neurosyphilis.
e. Serologic testing for suspected congenital syphilis is generally not rewarding.

66. Which one of the following concerning acquired ocular syphilis is false?
a. The eye may be involved in either secondary or tertiary syphilis.
b. Ocular findings may include iris nodules or vascularized papules.
c. The end stage of retinal vasculitis and inflammation may resemble retinitis pigmentosa.
d. Treatment of syphilitic uveitis is identical to that of neurosyphilis.
e. The fluorescent treponemal antibody-absorption (FTA-ABS) is a useful measure of disease response to therapy.

67. Which one of the following concerning Lyme disease is false?
a. The earliest eye finding is typically a follicular conjunctivitis.
b. The most common eye finding is a chronic iridocyclitis with vitreous cells.
c. Current serologic tests for Lyme disease are approaching >90% sensitivity.
d. The other organ systems commonly affected by Lyme disease include skin, central nervous system, cardiovascular, and musculoskeletal systems.
e. Recommended therapy includes either oral tetracycline or oral penicillin, with intravenous antibiotics reserved for neurologic involvement or multiple recurrences.

68. Which one of the following concerning ocular tuberculosis (TB) is true?
a. TB is the second most common cause of uveitis in the United States.
b. The ocular inflammation associated with TB reflects a hypersensitivity reaction without active infection.
c. Skin testing is valuable in the diagnosis of tuberculous uveitis but may need to be repeated with higher concentrations of purified protein derivative (PPD).
d. Eye disease is never seen in the setting of a normal chest x-ray.
e. Treatment of choice for tuberculous uveitis is topical and/or systemic corticosteroid.
69. Which one of the following concerning pars planitis is false?
   a. Most patients have unilateral involvement.
   b. Most patients are younger than 30 years.
   c. The most common cause of visual reduction is cystoid macular edema (CME).
   d. It may be associated with systemic conditions such as Lyme disease, sarcoidosis, and multiple sclerosis.
   e. Systemic immunosuppressives and pars plana vitrectomy may play a role in disease control.

70. T or F The most frequent source of contaminating organisms in postoperative endophthalmitis is incompletely sterilized surgical instrumentation.

71. T or F Pain is a sine qua non of endophthalmitis.

72. The genus of organisms most commonly isolated in bleb-associated endophthalmitis is:
   a. *Staphylococcus*.
   b. *Neisseria*.
   c. *Propionibacterium*.
   d. *Streptococcus*.
   e. *Enterococcus*.

73. T or F Postoperative endophthalmitis develops only after penetrating ocular surgery.

74. The organism(s) implicated in the pathogenesis of chronic postoperative endophthalmitis include:
   1. *Staphylococcus epidermidis*.
   2. *Candida* species.
   4. *Serratia* species.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

75. Which endophthalmitis has the worst prognosis?
   a. acute postoperative endophthalmitis.
   b. chronic postoperative endophthalmitis.
   c. posttraumatic endophthalmitis.
   d. bleb-associated endophthalmitis.
   e. endogenous *Candida* endophthalmitis.

76. The organism responsible for approximately 25% of posttraumatic endophthalmitis is:
   a. *Staphylococcus aureus*.
   b. *Streptococcus pneumoniae*.
   c. *Aspergillus*.
   d. *Acanthamoeba*.
   e. *Bacillus cereus*.

77. The organism with the poorest prognosis in posttraumatic endophthalmitis is:
   a. *Staphylococcus aureus*.
   b. *Streptococcus pneumoniae*.
   c. *Aspergillus*.
   d. *Acanthamoeba*.
   e. *Bacillus cereus*.

78. Which of the following medications acts by preventing synthesis of viral DNA from RNA?
   1. zalcitabine.
   2. zidovudine.
   3. didanosine.
   4. ritonavir.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

79. Which one of the following concerning fungal endophthalmitis is false?
   a. *Candida* is the most common etiology.
   b. The classic vitritis follows an earlier retinochoroiditis.
   c. Most patients with endogenous *Candida* endophthalmitis will have positive blood cultures.
   d. Approximately 10% of patients with candidemia will eventually develop endophthalmitis.
   e. *Candida* endophthalmitis is an uncommon manifestation of acquired immunodeficiency syndrome (AIDS).

80. T or F One helpful finding distinguishing true infectious endophthalmitis from exaggerated inflammation following trauma or intraocular surgery is vitritis that is out of proportion to anterior chamber reaction in the former.

81. In general, signs and/or symptoms typical of the onset of posterior uveitis include:
   1. floaters.
   2. ciliary flush.
   3. scotomata.
   4. brow pain.
Questions

82. Which one of the following concerning toxoplasmosis is true?
   a. The manifestations are always strictly posterior.
   b. The retinal vessels are always spared in ocular toxoplasmosis.
   c. Enzyme-linked immunosorbent assay (ELISA) testing for antitoxoplasma antibodies is important in the diagnosis of atypical lesions.
   d. Vitritis is a *sine qua non* of ocular toxoplasmosis.
   e. The definitive hosts of *Toxoplasma gondii* are mainly small rodents.

83. Potential adverse affects of the pharmacologic management of toxoplasmosis include all of the following except:
   a. pseudomembranous colitis.
   b. Stevens-Johnson syndrome.
   c. aggravation of diabetes mellitus.
   d. microcytic anemia.
   e. aplastic anemia.

84. Which one of the following regarding onchocerciasis is false?
   a. *Onchocerca volvulus* is transmitted by the bite of the *Simulium* black fly.
   b. Skin findings are rare in this condition.
   c. Microfilariae, released by adult worms, penetrate the eye by both direct invasion and hematogenous spread.
   d. Microfilariae may be seen swimming in the anterior chamber and may induce a severe anterior uveitis with glaucoma and cataract.
   e. Chorioretinal and optic atrophy are common in advanced disease.

85. **T or F** Like onchocerciasis, the pathology of cysticercosis is often dramatically worsened by death of the organism.

86. Which of the following constitute risk factors for *Candida* endophthalmitis?
   1. recent total parenteral nutrition.
   2. intravenous drug abuse.
   3. recent chemotherapy.
   4. acquired immunodeficiency syndrome (AIDS).

87. Which one of the following concerning cytomegalovirus (CMV) infection is false?
   a. The congenital form may be heralded by fever, pneumonia, or hepatosplenomegaly.
   b. The eye findings in congenital disease include cataract and peripheral retinal lesions, both atrophic and hyperpigmented.
   c. Retinal detachment is uncommon in CMV retinitis.
   d. Posterior segment involvement generally starts as a retinitis and secondarily involves the choroid.
   e. Viral inclusions may be seen both within the nucleus and cytoplasm of affected retinal cells.

88. **T or F** The retinitis of the maternal rubella syndrome is generally benign.

89. **T or F** Like congenital syphilitic retinopathy, congenital measles (rubeola) retinopathy is almost always benign.

90. Which one of the following concerning acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is false?
   a. The condition has only ocular manifestations.
   b. It typically affects patients younger than 30 years and has no sex predilection.
   c. There may be a mild or moderate anterior nongranulomatous uveitis.
   d. The fundus lesions are usually bilateral, multiple, and with minimal elevation.
   e. Angiographic findings include early hypofluorescence and late hyperfluorescence.

91. Which one of the following concerning necrotizing herpetic retinitis (acute retinal necrosis) is false?
   a. Anterior segment inflammation is variable.
   b. Posterior segment inflammation is generally heavy.
   c. The periphery of the retina is affected earlier and more severely than the posterior pole.
   d. Retinal detachment occurs in up to three-quarters of cases.
   e. Like other viral retinitides, affected patients are usually immunosuppressed.
92. **T or F** *Nocardia asteroides* is a fungus that can cause uveal inflammation as part of a systemic hematogenously spread infection dominated by the presence of multiple abscesses.

93. Which of the following concerning ocular toxocariasis is/are true?

1. The definitive host for the parasite is the dog or cat.
2. Ingested ova initially take up residence in the liver and lung.
3. Manifestations may include chronic endophthalmitis, localized macular granuloma, or localized peripheral granuloma.
4. Inflammation is usually unilateral.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

94. Which of the following concerning the presumed ocular histoplasmosis syndrome (POHS) is/are true?

1. The maculopathy generally precedes the formation of peripheral “histo spots.”
2. The vitritis associated with the condition may decrease vision.
3. Fundus lesions in their acute phase represent a retinitis with a secondary choroidal reaction.
4. A patient with a macular histo spot has about a one in four chance of active maculopathy over the next 3 years.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

95. Which of the following concerning sympathetic ophthalmia (SO) is/are true?

1. The anterior uveitis associated with SO is nearly always nongranulomatous.
2. The reported interval between injury or surgery and the onset of SO can be as short as 9 days or as long as 50 years, but it usually occurs 4 to 8 weeks after injury.
3. The histopathologic features of the uveitis are consistent with discrete granulomatous inflammation.
4. Granulomatous optic neuritis is commonly seen.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

96. Which of the following findings in a patient with bilateral granulomatous uveitis establish(es) the diagnosis of Vogt-Koyanagi-Harada (VKH) syndrome, rather than sympathetic ophthalmia (SO)?

   1. vitiligo.
   2. a history of previous vitrectomy.
   3. poliosis.
   4. histopathology revealing involvement of the choriocapillaris by the inflammatory process.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

97. Periocular steroid injection should be avoided in which of the following conditions?

1. pars planitis associated with sarcoidosis and cystoid macular edema (CME).
2. necrotizing scleritis associated with rheumatoid arthritis.
3. CME following cataract surgery.
4. ocular toxoplasmosis.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

98. Assuming all inflammation has been quiescent for several months, visually significant cataract associated with chronic/recurrent uveitis may be successfully managed with lens phacoemulsification and posterior chamber intraocular lens (PCIOL) implantation in which of the following situations?
1. an elderly man with idiopathic iritis.
2. a young adult with sarcoidosis.
3. a teenage boy with HLA-B27–associated uveitis.
4. a young girl with juvenile rheumatoid arthritis (JRA).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

99. Which of the following are common hematologic findings in patients with acquired immunodeficiency syndrome (AIDS)?
   1. CD4 lymphocytopenia.
   2. hypergammaglobulinemia.
   3. increased suppressor T cells, relative to helper T cells.
   4. granulocytopenia.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

100. What percentage of patients with full-blown acquired immunodeficiency syndrome (AIDS) will develop some ocular abnormality?
   a. 5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. at least 65%.

101. **T** or **F** Cytomegalovirus (CMV) retinitis is the most common ocular manifestation of human immunodeficiency virus (HIV) infection.

102. The most common intraocular infection associated with human immunodeficiency virus (HIV) infection is:
   a. toxoplasmosis.
   b. acute retinal necrosis secondary to herpes simplex virus (HSV).
   c. pneumocystis choroiditis.
   d. cytomegalovirus (CMV) retinitis.
   e. syphilitic uveitis.

103. **T** or **F** Cytomegalovirus (CMV) retinitis is typically seen in the later stages of acquired immunodeficiency syndrome (AIDS).

104. **T** or **F** Ocular toxoplasmosis in acquired immunodeficiency syndrome (AIDS) is similar in every way to other forms of acquired ocular toxoplasmosis except for being slightly more severe.

105. Which of the following is believed to be a significant risk factor for the development of pneumocystis choroiditis?
   a. active *Pneumocystis carinii* pneumonia (PCP).
   b. concurrent treatment with aerosolized pentamidine.
   c. severe cachexia.
   d. absolute lymphocyte count <500 cells/mm³.
   e. concurrent syphilitic uveitis.

106. Which of the following conditions developing in a healthy man in his 20s or 30s should raise suspicion of coincident human immunodeficiency virus (HIV) infection?
   1. acute retinal necrosis.
   2. syphilitic uveitis.
   3. herpes simplex keratitis.
   4. herpes zoster ophthalmicus (HZO).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

107. Which of the following foci may harbor the development of ocular Kaposi’s sarcoma?
   1. eyelid skin.
   2. choroid.
   3. conjunctiva.
   4. orbit.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

108. Which of the following medications acts by preventing proteolytic release of active human immunodeficiency virus (HIV) particles?
   1. zalcitabine.
   2. zidovudine.
   3. didanosine.
   4. ritonavir.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

109. Which agent is the drug of choice for prophylaxis of Pneumocystis carinii pneumonia (PCP) in human immunodeficiency virus (HIV)–positive patients?
a. oral trimethoprim-sulfamethoxazole.
b. oral dapsone.
c. aerosolized pentamidine.
d. oral pentamidine.
e. oral pyrimethamine.

110. According to National Institutes of Health (NIH) guidelines, oral zidovudine (ZDV, AZT) should be initiated at the time CD4 cell counts drop below what level in symptomatic human immunodeficiency virus (HIV)–positive patients?
a. 500/mm³.
b. 300/mm³.
c. 200/mm³.
d. 100/mm³.
e. 50/mm³.

111. Approximately what percentage of patients with documented candidemia will develop Candida endophthalmitis, defined as at least some degree of vitreous inflammation?
a. 0%.
b. 1%.
c. <5%.
d. >5%.
e. >10%.

112. Which of the following clinical features of newly diagnosed idiopathic pars planitis is most predictive of the future development of multiple sclerosis (MS)?
a. posterior subcapsular cataract.
b. 180 degrees of “snowbanking.”
c. retinal vascular sheathing.
d. angiographic optic disc edema.
e. neovascularization of the vitreous base.

113. Clinical features distinguishing the progressive outer retinal necrosis (PORN) syndrome from the acute retinal necrosis (ARN) syndrome include each of the following except:
a. initial involvement of the outer retina.
b. relative lack of intracocular inflammation.
c. relative lack of vasculitis.
d. earlier involvement of the macula.
e. better visual outcome.

114. One potential complication of cryotherapy for pars planitis is:
a. exacerbation of vitritis.
b. exacerbation of posterior subcapsular cataract.
c. exacerbation of cystoid macular edema (CME).
d. rhegmatogenous retinal detachment.
e. phthisis.

115. Which of the following are therapeutic options for pars planitis?
a. subtenon steroid.
b. oral steroids.
c. peripheral retinal cryoablation.
d. peripheral scatter laser treatment.
e. all of the above.

116. Initial evaluation of a patient with newly diagnosed pars planitis should include each of the following except:
a. serologic testing for toxoplasma.
b. serologic testing for syphilis.
c. tuberculin skin testing.
d. cranial computed tomography (CT) or magnetic resonance imaging (MRI).
e. chest x-ray.

117. Each of the following regarding sympathetic ophthalmia (SO) is true except:
a. The incidence following penetrating trauma is approximately 0.1%.
b. The incidence following elective intraocular surgery is approximately 0.01%.
c. An exciting eye with better visual potential than the sympathizing eye should not be removed.
d. The disease has a biphasic peak age prevalence.
e. Initial treatment usually includes systemic cytotoxic agents.

118. T or F Adaptive immunity typically activates complement by the classic pathway, whereas innate immunity activates it by the alternative pathway.

119. T or F The choroid has the highest blood flow rate in the body.

120. T or F Isolated, rare cases of human immunodeficiency virus (HIV) transmission as a result of corneal transplantation have been reported to occur.
Questions

121. What percentage of patients with primary central nervous system lymphoma (PCNSL) have ocular involvement?
   a. 5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

122. T or F Systemic corticosteroids and immunosuppressive agents initially reduce the appearance of intraocular inflammation in patients with ocular primary central nervous system lymphoma (PCNSL).

123. In men, choroidal metastases most commonly derive from a primary__________tumor.
   a. lung.
   b. breast.
   c. colon.
   d. prostate.
   e. testicular.

124. In women, choroidal metastases most commonly derive from a primary__________tumor.
   a. lung.
   b. breast.
   c. colon.
   d. cervical.
   e. ovarian.

125. Retinal metastases are most commonly from a primary:
   a. breast tumor.
   b. lung tumor.
   c. gastrointestinal tumor.
   d. cutaneous melanoma.
   e. prostate tumor.

126. What is the treatment of choice for uncomplicated acute retinal pigment epitheliitis (Krill’s disease)?
   a. observation.
   b. intravitreal corticosteroids.
   c. oral corticosteroids.
   d. oral corticosteroids and antibiotics.
   e. intravenous corticosteroids and immunosuppressive agents.

127. Which of the following retinochoroidopathies is typically unilateral?
   a. multiple evanescent white dot syndrome (MEWDS).
   b. acute posterior multifocal placoid pigment epitheliopathy (APMPPE).
   c. serpiginous choroidopathy.
   d. birdshot chorioretinopathy.
   e. punctate inner choroiditis (PIC).

128. A young woman recently traveled to Mexico and now complains of a floating object in her vision and mild eye pain. An external photograph of her eye is shown in the figure below. This woman most likely suffers from:
   a. ocular cysticercosis.
   b. toxocariasis.
   c. diffuse unilateral subacute neuroretinitis (DUSN).
   d. onchocerciasis.
   e. toxoplasmosis.

Matching

129. Arrange the following immunoglobulin classes in order of serum concentration, from most abundant (1) to least abundant (5).
   a. IgA.
   b. IgD.
   c. IgE.
   d. IgG.
   e. IgM.

130. Various hypersensitivity reactions with ocular manifestations are lettered below. In the right column, the hypersensitivity reaction types are listed. Assign a reaction type to each condition.
   a. idiopathic scleritis. 1. type I.
   b. Mooren’s ulcer. 2. type II.
   c. tuberculous leprosy. 3. type III.
   d. giant papillary conjunctivitis (GPC). 4. type IV.
   e. interstitial keratitis (IK). 5. type V.
7: Uveitis

f. atopic keratoconjunctivitis.
g. sarcoidosis.
h. retinal vasculitis.
i. corneal graft rejection.
j. phlyctenulosis.

131. Match each iris nodule characteristic lettered below with the type of nodule listed.
a. occur at the pupillary border. 1. Busacca nodule.
b. are gray or white in appearance. 2. Koeppe nodule.
c. occur within the iris stroma. 3. both 1 and 2.
d. may be associated with posterior synechiae. 4. neither 1 nor 2.
e. may be associated with granulomatous or nongranulomatous uveitis.
f. are associated only with granulomatous uveitis.

132. Arrange the three etiologies of exogenous endophthalmitis in order of decreasing frequency (1 being the most frequent and 3 being the least frequent):
a. postoperative (cataract) endophthalmitis.
b. bleb-associated endophthalmitis.
c. posttraumatic endophthalmitis.

133. Select the retinochoroidopathy with each of the following characteristics.
a. presents with blurry central vision. 1. serpiginous choroidopathy.
b. presents with nyctalopia or decreased colored vision. 2. birdshot chorioretinitis.
c. extends contiguously from the disc. 3. both.
d. retinal vessels may be sheathed. 4. neither.
e. geography reveals pronounced perifoveal capillary leakage with cystoid macular edema (CME).
f. epiretinal membrane is common.
g. electroretinogram (ERG) is abnormal.
h. no clear human leukocyte antigen (HLA) association.
i. associated strongly with HLA-A29.
j. associated with choroidal neovascularization.

134. Characteristics associated with various cytotoxic agents lettered below. Match each numbered agent with its lettered characteristics. (Note that the agents may be assigned to more than one characteristic and that each characteristic may be associated with more than one agent.)
a. DNA cross-linking agent. 1. chlorambucil.
b. purine analog. 2. azathioprine.
c. folate analog. 3. methotrexate.
d. associated with hepatotoxicity. 4. cyclophosphamide.
e. associated with azoospermia. 5. cyclosporine.
f. associated with hemorrhagic cystitis.
g. associated with hemolytic anemia.
h. associated with renal failure.
i. useful in the treatment of sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) syndrome, and severe ocular cicatricial pemphigoid (OCP).
j. potent suppressor of interleukin-2 production.
k. useful in the treatment of mild to moderate OCP.
l. useful in the treatment of SO, VKH, Behçet’s disease, and corneal graft rejection.

135. Match each of the following anti–human immunodeficiency virus (HIV) agents with its most prominent adverse effect(s).
a. ritonavir. 1. cytopenia.
b. zidovudine (AZT). 2. severe pancreatitis.
c. didanosine (ddI). 3. peripheral neuropathy.
d. zalcitabine (ddC). 4. nausea, diarrhea.
Answers

1. False. Passive immunization (parenteral administration of antibody) confers immediate but short-lived protection, whereas active immunization (vaccination with active or inactivated immunogens) confers protection that lasts months to years.

2. c. Natural killer (NK) cells are a distinct class of lymphocytes that have the ability to lyse a wide variety of cell types. It is felt that they represent the front-line defense against infections and neoplasia. NK cells may be involved in ocular protection against cytomegalovirus (CMV) retinitis or herpes simplex ocular infections.

3. d. Killer, or K, cells require antibody to effect cell death through the so-called antibody-dependent cellular cytotoxicity (ADCC).

4. b. Typically, lipids and nucleic acids are not antigenic but may become so if coupled with proteins or polysaccharides.

5. e. Macrophages can initiate the immune cascade by "phagocytosing" antigen and presenting it to T cells. Macrophages are also known as antigen-presenting cells.

6. e. IgM peaks earlier and disappears earlier than IgG during the primary immune response.

7. IgG is the major antibody formed following exposure to an antigen that has previously been encountered.

8. True. Class I molecules are found on all nucleated human cells. The subregions A, B, and C within the human leukocyte antigen (HLA) system are responsible for their genetic coding.

9. False. Major histocompatibility complex (MHC) class II molecules are expressed on the surfaces of "antigen-presenting cells"—including macrophages, dendritic cells, and B cells—where they serve to stimulate CD4+ T cells and initiate cytokine release that regulates other cellular processes associated with immune responses. The Dr, Dp, and Dq regions code for these antigens.

10. a. The human leukocyte antigen (HLA) complex governs immune response and surveillance.

11. c. Intravenous drug use accounts for approximately 25% of cases of human immunodeficiency virus (HIV) transmission. Sexual intercourse accounts for 70% of cases.

12. c. Other human leukocyte antigens (HLA) associations include:
   - A29—Birdshot chorioretinitis
   - B7 and DR2—Presumed ocular histoplasmosis syndrome (POHS) and multiple sclerosis (MS)
   - B8—Sarcoidosis, intermediate uveitis
   - B27—Idiopathic iritis, psoriatic arthritis, inflammatory bowel disease, ankylosing spondylitis, Reiter’s syndrome
   - B44—Retinal vasculitis

B31—Behçet’s disease (in Asians only)

13. True. The carboxy termini form the constant portion of the antibody molecule.

14. c. IgM is made of five units of the size of one IgG.

15. d. IgG has the longest half-life, 21 to 23 days. IgA is made second at about 6 days, followed by IgM (5 days), IgD (3 days), and IgE (2 days).

16. d. IgG transfer occurs both passively and by active transport. (Minimal amounts of IgA also may cross by passive diffusion.)

17. b. IgA may exist in a dimeric form (two subunits), especially when secreted. IgM is produced as a pentamer (five subunits).

18. c. IgA and IgE may play a role in activating the “alternative” pathway, but neither directly binds complement components nor initiates the complement cascade like IgG and IgM.

19. True. Two IgA molecules require two extra, non-immunoglobulin proteins to form true secretory immunoglobulin. The first is the J-chain provided by the plasma cell. The dimer is secreted, and as it passes through the epithelium, a “secretory piece” is added to form definitive secretory immunoglobulin.

20. e. IgM or IgM-like immunoglobulins tend to be the only type present in organisms with the most “rudimentary” immune systems (relative to mammals).

21. a. IgM, IgG, and IgD can be demonstrated on the surface of “virgin” B-lymphocytes and are involved with binding antigen, leading to the activation of the cells, and confer the capability for anamnestic responses.

22. c. Following initial exposure to an antigen, IgG will be produced in great abundance and may thereafter participate in the reidentification of the antigen, as well as response to it. IgE, after initial sensitization, binds to mast cell surfaces. Cross-linking by antigen of bound IgE molecules leads to histamine release (type I hypersensitivity).

23. b. C3 is present in serum at a concentration of approximately 1 mg/mL.

24. a. Type I reactions involve cross-linking of IgE bound to mast cells and basophils. Type II reactions frequently feature IgG or IgM interaction with either cytotoxic cells or circulating immunoglobulins and antigens and the subsequent deposition of those complexes. (Note that some type II processes are not strictly antibody dependent.) Type III reactions result from antigen–antibody complex formation.

25. True. Cyclooxygenase inhibitors (nonsteroidal agents) block production of all three end products.
26. False. Although edema occurs as a result of the effects of vasoactive substances released during mast cell and basophil activation, inflammatory cell infiltration of the affected area does not occur in the acute phase of type I reactions.
27. False. Cytotoxic T cells and killer (K) cells may be involved without complement playing a role; also, macrophages may ingest target cells marked with antibody, again without complement involvement.
28. True. The Arthus reaction (type III) results from formation of antigen–antibody complexes in tissue. This is different from circulating immune complexes that formed in blood. Neutrophils are the key players in effecting subsequent tissue damage.
29. True. Interleukins are products of activated monocytes, lymphocytes, and activated fibroblasts, which in general seem to upregulate cellular immunity. Fibroblasts also produce interferons, as do lymphocytes, which may augment or suppress cellular immune response.
30. e. Graves’ disease and myasthenia gravis are excellent examples of type V hypersensitivity. In these conditions, antibodies react with cell surface receptors and either stimulate or depress cellular function.
31. b. All major immunoglobulins classes except IgD have been detected in human tears. IgA (secretory immunoglobulin) is the primary immunoglobulin in tears.
32. c. Wessely’s rings, also known as immune rings, are ring infiltrates of the corneal stroma, parallel to the limbus. Some corneal rings are probably formed as antigen from a corneal infiltrate encounters antibody from peripheral corneal blood vessels. The infiltrate generally contains complement factors and polymorphonuclear neutrophils (PMNs).
33. c. Up to 90% of patients with ankylosing spondylitis are positive for HLA-B27. HLA-B27 is also associated with several other diseases, although the chance that an HLA-B27–positive individual will have a seronegative spondyloarthropathy or eye disease is approximately 25%. Note that 85% to 95% of patients with Reiter’s syndrome are HLA-B27–positive as well. Fifty percent of patients with acute iritis may be HLA-B27–positive.
34. a. Dalen-Fuchs nodules are focal accumulations of epithelioid-like cells between Bruch’s membrane and the retinal pigment epithelium (RPE). They may include degenerated RPE cells. They are classically associated with sympathetic ophthalmia (SO) and Vogt-Koyanagi-Harada (VKH) syndrome. They also may be found in tuberculous choroiditis and sarcoidosis.
35. e. Idiopathic iridocyclitis is the most common cause of anterior uveitis, making up at least 10% of all uveitis cases. HLA-B27 iridocyclitis is the second most common cause, and juvenile rheumatoid arthritis (JRA) and herpes (simplex or zoster) follow in incidence.
36. c. Toxoplasmosis is the most common cause of posterior uveitis, accounting for up to 7.0% of total uveitis cases. Other causes of posterior uveitis include retinal vasculitis, necrotizing herpetic retinopathy, and idiopathic causes.
37. c. The differential diagnosis of diffusely distributed keratic precipitates includes Fuchs’ heterochromic iridocyclitis, and rarely sarcoidosis, syphilis, and toxoplasmosis. The diffuse distribution, along with a gelatinous, stellate appearance, makes the keratic precipitates (KPs) of Fuchs’ iridocyclitis distinctive.
38. a. Granulomas as well as Koepppe and Busacca nodules may appear as iris nodules in ocular sarcoidosis. Juvenile xanthogranuloma (JXG) features yellow iris nodules, appearing during childhood and associated with spontaneous hyphema. JXG most frequently involves the skin. Raised orange lesions are typical. In neurofibromatosis, Lisch nodules, actually clusters of nevus cells, are definitive. Cutaneous neurofibromas, café-au-lait spots, and Lisch nodules are all important diagnostic criteria. Pseudoxanthoma elasticum is associated with “chicken skin” of the head and neck, but no iris nodules.
39. d. Intermediate uveitis accounts for 15% of all uveitis cases, and is associated with several systemic disorders including sarcoidosis, multiple sclerosis (MS), Lyme disease, tuberculosis (TB), and syphilis. Idiopathic intermediate uveitis, or pars planitis, accounts for >80% of cases. Eighty percent of pars planitis cases are bilateral. Macular edema, followed by cataract, is the most consistent cause of decreased vision in pars planitis. Cystoid macular edema (CME) may complicate anterior or posterior uveitis, but much less frequently.
40. a. The human leukocyte antigens (HLA) are determined by a series of four gene loci located on chromosome 6. HLA-B27 is present in 1.4% to 6% of the general population.
41. b. Ritonavir is a protease inhibitor which can increase CD8+ lymphocyte counts.
42. e. All of the diseases listed, along with reactive arthritis are known as the seronegative spondyloarthropathies, a group of autoimmune disease strongly associated with HLA-B27 positivity and acute anterior uveitis. The term seronegative refers to rheumatoid factor (RF); patients with these diseases by definition do not have a positive RF.
43. c. Up to 25% of individuals with HLA-B27 develop sacroiliac disease. Symptoms of sacroiliac disease may be subtle. Personal or family history of back problems in patients with iritis should prompt the physician to obtain sacroiliac radiographs.
44. d. Asymptomatic sacroiliac disease can be seen in patients with HLA-B27 spondylitis, particularly in young men. Because irreversible damage may occur before the onset of significant symptoms and simple physical
Answers

45. d. The most common eye finding in Reiter's syndrome is a nonspecific conjunctivitis. Nongranulomatous iritis, which can be bilateral and chronic, is less common. Keratoderma blennorragicum (answer e.) and circinate balanitis are also diagnostic criteria for the disease.

46. True. Although it is associated with both forms of inflammatory bowel disease, iritis occurs more commonly in patients with ulcerative colitis. Less than 15% of patients with ulcerative colitis and <5% of patients with Crohn’s disease develop acute anterior uveitis.

47. False. Acute anterior uveitis is associated with psoriatic arthritis, but usually not with psoriasis without arthritis.

48. c. Behçet’s disease is much more common among Japanese and individuals from eastern Mediterranean countries. Posterior uveitis, which can include retinal vasculitis, retinal hemorrhages, and retinal necrosis, is more common than anterior uveitis in Behçet’s disease.

49. True. Mild anterior uveitis with nonspecific symptoms is common in glaucomatocyclitic crisis (Posner-Schlossmann syndrome). Although the symptoms may be mild, intraocular pressure (IOP) may be markedly elevated during the recurrent episodes, which may last several days.

50. False. Older anterior chamber intraocular lenses, some of which were poorly sized or manufactured, have been found to cause the uveitis-glaucoma-hyphema syndrome due to chronic irritation of the iris root. The condition also has been rarely described with more modern posterior chamber lens styles.

51. d. Aspirin is the drug of choice for Kawasaki’s disease. Although most patients recover without complication, approximately 3% of children with Kawasaki’s disease develop acute coronary arteritis, which may lead to myocardial infarction (MI) and death. Corticosteroids are contraindicated in Kawasaki’s because of the increased risk of coronary aneurysm formation.

52. True. Herpetic uveitis is more commonly associated with elevation of intraocular pressure (IOP) than other types of uveitis. The differential diagnosis of uveitic glaucoma also includes sarcoidosis, zoster, Fuchs’ iridocyclitis, and rarely toxoplasma, syphilis, and sympathetic ophthalmitis (SO).

53. False. Segmental iris atrophy is more characteristic of herpes zoster (caused by a segmental iris vasculitis). With herpes simplex, patchy iris atrophy near the pupillary margin is more common.

54. c. Inflammation of the uveal tract lasting <6 weeks is defined as chronic uveitis.

55. d. Rheumatoid factor is typically absent in patients with uveitis secondary to juvenile rheumatoid arthritis (JRA).

Also, patients with polyarticular JRA are less likely to develop uveitis than those with pauciarticular (fewer than five joints) or monoarticular disease.

56. True. The iritis in juvenile rheumatoid arthritis (JRA) has been discovered in routine school eye examinations. The eye can be white and quiet, and the symptoms are often mild.

57. True. A chronic iridocyclitis indistinguishable from juvenile rheumatoid arthritis (JRA), but without arthritis, occurs primarily in young girls.

58. d. For unknown reasons, synechiae are unusual in Fuchs’ heterochromic iridocyclitis. Some experts question the “inflammatory” nature of this fascinating disorder. Visual prognosis is usually good after cataract extraction.

59. True. The lighter colored eye is usually (but not always) the eye affected with Fuchs’ heterochromic iridocyclitis. (Chronic inflammation causes stromal atrophy and melanocyte loss, leading to the lighter color.) In some brown eyes, however, iris stromal atrophy causes the involved eye to appear more brown, or darker.

60. False. Cataracts and glaucoma are more common long-term complications in Fuchs' heterochromia.

61. e. Posterior and anterior synechiae can be extensive in sarcoidosis. Although the iridocyclitis in sarcoidosis is classically granulomatous, it also can be nongranulomatous.

62. a. An elevated angiotension-converting enzyme (ACE) level occurs in approximately two thirds of patients with sarcoidosis, and an abnormal chest x-ray (hilar and/or mediastinal adenopathy) is very likely to be found. Elevated serum lysozyme is more sensitive but less specific. Gallium scan also may be helpful in the diagnosis. The antineutrophil cytoplasmic antibody (ANCA) assay is useful in diagnosing Wegener’s granulomatosis, not sarcoidosis.

63. False. Caseation is the hallmark of tuberculosis. Sarcoidosis features noncaseating granulomas.

64. False. Parotid gland infiltration compresses the facial nerve as an innocent bystander (remember that the terminal branches of the facial nerve arborize within the substance of the parotid gland). The Heerfordt-Waldenström syndrome describes fever, parotid enlargement, anterior uveitis, and facial nerve palsy secondary to sarcoidosis.

65. c. Interstitial keratitis (IK) usually produces intense pain and photophobia. The immune response in IK is felt to be an immune response to treponemal antigens (and not live organisms). Standard regimens for neurosyphilis are sufficient to treat luetic IK. Although results of the rapid plasma reagin (RPR) and Venereal Disease Research Laboratory (VDRL) tests may be negative in congenital syphilis, those of the fluorescent treponemal antibody-absorption (FTA-ABS) are usually positive.
66. c. Although nontreponemal tests such as the Venereal Disease Research Laboratory (VDRL) and rapid plasma reagin (RPR) titers decrease with successful syphilis treatment, the fluorescent treponemal antibody-absorption (FTA-ABS) titer usually does not decrease after treatment. Ocular inflammation (e.g., uveitis, chorioretinitis) secondary to syphilis should be treated as neurosyphilis.

67. c. Lyme immunofluorescent antibody titers and enzyme-linked immunosorbent assay (ELISA) for IgM and IgG are positive in only 40% to 60% of cases.

68. c. Tuberculosis is an uncommon but increasingly frequent cause of uveitis in the United States. Tuberculous bacilli may be found histopathologically in eyes with tuberculous uveitis. Tuberculous uveitis may be present even with a normal purified protein derivative (PPD) and normal chest x-ray. For these cases, a second strength (250 tuberculin units) skin test may be positive. Systemic corticosteroids may cause a dangerous flare-up in otherwise quiescent tuberculosis.

69. a. Eighty percent of cases of “peripheral” or intermedia uveitis are bilateral. Vitrectomy may be helpful in clearing media opacities and alleviating vitreous traction, but chronic cystoid macular edema (CME) is often vision limiting. Hypotony caused by chronic ciliary body inflammation aggravates this.

70. False. Postoperative endophthalmitis usually results from wound contamination by the patient’s lids and conjunctiva, generally with endogenous flora.

71. False. Three common signs of endophthalmitis are decreased vision, hypopyon, and vitritis. Pain is variable; if endophthalmitis is painful, the resolution of pain may indicate improvement of the endophthalmitis.

72. d. Acute bleb-associated endophthalmitis can occur at any time following successful filtration surgery. Pneumococcus (Streptococcus pneumoniae) and Haemophilus influenzae are the most frequent pathogens.

73. False. Although prolonged, complicated, invasive surgeries have a higher incidence of postoperative endophthalmitis, even surgeries that do not include ocular penetration, such as pterygium excision and strabismus surgeries, may be associated with endophthalmitis.

74. a. Serratia species produce severe acute postoperative endophthalmitis. Certain organisms are clearly implicated in chronic postoperative endophthalmitis and are associated with typical time courses:
   - Staphylococcus epidermidis, within 6 weeks
   - Candida species, 1 to 3 months
   - Propionibacterium acnes, 2 months to 2 years.

75. c. Posttraumatic endophthalmitis incurs a poor prognosis, with <10% retaining vision better than 20/400.

76. c. Posttraumatic endophthalmitis has a uniquely high percentage of Bacillus species, especially B. cereus, represented etiologically. Estimates have ranged from 20% to 25%, and the organisms seem to be particularly associated with retained metallic foreign bodies, as well as farm or soil-related injuries. B. cereus endophthalmitis can be extremely fulminant. The incidence of Staphylococcus epidermidis endophthalmitis is slightly greater (about 30% of posttraumatic endophthalmitis).

77. c. Bacillus cereus may be the single most destructive organism encountered in ocular infections. The organism’s enzymes and exotoxins can produce unsalvageable destruction within 24 hours. Of interest, B. cereus and Clostridium are two organisms capable of producing systemic, constitutional symptoms from endophthalmitis.

78. a. This group of drugs, the reverse transcriptase inhibitors, was the first available and front-line set of agents in the fight against human immunodeficiency virus (HIV). Ritonavir is a protease inhibitor.

79. c. Most of the fungal endogenous endophthalmitis occurs without evidence of fungemia.

80. True. Considerable inflammation can occur postoperatively, but vitritis out of proportion to anterior chamber reaction should provoke suspicion of infectious endophthalmitis.

81. b. Gradually decreased vision, floaters, and scotomata are common symptoms of posterior uveitis. Ciliary flush and spasm with brow-ache are more typical of anterior uveitis.

82. c. Granulomatous inflammation of the anterior segment can occur in toxoplasmosis. Perivasculitis near active retinal lesions is common (kyrieleis arteriolitis). The classic lesion of toxoplasmosis is exudative focal retinitis. The definitive host for Toxoplasma gondii is the cat, where it is found as an intestinal parasite. (The gondi is a small South American rodent, which is an important intermediate host in that region of the world.)

83. d. Clindamycin is clearly associated with pseudomembranous colitis. Sulfur drugs can cause Stevens-Johnson syndrome, as well as either hemolytic or aplastic anemia. Pyrimethamine can cause aplastic anemia (hence, the concurrent use of folic acid). Steroid therapy can aggravate diabetes.

84. b. The larvae of Onchocerca volvulus form subcutaneous nodules when they develop into mature worms. This is one manifestation of onchodermatitis.

85. True. Death of the larvae produces a severe inflammatory reaction, with granulomatous inflammation seen around the necrotic organism.

86. a. Iatrogenic immunosuppression, intravenous drug abuse, and in dwelling intravenous catheters for hyperalimentation are risk factors for candidal infections. Candida endophthalmitis is less common in acquired immunodeficiency syndrome (AIDS) (mucocutaneous candidasis is common).

87. c. Ocular infection by cytomegalovirus (CMV) may cause exudative or rhegmatogenous retinal detachments, with holes in the area of retinal necrosis.
88. True. Vision and electrophysiologic testing are usually normal after rubella retinitis.
89. False. Congenital measles can cause a retinitis with blindness to 12 days after the measles rash appears. Infants usually recover fully, but some patients progress to secondary pigmented degeneration with poor prognosis. Similarly, although congenital syphilitic retinopathy can be associated with normal vision, it can also cause extensive retinopathy leading to visual loss.
90. a. Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is believed to follow a prodromal influenza-like illness and has been associated with a cerebral vasculitis.
91. False. Systemic infection with Nocardia is characterized by pneumonia and disseminated abscesses. Nocardia is a filamentary bacterium. Ocular involvement results from hematogenous spread.
92. False. Toxocara canis is a helminth that will penetrate the intestinal wall and take up residence in the liver and lungs. From there, larvae can disseminate to any organ, including the eye. Eye involvement is usually unilateral.
93. a. Peripheral histo spots begin to appear around adolescence. The maculopathy usually does not appear until the 20s. The early stage of the disease is thought to be a choroiditis. Vitreous cells are not seen in presumed ocular histoplasmosis syndrome (POHS). Visual complaints are caused by the maculopathy.
94. d. Sympathetic ophthalmia (SO) is a granulomatous panuveitis. Histologically, the granulomatous inflammation of the uvea is diffuse. The granulomatous process can extend into scleral canals and the optic nerve.
95. d. Although vitiligo and poliosis (known as the Sugiura sign) are classic for Vogt-Koyanagi-Harada (VKH) syndrome, they also have been reported in sympathetic ophthalmia (SO). The same is true of sensorineural hearing loss and cerebrospinal fluid (CSF) pleocytosis. VKH syndrome also may feature other central nervous system signs, such as mual rigidity, fever, coma, and seizures. In VKH syndrome, the chronic diffuse granulomatous uveitis involves the choriocapillaris, whereas in SO this layer is spared. Obviously, the two diseases overlap considerably. SO must be considered in patients with a history of previous eye surgery.
96. c. Conjunctival steroid injection should not be used in cases of infectious uveitis such as toxoplasmosis, because of their potential to aggravate Toxoplasma retinitis. They can also lead to further melting and perforation in necrotizing scleritis. Longer-acting steroids (e.g., triamcinolone) given by periocular injection have the potential for raising intraocular pressure (IOP) for an extended period of time.
97. a. If inflammation has been well-controlled for at least 3 months before surgery, extracapsular cataract extraction with posterior chamber intraocular lens (IOL) implantation may be successful in many types of uveitic cataract. Young patients with cataract secondary to chronic uveitis associated with juvenile rheumatoid arthritis (JRA) should not undergo IOL implantation after cataract extraction. Persistent postoperative uveitis aggravated by an IOL can be devastating.
98. a. Patients with acquired immunodeficiency syndrome (AIDS) exhibit absolute lymphocytopenia, elevated immunoglobulin (especially IgA and IgG), and increased suppressor T-cell counts, particularly relative to helper T cells. Although there is an absolute lymphocytopenia, there may be no leukocytopenia or granulocytopenia.
99. c. Over 65% of patients with acquired immunodeficiency syndrome (AIDS) develop some ocular abnormality. Human immunodeficiency virus (HIV) retinopathy which occurs in >50% of cases is the most common ocular finding in patients with AIDS. One series reported that up to 92% of patients with AIDS will develop cotton-wool spots.
100. d. Cytomegalovirus (CMV) retinitis, along with Pneumocystis carinii pneumonia and Kaposi’s sarcoma, was one of the infections recognized early in the course of the epidemic as a defining feature of the disease.
101. True. The occurrence of cytomegalovirus (CMV) retinitis in a patient with acquired immunodeficiency syndrome (AIDS) is thought to confer a poor prognosis because many patients die within months after onset of retinitis. Yet, the mean survival of patients with AIDS and newly diagnosed CMV retinitis has increased as a result of newer therapies (e.g., highly active antiretroviral therapy [HAART], protease inhibitors). CMV retinitis usually occurs with CD4 counts <50 cells/mm³.
102. False. Usually, cases of ocular toxoplasmosis in patients with acquired immunodeficiency syndrome (AIDS) rarely show the typical fundus scar of a previous infection. Primary ocular toxoplasmosis, associated with central nervous system (CNS) toxoplasmosis, is more common in patients with AIDS. Thus, patients with AIDS diagnosed with ocular toxoplasmosis should undergo brain magnetic resonance imaging (MRI) to rule out further CNS involvement.
7: Uveitis

105. b. Pneumocystis choroiditis is seen particularly in patients receiving aerosolized pentamidine. Histopathologically, the lesions in the choroid contain cysts or trophozoites of *Pneumocystis carinii*. IV TMP-SMX therapy is the treatment of choice for *Pneumocystis carinii* pneumonia (PCP) choroiditis.

106. c. The development of herpes zoster ophthalmicus (HZO) in a young and otherwise healthy patient should raise the suspicion of immunocompromise (leukemia, chemotherapy, acquired immunodeficiency syndrome [AIDS]). Sexually transmitted diseases tend to occur together. This is particularly true of syphilis and human immunodeficiency virus (HIV). If a clinician obtains serologic studies for one disorder (lues or AIDS), he or she should strongly consider testing for the other as well (joint fluorescent treponemal antibody-absorption [FTA-ABS] and anti-HIV titers).

107. b. Kaposi’s sarcoma may be noted on the eyelid skin or conjunctiva. Skin lesions usually appear as nontender, elevated, purple nodules. Conjunctival involvement is manifested by red subconjunctival masses.

108. d. The protease inhibitors have shown great promise in actually reducing viral load systemically.

109. a. Bactrim is effective and has a relatively low cost. Unlike aerosolized pentamidine, it is a systemic treatment and may also be effective for treating extrapulmonary pneumocystis (e.g., choroidal).

110. a. Progression of the disease (to acquired immunodeficiency syndrome [AIDS]) was delayed in patients treated prophylactically.

111. e. Treatment is usually started if cystoid macular edema (CME) is evident, or if visual acuity is <20/40.

112. d. Identifiable causes of pars planitis include syphilis, tuberculosis, Lyme disease (rare), sarcoidosis, toxocariasis, and intermediate uveitis associated with multiple sclerosis. However, most cases are idiopathic.

113. e. Systemic corticosteroids are generally used initially, followed by cyclosporine or cytotoxic drugs as second-choice options.

114. True.

115. True. *Mycobacterium tuberculosis*, which is highly aerobic, can produce choroidal lesions.

116. False. Although human immunodeficiency virus (HIV) has been detected in corneal epithelium, as of 2004 there has never been a reported case of HIV transmission from corneal transplantation.

117. c. Up to 25% of patients with primary central nervous system lymphoma (PCNSL) will have ocular involvement.

118. True. Although systemic corticosteroids and immunosuppressive agents can reduce intraocular inflammation initially, they will eventually fail. Unfortunately, the use of these agents can sometimes cause vitrectomy specimens to be nondiagnostic.

119. a. Choroidal metastases are most commonly from a primary lung tumor in men.

120. b. Choroidal metastases are most commonly from a primary breast tumor in women.

121. d. Retinal metastases are rare. Cutaneous melanoma is the most common primary tumor.

122. a. Krill’s disease usually subsides without any treatment over 2 to 3 months.

123. a. Eighty percent of multiple evanescent white dot syndrome (MEWDS) cases are unilateral. Visual prognosis is excellent, and there is usually no treatment required. All of the other answers are typically bilateral.

124. a. This is a classic case of ocular cysticercosis. Surgical removal of the larvae is warranted. Praziquantel and laser therapy can also be used but risk worsening of the intraocular inflammation.

125. 1. IgG (1.0 to 1.4 g/100 mL).

126. 2. IgA (0.2 to 0.3 g/100 mL).

127. 3. IgM (0.04 to 0.15 g/100 mL).

128. 4. IgD (about 0.003 g/100 mL).

129. 5. IgE (about 0.00007 g/100 mL).

130. a. 3, b. 2, c. 4, d. 1, e. 4, f. 1, g. 4, h. 3, i. 4, j. 4. Note that corneal graft rejection may involve type II hypersensitivity mechanisms as well.

131. a. 2, b. 3, c. 1, d. 2, e. 2, f. 1. One suggested mnemonic is P for Pupil margin and for KoePpe.

132. 1. c, 2, b, 3. a. Historical data suggest a 7% incidence of endophthalmitis following penetrating ocular trauma. Older surveys estimate the incidence of
bleb-related infections to be approximately 1%, although more recent data suggest a lower rate, perhaps approaching that following cataract extraction, 0.1%.

133. a. 3, b. 2, c. 1, d. 3, e. 2, f. 2, g. 2, h. 1, i. 2, j. 3. Blurry vision is more likely to be the presenting symptom in serpiginous choroidopathy, because of cystoid macular edema (CME). Serpiginous lesions may spread contiguously, but birdshot lesions are usually scattered. Retinal vessels may be sheathed in both conditions. In birdshot chorioretinitis, angiography reveals pronounced perifoveal capillary leakage and CME. Epiretinal membranes are common in birdshot chorioretinitis, and the electoretinogram (ERG) is reduced or extinguished late in its course. HLA-A29 has been detected in ~90% of patients with birdshot, whereas there is no clear human leukocyte antigen (HLA) association seen with serpiginous choroidopathy (one report showed an association with HLA-B7). Both serpiginous choroidopathy and birdshot chorioretinitis may be associated with choroidal neovascularization.

134. a. 4, b. 2, c. 3, d. 3, e. 1, f. 4, g. 6, h. 5, i. 4, j. 5, k. 6, l. 5. Alkylating agents function by cross-linking DNA. Alkylating agents in this list are cyclophosphamide and chlorambucil. Methotrexate (and rarely cyclosporine and chlorambucil) is associated with hepatotoxicity. Sperm banking is recommended before chlorambucil therapy. Dapsone is associated with hemolytic anemia in patients with glucose-6-phosphate dehydrogenase (G6PD) deficiency. Cyclosporine is strongly associated with renal failure. Cyclophosphamide is useful in the treatment of Vogt-Koyanagi-Harada (VKH) syndrome, ocular cicatricial pemphigoid (OCP), and sympathetic ophthalmia (SO). Cyclosporine is a potent suppressor of interleukin-2 production and is useful in the treatment of SO, VKH, Behçet’s disease, and corneal graft rejection. Dapsone is useful in the treatment of mild to moderate OCP.

135. a. 4, b. 1, c. 2, d. 3.

■ Suggested Readings


Glaucoma, Lens, and Anterior Segment

Questions

1. Which of the following is/are primary determinants of intraocular pressure (IOP)?
   1. episcleral venous pressure.
   2. rate of aqueous humor secretion.
   3. aqueous humor outflow facility.
   4. relative pupillary block.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

2. Which of the following statements about the production of aqueous humor is/are true?
   1. The active transport component is independent of intraocular pressure (IOP), whereas the ultrafiltration component decreases as IOP decreases.
   2. Aqueous humor formation is probably based on active ionic secretion with secondary passive fluid movement.
   3. Aqueous humor formation is entirely dependent on adenosine triphosphatase (ATPase) activity.
   4. Aqueous humor formation diminishes considerably during the night.
   a. 1, 2, and 3.
   b. 2 and 3.
   c. 2 and 4.

3. T or F Approximately 1% of the anterior chamber volume turns over every minute.

4. Which of the following factors is/are associated with a decrease in aqueous humor formation?
   1. inflammation.
   2. surgery.
   3. ocular trauma.
   4. age.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

5. T or F The trabecular meshwork (TM) functions like a one-way valve.

6. Uveoscleral outflow accounts for what percentage of total aqueous outflow facility?
   a. <2%.
   b. <5%.
   c. 5% to 10%.
   d. 10% to 50%.
   e. 75% to 90%.

7. T or F Miotics decrease uveoscleral outflow facility.
Questions

8. The mean value for outflow facility in normal eyes is:
   a. 0.05 μL/min/mm Hg.
   b. 0.15 μL/min/mm Hg.
   c. 0.28 μL/min/mm Hg.
   d. 0.48 μL/min/mm Hg.
   e. 1.00 μL/min/mm Hg.

9. T or F The normal range for episcleral venous pressure is 4 to 8 mm Hg.

10. T or F In chronic elevation of episcleral venous pressure, each 1 mm Hg alteration in episcleral venous pressure will lead to a 1 mm Hg alteration in intraocular pressure (IOP).

11. T or F Intraocular pressure (IOP) is distributed along a gaussian curve.

12. T or F The typical range of diurnal fluctuation in intraocular pressure (IOP) ranges from 2 to 6 mm Hg.

13. T or F The timing of diurnal intraocular pressure (IOP) fluctuations varies from person to person.

14. T or F The general pattern of diurnal fluctuation in intraocular pressure (IOP) in a patient with open-angle glaucoma shows a consistently elevated IOP with less variability.

15. Which one of the following statements about Goldmann applanation tonometry is false?
   a. It is on the basis of the Fick principle, which holds that the pressure inside an ideal sphere is equal to the force required to flatten the sphere divided by the area of flattening.
   b. The diameter of flattening, 3.06 mm, is based on counterbalancing the corneal resistance and the capillary attraction of tears for the tonometer head.
   c. The intraocular pressure (IOP) on the Goldmann scale is equal to the force required to flatten the cornea multiplied by 10.
   d. Like Schiotz tonometry, applanation tonometry reflects ocular rigidity.
   e. Goldmann tonometer tip alignment is important in accurately determining IOP in eyes with high degrees of corneal astigmatism.

16. T or F The Perkins tonometer may only be used on supine patients.

17. Which of the following may be accompanied by a falsely low Goldmann intraocular pressure (IOP) measurement?
   1. increased corneal thickness.
   2. corneal edema.
   3. severe corneal scarring.
   4. a history of refractive surgery.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

18. Angle closure secondary to plateau iris is most often secondary to what primary underlying anatomic derangement?
   a. abnormally concave corneoscleral limbus.
   b. abnormally thickened peripheral iris stroma.
   c. forward displacement of the ciliary processes.
   d. hypertrophic iris pigment epithelium.
   e. increased relative pupillary block.

19. The anterior chamber angle structures may not be directly visualized because:
   a. There is total internal reflection at the aqueous-corneal endothelial interface.
   b. There is excessive scattering of light reflected from the angle off the iris.
   c. Opaque sclera overhangs the anterior chamber angle sufficiently to preclude direct visualization.
   d. There is total internal reflection at the cornea-air interface.
   e. The anterior chamber angle can be directly visualized, with a sufficiently oblique angle of view.

20. Which of the following gonioscopic methods is/are direct?
   1. Goldmann.
   2. Richardson.
   a. 1, 2, and 3.
   b. 1 and 3.
21. Which method of gonioscopy is considered best for evaluating a patient with potential traumatic (angle-recession) glaucoma?
   a. Goldmann.
   b. Koeppe.
   c. Zeiss.
   d. Sussman.
   e. any of the above.

22. Which gonioscopic method is least likely to distort the anterior chamber anatomy and lead to an incorrect diagnosis?
   a. Goldmann.
   b. Koeppe.
   c. Zeiss.
   d. Sussman.
   e. any of the above.

23. T or F Digital pressure on a Goldmann lens may tend to narrow the angle.

24. T or F Compression gonioscopy with the Goldmann contact lens is useful in distinguishing appositional angle closure from synechial angle closure.

25. T or F The superior portion of the angle is almost always the easiest for distinguishing landmarks.

26. T or F A grade IV angle is less likely to undergo spontaneous closure than a grade I angle.

27. Which of the following conditions may be associated with blood in Schlemm’s canal?
   1. carotid-cavernous fistula.
   2. severe thyroid eye disease.
   3. excessive digital pressure on a Goldmann gonioscopic lens.
   4. ocular hypotony.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

28. Abnormally heavy trabecular meshwork (TM) pigmentation is associated with:
   1. pseudoexfoliation.
   2. pigment dispersion.
   3. previous trauma or surgery.
   4. anterior segment dysgenesis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

29. T or F The mechanical theory of glaucomatous damage of the optic nerve states that pressure-induced distortion of optic nerve head blood vessels leads to hypoperfusion and axonal loss.

30. Several investigators have provided evidence that a gene responsible for some cases of juvenile-onset primary open-angle glaucoma (POAG) resides on chromosome:
   a. 1.
   b. 3.
   c. 8.
   d. 17.
   e. X.

31. Which one of the following statements about perimetry is true?
   a. In static perimetry, the stimulus intensity is held constant (static) and moved centrally until it is detected.
   b. Kinetic perimetry is most useful for quantifying and tracking visual field changes in a patient with established glaucoma.
   c. Early, specific signs of glaucoma include generalized constriction of isopters and baring of the blind spot.
   d. For a visual field defect to be classified as glaucomatous, it should have corresponding optic nerve head abnormalities.
   e. Vertical steps, that is, sensitivity discrepancy across the vertical meridian, are sensitive, early signs of glaucomatous visual field loss.

32. A 64-year-old African American man with a history of open-angle glaucoma presents with a Goldmann visual field documenting a superior nasal step to the I4e isopter in the right eye.
When he returns with a deteriorated visual field one year later, the most likely form of deterioration is:

a. an inferior Bjerrum scotoma.
b. a superior paracentral scotoma.
c. an inferior nasal step.
d. encroachment of his superior nasal step toward fixation.
e. a superotemporal wedge defect.

33. Which one of the following visual field patterns will most quickly progress to loss of fixation?

a. central 5-degree island.
b. a large superior nasal step encroaching on fixation (<10 degrees).
c. superior and inferior nasal steps encroaching to 20 degrees.
d. superior and inferior paracentral scotomas with no other defects.
e. split fixation to the I4e isopter.

34. T or F Static perimetry is considered suspicious for visual field defects if a single testing point is depressed by 10 dB or a cluster of three adjacent points is depressed by 5 dB from a population of age-matched normal patients.

35. Which of the following features would raise doubts about the diagnosis of glaucomatous optic nerve damage?

1. pallor out of proportion to cupping.
2. markedly asymmetric color visual loss.
3. field defects obeying the vertical meridian.
4. presence of afferent pupillary defect.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

36. Which of the following statements about open-angle glaucoma is/are true?

1. The particulate glaucoma constitute most of the open-angle glaucomas.
2. Glaucoma is the leading cause of irreversible blindness in African American patients.
3. There is an association of open-angle glaucoma with hyperopia.
4. There is a bidirectional association of diabetes and glaucoma.

37. A 54-year-old man presents with intraocular pressures (IOPs) of 24 mm Hg in the right eye and 26 mm Hg in the left eye. His visual fields and optic nerve heads appear normal. Which of the following clinical features might prompt prophylactic topical medical therapy?

1. elevated pachymetry measurements.
2. family history of glaucoma.
3. elevated nerve fiber layer thickness measurements using optical coherence tomography.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

38. Which of the following conditions might be misdiagnosed as normal-tension glaucoma on initial evaluation?

1. parasellar tumors.
2. methanol toxicity.
3. arteritic anterior ischemic optic neuropathy (AAION).
4. primary open-angle glaucoma (POAG).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

39. T or F Therapy for normal-tension glaucoma generally is more aggressive, with a lower target intraocular pressure (IOP) than therapy for primary open-angle glaucoma (POAG).

40. Features that distinguish pseudoexfoliation glaucoma from primary open-angle glaucoma (POAG) include:

1. greater sensitivity to laser therapy.
2. greater degree of interocular asymmetry.
3. degree of trabecular meshwork (TM) pigmentation.
4. the age of affected patients.
41. Which of the following statements about pigmentary glaucoma is/are true?
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

42. Which of the following glaucoma is least likely to respond to medical therapy alone?
   a. pseudoexfoliation.
   b. pigmentary glaucoma.
   c. phacolytic glaucoma.
   d. lens particle glaucoma.
   e. phacoantigenic glaucoma.

43. T or F Fuchs' heterochromic iridocyclitis is a typical rubeotic glaucoma culminating in secondary angle closure.

44. T or F Acid burns of the ocular adnexa are more likely to be associated with glaucoma than alkali burns.

45. T or F Hemolytic and ghost cell glaucoma both feature acute pressure increases following intraocular hemorrhage.

46. Topical corticosteroids should be used with caution in patients with:
   a. open-angle glaucoma.
   b. a family history of open-angle glaucoma.
   c. diabetes.
   d. concurrent fungal keratitis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

47. T or F The most common cause of angle closure is pupillary block.

48. T or F Angle closure is three to four times more common in women than in men.

49. T or F Most primary angle-closure glaucoma develops as the pupil initially dilates to mid-position.

50. Specific signs of previous angle-closure glaucoma include:
   a. small white opacities immediately beneath the anterior capsule of the lens.
   b. optic disc cupping.
   c. patchy iris stromal atrophy.
   d. patchy iris pigment epithelial loss.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

51. Which of the following modalities may be successful in breaking an attack of angle-closure glaucoma with pupillary block?
   a. compression gonioscopy.
   b. laser iridotomy.
   c. topical miotic treatment.
   d. oral hyperosmotic agents.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

52. T or F In primary angle-closure glaucoma, once the attack is broken with laser iridotomy in the affected eye, in the absence of peripheral synechialization, no further intervention is required for the patient.

53. Which of the following medications may be associated with the induction or aggravation of angle-closure glaucoma?
   1. pilocarpine.
   2. oral antihistamines.
Questions

3. cyclopentolate.
4. aspirin.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

54. The patient with bilaterally narrow anterior chamber angles and normal intraocular pressure (IOP) should probably undergo which of the following tests?
   a. thymoxamine test.
b. topical steroid challenge.
c. oral water challenge.
d. the prone-dark room test.
e. careful, depressed, dilated examination.

55. A patient with bilaterally narrow anterior chamber angles and elevated intraocular pressure (IOP) should probably undergo which of the following tests?
   a. thymoxamine test.
b. topical steroid challenge.
c. oral water challenge.
d. the prone-dark room test.
e. careful, depressed, dilated examination.

56. Which of the following clinical features might lead to a suspicion of plateau iris in a patient with angle closure?
   1. deep anterior chamber centrally.
   2. a young patient with myopia.
   3. a flat iris plane.
   4. a small anterior segment.

57. Which of the following conditions may be associated with pupillary block and secondary angle closure?
   1. previous episodes of a flat anterior chamber.
   3. neovascular glaucoma.
   4. pseudophakia.

58. Which of the following statements about the irido-corneal endothelial (ICE) syndromes is/are true?
   1. They are almost always unilateral and affect women more frequently than men.
   2. In Chandler’s syndrome, there may be corneal edema with only modestly elevated or normal intraocular pressure (IOP).
   3. Essential iris atrophy features stretch and atrophic iris holes with corectopia.
   4. The degree of IOP elevation directly reflects the amount of angle synechialization.

59. T or F The pigmented lesions in the Cogan-Reese syndrome represent iris nevi.

60. Which of the following statements about malignant glaucoma (aqueous misdirection syndrome) is/are true?
   1. It most commonly arises following intraocular surgery in patients with a prior history of angle closure.
   2. A feature differentiating malignant glaucoma from primary angle closure is the depth of the central portion of the anterior chamber.
   3. Definitive treatment usually alters the nature of the anterior hyaloid face.
   4. The initial medical agent of choice is pilocarpine.

61. T or F Fibrous downgrowth is generally more aggressive than epithelial downgrowth.

62. T or F In angle-closure glaucoma following scleral buckling procedures, a peripheral iridotomy is curative.
63. T or F The mechanism of secondary angle closure following panretinal photocoagulation and that seen following central retinal vein occlusion (CRVO) are partially similar in pathophysiology.

64. Which of the following statements about infantile glaucoma is/are true?
   1. Approximately 50% of cases are primary (i.e., no associated ocular or systemic conditions).
   2. Sixty percent are diagnosed within the first 6 months, and 80% within the first year.
   3. Two thirds of cases are bilateral, and two thirds of the cases affected are boys.
   4. In many cases, gonioscopy reveals an abnormally anterior iris insertion.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

65. Findings in an eye with infantile glaucoma, which are not seen in eyes with adult forms of glaucoma include:
   1. Haab striae.
   2. Corneal edema.
   3. Enlarged corneal diameter.
   4. Optic nerve cupping.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

66. Which of the following agents diminish intraocular pressure (IOP) during general anesthesia?
   1. Halothane.
   2. Enflurane.
   3. Isoflurane.
   4. Ketamine.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

67. T or F Like adult forms of glaucoma, infantile glaucoma is often successfully controlled with medical therapy.

68. The proper pediatric dose of acetazolamide (Diamox) is:
   a. 15 mg/kg/day in one dose.
   b. 15 mg/kg/day in three or four divided doses.
   c. 5 mg/kg/day in one dose.
   d. 5 mg/kg/day in three or four divided doses.
   e. 250 mg/day in three or four divided doses.

69. Which of the following medications would be contraindicated in a patient with a history of paroxysmal tachycardia?
   1. Dipivefrin.
   2. Timolol.
   3. Epinephrine.
   4. Pilocarpine.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

70. Which agents are relatively contraindicated in patients with a history of depression or other psychiatric illness?
   1. Pilocarpine.
   2. Timolol.
   3. Dipivefrin.
   4. Acetazolamide.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

71. Which of the following agents should be used with caution in a patient with narrow angles?
   1. Timolol.
   2. Epinephrine.
   3. Acetazolamide.
   4. Apraclonidine.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
Questions

72. The agent most likely to cause topical sensitization and medicamentous is:
   a. timolol.
   b. betaxolol.
   c. epinephrine.
   d. pilocarpine.
   e. apraclonidine.

73. Which of the following miotics are direct-acting parasympathomimetic agents?
   1. echothiophate.
   2. carbachol.
   3. demecarium.
   4. pilocarpine.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

74. Which of the following miotics are indirect-acting agents?
   1. echothiophate.
   2. carbachol.
   3. demecarium.
   4. pilocarpine.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

75. T or F In general, miotics act to lower intraocular pressure (IOP) by moving peripheral iris out of the anterior chamber angle, thereby enhancing outflow facility.

76. Which of the following complications of miotic administration are more likely with the indirect agents?
   1. cataractogenesis.
   2. punctal stenosis.
   3. retinal tears and detachment.
   4. bradycardia.

77. The side effect of carbonic anhydrase inhibitors that is most commonly encountered is:
   a. gastrointestinal distress.
   b. kidney stones.
   c. anemia.
   d. hypocalcemia.
   e. paresthesias.

78. T or F Intravenous mannitol is more effective than intravenous urea in lowering intraocular pressure (IOP).

79. T or F Osmotic agents are more effective in inflamed eyes, because the blood ocular barrier has been disrupted.

80. Which one of the following statements about Nd:YAG laser iridotomy is true?
   a. The incidence of spontaneous closure of a previously patent iridotomy is higher with Nd:YAG iridotomy than with argon iridotomy.
   b. The Nd:YAG laser is generally more effective for heavily pigmented irides when used alone (without argon laser pretreatment).
   c. The most frequent significant complication of Nd:YAG peripheral iridotomy is cystoid macular edema (CME).
   d. Malignant glaucoma has been reported as a sequela of Nd:YAG laser iridotomy.
   e. Unlike argon iridotomies, Nd:YAG laser iridotomy rarely induces elevation in intraocular pressure (IOP) that might threaten vision.

81. T or F In infantile glaucoma, the treatment of choice in the setting of a cloudy cornea is goniotomy.

82. The correct spot size for argon laser trabeculoplasty (ALT) is:
   a. 50 \mu m.
   b. 100 \mu m.
   c. 200 \mu m.
   d. 250 \mu m.
   e. 500 \mu m.
83. What proportion of patients with open-angle glaucoma enjoy a substantial decrease in intraocular pressure (IOP) during the first year following argon laser trabeculoplasty (ALT)?
   a. 20%.
   b. 40%.
   c. 50%.
   d. 80%.
   e. 90%.

84. Relative to patients with primary open-angle glaucoma (POAG), which of the following conditions is associated with an equal or better response to argon laser trabeculoplasty (ALT)?
   1. aphakic glaucoma.
   2. pigmentary glaucoma.
   3. uveitic glaucoma.
   4. pseudoxfoliative glaucoma.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

85. A 54-year-old white woman presents with glaucomatous optic nerve head changes in each eye and split fixation in her right eye, consistent with her disc findings. Review of her record reveals that she has progressively lost visual field and neural rim tissue while running intraocular pressures (IOPs) in her early teens. Gonioscopy has been documented as normal repeatedly. She is currently on maximal tolerated medical therapy and reports subjective decrease in vision in her right eye. A surgical intervention in the right eye is felt to be the next indicated maneuver. Which procedure would be the one of choice?
   a. iridoplasty.
   b. surgical peripheral iridectomy.
   c. trabeculectomy.
   d. trabeculectomy with a seton.
   e. cyclocryotherapy or cyclophotocoagulation.

86. A 63-year-old patient presents after 4 days following trabeculectomy for early suture lysis. She complains of severe pain and decreased vision in her operated eye. Examination discloses visual acuity that is slightly lower than her last postoperative checkup, a diffusely shallow anterior chamber, and no view of the posterior segment. There is no excessive anterior chamber inflammation, and there are no vitreous cells observed. The most important conditions to be considered in this clinical circumstance include:
   1. endophthalmitis.
   2. malignant glaucoma.
   3. corneal ulcer.
   4. suprachoroidal hemorrhage.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

87. The most helpful diagnostic interventions at this point would include:
   1. anterior chamber paracentesis.
   2. tonometry.
   3. corneal scraping and culture.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

88. T or F The lens arises embryologically from surface ectoderm.

89. Microspherophakia is most often one component of:
   a. Peters' anomaly.
   b. Marfan’s syndrome.
   c. Lowe's syndrome.
   d. Alport’s syndrome.
   e. Weill-Marchesani syndrome.

90. T or F Since microspherophakia may lead to an angle-closure glaucoma with pupillary block, treatment with miotics is helpful until iridotomy can be performed.

91. T or F Congenital lens dislocations are commonly inherited on an autosomal dominant basis.

92. T or F Dilated examination of a patient with Marfan’s syndrome frequently reveals a subluxated lens, upward and outward.
93. Dilated examination of a woman's eye reveals a
dense white spot on the vitreal surface of the poste-
rior capsule just inferonasal to the center of the
posterior capsule. The patient should be advised
that:
   a. cataract formation with visual loss is imminent.
b. she should have a glucose tolerance test
   immediately.
c. she should have urinalysis performed to detect
   hematuria and proteinuria.
d. she has a benign finding with no significant
   implications.
e. all her first-degree relatives should undergo
   ophthalmic examination for visually significant
   cataract.

99. Which of the following conditions is/are associ-
ated with cataracts in juveniles and young adults
(with no history of ocular trauma)?
   1. myotonic dystrophy.
   2. neurofibromatosis type 1.
   3. atopic dermatitis.
   4. galactosemia.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

94. T or F In young children, anterior polar
cataracts are typically unilateral, whereas nuclear
cataracts are typically bilateral.

95. Which of the following forms of infantile or
congenital cataract is least likely to affect vision
seriously?
   a. complete.
b. nuclear.
c. lamellar.
d. capsular.
e. polar.

96. Which of the following is usually not
associated with pediatric cataracts?
   a. Turner’s syndrome.
b. Marfan’s syndrome.
c. aminoaciduria.
d. maternal rubella infection.
e. all of the above conditions are associated with
   pediatric cataracts.

97. T or F The key feature differentiating true
galactosemia and galactokinase deficiency is the
presence of cataract in the former.

98. Which of the following syndromes featuring con-
genital or infantile cataract may be associated with
facial abnormalities?
   1. craniosynostosis.
   2. Hallermann-Streiff syndrome.
   1. incomplete pupillary dilation.
   2. abnormally weak lens zonules.
   3. a thickened or “tough” anterior capsule.
   4. abnormally thin sclera.
8: Glaucoma, Lens, and Anterior Segment

105. A 73-year-old man reports to the office on the first day following his cataract extraction complaining of severe eye pain. Visual acuity is counting fingers at 3 feet in the involved eye. Slit-lamp examination reveals a diffusely shallow anterior chamber and corneal edema without hypopyon. The next step should be:
a. dilated fundus examination.
b. tonometry.
c. gonioscopy.
d. pachymetry.
e. B-mode ultrasonography.

106. After obtaining an applanation intraocular pressure (IOP) of 42 mm Hg and attempting to examine the fundus unsuccessfully, ultrasonography is performed and reveals a normal posterior segment. Gonioscopy reveals a completely closed angle. The next intervention should be:
a. surgical revision of the wound.
b. posterior sclerotomy.
c. peripheral iridotomy.
d. dilation with potent cycloplegics.
e. medical treatment with potent miotics.

107. The intervention in question 106 fails. What is the next step?
a. surgical revision of the wound.
b. posterior sclerotomy.
c. peripheral iridotomy.
d. dilation with potent cycloplegics.
e. medical treatment with potent miotics.

108. Vitreous prolapse into the anterior chamber following lens phacoemulsification with posterior chamber intraocular lens implantation (Phaco/PCIOL) may lead to which of the following complications?

1. localized or diffused corneal edema.
2. open-angle glaucoma.
3. cystoid macular edema (CME).
4. retinal detachment.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

109. Recurrent hyphema weeks to months following uneventful lens phacoemulsification with posterior chamber intraocular lens implantation (Phaco/PCIOL) is a rare syndrome whose etiology may be discerned by:
a. tonometry.
b. gonioscopy.
c. dilated fundus examination.
d. wound exploration.
e. ultrasonography.

110. The incidence of visually significant cystoid macular edema (CME) following uncomplicated extracapsular cataract surgery is approximately:
a. <1%.
b. 1% to 2%.
c. 3% to 5%.
d. 5% to 10%.
e. 15% to 20%.

111. T or F The incidence of angiographic cystoid macular edema (CME) is considerably higher than that of visually significant CME.

112. Independent risk factors for retinal detachment following cataract extraction include:
1. retinal detachment in the contralateral eye.
2. myopia.
3. lattice retinal degeneration.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

113. Which one of the following statements about postoperative endophthalmitis is true?
a. It most commonly presents 7 to 10 days postoperatively, and the most common etiologic agent is Streptococcus.
b. It most commonly presents 2 to 5 days postoperatively, and the most common etiologic agent is Streptococcus.
c. It most commonly presents 2 to 5 days postoperatively, and the most common etiologic agent is Staphylococcus.
d. It most commonly presents 7 to 10 days postoperatively, and the most common etiologic agent is Staphylococcus.
e. Agents responsible for these infections are generally part of the surgeon’s normal flora.

114. **T or F** The style of anterior chamber intraocular lens implant (ACIOL) currently preferred to minimize complications features closed loops with three or four footplates.

115. Which of the following complications is/are more frequently seen following cataract surgery in children than in adults?

1. opacification of posterior capsule.
2. glaucoma.
3. retinal detachment.
4. cystoid macular edema (CME).
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

116. **T or F** Correction of aphakia with spectacles is less successful in children than in adults.

117. The primary ocular manifestation of electrocution injury is:
   a. madarosis.
b. superficial punctate keratitis.
c. iritis.
d. cataract.
e. pigmentary retinopathy.

118. The initial step in management of chemical burns must be:
   a. debridement of foreign particles.
b. irrigation.
c. measurement of intraocular pressure (IOP).
d. slit-lamp examination.
e. fundus examination.

119. Which one of the following statements about traumatic hyphema is false?
   a. Complete hyphemas can resolve without sequelae if not associated with other secondary complications.
   b. The incidence of complications from hyphema increases with rebleeding.
c. It most commonly occurs in young men.
d. Optic nerve atrophy may develop more quickly and at lower intraocular pressure (IOP) in patients with sickle cell disease or trait.
e. Corneal blood staining generally clears rapidly once IOP is normalized.

120. Which of the following constitutes indication(s) for intervention (medical or surgical) in lens subluxation or dislocation?
   1. polyopia.
   2. pupillary block.
   3. corneal-lenticular touch.
   4. poor uncorrected visual acuity.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

121. **T or F** Intraocular copper of >85% to 90% purity generally causes an acute inflammatory response, whereas concentrations less than this are generally inert.

122. **T or F** Iron has an affinity for epithelial cells, whereas copper tends to deposit in basement membranes.

123. Features of ocular siderosis include all of the following except:
   a. deposits in Descemet’s membrane identical to the Kayser-Fleischer ring in Wilson’s disease.
b. iris heterochromia.
c. brunescent cataract.
d. mydriasis.
e. loss of the b-wave on electroretinography.

124. A 37-year-old man presents with gradual loss of vision affecting his right eye. His past medical and ocular histories are unremarkable. Visual acuity is 20/400 in the right eye and 20/15 in the left eye. The neuromuscular examination is normal. Slit-lamp examination reveals stellate keratic precipitates on the right corneal endothelium, along with a “washed-out” appearing iris stroma, and
125. A 48-year-old man presents complaining of difficulty reading. Subjective refraction reveals +0.25 in both eyes, giving 20/15 in both eyes. During the completion of the routine examination, intraocular pressure (IOP) is measured as 34 mm Hg in the right eye and 16 mm Hg in the left eye. Gonioscopy of the right eye reveals prominent, engorged vessels that bridge the angle without synechiae. Gonioscopy of the left eye is normal. The right fundus cannot be clearly seen because of the cataract. Which one of the following is true about this patient?

a. There was probably an episode of intense intraocular inflammation affecting the right eye in the past.
b. Antibody titers to herpes zoster virus are likely to be elevated in serum and aqueous humor.
c. A classic sign of this disorder is hyphema occurring at the beginning of filtration surgery.
d. The glaucoma is caused by typical rubeosis and secondary angle closure.
e. Cataract surgery will cure the glaucoma.

128. A dense white cataract, as shown in the figure below left. There are trace cells in the aqueous. The left eye is normal (as shown in the figure below right). Intraocular pressures (IOPs) are 32 in the right eye and 12 in the left eye. Gonioscopy of the right eye reveals prominent, engorged vessels that bridge the angle without synchiae. Gonioscopy of the left eye is normal. The right optic nerve shows evidence of inferior rim excavation. Which one of the following statements about these findings is false?

a. This entity is more common in men.
b. The incidence and severity of the glaucoma in this condition are correlated with the extent of angle abnormalities.
c. Careful slit-lamp examination may reveal iris abnormalities in the right eye.
d. The same pathophysiologic process will usually affect the left eye within the next 3 to 5 years.
e. The patient should undergo careful, dilated, depressed retinal examination.
126. Which one of the following statements about antimetabolite glaucoma therapy is false?
   a. Mitomycin-C (MMC) directly interacts with DNA, blocking RNA and subsequent protein synthesis in all metabolically active cells.
   b. The antiproliferative effects of MMC are generally more potent and longer-lasting than those of a comparable dose of 5-fluorouracil (5-FU).
   c. Great care should be taken to ensure adequate exposure of the bleb wound edges to MMC in order to gain the maximal antifibrotic effect.
   d. Intraoperative dosage of MMC varies from 0.2 to 0.5 mg/cc for 1 to 4 minutes of exposure, depending on the specific clinical situation.
   e. 5-FU is associated with a greater incidence of postoperative hypotony and surface-related complications than MMC.

127. Which of the following complications of filtration surgery is more likely with inferiorly placed blebs?
   a. choroidal hemorrhage.
   b. cataract.
   c. endophthalmitis.
   d. cystic bleb encapsulation.
   e. aqueous misdirection syndrome.

128. T or F Tonopen readings of intraocular pressure (IOP) are less accurate than Goldmann applanation tonometry in eyes status post laser-assisted in situ keratomileusis (LASIK).

129. T or F Blood enters Schlemm’s canal during gonioscopy when episcleral venous pressure exceeds intraocular pressure (IOP).

130. Which of the following topical steroids given to a patient will likely lead to the highest rise in intraocular pressure (IOP)?
   a. medrysone 1%.
   b. prednisolone 1%.
   c. fluorometholone 0.1%.
   d. dexamethasone 0.1%.
   e. hydrocortisone 1%.

131. All of the following are ocular side effects of the antiepileptic topiramate except:
   a. myopia.
   b. suprachoroidal effusion.
   c. scleritis.
   d. open-angle glaucoma.
   e. blurred vision.

132. What is the most common type of congenital cataract?
   a. complete.
   b. nuclear.
   c. lamellar.
   d. cerulean.
   e. capsular.

133. Which of the following statements about selective laser trabeculoplasty (SLT) is false?
   a. Unlike argon laser trabeculoplasty (ALT), the procedure can be repeated for significant pressure lowering effects.
   b. SLT disrupts intraocular tissue architecture significantly more than ALT.
   c. SLT and ALT have comparable intraocular pressure (IOP)–lowering effects.
   d. SLT targets the melanin-containing cells within the trabecular meshwork (TM).
   e. Patients can receive up to two SLT treatments every year.

134. T or F Generally, uncomplicated cataract surgery raises intraocular pressure (IOP) by 1 to 2 mm Hg.

135. Which of the following treatments should not be advised for no light perception (NLP) eyes?
   a. topical steroids to reduce inflammation.
   b. enucleation for blind, painful eyes.
   c. ciliary body ablation to treat high intraocular pressure (IOP).
   d. retrobulbar alcohol injection for blind, painful eyes.
   e. frequent artificial tear use.

136. Which of the following is not a common side effect of topical prostaglandin analogs?
   a. eyelash hypertrichosis.
   b. conjunctival hyperemia.
   c. darkening of periocular skin.
   d. cystoid macular edema (CME).
   e. iris atrophy/iris transillumination defects.

137. T or F Spontaneous hyphema in an infant is pathognomonic for juvenile xanthogranuloma (JXG).
138. Which of the following is generally not a cause of increased episcleral venous pressure?
- a. thyroid ophthalmopathy.
- b. orbital varix.
- c. superior vena cava syndrome.
- d. Sturge-Weber syndrome.
- e. contralateral carotid artery stenosis.

139. For each of the glaucoma medications that follows, indicate whether it acts to lower intraocular pressure (IOP) by diminishing aqueous humor production, by increasing aqueous humor outflow, or by another mechanism.
- a. timolol.
- b. epinephrine.
- c. acetazolamide.
- d. levobunolol.
- e. apraclonidine.
- f. pilocarpine.
- g. betaxolol.
- h. phospholine.
- i. latanoprost.
- j. dipivefrin.
- k. mannitol.

140. Indicate the proper association for each of the following symptoms or signs:
- a. glaucoma.
- b. polar cataracts and lenticonus.
- c. hematuria.
- d. X-linked inheritance pattern.
- e. progressive sensorineural hearing loss.
- f. vestibular dysfunction.
- g. areflexia.
- h. renal tubular acidosis with rickets.
- i. pigmentary retinopathy.

141. Match the cataract morphologies lettered below with the systemic association numbered below.
- a. sunflower cataract.
- b. oil-droplet cataract.
- c. snowflake cataract.
- d. “Christmas tree” cataract.

- 1. diabetes mellitus type 1.
- 2. myotonic dystrophy.
- 4. galactosemia.
Answers

1. a. As with any system of fluid dynamics, the equation pressure = flow × resistance defines the relationship between these variables. In the eye, the driving pressure for aqueous outflow equals the difference between intraocular pressure (IOP) and episcleral venous pressure (V_e). The outflow facility is the reciprocal of resistance. Therefore,
   \[ IOP - V_e = (\text{aqueous flow/outflow facility}) \]
   so,
   \[ IOP = (\text{aqueous flow/outflow facility}) + V_e. \]
   Aqueous flow, outflow facility, and episcleral venous pressure are thus the primary determinants of IOP. Relative pupillary block can affect IOP by decreasing outflow facility, but is not a primary determinant of IOP.

2. c. A pressure-independent active transport mechanism and a pressure-dependent ultrafiltration component both contribute to aqueous formation. The active transport component is felt to be the primary mechanism of aqueous production. This requires adenosine triphosphatase (ATPase) activity and creates a concentration gradient of certain ions such as sodium chloride or bicarbonate, followed by a passive fluid movement. Ultrafiltration, which is independent of ATP, generally decreases as intraocular pressure (IOP) increases. A diurnal rhythm of aqueous flow has been found to exist, with flow being lower at night. Most people have their highest pressure during the morning hours.

3. True. The anterior chamber volume is approximately 200 to 250 microliters, and the rate of aqueous formation is approximately 2 to 3 μL/min.

4. c. Increasing age, ocular injury or inflammation, and ocular surgery can all decrease the rate of aqueous humor formation.

5. True. The trabecular meshwork (TM) collapses when intraocular pressure (IOP) is low, thereby reducing backflow and functioning to an extent as a one-way valve. Also, even at normal IOP, blood cells cannot reflux out of the TM into the anterior chamber.

6. d. Estimates of this value actually vary, with some being as low as 10%, and some as high as 50%. Uveoscleral outflow is pressure-independent.

7. True. Miotics lower intraocular pressure (IOP) by increasing trabecular outflow facility. Cycloplegic agents, epinephrine, and prostaglandin analogs have been shown to increase uveoscleral flow, whereas miotics decrease uveoscleral flow.

8. c. Outflow facility in normal eyes ranges from 0.22 to 0.28 μL/min/mm Hg, and decreases with age, ocular surgery, and trauma. Patients with glaucoma often have decreased outflow facility.

9. False. The normal range of episcleral venous pressure is approximately 8 to 12 mm Hg.

10. False. Acute elevation in episcleral venous pressure generally leads to an abrupt increase in intraocular pressure (IOP) of roughly equal magnitude. Chronic elevation of episcleral venous pressure has a more complex effect on IOP and may result in elevations that are greater, lesser, or equal in magnitude to the increase in episcleral venous pressure.

11. False. The distribution of intraocular pressure (IOP) cannot be predicted by the normal (gaussian) distribution. The use of the mean +/- 2 standard deviations as the range of "normal IOP" in the general population is not valid because the actual distribution is skewed toward higher IOP.

12. True. More than 10 mm Hg of diurnal fluctuation is suggestive of glaucoma.

13. True.

14. False. Diurnal intraocular pressure (IOP) fluctuation in normal eyes ranges from 2 to 6 mm Hg. Glaucomatous eyes often show a wider range of diurnal variation, sometimes exceeding 20 mm Hg (of fluctuation). The peak IOP in most individuals occurs during the day, although morning, afternoon, and evening peaks have been demonstrated. Measurement of IOP at different times during the day is useful in evaluating patients with apparent glaucomatous optic nerve damage, but with no documented elevation of IOP. Furthermore, IOP in patients with glaucoma is not always similar in both eyes; hence, the use of monocular topical pharmacotherapy trials may not necessarily prove or disprove efficacy of a particular agent.

15. d. Unlike Schiotz tonometry, applanation tonometry displaces a very small volume of aqueous humor from the eye and does not significantly increase intraocular pressure (IOP). For this reason, applanation measurements are essentially independent of ocular rigidity.

16. False. The Perkins tonometer is counterbalanced so that it can be used in the erect or supine position. The Tonopen, a portable electronic applanation device, can also be used in the erect or supine position.

17. c. Goldmann tonometry may be falsely low in patients with corneal edema and patients who have had laser-assisted in situ keratomileusis (LASIK). Digital pressure on the globe or applanation over a corneal scar can lead to artificially high intraocular pressure (IOP) measurements. Patients with increased central corneal thickness may have artificially high IOP measurements.

18. c. Anterior displacement of the ciliary processes pushes the peripheral iris forward, which subsequently narrows the anterior chamber and blocks the trabecular meshwork (TM). This has been confirmed with anterior segment...
8. Glaucoma, Lens, and Anterior Segment

ultrasonography. Patients who have plateau iris syndrome (e.g., resistant to laser peripheral iridotomy) should be treated with long-term miotics and possibly iridoplasty.

19. d. The anterior chamber angle cannot be viewed directly through the cornea because there is total internal reflection of light at the cornea–air interface. Gonioscopy replaces this interface with a new cornea–lens interface. The difference in refractive indices is reduced, so total internal reflection does not occur.

20. c. Koeppe and Richardson lenses are used in direct gonioscopy, whereas the Goldmann and Zeiss lenses are used in indirect gonioscopy. Direct gonioscopy is useful during intraocular surgery (e.g., goniotomy).

21. b. Koeppe gonioscopy is considered best for evaluating a patient with potential angle recession because this system allows easier comparison of one eye with the fellow eye, or one portion of the angle with another.

22. b. The Koeppe lens is least likely to distort the anterior chamber anatomy. Posterior pressure on the Goldmann lens may falsely narrow the angle by indenting the sclera. Posterior pressure with the Zeiss lens will push the aqueous from the center to the periphery of the anterior chamber (AC).

23. True. See answer 22.

24. False. Because of their smaller diameters, Zeiss and Sussman lenses may be used in indentation gonioscopy. This maneuver distinguishes appositional angle closure from synechial angle closure by artificially deepening the anterior chamber with digital pressure on the lens. Goldmann lenses are too large for this.

25. False. The inferior angle (viewed through the superior mirror on a Zeiss goniprisp lens) is wider and is thought to be the easiest portion for distinguishing landmarks.

26. True. Grade IV describes a 45-degree angle between the surface of the trabecular meshwork (TM) and the iris, whereas grade I describes a 10-degree angle. Thus, a grade IV angle is less likely to undergo spontaneous closure.

27. c. Blood may enter Schlemm’s canal when episcleral venous pressure exceeds intraocular pressure (IOP). This may be seen in all of the mentioned conditions.

28. a. Heavy trabecular meshwork (TM) pigmentation is not a feature of anterior segment dysgenesis. It should suggest the differential diagnosis of pseudoxfoliation, pigment dispersion, previous inflammation or surgery, or uveal melanoma.

29. False. This is actually the vascular or ischemic theory of glaucomatous damage. The mechanical theory states that elevated intraocular pressure (IOP) causes pressure-induced damage of optic nerve axons by compression at the lamina cribrosa.

30. a. This form of the disease is characterized by markedly elevated pressures, often >40 mm Hg, and poor response to medication. Other cloned genes known to be associated with glaucoma include PAX6 (11p13, aniridia), CYP1B1 (2p21, congenital glaucoma), PITX2 (4q25, Axenfeld-Rieger syndrome), LMX1B (9q34, glaucoma associated with nail-patella syndrome).

31. d. If a patient has glaucomatous visual field–type defects, corresponding optic nerve head abnormalities should exist. Otherwise, alternative etiologies should be considered. In static perimetry, the stimulus is of variable intensity and is kept stationary (static) until it is noticed by the patient. Baring of the blind spot and generalized constriction are not very specific and can be produced by miosis, uncorrected refractive error, aging, and cataract.

32. d. Areas of retina and/or optic nerve damaged by glaucoma are believed to be more vulnerable to ongoing damage at lower intraocular pressures (IOP), thereby, field defects tend to become more severe with time. New defects also may appear, of course, but generally accompany progression of previous defects.

33. e. Split fixation is the presence of visual field loss that comes close to fixation. A typical pattern of progression is (i) loss near fixation (paracentral scotoma) to (ii) split fixation to (iii) loss of fixation. Thus, the eye at greatest risk is not one with a 5-degree central field, but one with the split–fixation in the horizontal meridian.

34. True. Note that the points at the extreme periphery of the visual field are often depressed because of malpositioning of the near add and should be interpreted cautiously (so-called rim defects).

35. a. It is important to correlate changes in visual field with changes in the optic disc. The following should raise suspicion about the diagnosis of glaucoma: (i) an optic disc that is less cupped than would be expected for observed field loss, (ii) pallor of the disc that is more impressive than the cupping, (iii) markedly asymmetric dyschromatopsia, or (iv) visual field defects uncharacteristic for glaucoma (e.g., respecting the vertical meridian). A relative afferent pupillary defect (APD) can occur with glaucoma.

36. c. Primary open-angle glaucoma (POAG) is the most common of all glaucomas in the United States (60% to 70%), affecting 0.5% to 1% of people over the age of 40. POAG occurs more frequently in African Americans than in whites and is the leading cause of blindness in African Americans. Patients with glaucoma are more likely to have diabetes, and patients with diabetes are more likely to have glaucoma. Angle-closure glaucoma is clearly associated with hyperopia. POAG is strongly associated with myopia.

37. c. Treatment of the suspected patient with glaucoma should be limited to patients with a high risk of ultimate damage to the optic nerve caused by elevated intraocular pressure (IOP). These risk factors include elevated IOP, positive family history of glaucoma, myopia, diabetes
mellitus, cardiovascular disease, race (i.e., African American), asymmetric cupping, large cups, and early nonspecific visual field changes. The Ocular Hypertension Treatment Study has demonstrated that individuals with ocular hypertension and thin corneas are more likely to develop glaucoma. Decreased nerve-fiber layer thickness readings using optical coherence tomography (OCT) would make one suspicious for optic nerve damage.

38. e. A diagnosis of normal tension can be made only after other causes of optic neuropathy are eliminated. Although difficult to prove, the situation that may be the one most frequently misdiagnosed as “normal-tension” glaucoma is primary open-angle glaucoma (POAG) in which intraocular pressure (IOP) fluctuations have obscured the actual nature of the disease. In addition, there are other disorders that can feature pseudoglaucomatous optic nerve cupping, namely: (i) chiasmal compression, (ii) arteritic anterior ischemic optic neuropathy (AAION), (iii) toxic optic neuropathies (such as methanol toxicity), and (iv) hypotension (“shock” optic neuropathy).

39. True. Some experts feel that therapy should be instituted only when progression has been documented. The goal of therapy should be an intraocular pressure (IOP) as low as practical.

40. a. In pseudoexfoliation, fibrillar material is deposited in the anterior segment of the eye. Patients with this glaucoma are often resistant to medical therapy, but laser trabeculoplasty is often very effective. Pseudoexfoliation with glaucoma also differs from primary open-angle glaucoma (POAG) in that it is often monocular or asymmetric and has greater pigmentation of the trabecular meshwork (TM), as well as pigment deposited anterior to Schwalbe line (Sampaiolesi line). There is considerable overlap in the age range of patients affected by each disorder.

41. c. The sine qua non of this condition is radial defects in the iris pigment epithelium. Krukenberg spindles are less specific. Pigmentary glaucoma usually occurs in young myopic men, typically in their third or fourth decade of life. For obscure reasons, women with the disease tend to be older than men. Exercise or pupillary movements may induce a shower of iris pigment release, with resultant increased intraocular pressure (IOP).

42. c. Phacolytic glaucoma results when mature or hypermature cataracts leak high-molecular-weight proteins through microscopic defects in the capsule. A resultant macrophage response clogs the trabecular meshwork (TM). Although medication is used for short-term intraocular pressure (IOP) control, definitive therapy requires cataract extraction. Other lens-induced conditions may respond to topical steroid. Lens particle glaucoma occurs when lens cortex material deposits along the TM and can be seen after cataract surgery. Medical therapy to lower IOP and reduce inflammation can help.

43. False. Fuchs’ heterochromic iridocyclitis is considered a secondary open-angle glaucoma. The glaucoma can be difficult to control and does not parallel the degree of inflammation. The ruberosis in this condition is odd—the vessels are particularly prone to bleed, but do not induce synechialization.

44. False. Alkali penetrates ocular tissues rapidly, unlike acid. The glaucoma that results is probably multifactorial, with primary structural damage to the trabecular meshwork (TM), with sclerosis and shrinking accounting for chronic hypertension.

45. False. In hemolytic glaucoma, hemoglobin-laden macrophages block the trabecular meshwork (TM), whereas in ghost cell glaucoma, rigid, degenerated khaki-colored red blood cells from the vitreous enter the anterior chamber and obstruct the TM. Hemolytic glaucoma may occur within days of hemorrhage, whereas ghost cell glaucoma is seen weeks to months later.

46. c. All of the patients listed are at risk of developing a steroid response or increasing the virulence of current infection.

47. True. Pupillary block is obstruction of flow of aqueous from posterior to anterior caused by a functional block between the lens and iris. It is the most frequent cause of angle-closure glaucoma.

48. True. Pupillary block is more common in women than in men.

49. False. Generally, it occurs after full dilation, as the pupil shrinks to mid-position. This is the region of maximal iris-lens contact.

50. b. The increased intraocular pressure (IOP) that occurs during an attack of angle closure can cause ischemia of the iris and may produce stromal atrophy. Small anterior subcapsular lens opacities, or glaukomflecken, also may develop as a direct result of pressure-induced lens epithelial death. Optic disc cupping may be seen if the attack is prolonged or severe, but this is not specific.

51. e. The major goal of treatment of pupillary block is to relieve the block. Compression gonioscopy may “burp” aqueous through the block and open the angle. In some cases, low-dose pilocarpine (1%) can pull the peripheral iris away from the trabecular meshwork (TM) and break a mild attack of angle-closure. When the intraocular pressure (IOP) is highly elevated, however, the iris may not respond to miotics. In such cases, IOP should first be lowered by other medications, such as beta blockers, carbonic anhydrase inhibitors, and/or hyperosmotic agents. Once the IOP is controlled, iridotomy is indicated. Laser iridotomy is the treatment of choice; if this cannot be accomplished, surgical iridectomy should be considered.
52. False. Angle-closure glaucoma is often a bilateral disease, and examination and treatment of the fellow eye with iridotomy must be considered.

53. a. Both mydriatics and miotics can precipitate angle-closure in eyes with shallow anterior chambers. This is true for both topical medications and systemic drugs that affect the pupil. Examples include antihistamines, which can have anticholinergic activity.

54. d. Angle-closure develops in only a small number of patients with narrow anterior chambers. A number of provocative tests exist to attempt to cause angle closure in susceptible patients. Perhaps the most predictive is the prone–darkroom test. Intraocular pressure (IOP) is measured before and after 30 to 60 minutes of total darkness attained with the patient prone. Darkness will induce pupillary dilation, and prone positioning will move the lens forward. Both tend to increase pupillary block. None of these tests, however, has been evaluated in a prospective study.

55. a. The patient with very narrow angles and elevated pressure may have “mixed mechanism” glaucoma with partial angle closure caused by pupillary block superimposed on open-angle glaucoma. To determine if an angle-closure component is present, the effect of minimizing pupillary block on intraocular pressure (IOP) must be determined. Cholinergic miotics (pilocarpine) will cause miosis and lessen pupillary block and will also exert traction on the trabecular meshwork (TM). A decrease in pressure after thymoxamine (lessened pupillary block) implies partial angle closure, and iridotomy is indicated. No change in IOP after thymoxamine-induced miosis implies that an iridotomy may not be helpful.

56. a. A small anterior segment is not associated with plateau iris.

57. c. Ectopia lentis increases the risk of pupillary block. It may occur in association with Marfan’s syndrome or as an isolated condition. Neovascular glaucoma typically starts as an open-angle glaucoma with progressive anterior synechialization leading to closure without any obstruction to flow of aqueous through the pupil. The same is true of synechialization following a flat chamber.

58. a. The glaucoma associated with iridocorneal endothelial (ICE) syndrome is often worse than predicted by the extent of synechiae, likely because of clinically undetectable endothelialization of the angle.

59. False. The “nevi” are actually nodular collections of stromal melanocytes, not nevus cells.

60. a. Initial treatment of ciliary block or malignant glaucoma includes cycloplegia. Vitrectomy in phakic eyes is used to alter the anterior vitreous face. In aphakic or pseudophakic eyes, the anterior vitreous can be disrupted by laser treatment. In primary angle closure, the central chamber is deeper than the peripheral angle caused by pupillary block. In malignant glaucoma, vitreous enlargement by aqueous humor causes the entire chamber to become shallow.

61. False. Epithelial downgrowth is usually more aggressive than fibrous downgrowth.

62. False. Peripheral iridotomy does not help relieve the glaucoma associated with scleral buckling because the mechanism of angle closure is not pupillary block. Obstruction of venous outflow produces choroidal effusions that cause anterior rotation of the ciliary body and secondary angle closure. Another mechanism of angle closure following buckling surgery is neovascular glaucoma caused by buckle-induced ocular ischemia. Medical management is not effective during the acute period, the buckle may need to be repositioned or removed.

63. True. Following heavy panretinal photocoagulation, choroidal effusion may cause anterior rotation and swelling of the ciliary body resulting in closure of the angle. In central retinal vein occlusion (CRVO), there may be transudation of serum into the vitreous, driven by the elevated intravascular pressure. This hydration causes vitreous swelling with subsequent secondary angle closure. There may also be ciliary body swelling associated with CRVO.

64. c. The pathophysiology of this disorder has not been elucidated, but gonioscopy suggests maldevelopment of the anterior chamber angle, often with abnormally anterior iris insertion. Infants with congenital glaucoma may also have an impermeable trabecular meshwork (TM).

65. b. Haab striae are tears in Descemet’s membrane resulting from corneal stretching induced by high intraocular pressure (IOP). The same phenomenon leads to enlarged corneal diameter. Corneal edema and optic nerve cupping can be seen in glaucoma in adults.

66. a. Among these general anesthetic agents, ketamine alone is associated with increases in intraocular pressure (IOP) (primarily at high doses). Succinylcholine also can elevate IOP. The other agents usually cause a decrease.

67. False. Infantile glaucoma represents a developmental anomaly of the angle structures, with either an intrinsic defect of the trabecular meshwork (TM) or mechanical obstruction of the TM by a membrane. It is not amenable to long-term medical management, although aqueous suppressants are useful as a temporizing measure before surgery.

68. b. Caution must be exercised in the use of carbonic anhydrase inhibitors for small children because of their susceptibility to weight loss, lethargy, and metabolic acidosis.

69. b. Although their use has been in decline, epinephrine and dipivefrin are beta-2 adrenergic agonists
with potentially serious systemic side effects. Propine is a prodrug that is converted to epinephrine by corneal enzymes. Systemic absorption of epinephrine is dramatically reduced, but the drug is still relatively contraindicated in this circumstance, as is epinephrine itself. Timolol, as a beta antagonist, is not a cardiotonic (quite the opposite). The same is true of pilocarpine.

70. c. Beta antagonists, in general, can cause fatigue, dizziness, and depression at therapeutic doses. Carbonic anhydrase inhibitors share an ability to cause lethargy, malaise, and depression. Either of these groups should be used with caution in patients with a history of significant depression.

71. c. There is a danger of precipitating angle closure in individuals with very narrow angles through the use of any medication with mydriatic action. In the case of plateau iris with narrow angles, dilation causes the peripheral roll of iris to “bunch up” and obstruct the trabecular meshwork (TM). With narrow angles, mydriasis may increase pupillary block, most commonly as the agent is wearing off.

72. c. Epinephrine has a well-established tendency to provoke irritation and allergic responses. More than one fifth of patients will eventually experience an adverse local reaction with prolonged use.

73. c. Direct-acting miotics interact directly with the acetylcholine receptor, whereas indirect-acting agents increase the activity of native acetylcholine at the synaptic junction (by blocking its enzymatic degradation). Pilocarpine is a purely direct agent, whereas carbachol is felt to exhibit both direct and indirect effects.

74. a. See answer 73. Currently, the only commercially available purely indirect parasympathomimetic is demecarium. Indirect parasympathomimetics can also be used against eyelid lice infestations because of their potent insecticidal effects.

75. False. This statement is true only for the unusual patient with plateau iris. The primary mode of action is through ciliary muscle contraction. By way of its insertion on the scleral spur, ciliary muscle tightening puts traction on the trabeculum, increasing conventional outflow.

76. a. Indirect agents, along with the strongest direct agents, tend to have the most pronounced systemic and ocular side effects. Bradycardia is never seen with any of the miotics.

77. c. Changes in urine pH secondary to carbonic anhydrase inhibitors can predispose a patient to calcium oxalate and calcium phosphate nephrolithiasis. Aplastic anemia is a rare but potentially lethal side effect related to the sulfite derivation of the drugs. Gastrointestinal distress occurs, but not most commonly. Hypokalemia may occur as a result of the effects on renal ion transport, but hypocalcemia is not seen. Paresthesias are reported by most patients taking these potent agents.

78. True. Mannitol is distributed only in the blood compartment, whereas urea moves freely in total body water. As a result, mannitol can generate a greater osmotic gradient than urea because its intravascular concentration remains greater. The prevalence of intravenous urea as a hyperosmotic agent to control intraocular pressure (IOP) is decreasing in part because of its risk for tissue necrosis with extravasation.

79. False. The blood–aqueous barrier allows formation of the gradient necessary to draw fluid from the vitreous. Interruption of this gradient decreases the effectiveness and duration of osmotic effects.

80. d. Argon iridotomies are more likely to close spontaneously than neodymium supported by yttrium–aluminium–garnet (Nd:YAG) iridotomies. Argon laser is often useful for pretreating dark irides before Nd:YAG iridotomy because Nd:YAG iridotomy on heavily pigmented irides is difficult. The most common complication of both Nd:YAG and argon laser iridotomies is acute glaucoma. Malignant glaucoma has been reported after a variety of seemingly benign ocular laser procedures.

81. False. Goniotomy requires a clear cornea for viewing of the needle knife. Trabeculotomy is the treatment of choice because an external incision is used and the trabeculotomy is rotated into the anterior chamber.

82. a. Argon laser trabeculoplasty (ALT) uses a 50-micron beam with variable power to produce blanching or a tiny bubble at the anterior pigmented edge of the trabecular meshwork (TM). This requires 180 degrees of treatment, rather than 360 degrees. Outflow facility typically improves following successful ALT.

83. d. Studies suggest that approximately 80% of the patients experience a significant decrease in intraocular pressure (IOP) 1 year after argon laser trabeculoplasty (ALT). ALT generally reduces IOP by 20% to 25%.

84. c. Argon laser trabeculoplasty (ALT) response appears to be better for pigmentary glaucoma and pseudoxfoliation and poorer for inflammatory diseases, recessed angles, membranes in angles, young patients with developmental defects, and aphakic eyes.

85. c. Given the extent of the glaucomatous damage as indicated by split fixation and progression of both fields and disc at low–normal pressures, the maximal decrease in intraocular pressure (IOP) is necessary. Primary filtration presents the best choice. Iridoplasty is usually reserved for eyes with plateau iris syndrome (or, sometimes angle closure). Peripheral iridectomy would not be appropriate because the irises appear normal, and the likelihood of excellent pressure control after combined procedure is not high enough. Cyclocryotherapy and cyclophotocoagulation are reserved for end-stage disease. Setons (Molteno, Baerveldt, Denver–Krupin)
are reserved for special cases because of a higher rate of complication.

86. c. Posttrabeculectomy pain and decreased vision require evaluation for endophthalmitis, malignant glaucoma, and suprachoroidal hemorrhage. However, a clear anterior chamber and the lack of vitreous cells make endophthalmitis less likely.

87. c. To distinguish malignant glaucoma from suprachoroidal hemorrhage that usually occurs in a setting of hypotony, tonometry and ultrasonography would be most helpful.

88. True. Optic vesicle formation influences surface ectoderm to form the lens plate, which then forms the lens.

89. e. Microspherophakia, as its name implies, is characterized by a lens with small diameter and a spherical shape. High ametropia is often found as well. It may occur as an isolated hereditary disorder, but most commonly accompanies Weill-Marchesani syndrome, along with short stubby fingers, broad hands, and decreased joint mobility.

90. False. Spherical shape increases relative pupillary block by increasing the degree of iris-lens contact. Miotics further exacerbate this by shifting the iris-lens diaphragm forward and further increasing iris-lens contact (as the lens becomes even more spherical). Cycloplegia moves the diaphragm backward and flattens the lens. It is the medical treatment of choice.

91. True. Simple ectopia lentis is usually transmitted in an autosomal dominant fashion.

92. True. Superotemporal lens subluxation is typical of Marfan’s syndrome. In homocystinuria, the subluxation is typically inferonasal.

93. d. Mittendorf’s dot is a remnant of the posterior tunica vasculosa lentis and results in a white dot inferonasally on the posterior capsule of the lens. It is meaningless visually.

94. False. Anterior polar cataracts are usually bilateral and usually do not impair vision. Nuclear cataracts are usually bilateral and often cause severe visual impairment.

95. d. Capsular cataracts are opacities of the anterior lens capsule and epithelium that do not usually affect vision.

96. e. Lowe’s syndrome is associated with aminoaciduria. All of the above answers are associated with pediatric cataracts.

97. False. Cataracts may occur both in galactosemia (galactose-1-phosphate uridyltransferase deficiency) and galactokinase deficiency. Galactosemia, however, also can feature hepatosplenomegaly, mental retardation, and other systemic manifestations.

98. a. In the craniosynostoses, small orbits, proptosis, and cranial changes affect facial structure. Patients with Hallermann-Streiff syndrome typically have mandibular hypoplasia with “bird face.” Stickler’s syndrome may be associated with many facial abnormalities, including maxillary and mandibular hypoplasia, epicanthus, a long philtrum, and the Pierre Robin’s anomaly. Alport’s syndrome features hereditary nephritis and cataract with normal facies.

99. b. Neurofibromatosis type 2, not type 1, may be associated with posterior subcapsular cataracts (and acoustic neuromas as well). Galactosemia usually results in early death unless galactose is removed from the diet.

100. False. Surgical removal of the lens does not seem to change the course of pseudoexfoliation glaucoma.

101. False. A younger patient’s lens epithelial cells are more actively growing than an older patient’s and are thus more susceptible to radiation damage.

102. a. This history is most consistent with nuclear sclerosis. In contrast, patients with posterior subcapsular cataract (PSC) will notice difficulty primarily with reading vision, particularly in bright ambient lighting.

103. True. Glare can be disabling to patients with posterior subcapsular cataract (PSC). Thus, standard dark room visual acuity may overestimate their functional acuity. This is the basis of the brightness acuity test (BAT), which quantifies visual acuity under low and high ambient illumination.

104. a. Abnormally thin sclera is not a feature of pseudoexfoliation syndrome. The other three findings are recognized hazards of cataract surgery in patients with pseudoexfoliation.

105. b. High intraocular pressure (IOP) secondary to angle closure may be causing severe eye pain and decreased visual acuity. Hypotony (wound leak, cyclodialysis) could precipitate painful choroidal hemorrhage.

106. c. Because pupillary block is probably present, an iridotomy should be performed. Miotics tend to increase postoperative inflammation and should be avoided here.

107. d. Failure to relieve postoperative angle closure with iridotomy suggests malignant glaucoma, which often responds to potent cycloplegics. If medical management fails, laser treatment to open the anterior hyaloid face, or even pars plana vitrectomy, is necessary.

108. e. Any of the complications can occur with vitreous prolapse into the anterior chamber (AC).

109. b. A recurrent or delayed hyphema after uncomplicated cataract surgery is usually caused by vascularization of the wound or an implant that is rubbing against the iris. Gonioscopy is useful in identifying these vessels. Laser photocoagulation can then be used to ablate the offending vessels.

110. b. Although angiographically detectable cystoid macular edema (CME) is present in 10% to 20% of patients following extracapsular surgery, visual loss only occurs in 1% to 2%. For intracapsular surgery, the percentages are 40% to 60% and 2% to 10%, respectively.

111. True. Angiographic cystoid macular edema (CME) does not necessarily lead to decreased visual acuity.

112. e. All of the above answers are potential risks for retinal detachment following cataract extraction.
113. c. Postoperative endophthalmitis is usually caused by contamination with bacteria from the patient's normal lid flora. *Staphylococcus epidermidis* is the most common agent.

114. False. Anterior chamber intraocular lenses with closed support loops are associated with a higher incidence of complications. They are no longer used because of this reason.

115. a. Posterior capsule opacification, glaucoma (angle closure caused by excessive inflammation), and retinal detachment (caused by vitreous loss) are more common postoperative complications in children undergoing cataract extraction. The incidence of cystoid macular edema (CME) is lower in children than in adults.

116. False. The plasticity of the visual system in children allows them to adapt to the various distortions inherent in aphakic spectacles more readily than adults.

117. d. Electrical injuries to the eye most often involve the cortex of the lens, producing a cataract, typically involving changes in the anterior subcapsular cortex.

118. b. The initial treatment of chemical burns is copious irrigation of the affected eye. This should take place at the site of injury if possible; tap water or any nontoxic liquid may be used.

119. e. Corneal blood staining is a complication of hyphema that may take years to clear. The blood staining clears in a centripetal pattern (starting at the periphery). African Americans with hyphema should be checked for sickle cell disease or trait because the sickled cells may become trapped in the trabecular meshwork (TM), increasing the intraocular pressure (IOP). Furthermore, the optic nerve is more susceptible to atrophy in patients with sickle cell disease, even at relatively mild elevations of IOP.

120. a. Polyopia, pupillary block, and corneal-lenticular touch are indications for medical or surgical intervention in lens subluxation (which can be accomplished with the implantation of a Morcher endcapsular ten- sion ring). Decreased vision often can be corrected solely with an aphakic refraction.

121. False. Although intraocular foreign bodies containing >90% copper generally cause an acute inflammatory response (including intraocular necrosis), concentrations of 70% to 90% cause chalcosis leading to a copper ring (identical to the Kayser-Fleischer ring in Wilson's disease) and sunflower cataract. Concentrations <70% can be tolerated; however, other factors such as location and fibrous encapsulation modulate the tissue reaction.

122. True. Copper accumulates in Descemet's membrane, the lens capsule, and other basement membranes, whereas iron accumulates in basal epithelial cells.

123. a. The deposit of metal in Descemet's membrane occurs with copper foreign bodies, not siderosis bulbi.

124. c. The constellation of heterochromia iridis (compare the two figures carefully), gelatinous–stellate keratic precipitates, mild anterior chamber reaction, distinctive rubecue, and ipsilateral cataract and glaucoma is nearly pathognomonic for Fuchs' heterochromatic iridocyclitis. If inflammation is severe, another diagnosis should be considered. The iris atrophy in Fuchs' is generally diffuse and stromal, whereas that of herpes zoster iritis is typically sectoral with pigment epithelial involvement. The rubecue is distinctive because the vessels are typically quite fine and rarely induce synechiae or angle closure. The vessels are also quite fragile, and spontaneous or iatrogenic hyphema (as the paracentesis is performed at filtration surgery) is a classic sign. The mechanism for the glaucoma is poorly understood. Cataract surgery is usually indicated for visual rehabilitation; it has little or no effect on the glaucoma. In fact, in many patients, intraocular pressure (IOP) does not increase for many months or years after cataract extraction.

125. d. Assuming the left angle is normal, this patient can be diagnosed with angle recession (posttraumatic) glaucoma affecting the right eye. Because men are victims of ocular trauma far more frequently than women, this glaucoma is more common in men. The lifetime risk for developing glaucoma seems to be correlated with the amount of angle recession and is estimated to be approximately 10% in patients with 180 degrees of involved angle. Iris sphincter tears, Vossius's ring, and posterior subcapsular cataract all may be seen in conjunction with the disorder. Retinal dialysis must be ruled out in the posttraumatic period by a dilated retinal examination with 360 degree scleral depression. Although the disorder may be bilateral if the contralateral eye is traumatized, it is much more commonly unilateral.

126. c. Typically, the surgeon should fastidiously avoid contact between the Mitomycin-C (MMC)–containing sponge and the wound edges to reduce the risk of postoperative wound leaks and hypotony. This is one of the main reasons why most MMC filters are performed with a limbus-based flap. MMC is associated with a lower incidence of hypotony and corneal–surface disorders than 5-fluorouracil (5-FU).

127. c. This is probably caused by pooling of infectious agents in the tear film inferiorly.

128. False. The Tonopen appears to have more accurate pressure readings in patients after laser-assisted in situ keratomileusis (LASIK) surgery. Goldmann readings can be artificially low after LASIK.

129. True. This is a normal phenomenon that can occur with compression of the episcleral veins during gonioscopy.

130. d. Compared to the other medications listed, dexamethasone 0.1% is associated with the highest rise in intraocular pressure (IOP) and is the most potent steroid of those listed.
8: Glaucoma, Lens, and Anterior Segment

131. d. Topiramate (Topamax) is associated with acute secondary angle-closure glaucoma. Most cases are bilateral, and can lead to blindness if not treated or recognized. All of the findings are reversible if recognized early and the drug is discontinued. Peripheral iridectomy is ineffective for this type of angle-closure glaucoma.

132. c. Lamellar cataracts are the most common type of congenital cataracts and may occur as a result of toxic influence to the fetus or may be inherited as an autosomal dominant trait.

133. b. Selective laser trabeculoplasty (SLT) is a relatively new procedure with effects for lowering intraocular pressure (IOP) comparable to argon laser trabeculoplasty (ALT). It has less disruptive effects on intraocular tissue than ALT.

134. False. Generally, uncomplicated cataract surgery (Phaco/PCIOL) lowers intraocular pressure (IOP) by 1 to 2 mm Hg.

135. c. Ciliary body ablation should not be considered because of the risk of sympathetic ophthalmia.

136. c. Iris atrophy is not a common side effect of the prostaglandin analogs (e.g., latanoprost/travoprost). All of the other answers listed are potential side effects of topical prostaglandin analogs.

137. False. Juvenile xanthogranuloma (JXG) can cause spontaneous hyphemas. However, in any infant that presents with “spontaneous hyphema,” an ophthalmologist must consider child abuse. Other causes of spontaneous hyphema include bleeding diatheses, leukemia, and retinoblastoma.

138. c. All of the other answers can cause increased episcleral venous pressure.

139. a. 1, b. 3, c. 1, d. 1, e. 1, f. 2, g. 1, h. 2, i. 3, j. 2, k. 3. Mannitol is an osmotic diuretic that decreases vitreous volume by creating an osmotic gradient between the gel and plasma.

140. a. 1, b. 2, c. 2, d. 1, e. 2, f. 4, g. 1, h. 1, i. 2. The retinopathy of Alport’s syndrome is generally mild and nonprogressive and resembles fundus albipunctatus clinically.

141. a. 3, b. 4, c. 1, d. 2.

Suggested Readings


Cornea, External Disease, and Refractive Surgery

Questions

1. The cardinal signs of inflammation include all of the following except:
   a. redness.
   b. irreversible architectural disruption.
   c. tenderness or pain.
   d. loss of function.
   e. warmth.

2. T or F The key difference between a toxic agent and an immune agent is that cellular damage by the former is independent of previous exposure.

3. T or F Macrophages and polymorphonuclear leukocytes (PMNs) incite similar degrees of local tissue damage in acute inflammatory reactions.

4. Which of the following markers of ocular inflammation are relatively nonspecific in the determination of etiology?
   1. conjunctival papillae.
   2. conjunctival follicles.
   3. chemosis.
   4. giant conjunctival papillae.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

5. Conjunctival papillae can be seen in inflammation of the:
   1. conjunctival fornices.
   2. tarsal conjunctiva.
   3. bulbar conjunctiva.
   4. limbal conjunctiva.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

6. A conjunctival inflammatory response characterized by multiple polygonal nodules with central fibrovascular cores is consistent with a:
   a. follicular response.
   b. papillary response.
   c. phlyctenular response.
   d. giant papillary response.
   e. ligneous response.

7. Connective tissue septa in the subepithelial space are responsible for the development of which inflammatory morphology?
   a. papillae.
   b. follicles.
   c. phlyctenules.
   d. Herbert's pits.
   e. giant papillae.
8. The least reliable location of a conjunctival papillary response for etiologic interpretation is the:
   a. inferior fornix.
   b. superior fornix.
   c. superior edge of superior tarsus.
   d. inferior edge of superior tarsus.
   e. limbus.

9. T or F The definition of a giant papilla is a papilla > 5 mm in diameter.

10. The differential diagnosis of a true giant papillary conjunctivitis (GPC) includes all of the following except:
    a. contact lens-related conjunctivitis.
    b. trachoma.
    c. atopic keratoconjunctivitis.
    d. ocular prosthesis-related conjunctivitis.
    e. vernal keratoconjunctivitis.

11. Clumps of calcific degeneration and eosinophils at the limbus are termed:
    a. Herbert’s pits.
    b. The von Arlt line.
    c. Fuchs’ spots.
    d. Horner-Trantas dots.
    e. Cogan’s patches.

12. Rounded, depressed regions of necrotic limbal follicles are termed:
    a. Herbert’s pits.
    b. The von Arlt line.
    c. Fuchs’ spots.
    d. Horner-Trantas dots.
    e. Cogan’s patches.

13. T or F The various forms of giant papillary conjunctivitis (GPC) are indistinguishable clinically.

14. T or F With adequate treatment, the clinical symptoms of contact lens-related giant papillary conjunctivitis (GPC) generally remit before the papillae disappear.

15. Mild contact lens-related giant papillary conjunctivitis (GPC) may be differentiated from an infectious follicular conjunctivitis by:
    1. the presence of itching in the former.
    2. the presence of mucous and serous discharge in the latter.

16. Rounded, gelatinous–appearing lesions of the conjunctiva with vessels at the periphery, but never within the nodular substance, are called:
    a. follicles.
    b. papillae.
    c. phlyctenules.
    d. giant papillae.
    e. pseudomembranes.

17. Follicular conjunctivitides are typically more severe inferiorly than superiorly except for:
    a. adult inclusion conjunctivitis.
    b. epidemic keratoconjunctivitis (EKC).
    c. Parinaud’s syndrome.
    d. trachoma.
    e. medicamentosa.

18. By definition, a conjunctivitis may be termed “chronic” if it lasts longer than:
    a. 2 weeks.
    b. 4 weeks.
    c. 6 weeks.
    d. 8 weeks.
    e. 12 weeks.

19. The differential diagnosis for acute follicular conjunctivitis includes all of the following except:
    a. epidemic keratoconjunctivitis (EKC).
    b. herpes simplex keratoconjunctivitis.
    c. medicamentosa.
    d. trachoma.
    e. adult inclusion conjunctivitis.

20. The differential diagnosis for chronic follicular conjunctivitis includes all of the following except:
    a. epidemic keratoconjunctivitis (EKC).
    b. medicamentosa.
    c. Parinaud’s oculoglandular syndrome.
    d. benign folliculosis.
    e. trachoma.
Questions

21. Infectious etiologies of pseudomembranous or membranous conjunctivitis include all of the following except:
   a. *Gonococcus.*
   b. herpes simplex.
   c. adult inclusion conjunctivitis.
   d. diphtheria.
   e. *Candida.*

22. Immunologic etiologies of a pseudomembranous or membranous conjunctivitis include all of the following except:
   a. ocular cicatrical pemphigoid (OCP).
   b. vernal keratoconjunctivitis.
   c. ligneous keratoconjunctivitis.
   d. Stevens-Johnson syndrome.
   e. atopic keratoconjunctivitis.

23. T or F Microcystic corneal epithelial edema consists of fluid in the intracellular space, because of cellular hypoxia.

24. T or F Pathophysiologic factors critical for the development of a filamentary keratopathy include corneal stromal thickening and increased mucus production.

25. Entities to be considered in the differential diagnosis of a filamentary keratitis include all of the following except:
   a. ptosis.
   b. neurotrophic keratopathy.
   c. recurrent erosion syndrome.
   d. ocular cicatrical pemphigoid (OCP).
   e. medicamentosa.

26. Superficial opacification of the cornea in a horizontal fashion between the eyelid margins is best referred to as:
   a. superficial punctate keratitis.
   b. micropannus.
   c. band keratopathy.
   d. gross corneal pannus.
   e. interstitial keratitis (IK).

27. Cellular infiltrates that characterize corneal stromal inflammation can be derived from:
   a. limbal vasculature.
   b. aqueous humor.
   c. tears.

28. The predominant cell forms seen in nongranulomatous keratic precipitates (KPs) are:
   1. lymphocytes.
   2. epithelioid cells.
   3. polymorphonuclear leukocytes (PMNs).
   4. macrophages.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

29. The predominant cell forms seen in “mutton-fat” keratic precipitates (KPs) are:
   1. lymphocytes.
   2. epithelioid cells.
   3. polymorphonuclear leukocytes (PMNs).
   4. macrophages.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

30. Which of the following is not an advantage of laser-assisted in situ keratomileusis (LASIK) when compared to photorefractive keratectomy (PRK)?
   a. reduced incidence of stromal haze.
   b. more rapid epithelial healing.
   c. reduced incidence of postoperative endophthalmitis.
   d. all of the above.
   e. none of the above.

31. Which of the following is not an advantage of photorefractive keratectomy (PRK) when compared to laser-assisted in situ keratomileusis (LASIK)?
   a. reduced incidence of endophthalmitis.
   b. reduced incidence of corneal perforation.
   c. reduced postoperative pain.
   d. all of the above.
   e. none of the above.
32. A corneal ulcer, recalcitrant to routine treatment, is rescraped for special staining and cultures. The Gram stain is reported as growing moderate diphtheroids. Which special stain is most likely to be of value in determining the actual diagnosis?
   b. Warthin-Starry stain.
   c. methenamine silver stain.
   d. Giemsa stain.
   e. periodic acid-Schiff (PAS) stain.

33. Which culture media should be used in addition to routine broth to isolate the organism suspected in the previous question?
   1. blood agar.
   2. Sabouraud’s medium.
   3. Löwenstein-Jensen medium.
   4. blood agar with Staphylococcus aureus colonies.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

34. Antibiotics useful in the topical treatment of mycobacterial keratitis include:
   1. kanamycin.
   2. rifampin.
   3. amikacin.
   4. vancomycin.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

35. T or F To localize treponemes from infected tissues or secretions, the preparations must be fresh.

36. T or F The Treponema pallidum immobilization (TP1) test is as sensitive and specific as the fluorescent treponemal antibody-absorption (FTA-ABS) test.

37. T or F The fluorescent treponemal antibody-absorption (FTA-ABS) test is as sensitive and specific as the microhemagglutination of Treponema pallidum (MHA-TP) test.

38. A case of secondary acquired syphilis is adequately treated with parenteral penicillin. Which of the following blood tests would be expected to normalize?
   1. VDRL (Venereal Disease Research Laboratory).
   2. microhemagglutination of Treponema pallidum (MHA-TP) test.
   3. rapid plasma reagin (RPR).
   4. fluorescent treponemal antibody-absorption (FTA-ABS).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

39. T or F As culture methods have improved, these have become more reliable than serology in the diagnosis of Lyme disease.

40. The most common agent involved in mycotic ocular infections in the northern half of the United States is:
   a. Alternaria.
   b. Aspergillus.
   c. Fusarium.
   d. Penicillium.
   e. Candida.

41. The most common agent involved in mycotic ocular infections in the southern half of the United States is:
   a. Alternaria.
   b. Aspergillus.
   c. Fusarium.
   d. Penicillium.
   e. Candida.

42. The most basic difference between Candida and Fusarium is:
   a. Fusarium is a mold, and Candida is dimorphic.
   b. Fusarium is dimorphic, and Candida is a mold.
   c. Fusarium is a mold, and Candida is a yeast.
   d. Fusarium is a yeast, and Candida is a mold.
   e. Fusarium as a yeast has pseudohyphae, whereas Candida has true hyphae.

43. The most basic difference between Fusarium and Mucor is:
   a. Fusarium is a mold, and Mucor is dimorphic.
   b. Fusarium is dimorphic, and Mucor is a mold.
Questions

c. *Fusarium* is a mold, and *Mucor* is a yeast.
d. *Fusarium* is a yeast, and *Mucor* is a mold.
e. *Fusarium* has septate hyphae, whereas *Mucor* has nonseptate hyphae.

44. The most common ocular manifestation of cryptococcal infection is:
   a. membranous conjunctivitis.
   b. orbital cellulitis.
   c. lid abscesses.
   d. endogenous endophthalmitis.
   e. ulcerative keratitis.

45. T or F Dimorphic fungi are frequently encountered in mycotic keratitis.

46. Factors that increase the difficulty of laboratory identification of fungal pathogens include all of the following except:
   a. inclusion of cycloheximide in various media.
   b. fastidiousness of the fungal agents.
   c. discarding of plates before full identification of fungal species.
   d. discarding of plates before full exploration of fungal sensitivities.
   e. confusion of pathogenic fungal species as contaminants.

47. Which of the following statements about the epidemiology of herpes simplex virus (HSV) infections in humans is/are true:
   1. Type 1 typically causes recurrent perioral cold sores.
   2. Ocular herpes infections are split evenly between types 1 and 2.
   3. Type 2 typically causes genital disease.
   4. Acute and convalescent antibody titers are helpful in the diagnosis of acute and recurrent disease.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

48. T or F Intranuclear inclusion bodies of herpes simplex virus (HSV) epithelial infection are best highlighted with a Papanicolaou (Pap) smear.

49. T or F The multinucleated giant cells seen in epithelial herpes simplex virus (HSV) infection are best seen with Giemsa stain.

50. T or F The herpes zoster virus (HZV) produces the same inclusion bodies and giant cells as herpes simplex virus (HSV).

51. Which of the following Epstein–Barr virus (EBV) antibodies peak in serum level within the first 6 to 12 weeks of infection?
   1. viral capsid antigen immunoglobulin M (VCA-IgM).
   2. VCA-IgG.
   3. early antigen-diffuse (EA-D).
   4. Epstein–Barr nuclear antigen (EBNA).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

52. Which of the following Epstein–Barr virus (EBV) antibodies are detectable in serum for life following infection?
   1. viral capsid antigen immunoglobulin M (VCA-IgM).
   2. VCA-IgG.
   3. early antigen-diffuse (EA-D).
   4. Epstein–Barr nuclear antigen (EBNA).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

53. Which of the following is not true regarding laser-assisted subepithelial keratomileusis (LASEK)?
   a. There is no stromal flap created.
   b. The procedure may offer increased comfort for the patient as compared to laser-assisted in situ keratomileusis (LASIK).
   c. Ethanol 20% can be used to trephine the epithelium.
   d. The procedure has less haze associated with it than LASIK.
   e. None of the above.
54. Biologic features of chlamydiae that render them closer to bacterial than viral life forms include all of the following except:
   a. nucleic acid content.
   b. mechanism of replication.
   c. cell wall properties.
   d. full complement of organelles.
   e. response to certain antimicrobial agents.

55. T or F Accurate diagnosis of ocular chlamydial infection relies on recovery and culture of live organism.

56. Acceptable alternatives for adequate treatment of ocular chlamydial infection include:
   1. erythromycin orally for 1 week.
   2. rifampin orally for 3 weeks.
   3. erythromycin ointment for 3 weeks.
   4. tetracycline orally for 3 weeks.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

57. Chlamydial serotypes associated with trachoma in humans include:
   1. A.
   2. E.
   3. C.
   4. L.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

58. Which one of the following statements about Acanthamoeba and its ocular manifestations is false?
   a. The organism, particularly in its cyst form, is very hearty.
   b. Culture technique generally involves non-nutrient agar overlaid with Escherichia coli.
   c. Diagnostic yield from conjunctival fornix culture is high (75%).
   d. Calcofluor white is the stain of choice and requires fluorescence microscopy.
   e. The organism may induce an optic neuritis.

59. At what point do most flap folds occur after laser-assisted in situ keratomileusis (LASIK)?
   a. within 1 week.
   b. after 2 weeks.
   c. after 1 month.
   d. after 6 months.
   e. after 1 year.

60. Which one of the following statements about louse infections of the eye is false?
   a. Pediculus capitis and Phthirus pubis are the only organisms that infect the periocular structures.
   b. Ocular irritation is due to injection of toxic louse saliva into lid tissue.
   c. Sexual contact is felt to be the significant mode of transmission.
   d. The same organisms that infect eyelashes infect pubic hairs.
   e. Eradication of organisms depends on suffocation, either by bland ointments or paralytic medications such as eserine.

61. Which of the following statements about arachnid infections of the eye is/are true?
   1. The most common genus involved is Demodex.
   2. These organisms are normal commensals.
   3. The classic slit-lamp sign is sleeving of eyelash bases.
   4. Antiparasitic therapy rests on suffocation of the organisms with bland ointments or paralytic medications (eserine).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

62. Members of the normal flora of human conjunctiva include all of the following except:
   a. Staphylococcus epidermidis.
   b. diphtheroid organisms.
   c. Streptococcus viridans.
   d. Bacillus cereus.
   e. Staphylococcus aureus.

63. T or F In an adult inclusion conjunctivitis, the likelihood of finding cytoplasmic or intranuclear inclusions is high.
64. T or F In neonatal inclusion conjunctivitis, the likelihood of finding cytoplasmic or intranuclear inclusions is high.

65. Which of the following procedures has not been approved by the U.S. Food and Drug Administration (FDA) for surgical correction of hyperopia?
   a. laser-assisted in situ keratomileusis (LASIK).
   b. thermokeratoplasty using noncontact holmium-YAG laser.
   c. thermokeratoplasty using contact holmium-YAG laser.
   d. conductive keratoplasty.
   e. photorefractive keratectomy (PRK).

66. The method of choice for documenting intracytoplasmic inclusion bodies is:
   a. thioglycolate broth.
   b. Giemsa stain.
   c. blood agar with Staphylococcus aureus cultures.
   d. Sabouraud’s agar.
   e. Ziehl-Neelsen stain.

67. Which of the following agents has in vivo activity against herpes simplex virus (HSV)?
   1. idoxuridine.
   2. vidarabine.
   3. trifluridine.
   4. acyclovir.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

68. Which of the following antivirals has in vivo activity against herpes zoster virus (HZV)?
   1. idoxuridine.
   2. vidarabine.
   3. trifluridine.
   4. acyclovir.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

69. Which of the following antivirals has an ophthalmic ointment preparation?
   1. idoxuridine.
   2. trifluridine.
   3. vidarabine.
   4. acyclovir.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

70. Which of the following antivirals blocks viral replication by interference with DNA synthesis?
   1. idoxuridine.
   2. trifluridine.
   3. vidarabine.
   4. acyclovir.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

71. T or F Hypersensitivity to one antiviral does not imply hypersensitivity to all antivirals.

72. T or F Resistance to one antiviral does not imply resistance to all antivirals.

73. Which of the following antivirals has a mechanism of action specific for herpes viruses, with scarcely any effect on host cells?
   1. idoxuridine.
   2. trifluridine.
   3. vidarabine.
   4. acyclovir.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

74. T or F The drug of choice in the treatment of an indolent, steroid-resistant, infectious herpetic ulcer is vidarabine.
75. Which of the following is false regarding radial corneal incisions?
   a. Radial incisions cause a local flattening of the cornea.
   b. Radial incisions cause flattening 90 degrees away from the meridian of the incision.
   c. Radial incisions have a greater effect as they become closer to the visual axis.
   d. Radial incisions have a greater effect the larger they are (up to 11 mm).
   e. All of the above.

76. All of the following have been described as signs of antiviral toxicity except:
   a. punctate epithelial keratopathy.
   b. follicular conjunctivitis.
   c. anterior uveitis.
   d. indolent corneal ulceration.
   e. preauricular lymphadenopathy.

77. The drug of choice for presumed filamentous keratomycosis is:
   a. topical amphotericin.
   b. oral ketoconazole.
   c. topical clotrimazole.
   d. topical natamycin.
   e. topical flucytosine.

78. Which one of the following statements about the polyene class of antifungal agents is false?
   a. They are classified by the number of double bonds present.
   b. Amphotericin has the greatest topical activity against filamentous fungi.
   c. The corneal epithelium is a significant barrier to penetration of natamycin.
   d. Acceptable intraocular concentrations of polyene antifungals can only be achieved with direct intravitreal injection.
   e. Topical amphotericin must be stored in dark glass or opaque containers.

79. Which one of the following antifungal agents is most likely to encounter fungal resistance?
   a. natamycin.
   b. flucytosine.
   c. amphotericin.
   d. miconazole.
   e. thimerosal.

80. The drug of choice for Aspergillus keratitis is:
   a. clotrimazole.
   b. flucytosine.
   c. natamycin.
   d. amphotericin.
   e. tolnaftate.

81. Which of the following organ systems is most likely to be the target of toxicity from the polyene class of antifungal agents?
   a. central nervous system (CNS).
   b. cardiovascular.
   c. renal.
   d. hematopoietic.
   e. hepatic.

82. Which of the following organ systems is most likely to be the target of the imidazole class of antifungal agents?
   a. central nervous system (CNS).
   b. cardiovascular.
   c. renal.
   d. hematopoietic.
   e. hepatic.

83. Which of the following antifungal agents is/are in the same class?
   1. flucytosine.
   2. natamycin.
   3. miconazole.
   4. amphotericin.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

84. Which of the following antifungal agents is/are in the same class?
   1. ketoconazole.
   2. miconazole.
   3. clotrimazole.
   4. nystatin.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
Questions

85. Agents recognized to be useful in the medical therapy for *Acanthamoeba* keratitis include:
   1. propamidine.
   2. clotrimazole.
   3. neomycin.
   4. pentamidine.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

86. The cellular element generally responsible for inflammatory corneal damage is the:
   a. macrophage.
   b. lymphocyte.
   c. polymorphonuclear leukocyte (PMN).
   d. T lymphocyte.
   e. eosinophil.

87. Which of the following statements regarding intracorneal rings (ICRs) is false?
   a. One advantage of ICR implantation is the reversible nature of the procedure.
   b. The cornea's positive asphericity is maintained with the procedure.
   c. Unfortunately, the most common complication, corneal perforation, can lead to severe visual loss.
   d. More than 90% of patients have <1D of change in refraction at 1-year postoperative follow-up.
   e. Decreased corneal sensation can be a postoperative complication.

88. T or F Phosphate preparations of topical steroids are generally more soluble in water than acetate preparations and may be dispensed in solution rather than suspension.

89. The topical steroid preparation with the greatest antiinflammatory activity within the cornea is:
   a. prednisolone phosphate 1.0%.
   b. dexamethasone phosphate 1.0% ointment.
   c. prednisolone acetate 1.0%.
   d. dexamethasone alcohol 0.1% suspension.
   e. fluorometholone alcohol 0.1% suspension.

90. The ocular corticosteroid that is least likely to induce intraocular pressure (IOP) elevation is:
   a. prednisolone phosphate.
   b. prednisolone acetate.
   c. dexamethasone alcohol.
   d. fluorometholone.
   e. medrysone.

91. T or F Topical corticosteroid treatment can lead to reactivation of herpes simplex virus (HSV) keratitis in a quiet eye with a history of previous herpetic disease.

92. Topical corticosteroids should probably be withheld in any eye with:
   1. active dendritic epithelial keratitis.
   2. disciform keratitis.
   3. presumed fungal keratitis.
   4. pseudodendritic epithelial keratitis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

93. Etiologies recognized in the pathogenesis of conjunctival papilloma include:
   1. immunologic.
   2. neoplastic.
   3. toxic.
   4. infectious.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 3.
   d. 4 only.
   e. 1, 2, 3, and 4.

94. Which of the following statements regarding intraocular lens (IOL) power calculations after laser-assisted *in situ* keratomileusis (LASIK) surgery is true?
   a. IOL power can be readily calculated using manual keratometry and the Sanders-Retzlaff-Kraff (SRK) formula with a different A-constant.
   b. IOL power can be readily calculated using a topographic keratometry device.
   c. IOL power can be easily calculated using only the preoperative keratometry readings.
d. Patients postoperative to LASIK surgery who undergo IOL implantation using standard IOL calculations will likely be hyperopic postoperatively.
e. Patients postoperative to LASIK surgery who undergo IOL implantation using standard IOL calculations will likely be myopic postoperatively.

95. A patient presents 6 hours after receiving a prescription for topical antibiotic for a suspected bacterial blepharoconjunctivitis. The patient complains of itching and tearing, and an examination documents severe chemosis and mild hyperemia. The most likely diagnosis is:
   a. antibiotic resistance.
   b. toxic follicular conjunctivitis.
   c. toxic papillary conjunctivitis.
   d. anaphylactoid reaction.
   e. contact allergic reaction.

96. A patient presents approximately 6 days after daily use of antibiotic ointment following cataract surgery. The patient describes a gradual onset of scaling and itching of the skin and increasing redness of the eye. The hypersensitivity pattern most likely at play is:
   a. type I.
   b. type II.
   c. type III.
   d. type IV.
   e. type V.

97. Which type of hypersensitivity conjunctivitis takes the longest to develop?
   a. toxic follicular conjunctivitis.
   b. toxic papillary conjunctivitis.
   c. contact dermatitis with conjunctivitis.
   d. anaphylactoid reaction.
   e. no clear time pattern can be assigned to any of the reactions.

98. T or F Most patients who develop contact lens–induced papillary conjunctivitis develop giant papillae.

99. Presenting symptoms compatible with the diagnosis of contact lens–induced papillary conjunctivitis include:
   1. itching.
   2. mucoid discharge.

100. T or F The incidence of giant papillary conjunctivitis (GPC) is greatest with soft contact lenses (SCL) and least with polymethylmethacrylate (PMMA) lenses.

101. Manipulations in the contact lens regimen that are helpful in the control of contact lens-induced papillary conjunctivitis include:
   1. change of contact lens brand or style.
   2. change to heat sterilization methods.
   3. use of topical mast cell inhibitors.
   4. decreasing the frequency of enzyme treatments.

102. The classic clinical sign of staphylococcal blepharoconjunctivitis is:
   a. inferior follicular conjunctivitis.
   b. scurf.
   c. limbal papillae.
   d. collarettes.
   e. sleeving of lashes.

103. Symptoms consistent with staphylococcal blepharoconjunctivitis include which of the following:
   1. red eyes.
   2. epiphora.
   3. photophobia.
   4. no symptoms.
104. T or F Culture techniques offer nothing diagnostically or therapeutically in staphylococcal blepharoconjunctivitis.

105. Which of the following, when combined with lid hygiene, is not an appropriate treatment regimen for staphylococcal blepharoconjunctivitis?

- a. bacitracin ointment plus prednisolone 0.125% twice a day.
- b. warm compresses twice a day.
- c. warm compresses followed by bacitracin twice a day.
- d. prednisolone 0.125% twice a day.
- e. warm compresses followed by bacitracin ointment and prednisolone 0.125% twice a day.

106. Which of the following may be direct manifestations of ocular infection?

1. hordeolum.
2. chalazion.
3. meibomitis.
4. phlyctenulosis.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

107. The two organisms most frequently involved in phlyctenulosis are:

- a. Coccidioides immitis and Mycobacterium tuberculosis.
- b. Coccidioides and Staphylococcus.
- c. Chlamydia trachomatis and Staphylococcus.
- d. Mycobacterium tuberculosis and Staphylococcus.
- e. Coccidioides and Candida.

108. Phlyctenules are a manifestation of what type hypersensitivity?

- a. type I.
- b. type II.
- c. type III.
- d. type IV.
- e. type V.

109. Bacterial conjunctivitides in a healthy host are self-limited with the exception(s) of:

1. Neisseria gonorrhoeae.
2. Haemophilus species.
3. Neisseria meningitidis.
4. Staphylococcus.

110. The most common cause of hyperacute purulent conjunctivitis is:

- a. Haemophilus aegyptius.
- b. Neisseria meningitidis.
- c. Streptococcus pneumoniae.
- d. Neisseria gonorrhoeae.
- e. Haemophilus influenzae.

111. The only bacterial conjunctivitis that routinely leads to preauricular lymphadenopathy is:

- a. Haemophilus aegyptius.
- b. Neisseria meningitidis.
- c. Streptococcus pneumoniae.
- d. Neisseria gonorrhoeae.
- e. Haemophilus influenzae.

112. Which of the following would be an effective treatment of culture-proven gonococcal conjunctivitis?

- a. topical penicillin G four times daily for 14 days.
- b. topical penicillin G four times daily with doxycycline 100 mg orally twice daily for 1 week.
- c. topical penicillin G four times daily and topical tetracycline four times daily for 1 week.
- d. ceftriaxone 1 g intramuscularly daily for 5 days.
- e. ceftriaxone 1 g intramuscularly daily for 5 days and doxycycline orally twice daily for 3 weeks.

113. A pearly eyelid nodule with a central crater and associated inferior follicular conjunctivitis is the classic picture for:

- a. phlyctenulosis.
- b. staphylococcal blepharoconjunctivitis.
- c. phthiriasis.
- d. primary herpes simplex dermatitis.
- e. molluscum contagiosum.

114. Classic epidemic keratoconjunctivitis (EKC) is typically caused by:

- a. enterovirus type 70.
- b. adenovirus types 3 and 7.
c. Newcastle virus.
d. adenovirus types 8 and 19.
e. coxsackievirus A24.

115. **T or F** Late cicatrization excludes the diagnosis of epiolemic keratoconjunctivitis (EKC).

116. The most important element in the management of a patient with epidemic keratoconjunctivitis (EKC) is:
   a. topical bacitracin ointment.
   b. topical trifluridine drops.
   c. warm compresses.
   d. topical prednisolone.
   e. fastidious hygiene.

117. **T or F** The key feature distinguishing acute hemorrhagic conjunctivitis from epidemic keratoconjunctivitis (EKC) is the presence of subconjunctival hemorrhage in the former.

118. Which one of the following statements about trachoma is false?
   a. Serotypes A through C are responsible.
   b. Blinding complications of the infection are due to the pronounced hypersensitivity responses to *Chlamydia*.
   c. In the MacCallan classification, stages I and II feature follicles with no scarring, whereas stages III and IV have pronounced subepithelial fibrosis.
   d. The World Health Organization (WHO) has a simplified grading system based on the presence or absence of follicles, intense inflammation, scarring, trichiasis, or corneal opacification.
   e. Treatment with antibiotics should be prolonged, and may require retreatment with multiple agents because of resistance.

119. Which one of the following statements about inclusion conjunctivitis is false?
   a. Except for transmission in swimming pools, the adult form is generally sexually transmitted.
   b. Unlike trachoma, adult inclusion conjunctivitis affects the inferior conjunctiva more than the superior conjunctiva.
   c. Unlike epidemic keratoconjunctivitis (EKC), inclusion conjunctivitis is never membranous.
   d. Unlike EKC, associated keratitis may be localized to the superior one third of the cornea.
   e. Unlike EKC, corneal neovascularization may be seen.

120. Successful long-term management of adult inclusion conjunctivitis includes:
   a. recurrent topical tetracycline.
   b. recurrent oral tetracycline therapy.
   c. chronic daily use of a mild topical steroid preparation.
   d. conjunctival transplantation.
   e. examination with treatment, if necessary, of personal contacts.

121. Important differences between neonatal inclusion conjunctivitis and adult inclusion conjunctivitis include all of the following except:
   a. more prominent follicular response in neonates.
   b. more discharge in neonates.
   c. pseudomembranes or membranes in neonates.
   d. more prominent cytoplasmic inclusion bodies in neonates.
   e. better response to topical therapy in neonates.

122. **T or F** Because of its response to topical therapy, neonatal inclusion conjunctivitis can be treated by this route alone.

123. The spectrum of atopic ocular disease includes all of the following except:
   a. seasonal allergic conjunctivitis.
   b. phlyctenulosis.
   c. atopic keratoconjunctivitis.
   d. giant papillary conjunctivitis (GPC).
   e. vernal keratoconjunctivitis.

124. Which of the following disorders is most likely to respond quickly to topical antihistamine therapy?
   a. seasonal allergic conjunctivitis.
   b. phlyctenulosis.
   c. atopic keratoconjunctivitis.
   d. giant papillary conjunctivitis (GPC).
   e. vernal keratoconjunctivitis.

125. For which disorder is chronic use of systemic antihistamine most important?
   a. seasonal allergic conjunctivitis.
   b. phlyctenulosis.
Questions

c. atopic keratoconjunctivitis.
d. giant papillary conjunctivitis (GPC).
e. vernal keratoconjunctivitis.

126. Clinical features distinguishing seasonal allergic conjunctivitis from vernal keratoconjunctivitis include:

1. prominent itching.
2. response to topical antihistamine.
3. seasonal occurrence.
4. the presence of giant papillae.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

127. Which of the following statements about limbal vernal keratoconjunctivitis is/are true?

1. In African Americans, it is more common than palpebral vernal conjunctivitis.
2. This variety is more commonly associated with shield corneal ulcer than the palpebral form.
3. Collections of cellular debris and eosinophils may accumulate at the surface of limbal papillae.
4. Long-term management is centered around daily corticosteroid use.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

128. Features distinguishing atopic keratoconjunctivitis from vernal keratoconjunctivitis include:

1. age range of typically affected patients.
2. seasonal variations of incidence.
3. presence of extensive conjunctival and corneal scarring.
4. presence of eosinophils in conjunctival scrapings.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.

d. 4 only.
e. 1, 2, 3, and 4.

129. To secure the diagnosis of atopic keratoconjunctivitis, it is critical to inquire about a previous or active history of:

a. asthma.
b. sinusitis.
c. vesicular rash consistent with herpes simplex virus (HSV).
d. eczema.
e. aspirin hypersensitivity.

130. Clinical findings consistent with a diagnosis of Reiter’s syndrome include all of the following except:

a. keratoderma blennorrhagicum.
b. severe retinal vasculitis.
c. nonerosive oligoarthritis.
d. papillary conjunctivitis with corneal neovascularization.
e. acute nongranulomatous iridocyclitis.

131. An obese 35-year-old man presents to an ophthalmologist complaining of increasing redness and irritation of his left eye, progressive over the previous 4 to 6 months. Examination discloses mildly edematous and erythematous left eyelids with mild conjunctival infection and scant mucus discharge. The conjunctival findings are much more prominent superiorly. The right eye appears normal. With this patient, the critical historical feature to inquire about is:

a. use of over-the-counter eye medications.
b. any history of previous sexually transmitted diseases.
c. which side of his body he generally chooses to sleep on.
d. any previous history of allergic disorders.
e. any previous history of arthritic disorders.

132. T or F One feature that characterizes infectious marginal keratitis is tendency for centripetal spread.

133. The two leading causes of corneal blindness in the United States are:

a. trachoma and trauma.
b. trachoma and herpes simplex.
9: Cornea, External Disease, and Refractive Surgery

c. trauma and herpes simplex.
d. trachoma and onchocerciasis.
e. trauma and onchocerciasis.

134. T or F The most common means of developing a type 1 herpes simplex virus (HSV) genital infection is by orogenital sexual activity.

135. T or F The most common means of developing a type 2 herpes simplex virus (HSV) ocular infection is by orogenital sexual activity.

136. Latent type 1 herpes simplex virus (HSV) (responsible for recurrent orofacial infection) generally resides in the:
a. oculomotor nucleus.
b. gasserian ganglion.
c. geniculate ganglion.
d. sphenopalatine ganglion.
e. superior cervical ganglion.

137. Clinical features differentiating primary from recurrent herpes simplex virus (HSV) infection include:
   1. prominent follicular membranous conjunctivitis.
   2. preauricular lymphadenopathy.
   3. duration and size of corneal dendrites.
   4. vesicular blepharitis.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

138. T or F Decreased corneal sensation is a sensitive and specific sign of recurrent herpetic keratitis.

139. Which of the following statements about the treatment of recurrent herpetic epithelial keratitis is/are true?
   1. Debridement of active dendrites may be of value.
   2. No improvement in epithelial disease after 7 to 10 days of therapy indicates resistance to the selected medication.
   3. Oral acyclovir may play a role in the treatment of recurrent epithelial herpes simplex virus (HSV) keratitis.
   4. Topical steroids in low doses are recommended to minimize corneal scarring.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

140. Potential outcomes of overtreatment with topical antivirals for herpes simplex virus (HSV) keratitis include:
   1. sterile corneal ulceration.
   2. pseudodendrites.
   3. punctate keratitis with photophobia.
   4. bacterial superinfection.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

141. Two weeks after initial diagnosis and topical therapy for herpes simplex virus (HSV) epithelial keratitis, a patient returns with a 4-mm, oval, central epithelial defect with smooth, rolled edges. Factors that may be important in the pathogenesis of this finding include:
   1. underlying stromal inflammation.
   2. overuse of prescribed topical antivirals.
   3. impaired corneal sensation.
   4. active intraepithelial virus replication.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

142. T or F The descriptor “disciform” is specific for autoimmune keratitis due to herpes simplex virus (HSV) infection.

143. Which of the following is/are seen as part of the spectrum of herpes simplex virus (HSV) disciform keratitis?
   1. Descemet’s folds.
   2. mild anterior uveitis with keratic precipitate (KP).
   3. glaucoma.
   4. peripheral anterior synchiae.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
Questions

144. A circular, superficial distribution of neutrophils around an area of corneal edema or inflammation is called:
   a. Wessely’s ring.
   b. disciform keratitis.
   c. corneal abscess.
   d. metaherpetic ulcer.
   e. ring ulcer.

145. The differential diagnosis of the lesion described in question 144 includes all of the following except:
   a. herpes simplex virus (HSV).
   b. Reiter’s syndrome.
   c. Acanthamoeba keratitis.
   d. anesthetic keratopathy.
   e. Behçet’s disease.

146. T or F Topical steroids play a role in the management of all cases of disciform keratitis.

147. T or F Herpetic interstitial keratitis (IK) is generally more aggressive than disciform keratitis.

148. Which of the following are justifications for the use of oral acyclovir in the management of herpes simplex ocular infection?
   1. primary infection.
   2. concomitant chronic oral steroid therapy for a nonrelated disorder.
   3. recalcitrant epithelial or stromal keratitis.
   4. systemic immune deficiency.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

149. T or F Cutaneous vesicles at the tip of the nose (the Hutchinson sign) indicate a high probability of ocular involvement by herpes simplex virus (HSV).

150. T or F All cases of herpes zoster ophthalmicus (HZO) represent reactivation of latent trigeminal ganglion infection.

151. T or F The dermatitis associated with herpes zoster reactivation is generally more protracted than the ocular inflammation.

152. Medications useful in the control of postherpetic neuralgia include:
   1. cimetidine orally for the first 7 days of active dermatitis.
   2. oral tricyclic antidepressant.
   3. acyclovir.
   4. prednisone orally, started 7 to 10 days after eruption of active dermatitis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

153. All of the following features of corneal dendrites favor the diagnosis of herpes zoster ophthalmicus (HZO) except:
   a. a large, frequently branching dendrite.
   b. a dendrite with overlying plaque of epithelial cells.
   c. a dendrite with no terminal bulb.
   d. coarse, ropy dendrites with blunt ends.
   e. a dendrite with dull fluorescein and no rose bengal staining.

154. Which of the following are systemic risk factors for the development of bacterial keratitis?
   1. drug abuse.
   2. aging.
   3. vitamin deficiency.
   4. diabetes mellitus.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

155. Local factors that may increase the risk of bacterial keratitis include:
   1. dry eye syndrome.
   2. impaired corneal sensation.
   3. recurrent erosions.
   4. chronic topical steroid therapy.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
156. T or F Compared with Gram-negative organisms, Gram-positive organisms tend to produce focal infiltrates.

157. T or F Compared to Gram-positive organisms, Gram-negative organisms are more likely to produce fulminant liquefactive corneal necrosis.

158. Which of the following constitute independent risk factors for the development of Acanthamoeba keratitis?
   1. diabetes mellitus.
   2. frequent use of hot tubs.
   3. previous history of a corneal transplant.
   4. contact lens use.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

159. Which of the following features of contact lens use constitutes the greatest risk for the development of Acanthamoeba keratitis?
   a. homemade saline storage solutions.
   b. use of solutions containing thimerosal.
   c. heat sterilization techniques.
   d. chemical sterilization techniques.
   e. insufficient enzyme treatments.

160. Which clinical feature is nearly universal in Acanthamoeba keratitis?
   a. tearing.
   b. pseudoptosis.
   c. severe pain.
   d. ring infiltrate.
   e. hypopyon.

161. Which of the following may be considered independent risk factors for the development of fungal keratitis?
   1. prolonged use of topical corticosteroids.
   2. prolonged use of broad-spectrum topical antibiotics.
   3. corneal trauma.
   4. previous history of herpetic keratitis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

162. Leading causes of interstitial keratitis (IK) in the United States include all of the following except:
   a. sarcoidosis.
   b. lepromatous leprosy.
   c. Cogan’s syndrome.
   d. herpes zoster virus (HZV).
   e. syphilis.

163. T or F Interstitial keratitis (IK) is more frequently seen as a complication of congenital syphilis than acquired syphilis.

164. T or F Compared with congenital syphilis, tuberculous interstitial keratitis (IK) is more frequently unilateral and sectoral.

165. Cogan’s syndrome is frequently associated with which systemic disorder?
   a. polyarteritis nodosa.
   b. Wegener’s granulomatosis.
   c. rheumatoid arthritis.
   d. adenocarcinoma, usually bronchogenic.
   e. systemic lupus erythematosus (SLE).

166. Which one of the following statements about Thygeson’s superficial punctate keratitis is false?
   a. There may be a human leukocyte antigen (HLA) association.
   b. The presenting symptom is typically photophobia or tearing.
   c. There is usually an associated follicular conjunctivitis.
   d. The corneal deposits may resemble those of epidemic keratoconjunctivitis (EKC).
   e. Topical steroids have been used for symptomatic relief, but may prolong the natural history of the disorder.

167. There is a definite association of superior limbic keratoconjunctivitis (SLK) with:
   a. valvular heart disease.
   b. thyroid disease.
   c. inflammatory bowel disease.
   d. systemic lupus erythematosus (SLE).
   e. bacillary dysentery.
Questions

168. Modalities helpful in long-term control of superior limbic keratoconjunctivitis (SLK) include:
   1. topical silver nitrate.
   2. cautery of bulbar conjunctiva.
   3. pressure patching or contact lenses.
   4. topical corticosteroids.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

169. T or F The approach to superior limbic keratoconjunctivitis (SLK) in a patient with a history of soft contact lens (SCL) use is identical to that for a patient with no such history.

170. Which class of chemicals constitutes the greatest threat for ocular injury?
   a. solvents.
   b. petroleum products.
   c. acids.
   d. alkalis.
   e. detergents.

171. T or F The pathophysiology of alkali burns involves protein denaturation and precipitation of previously soluble tissue protein.

172. T or F The tissue damage due to acid burns tends to be self-limited by tissue buffering and barrier effects of the tissue damage itself.

173. T or F Glaucoma is more likely to be a long-term sequela of alkali burns than acid burns.

174. Clinical features to be observed at the initial evaluation of a chemical burn include:
   1. tear pH.
   2. extent of epithelial defects.
   3. extent of limbal ischemia.
   4. corneal clarity.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

175. After thorough and copious irrigation of the conjunctival fornices, the next most important step in initial management of a patient with a chemical burn is:
   a. topical steroid agents.
   b. topical antibiotic agents.
   c. debridement of any foreign bodies.
   d. topical ascorbate.
   e. topical citrate.

176. The primary goal in intermediate therapy for chemical burns is:
   a. normalization of intraocular pressure (IOP).
   b. reestablishment of limbal blood flow.
   c. control of intraocular inflammation.
   d. normalization of tear and conjunctival pH.
   e. reepithelialization of the corneal surface.

177. T or F The best antibiotic prophylaxis following a chemical burn is a broad-spectrum antibiotic such as gentamicin.

178. T or F The optimal time for the administration of topical corticosteroids for dampening inflammation is 7 to 14 days following chemical injury.

179. T or F Long-term management following severe ocular chemical injury involves reestablishment of healthy epithelial surfaces with conjunctival and/or corneal transplants.

180. Which one of the following statements about episcleritis is false?
   a. Both nodular and diffuse forms have been described.
   b. Most cases are sectoral.
   c. Most cases will be recurrent.
   d. The condition may lead to scleritis if not promptly treated.
   e. Treatment consisting of topical steroids or nonsteroidal agents is generally ameliorative.

181. Features distinguishing episcleritis from scleritis include all of the following except:
   a. the presence of pain.
   b. the color of affected sclera.
   c. response to topical phenylephrine.
   d. the presence of episcleral versus scleral edema.
   e. association with systemic connective tissue disorders.

182. The most benign form of scleritis is:
   a. diffuse anterior scleritis.
   b. nodular anterior scleritis.
c. necrotizing scleritis with inflammation.
d. scleromalacia perforans.
e. posterior scleritis.

183. The scleritis associated with the gravisest systemic
prognosis is:
a. nodular diffuse anterior scleritis.
b. nodular anterior scleritis.
c. necrotizing scleritis with inflammation.
d. scleromalacia perforans.
e. posterior scleritis.

184. The scleritis most likely to be associated with
rheumatoid arthritis is:
a. diffuse anterior scleritis.
b. nodular anterior scleritis.
c. necrotizing scleritis with inflammation.
d. scleromalacia perforans.
e. posterior scleritis.

185. The scleritis most likely to present with proptosis
and visual loss is:
a. nodular diffuse anterior scleritis.
b. nodular anterior scleritis.
c. necrotizing scleritis with inflammation.
d. scleromalacia perforans.
e. posterior scleritis.

186. T or F Posterior uveitis (i.e., vitreous cells, pa-
pillitis, macular edema with exudates) is common
in scleritis; anterior uveitis is rare.

187. T or F The pathophysiology of scleritis also may
be manifest in the cornea as either a sclerosing or
a lytic marginal keratitis.

188. Infectious scleritis may be seen due to all of the
following except:
a. Chlamydia.
b. syphilis.
c. tuberculosis.
d. herpes zoster.
e. leprosy.

189. Systemic disease associations for scleritis include:
1. rheumatoid arthritis.
2. Wegener’s granulomatosis.
3. polyarteritis nodosa.
4. inflammatory bowel disease.

190. Agents helpful in the medical management of au-
toinflammatory sclerokeratitis include all of the
following except:
a. topical indomethacin.
b. oral prednisone.
c. subtenon’s injection of corticosteroid.
d. cyclophosphamide.
e. cyclosporine A.

191. The most common cause of acute, painful en-
largement of the lacrimal gland is:
a. sarcoidosis.
b. Sjögren’s syndrome.
c. bacterial dacryoadenitis.
d. leprosy.
e. herpes zoster virus (HZV).

192. The most common cause of painless, bilateral
enlargement of lacrimal glands is:
a. sarcoidosis.
b. Sjögren’s syndrome.
c. bacterial dacryoadenitis.
d. leprosy.
e. herpes zoster virus (HZV).

193. Mikulicz’s syndrome refers to the combination of
chronic dacryoadenitis with:
a. rheumatoid arthritis.
b. enlargement and inflammation of the parotid
glands.
c. keratoconjunctivitis sicca.
d. systemic lupus erythematosus (SLE).
e. dacryocele.

194. The treatment of choice for the most common
cause of chronic canaliculitis is:
a. topical tetracycline for 2 weeks.
b. oral tetracycline for 3 weeks.
c. surgical evacuation of the canaliculus.
d. topical corticosteroids.
e. oral acyclovir for 2 weeks.

195. Chronic asymptomatic dacryocystitis is most
frequently caused by:
a. Staphylococcus aureus.
b. Streptococcus pneumoniae.
Questions

c. *Staphylococcus epidermidis*.
d. *Haemophilus influenzae*.
e. *Pseudomonas aeruginosa*.

196. A patient presents with a tender mass below the medial canthal tendon and mucopurulent discharge from the inferior canaliculus. One week of oral antibiotic treatment and warm compresses lead to an increase in size and fluctuance of the mass. The next step in treatment should be:

a. increasing the frequency of warm compresses.
b. change in antibiotic agents.
c. increasing the frequency of dosage of the antibiotic agent.
d. probing and irrigation of the nasolacrimal system.
e. incision and drainage of the fluctuant mass.

197. A 2-month-old infant with unilateral epiphora in the left eye is brought to the ophthalmologist by her parents. Gentle compression of the lacrimal sac produces reflux of mucus from the canaliculi, but only on the left. There is obviously increased tear flow on the left as well. The next step should probably be:

a. warm compresses to the left eye.
b. probing and irrigation of the nasolacrimal system on the left.
c. reassurance with once daily antibiotic ointment and gentle medial canthal massage.
d. incision and drainage of the lacrimal sac.
e. oral antibiotics.

198. A 13-month-old infant with chronic epiphora and discharge in the left eye is brought to the ophthalmologist by his parents. Gentle massage of the medial canthal area produces a reflux of mucus from the canaliculi. The next step in management should be:

a. warm compresses to the left eye.
b. probing and irrigation of the nasolacrimal system on the left.
c. reassurance with once daily antibiotic ointment and gentle medial canthal massage.
d. incision and drainage of the lacrimal sac.
e. oral antibiotics.

199. T or F Following unsuccessful probing of a congenitally impatent nasolacrimal system, the next therapeutic step should be dacryocystorhinostomy.

200. The epithelium of eyelid skin is:

a. keratinizing stratified squamous.
b. nonkeratinizing stratified squamous.
c. keratinizing stratified columnar.
d. nonkeratinizing stratified columnar.
e. keratinizing pseudostratified.

201. Which of the following with regard to wavefront analysis and wavefront aberrations is true?

a. Wavefront analysis is only used to describe reference spheres and cannot be used in patients with irregular astigmatism.
b. A penetrating keratoplasty (PKP) using eight interrupted sutures will produce a four-leaf clover.
c. Spherical aberration occurs when central rays focus more in front of peripheral rays, leading to night myopia in some patients postoperative to laser-assisted *in situ* keratomileusis (LASIK).
d. An eye with no astigmatism will have no wavefront aberration.
e. None of the above.

202. T or F Corneal haze is a common complication of lamellar keratoplasty.

203. In which of the following conditions would lamellar keratoplasty be least efficacious?

a. Terrien’s marginal degeneration.
b. desmetocele.
c. pellucid marginal degeneration.
d. Salzmann’s nodular degeneration.
e. Fuchs’ endothelial dystrophy.

204. Features differentiating epibulbar epithelium from epidermal epithelium include:

1. lack of rete ridges.
2. lack of a granular layer.
3. presence of goblet cells.
4. lack of a prickle cell layer.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

205. Features differentiating conjunctival from corneal epithelium include:

1. lack of rete ridges.
2. lack of granular cell layer.
3. lack of a keratin layer.
4. presence of goblet cells.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

206. Which of the following statements accurately represents Munnerlyn’s formula?
   a. The ablation depth (in microns) is equal to the diopters of myopia divided by 3 and multiplied by the square of the optical zone (in millimeters).
   b. The ablation depth (in microns) is equal to the diopters of myopia multiplied by 3 and divided by the square of the optical zone (in millimeters).
   c. The ablation depth (in microns) is equal to the diopters of myopia divided by 5 and multiplied by the cube of the optical zone (in millimeters).
   d. The ablation depth (in microns) is equal to the diopters of myopia multiplied by 3 and divided by the cube of the optical zone (in millimeters).
   e. The ablation depth (in microns) is equal to the diopters of myopia multiplied by 2 and added to the square of the optical zone (in millimeters).

207. T or F A melanophage is a melanocyte that has differentiated into a phagocytic cell.

208. Which of the following is/are common features of conjunctival papillomas?
   1. hyperkeratosis.
   2. acanthosis.
   3. parakeratosis.
   4. anaplasia.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

209. Which of the following eyelid or conjunctival lesions may assume a papillomatous growth pattern?
   1. seborrheic keratosis.
   2. actinic keratosis.
   3. verruca.
   4. squamous cell carcinoma.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

210. A 33-year-old man presents to an ophthalmologist complaining of a “growth” on his eyelid. He maintains that the lesion developed over the preceding 4 weeks and is nontender. He produces a driver’s license photo from 4 months earlier, which shows normal eyelids. Examination discloses a 3.5-cm round elevated lesion of the right lower eyelid with a central depressed area and debris within. There is no pigmentation. The most likely diagnosis is:
   a. seborrheic keratosis.
   b. actinic keratosis.
   c. keratoacanthoma.
   d. basal cell carcinoma.
   e. squamous cell carcinoma.

211. T or F If diagnostic suspicion proves correct for the patient in question 210, biopsy will often reveal severe dysplasia.

212. T or F If diagnostic suspicion proves correct for the patient in question 210, biopsy may reveal significant inflammation.

213. T or F If the patient in question 210 is not troubled by the cosmetic features, he may be reassured and instructed to return for a follow-up examination in 3 to 6 months.

214. Which one of the following statements about seborrheic keratosis is false?
   a. It is a lesion most commonly seen in elderly people.
   b. There may be an inherited tendency for its development.
   c. Texturally, the lesion appears dry and scaly.
   d. Histopathologically, there is prominent dyskeratosis and hyperpigmentation in a papillary growth pattern.
   e. The lesion must be carefully distinguished from actinic keratosis.

215. The key differentiating feature between a dermoid cyst and an epidermoid cyst is:
   a. Dermoid cysts contain keratin within the cyst cavity.
   b. Dermoid cysts are solid.
Questions

c. Dermoid cysts contain sebum within the cyst cavity.
d. Dermoid cysts are morphologically identical, but follow trauma or surgery.
e. Dermoid cysts contain dermal appendages within the cyst wall.

216. **T or F** The lesions of molluscum contagiosum are similar to keratoacanthoma except that the former are generally more inflamed.

217. Features serving to differentiate between actinic keratosis and seborrheic keratosis include:
   1. hyperpigmentation.
   2. elastosis.
   3. chronic dermal inflammation.
   4. epithelial atypia.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

218. Which of the following growth patterns of basal cell carcinoma carries the worst prognosis?
   a. fibrosing.
   b. cystic.
   c. adenoid.
   d. adenocystic.
   e. nodular.

219. Which one of the following statements about basal cell carcinoma is true?
   a. Growth is typically explosively rapid.
   b. In 25% of the cases, the conjunctiva is primarily involved with secondary skin involvement.
   c. The upper eyelid is affected more frequently than the lower eyelid.
   d. Nuclei at the periphery of tumor cell nests retain polarity with palisading.
   e. There are usually numerous mitotic figures per high power field.

220. Which location of basal cell carcinoma carries the poorest prognosis?
   a. medial lower lid.
   b. lateral lower lid.
   c. lateral canthus.
   d. upper lid.
   e. medial canthus.

221. Basal cell carcinoma causes the most significant systemic morbidity and mortality by:
   a. hematogenous metastasis to the brain.
   b. local invasion of skull and central nervous system (CNS).
   c. lymphatic metastasis.
   d. hematogenous metastasis to the lung.
   e. hematogenous metastasis to liver.

222. Which one of the following statements about squamous cell carcinoma of the eyelid is false?
   a. It is less common than basal cell carcinoma.
   b. Chronic actinic exposure plays a role in its development.
   c. The upper eyelid is more frequently involved than the lower eyelid.
   d. Metastatic potential is greater than for basal cell carcinoma.
   e. Growth pattern is usually rapid.

223. A 68-year-old woman complains to her ophthalmologist that her stye just will not go away, despite 3 months of warm compresses and two surgical drainages. She undergoes full-thickness biopsy of her lower lid; a light-microscopic section is shown in the figure below. Which of the following is/are true regarding her situation?
   1. The disorder typically affects middle-aged or elderly people.
   2. Prompt drainage of the initial chalazion would have been curative.
   3. Grossly, the lesion may have an orange-yellow hue.
   4. Mohs' micrographic techniques can be curative at this stage.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
224. The biopsy specimen from the patient in the preceding question should also have undergone which one of the following histopathologic techniques?
   a. electron microscopy.
   b. cell surface marker studies.
   c. cellular adhesion studies.
   d. frozen section processing.
   e. serial section techniques.

225. All of the following features are often seen in sebaceous cell carcinoma except:
   a. epithelial xanthomatization.
   b. peripheral palisading of tumor cell nuclei.
   c. intraepithelial tumor cells.
   d. multifocal tumor cell nests.
   e. intraepithelial inflammatory cells.

226. Potential sites of origin of sebaceous cell carcinoma include:
   1. the glands of Zeis.
   2. the glands of the caruncle.
   3. the meibomian glands.
   a. 1, 2, and 3.
   b. 2 and 4.
   c. 1 and 3.
   d. 4 only.
   e. 1, 2, 3, and 4.

227. Adnexal tumors of hair follicle origin include all of the following except:
   a. syringoma.
   b. trichoepithelioma.
   c. trichilemmoma.
   d. pilomatrixoma.
   e. trichofolliculoma.

228. Which of the following tumors is most likely to calcify?
   a. syringoma.
   b. trichoepithelioma.
   c. trichilemmoma.
   d. pilomatrixoma.
   e. trichofolliculoma.

229. T or F The difference between ephelis and lentigo is an increased number of melanocytes in the latter.

230. T or F The difference between nevi and lentigo is modification of the melanocyte population in nevi.

231. Which one of the following statements about nevi is false?
   a. pigmentation and growth generally increase around the onset of puberty.
   b. With time, nevi tend to advance superficially, toward the surface epithelium.
   c. Junctional activity carries the greatest potential for malignant transformation.
   d. Subepithelial or dermal activity carries the least potential for malignant transformation.
   e. Nevi may be considered as hamartomatous abnormalities.

232. T or F One potential advantage of the femtosecond laser in laser-assisted in situ keratomileusis (LASIK) surgery is that there is no need for a microkeratome in order to create a stromal flap.

233. Each of the following lesions is related to chronic sun exposure except:
   a. squamous cell carcinoma of the eyelid.
   b. lentigo maligna.
   c. sebaceous cell carcinoma.
   d. basal cell carcinoma.
   e. malignant melanoma.

234. The textural term most frequently applied to the consistency of a plexiform neurofibroma of the upper eyelid is:
   a. micropebbly.
   b. “bag of marbles.”
   c. gravelly.
   d. “bag of worms.”
   e. corduroy.

235. Which one of the following statements about Kaposi's sarcoma is false?
   a. It may be considered the malignant counterpart of a pyogenic granuloma.
   b. The disorder is endemic in Central Africa.
   c. In the setting of normal immune regulation, the disease typically affects the lower extremities of older men.
   d. Radiation currently plays no role in the management of ocular Kaposi's sarcoma.
   e. The disorder is generally more aggressive and lethal in the immunocompromised individual.
Questions

236. A stye or external hordeolum must arise from:
   1. glands of Zeis.
   2. eccrine sweat glands.
   3. hair follicles.
   4. meibomian glands.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

237. T or F All styes (external hordeola) are infectious.

238. Features critical in the definition of chalazion include:
   1. no history of pain.
   2. involvement of sebaceous glands.
   3. insidious onset.
   4. granulomatous inflammation.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

239. Potential etiologies for multiple discrete eyelid nodules include all of the following except:
   a. juvenile xanthogranuloma.
   c. xanthelasma.
   d. lipoid proteinosis.
   e. syringoma.

240. T or F The key difference between epibulbar and adnexal dermoid tumors is the presence of all three cell lines (ectoderm, mesoderm, and endo-derm) in epibulbar dermoids.

241. A 32-year-old man from North Carolina presents to an ophthalmologist for routine examination. The ophthalmologist notes bilateral bulbar leuko-plakia at the nasal and temporal limbus. On further questioning, the patient reports that these lesions have been present for many years and that several of his siblings have similar findings. Examination of which of the following is most likely to confirm the probable diagnosis:
   a. medication history.
   b. history of sunlight exposure.

242. Epithelial neoplasms of the conjunctiva and cornea bear striking pathologic similarities to neoplasms of the:
   a. stomach.
   b. ovary.
   c. cervix.
   d. urinary bladder.
   e. colon.

243. Which one of the following is not a risk of clear lens exchange as a corrective treatment of high myopia?
   a. endophthalmitis.
   b. stromal flap dehiscence.
   c. intraocular lens (IOL) dislocation or decenteration.
   d. retinal detachment.
   e. ptosis.

244. The most common location of origin for corneal intraepithelial neoplasia is:
   a. the inferior fornix.
   b. the superior fornix.
   c. the limbus.
   d. the bulbar conjunctiva.
   e. the palpebral conjunctiva.

245. The key structure preventing local invasion of squamous cell carcinoma of the cornea is:
   a. epithelial basement membrane.
   b. Bowman’s zone.
   c. corneal stroma.
   d. Descemet’s membrane.
   e. endothelium.

246. Which one of the following statements about conjunctival nevi is false?
   a. Because of the absence of a dermal layer, conjunctival nevi are of the junctional variety only.
   b. Conjunctival nevi are frequently cystic.
   c. Because of sudden enlargement, mucus secretion within nevi can lead to the false impression of malignant transformation.
   d. Conjunctival nevi are more frequently amelanotic or lightly pigmented than skin nevi.
   e. Malignant transformation to melanoma is rare.
247. Which of the following statements regarding congenital melanosis oculi is accurate?
   a. Ocular melanocytosis is more common in African Americans and Asians, in whom malignant transformation is more common.
   b. Ocular melanocytosis is more common in whites, but malignant transformation to melanoma is more common in African Americans and Asians.
   c. Oculodermal melanocytosis is more common in Asians and African Americans, in whom malignant transformation is more common.
   d. Oculodermal melanocytosis is more common in African Americans and Asians, but malignant transformation is more common in whites.
   e. Oculodermal melanocytosis is equally common among whites, African Americans, and Asians.

248. Which of the following statements about primary acquired melanosis of the conjunctiva is/are true?
   1. The pigmented lesions represent proliferation of intraepithelial melanocytes.
   2. It is primarily a disorder of the middle-aged and elderly.
   3. The most troublesome sign (indicating potential malignant transformation) is nodular thickening.
   4. The most frequently involved region is the palpebral conjunctiva.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

249. T or F Biopsy of acquired melanosis of the conjunctiva should be undertaken only when absolutely necessary, and always excisionally, to avoid seeding and spread of tumor.

250. T or F The most important adjunctive therapy to excisional biopsy of conjunctival melanoma is radiotherapy.

251. Features suggestive of benign reactive lymphoid hyperplasia rather than conjunctival lymphoma include:
   1. presence of prominent vascularity.
   2. heterogeneous cell population (neutrophils, eosinophils, plasma cells, and numerous lymphocytes).
   3. presence of numerous follicles.
   4. nodular, fleshy appearance on bulbar conjunctiva.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

252. T or F Most conjunctival lymphomas are derived from T lymphocytes.

253. T or F The best indicator of conjunctival lymphoma (rather than a reactive lesion) is light-microscopic evidence of cellular atypia.

254. Which one of the following statements about abnormalities of corneal size is false?
   a. Megalocornea is defined as a cornea whose horizontal diameter is >13 mm.
   b. Most of the megalocornea is bilateral and seen in men.
   c. Megalocornea may be associated with systemic abnormalities such as Down’s syndrome, Marfan’s syndrome, or Alport’s syndrome.
   d. Microcornea is often transmitted familially.
   e. In isolated microcornea, the eye is generally myopic.

255. The anterior segment dysgeneses reflect developmental abnormalities related to what cell line?
   a. surface ectoderm.
   b. neuroectoderm.
   c. neural crest.
   d. mesoderm.
   e. endoderm.

256. Schwalbe’s ring represents the normal:
   a. peripheral termination of corneal epithelium.
   b. peripheral border between corneal and trabecular endothelium.
Questions

c. peripheral border between nonpigmented and pigmented trabecular meshwork.
d. peripheral termination of Bowman’s zone.
e. anterior insertion point of ciliary muscle.

257. An abnormally prominent Schwalbe line is referred to as:
a. posterior embryotoxon.
b. Rieger’s anomaly.
c. Peters’ anomaly.
d. Axenfeld’s anomaly.
e. internal ulcer of von Hippel.

258. A patient presents with unilateral glaucoma. Gonioscopy reveals an anteriorly displaced, prominent Schwalbe line with attached iris processes. The angle is otherwise open. This clinical picture is most consistent with:
a. posterior embryotoxon.
c. Peters’ anomaly.
d. iridocorneal endothelial (ICE) syndrome.
e. none of the above.

259. The brother of the patient described in question 258 is examined and also found to have elevated intraocular pressure (IOP). Gonioscopy on this patient reveals findings similar to those of the earlier patient, but with additional findings of iris stromal hypoplasia and polycoria. There are no other obvious systemic abnormalities noted. This patient’s clinical condition would be most correctly termed:
a. posterior embryotoxon.
c. Peters’ anomaly.
d. iridocorneal endothelial (ICE) syndrome.
e. none of the above.

260. All cases of Peters’ anomaly share which of the following features?

1. polycoria.
2. central absence of Descemet’s membrane and endothelium.
3. cataract or ectopia lentis.
4. corneal leukoma.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

261. T or F Approximately half of the patients with Peters’ anomaly have glaucoma and are bilaterally affected.

262. Which of the following statements about corneal birth trauma is/are true?

1. There are no means of distinguishing the findings from those of congenital glaucoma.
2. The presenting finding is typically corneal haziness (stromal edema) in the first postnatal week.
3. If corneal edema clears, there are no visual consequences and no permanent physical findings.
4. Corneal stromal edema may recur later in life.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

263. Which one of the following statements about pingueculae is false?

a. The agent most frequently implicated in the pathogenesis is ultraviolet light.
b. Histologically, accumulation of abnormal elastin material can be observed.
c. The nasal limbus is more frequently involved than the temporal limbus.
d. The lesions may calcify or become chronically inflamed.
e. Surgical excision is generally not pursued unless there are cosmetic or comfort issues.

264. Which of the following statements about pterygia is/are true?

1. Epidemiologically and histologically, pterygia are clearly extensions of pingueculae.
2. Corneal invasion is limited in depth by the epithelial basement membrane.
3. Mild inflammation and iron lines at the leading edge are typically seen.
4. Like pingueculae, surgical intervention is usually mandated for comfort.
a. 1, 2, and 3.
b. 1 and 3.
9: Cornea, External Disease, and Refractive Surgery

c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

265. Methods to diminish the recurrence rate of pterygia following excision include:
   1. conjunctival autotransplantation.
   2. beta-irradiation.
   3. topical mitomycin C.
   4. 5-fluorouracil (5-FU).
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

266. The stain of choice for suspected amyloid deposits of the external eye is:
   a. hematoxylin and eosin (H & E).
   b. Congo red.
   c. periodic acid–Schiff (PAS).
   d. alcian blue.
   e. mucicarmine.

267. A conjunctival deposit of amyloid is examined by biopsy and stained with Congo red. As a polarizing filter between the illuminating light and the specimen is rotated 90 degrees, the amyloid deposits seem to change from cherry red to apple green. This phenomenon is known as:
   a. birefringence.
   b. autofluorescence.
   c. metachromasia.
   d. fruit-looping.
   e. dichroism.

268. The most common form of conjunctival amyloidosis is:
   a. primary localized.
   b. primary systemic.
   c. heredofamilial.
   d. secondary localized.
   e. secondary systemic.

269. The most common type of eyelid amyloidosis is:
   a. primary localized.
   b. primary systemic.
   c. heredofamilial.
   d. secondary localized.
   e. secondary systemic.

270. Corneal forms of amyloidosis include all of the following except:
   a. limbal girdle of Vogt.
   b. primary gelatinous droplike dystrophy.
   c. lattice dystrophy type I.
   d. polymorphic amyloid degeneration.
   e. Meretoja’s syndrome.

271. Which one of the following statements about corneal arcus is false?
   a. The deposits generally begin in the interpalpebral fissure and spread superiorly and inferiorly with time.
   b. Incidence approaches 100% in patients older than 80 years.
   c. There is an increased incidence of arcus in African Americans.
   d. There is generally a lucent zone between the limbus and the peripheral edge of the arcus.
   e. Unilateral corneal arcus may be seen in the setting of contralateral high-grade carotid stenosis.

272. T or F Hassall-Henle bodies develop from a pathophysiologic mechanism identical to corneal guttae.

273. T or F Hassall-Henle warts are forerunners of corneal edema.

274. A 65-year-old woman is examined as part of a routine annual checkup. On retroillumination of the cornea, flecklike deposits in the deep corneal stroma are detectable centrally. Visual acuity is normal, and there are no other ocular findings. The most likely diagnosis is:
   a. Hassall-Henle bodies.
   b. cornea guttata.
   c. cornea farinata.
   d. central cloudy dystrophy.
   e. Fuchs’ dystrophy.

275. The most common etiology of calcific band keratopathy is:
   a. chronic ocular inflammation.
   b. systemic hypercalcemia.
   c. primary hereditary band keratopathy.
   d. renal failure.
   e. chronic mercurial exposure.
276. Clinically and histopathologically, the earliest calcium deposits in band keratopathy are located in the:
   a. horizontal peripheral cornea, Descemet’s membrane.
   b. vertical peripheral cornea, Descemet’s membrane.
   c. vertical peripheral cornea, Bowman’s zone.
   d. horizontal peripheral cornea, Bowman’s zone.
   e. central cornea, Descemet’s membrane.

277. The two most commonly encountered chemical compositions of band keratopathy are:
   a. urate and amyloid.
   b. urate and cholesterol.
   c. cholesterol and calcium.
   d. urate and calcium.
   e. cholesterol and amyloid.

278. **T** or **F** Pathophysiologic mechanisms at work in spheroidal corneal degeneration (climatic droplet keratopathy) are the same as that for pterygia and pingueculae.

279. Which of the following corneal degenerations are generally seen only in association with corneal neovascularization?
   1. Salzmann’s nodular degeneration.
   2. Spheroidal degeneration.
   3. Coats’ white ring.
   4. lipid keratopathy.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

280. A 63-year-old woman presents with a red, painful right eye. Examination discloses an ulcerative, circumferential marginal keratitis with a leading, undermined edge and early neovascularization. Which one of the following statements about the condition is false?
   a. There is dysregulation in both cellular and humoral immunity.
   b. A milder, less painful variant may be seen in young African American men.
   c. Medical management might include oral prednisone, cyclophosphamide or methotrexate.
   d. Surgical intervention might include conjunctival resection or lamellar keratoplasty.
   e. An evaluation for connective tissue disease is mandatory.

281. Which of the following conditions is not an absolute or relative contraindication to laser-assisted in situ keratomileusis (LASIK) surgery?
   a. a history of long-term topical corticosteroid use.
   b. a history of keratoconus.
   c. an expectation of 20/8 vision postoperatively.
   d. a history of long-term gas-permeable contact lens use.
   e. a history of being immunocompromised secondary to human immunodeficiency virus (HIV) infection.

282. Ulcerative marginal keratitis indistinguishable from Mooren’s ulcer may develop in patients with:
   1. acne rosacea.
   2. relapsing polychondritis.
   3. Wegener’s granulomatosis.
   4. polymyositis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

283. **T** or **F** There is a high spontaneous perforation rate in senile furrow degeneration.

284. **T** or **F** Phakic intraocular lens (IOLs) have been used to successfully treat both high hyperopia and high myopia.

285. **T** or **F** The most common classification for corneal dystrophies is by the biochemical nature of the corneal deposits.

286. The most common anterior corneal dystrophy is:
   a. Meesman’s dystrophy.
   b. Map-dot-fingerprint dystrophy.
   c. lattice dystrophy type 1.
   d. central cloudy dystrophy.
   e. Reis-Bückler dystrophy.
287. Which one of the following statements about anterior membrane dystrophy is false?
   a. Examination of family members may disclose a familial pattern.
   b. A disorder indistinguishable from the inherited form may be seen secondary to chronic blepharoconjunctivitis.
   c. Abnormalities in basement membrane production are manifest as map and fingerprint lines.
   d. Dots represent calcification of epithelial debris.
   e. Symptoms are generally related to defective epithelial adherence.

288. Treatment modalities useful in symptomatic anterior membrane dystrophy include all of the following except:
   a. epithelial debridement.
   b. copious lubrication.
   c. hypertonic saline ointments.
   d. stromal puncture.
   e. penetrating keratoplasty (PKP).

289. A 28-year-old patient presents to an ophthalmologist complaining of irritation and episodic blurry vision bilaterally. Slit-lamp examination reveals bubbles intraepithelially that are too numerous to count and that are visible only on retroillumination. These bubbles appear entirely transparent. A lamellar keratoplasty specimen should reveal:
   a. areas of reduplicated basement membrane and trapped epithelial cells.
   b. periodic acid–Schiff (PAS) staining of epithelially contained “peculiar substance.”
   c. focal areas of absence of basement membrane and fibrocellular invasion of Bowman’s zone.
   d. hyaline deposits in the anterior stroma.
   e. congophilic deposits in the anterior and mid-stroma.

290. Recurrent erosions are prominent features of which corneal dystrophies?
   1. anterior membrane dystrophy.
   2. Meesman’s dystrophy.
   3. Reis-Bückler dystrophy.
   4. central cloudy dystrophy.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

291. Which of the following corneal dystrophies is the most disabling visually?
   a. anterior membrane dystrophy.
   b. Reis-Bückler dystrophy.
   c. Meesman’s dystrophy.
   d. central cloudy dystrophy.
   e. pre-Descemet’s dystrophy.

292. A 29-year-old woman undergoes a routine ophthalmic examination. Visual acuity is normal. Slit-lamp examination discloses numerous crumblike deposits in the anterior corneal stroma, which are bilaterally symmetric and centrally densest. Intervening stroma is clear. Which one of the following is likely to be false?
   a. Careful recording of the woman’s clinical history may elicit a history of recurrent corneal erosions.
   b. Examination of family members will disclose similar findings.
   c. Visual acuity generally remains normal throughout life.
   d. Histopathologic review will reveal hyaline deposits on the Masson’s trichrome stain.
   e. Histopathologic review may reveal congophilic deposits resembling amyloid.

293. A 38-year-old man presents to the ophthalmologist complaining of gradual diminution in vision bilaterally. Visual acuity is 20/100 in both eyes. Slit-lamp examination reveals focal gray crumblike deposits in the stroma, densest centrally but extending to the limbus. Intervening areas of stroma have an ill-defined haze. Which one of the following statements about this condition is likely to be false?
   a. The patient’s siblings may be affected, but his offspring are unlikely to be.
   b. It is the stromal dystrophy most likely to be associated with recurrent erosions.
   c. It is the least common of the stromal dystrophies.
   d. A blood test may aid in the diagnosis.
   e. The stain of choice for the corneal biopsy specimen is alcian blue.

294. A 59-year-old woman presents to the ophthalmologist complaining of gradual loss of vision in each eye over the previous 5 years. Visual acuity is 20/80 in both eyes. Slit-lamp examination reveals multiple crumblike deposits in the anterior central
stroma along with refractile, branching, linear, stromal deposits centrally. The peripheral cornea is clear. Both eyes are symmetrically involved. Which one of the following statements is likely to be false?

a. Each of the patient’s children has a 50% chance of being affected with the same disorder.

b. Recurrent erosions are more likely to develop in this disorder than any of the other related disorders.

c. Recurrence in donor corneas following penetrating keratoplasty (PKP) is more likely with this disorder than any other related disorder.

d. The patient, on further questioning, will probably complain of a history of double vision or facial droop.

e. With polarizing filters on both sides of the specimen, light-microscopic evaluation of the patient’s corneal button following keratoplasty will reveal birefringence of the abnormal deposits.

295. Which of the stromal dystrophies, such as arcus and xanthelasma, may be associated with systemic hyperlipidemia?

a. granular dystrophy.

b. central crystalline dystrophy.

c. fleck dystrophy.

d. central cloudy dystrophy of Francois.

e. posterior amorphous stromal dystrophy.

296. Which one of the stromal dystrophies may be associated with keratoconus, atopy, or pseudoxanthoma elasticum?

a. granular dystrophy.

b. central crystalline dystrophy.

c. fleck dystrophy.

d. central cloudy dystrophy of Francois.

e. posterior amorphous stromal dystrophy.

297. Which one of the following stromal dystrophies is least likely to be associated with poor vision?

a. granular dystrophy.

b. lattice dystrophy.

c. macular dystrophy.

d. central cloudy dystrophy of Francois.

e. congenital hereditary stromal dystrophy.

298. T or F Pathophysiologically, the deposits representing corneal guttae are similar to macular drusen.

299. Which one of the following statements about Fuchs’ endothelial dystrophy is false?

a. At one end of the spectrum are corneal guttata; at the other are epithelial bullae.

b. Symptoms usually consist of blurry vision and pain, worsening in the evening.

c. Typically, stromal edema develops before epithelial abnormalities are noted.

d. Specular microscopy reveals larger, irregular cells (endothelial “polymegathism”) and decreased cell counts.

e. Penetrating keratoplasty (PKP) may need to be undertaken at the time of cataract surgery, although corneal symptoms are minimal or nonexistent.

300. An infant is born with bilaterally thickened, hazy corneas with epithelial edema. Corneal diameters are normal. There is associated nystagmus. Which one of the following statements about this condition is false?

a. Intraocular pressure (IOP) is likely to be elevated.

b. Another variant of the disorder exists in which there is no nystagmus and the onset is later.

c. Features distinguishing this disorder from congenital hereditary stromal dystrophy include corneal thickening and epithelial edema.

d. Descemet’s membrane may be thickened but there are no guttata.

e. This disorder reflects an abnormality in neural crest cell migration or differentiation.

301. A 28-year-old man presents claiming that “someone said my eyes look funny.” Visual acuity is normal bilaterally. Slit-lamp examination reveals multiple abnormalities on the posterior corneal surface, including groups of blisterlike deposits, scalloped banding, and irregular maplike grayish deposits on the endothelium with focal stromal edema. These findings are bilateral, as is corectopia, with the pupil drawn temporally. Careful questioning fails to reveal any family history of eye disorders. Which one of the following statements about this condition is probably false?

a. Careful examination of a sibling may reveal milder but similar findings.

b. Intraocular pressure (IOP) may be elevated.

c. Gonioscopy may reveal anterolaterally placed peripheral anterior synechiae.
9. Cornea, External Disease, and Refractive Surgery

d. Histopathologic findings of the eye, if re-viewed, would reveal an abnormally proliferative corneal endothelium with desmosomes and microvilli.
e. The most likely diagnosis is iridocorneal endothelial (ICE) syndrome.

302. If corneal transplantation were required on the eye of the patient in question 301, the pathology of the host corneal button would be strikingly similar to that seen in:
a. keratoconus.
b. epithelial downgrowth.
c. Fuchs’ dystrophy.
d. granular dystrophy.
e. macular dystrophy.

303. The most common finding in the contralateral eye of a patient with unilateral keratoconus is:
b. horizontal breaks in Descemet’s membrane.
c. fleck dystrophy.
d. myopia with high astigmatism.
e. Brushfield’s spots.

304. The time of greatest progression of keratoconus is during the:
a. first decade.
b. second decade.
c. third decade.
d. fourth decade.
e. the condition is generally static.

305. T or F Conductive keratoplasty has been approved by the U.S. Food and Drug Administration (FDA) for the treatment of low degrees of presbyopia.

306. A patient with known keratoconus presents to the ophthalmologist with a sudden decrease in vision and tearing from the right eye. Which one of the following statements about this situation is probably false?
a. The “tearing” represents spontaneous perforation and demands immediate surgery.
b. The corneal findings at the slit lamp may slowly resolve with time.
c. There may be considerable associated pain.
d. Typically, the condition is painless.
e. This process may accelerate scarring.

307. A patient presents to the ophthalmologist unhappy with his latest refraction. Examination discloses vision of 20/50, with each eye through his new pair of spectacles. This improves to 20/25 +, with a pinhole over either lens. Keratometry reveals 45.5D at 180 degrees and 53.5D at 90 degrees in each eye. The next logical intervention should be:
a. photorefractive keratectomy (PRK).
b. toric soft contact lens (SCL) fitting.
c. rigid contact lens fitting.
d. thermal keratoplasty.
e. penetrating keratoplasty (PKP).

308. T or F Keratoglobus usually offers a better visual prognosis either with or without penetrating keratoplasty (PKP) than keratoconus.

309. Contact lens fitting is usually most challenging for patients with:
a. keratoconus.
b. keratoglobus.
c. pellucid marginal degeneration.
d. posterior keratoconus.
e. microcornea.

310. At what point does diffuse lamellar keratitis post-laser-assisted in situ keratomileusis (LASIK) surgery typically manifest?
a. 6 to 12 hours postoperatively.
b. 24 to 48 hours postoperatively.
c. within the first week postoperatively.
d. within the first two weeks after LASIK.
e. within the first month after LASIK.

311. Which one of the following statements about tear deficiency states is false?
a. Prominent symptoms include blurry vision and pain with blinking.
b. Classic signs include ropy mucus discharge, corneal filaments, and punctate rose bengal staining in the exposure zone.
c. Pathophysiologically, the problem is a loss of adequate tear volume.
d. Patients may complain of epiphora.
e. Tests to be considered in the evaluation of potential tear deficiency include the Schirmer tests, rose bengal stain, and observation of tear breakup time.

312. T or F The purpose of topical anesthesia before the Schirmer testing is to reduce spontaneous blinking and prevent dislodging of the testing strips.
Questions

313. **T or F** Normal tear production during anesthetized Schirmer test is 10 mm or more after 5 minutes.

314. **T or F** An ophthalmologist can be released of his or her obligations to the patient immediately after surgery only when a patient undergoes a successful operation.

315. A patient presents with complaints typical for dry eye syndrome. Schirmer testing with and without anesthesia is normal. A hypothesis of tear deficiency state due to inadequate tear lipid or mucus layer would be best confirmed by:
   a. repeat the Schirmer testing.
   b. tear breakup time testing.
   c. tear osmolarity testing.
   d. rose bengal staining.
   e. tear lysozyme testing.

316. Which of the following historical features favor the diagnosis of a tear deficiency state?
   1. symptoms worse in the morning.
   2. symptoms aggravated by dry conditions.
   3. history of chalazia.
   4. symptoms aggravated in windy conditions.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

317. Classic Sjögren’s syndrome consists of keratoconjunctivitis sicca, xerostomia, and:
   a. eczema.
   b. arthritis.
   c. the Raynaud phenomenon.
   d. SS-A and SS-B autoantibodies.
   e. Hashimoto’s thyroiditis.

318. The critical difference between patients with primary and secondary Sjögren’s syndrome is:
   a. the presence of autoantibodies in the primary group.
   b. the presence of autoantibodies in the secondary group.
   c. salivary gland involvement.
   d. human leukocyte antigen (HLA)-Dw3 subtype in the primary group.
   e. association with systemic connective tissue disease in the secondary group.

319. Patients with primary Sjögren’s syndrome are at increased risk for subsequent development of:
   1. autoimmune thyroiditis.
   2. Waldenström’s macroglobulinemia.
   3. lymphoma.
   4. adenoid cystic carcinoma of the lacrimal gland.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

320. A patient with established dry eye syndrome is suffering persistent discomfort and blurry vision despite hourly topical artificial tears. The next most appropriate intervention might be:
   a. warm compresses to both eyes twice a day.
   b. swimmers’ goggles to be worn during the day.
   c. lateral tarsorrhaphy.
   d. a trial of temporary inferior punctal occlusion with collagen plugs.
   e. permanent thermal punctal occlusion for all four puncta.

321. Tear deficiency may play a pathophysiologic role in all of the following conditions except:
   a. corneal foreign body.
   b. ocular cicatricial pemphigoid (OCP).
   c. Bell’s palsy.
   d. use of oral contraceptives.
   e. previous herpes zoster keratitis.

322. **T or F** The distribution of keratopathy seen in long-standing Bell’s palsy is similar to that seen in primary Sjögren’s syndrome.

323. **T or F** The distribution of keratopathy seen in long-standing neurotrophic keratitis is similar to that seen in primary Sjögren’s syndrome.

324. A patient with a history of previous severe herpes zoster keratitis presents with Bell’s palsy.
Appropriate intervention at this point should include:

a. hourly artificial tears.
b. bandage contact lens. 
c. lateral tarsorrhaphy.
d. conjunctival flap. 
e. oral acyclovir.

325. Which one of the following statements about rosacea is false?
   a. It is more common in fair-skinned races.
   b. Facial lesions include telangiectasia, papules, pustules, and comedones.
   c. Nasal skin thickening (rhinophyma) is a late sign.
   d. Midfacial flushing may be enhanced with spicy food intake or hot beverages.
   e. Alcohol consumption can aggravate the disorder.

326. The most common ocular manifestation of rosacea is:
   a. chronic or recurrent meibomitis.
   b. marginal keratitis.
   c. episcleritis.
   d. peripheral keratitis.
   e. anterior uveitis.

327. T or F Tear deficiency state is more common in patients with rosacea than in patients with no evidence of the disorder.

328. Which of the following statements about the pathology of acne rosacea is/are true?
   1. Demodex infestation may play an inciting or aggravating role.
   2. Biopsy of involved tissues frequently reveals granulomatous inflammation.
   3. There may be irreversible obliteration of meibomian glands.
   4. Type IV hypersensitivity may play a role in the pathogenesis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

329. A 42-year-old man of Irish descent presents to the ophthalmologist complaining that his eyes have been red for several months. Examination discloses multiple brow and cheek telangiectasias with small papillary rash at the tip of the nose. All four eyelids are thickened with telangiectasias crossing the lid margin. There is focal meibomian gland loss. Both eyes have moderate conjunctival infection, and the right eye has a marginal infiltrate under intact epithelium at the inferior temporal limbus. An effective treatment strategy might include all of the following except:
   a. warm compresses to both eyes twice a day.
   b. topical bacitracin ointment to the eyelids twice a day.
   c. topical prednisolone 1.0% to the right eye every 2 to 4 hours.
   d. topical metronidazole for the skin findings.
   e. long-term oral doxycycline.

330. Which one of the following statements about erythema multiforme is false?
   a. The distinction between minor and major variants is the involvement of mucous membranes in the latter.
   b. The acute phase of the major type lasts longer than the minor type.
   c. Inflammation is generally confined to the dermis or submucosal stroma.
   d. Both major and minor varieties are self-limited and resolve uneventfully with proper supportive care.
   e. The inflammation is primarily angiocentric.

331. The most commonly implicated inciting agent in erythema multiforme is:
   a. recent use of penicillin.
   b. recent herpes simplex infection.
   c. recent mycoplasma infection.
   d. recent use of sulfonamides.
   e. recent use of anticonvulsants.

332. T or F Like the skin, the classic mucous membrane lesion of erythema multiforme resembles a target, or bull’s-eye.

333. Which one of the following statements about the acute care and long-term prognosis of erythema multiforme is false?
   a. Mortality approaches 20% in severe cases.
   b. The most frequent cause of death is secondary infection.
Questions

c. Systemic corticosteroids have substantially diminished mortality.
d. Aggressive ocular lubrication and lysis of symblepharons significantly reduce ultimate scarring.
e. The long-term ocular prognosis in toxic epidermal necrolysis is somewhat better than that of systemic erythema multiforme.

334. The mucous membrane most frequently involved in cicatricial pemphigoid is:
   a. oral.
   b. conjunctival.
   c. pharyngeal.
   d. esophageal.
   e. genitourinary.

335. T or F The pathophysiology of ocular cicatricial pemphigoid (OCP) is identical to that of Stevens-Johnson syndrome (erythema multiforme major).

336. Which one of the following statements about the clinical features of ocular cicatricial pemphigoid (OCP) is false?
   a. The disorder is rarely seen in patients younger than 30 years.
   b. Women are more commonly affected than men.
   c. The disease most typically presents as insidious, bilaterally asymmetric chronic conjunctivitis.
   d. Chronic use of topical ocular medications may induce a clinically identical picture.
   e. The most sensitive region of the eye to examine for early findings is the superior tarsus.

337. The classic histologic finding in ocular cicatricial pemphigoid (OCP) is:
   a. hypersensitivity angiitis in submucosal stroma.
   b. intraepithelial immunoglobulin and intraepithelial bullae.
   c. granulomatous destruction of epithelial basement membrane.
   d. complement and immunoglobulin bound to epithelial basement membrane.
   e. mast cells and eosinophils in the epithelium and subepithelial stroma.

338. T or F The initial drug of choice in all cases of ocular cicatricial pemphigoid (OCP) is dapsone.

339. Ocular findings consistent with vitamin A deficiency include:
   1. keratinization and bacterial superinfection of bulbar conjunctiva.
   2. tear deficiency state.
   3. deep white lesions of the peripheral retina.
   4. diffuse corneal necrosis with ulceration.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

340. Bilateral corneal ulceration should be presumed to be due to vitamin A deficiency until proved otherwise in patients with:
   1. cystic fibrosis.
   2. history of gastric bypass surgery.
   3. cirrhosis.
   4. patients with a history of heavy smoking but normal nutrition.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

341. A 32-year-old woman presents to the ophthalmologist with a complaint of recurrent pain and tearing of her right eye over the previous 1 to 2 months. Closer questioning discloses that her symptoms are virtually always present upon awakening and disappear after 2 or 3 hours. She denies any history of contact lens use. Which of the following statements is/are likely to be true?
   1. Careful questioning may reveal a history of corneal abrasion or trauma in the right eye.
   2. Careful examination of the right eye may disclose a focal abnormality in tear breakup.
   3. Careful examination of the left eye may reveal map or fingerprint abnormalities.
   4. This syndrome may be seen more frequently in patients with diabetes mellitus.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
342. Modalities accepted for treatment of the syndrome described in question 341 include all of the following except:
   a. pressure patching.
   b. copious lubricating ointments.
   c. five percent sodium chloride ointment at bedtime for 2 to 4 weeks.
   d. bandage contact lens.
   e. anterior stromal puncture with a 27-gauge needle.

343. Which one of the following statements about ligneous conjunctivitis is false?
   a. This is primarily a disorder of childhood.
   b. It is typically bilateral.
   c. It typically presents as an acute membranous conjunctivitis.
   d. Topical cyclosporine may be the most promising medical treatment.
   e. Surgical resection of involved conjunctiva is the definitive treatment.

344. Which of the following statements about the mucopolysaccharidoses (MPS) is/are true?
   1. Lysosomal enzyme defects lead to accumulation of metabolites within keratocytes.
   2. The MPS least likely to demonstrate corneal clouding is type III (Sanfilippo’s syndrome).
   3. Metabolites that accumulate include keratan sulfate, dermatan sulfate, and heparan sulfate.
   4. These disorders are all inherited on autosomal-recessive bases.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

345. The typical corneal finding in patients with Fabry’s disease is:
   a. corneal clouding.
   b. anterior membrane dystrophy.
   c. vortex keratopathy.
   d. corneal guttata.
   e. corneal neovascularization.

346. A 26-year-old woman presents for a routine ophthalmic examination. Slit-lamp examination discloses a vortex keratopathy bilaterally with telangiectatic conjunctival vessels. Her mother and father are both healthy. Which one of the following statements is false?
   a. A careful drug history should be taken.
   b. The patient should be warned of potentially lethal renal failure.
   c. Other slit-lamp findings might include subtle posterior subcapsular cataract.
   d. Fundus findings might include telangiectatic retinal vessels.
   e. Half of the patient’s brothers will be seriously affected by the same disorder.

347. A patient presents with photophobia and blurry vision. Examination discloses crystalline deposits throughout the entire stroma, densest peripherally. Findings that are important in determining the correct diagnosis include all of the following except:
   a. family history of eye findings.
   b. previous history of kidney transplant.
   c. history of peptic ulcer disease (PUD).
   d. presence of pigmentary retinopathy in each eye.
   e. serum immunoelectrophoresis.

348. The corneal findings in tyrosinemia most closely resemble those of:
   a. anterior membrane dystrophy.
   b. herpes simplex keratitis.
   c. Wilson’s disease.
   d. chronic indomethacin usage.
   e. ochronosis.

349. Which one of the following statements about vortex keratopathy is false?
   a. Drugs associated with the finding include amiodarone, indomethacin, chloroquine, and chlorpromazine.
   b. The findings in drug-induced vortex keratopathy are identical to those of Fabry’s disease.
   c. The pathophysiology of the deposits in drug-induced vortex keratopathy is identical to that of Fabry’s disease.
   d. The drug-induced varieties are accompanied by a pigmentary retinopathy.
   e. Cessation of drug therapy will usually lead to resolution of vortex keratopathy.
350. A patient is sent to an ophthalmologist by a gastroenterologist to “rule out Wilson’s disease.” The key part of the ophthalmologist’s examination should be:
   a. visual acuity measurement.
   b. slit-lamp examination.
   c. tonometry.
   d. gonioscopy.
   e. dilated funduscropy.

351. A Kayser-Fleischer ring may be seen in which of the following disorders?
   1. primary biliary cirrhosis.
   2. chronic active hepatitis.
   4. chalcosis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

352. T or F Photorefractive keratectomy (PRK) has been approved for the treatment of hyperopic astigmatism.

353. The most accurate characterization of donor endothelial cell counts following penetrating keratoplasty (PKP) is:
   a. no significant change.
   b. steady, progressive loss of endothelial cells forever.
   c. slow, steady increase in endothelial cell count over 10 to 15 years.
   d. rapid loss of endothelial cells over the first postoperative year, slow loss of endothelial cells over the next 10 to 15 years, with stable cell counts after 15 years.
   e. slow, progressive loss of endothelial cells over 10 to 15 years, with stabilization thereafter.

354. Currently, the most frequent indication for penetrating keratoplasty (PKP) in the United States is:
   a. keratoconus.
   b. bullous keratopathy following cataract extraction.
   c. Fuchs’ dystrophy.
   d. herpes simplex keratitis.
   e. corneal stromal dystrophy.

355. Which of the following are considered favorable prognostic factors for penetrating keratoplasty (PKP)?
   1. relatively young age.
   2. glaucoma.
   3. large graft size (>8.5 mm).
   4. no previous history of graft rejection.
   a. 1, 2, and 3.
   b. 2 and 4.
   c. 1 and 3.
   d. 4 only.
   e. 1, 2, 3, and 4.

356. Which of the following are considered unfavorable prognostic factors for penetrating keratoplasty (PKP)?
   1. considerable stromal vascularization.
   2. tear deficiency state.
   3. active intraocular inflammation.
   4. corneal hypesthesia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

357. The primary cause of poor vision following penetrating keratoplasty (PKP) for aphakic bullous keratopathy is:
   a. glaucoma.
   b. endophthalmitis.
   c. cystoid macular edema.
   d. graft rejection.
   e. retinal detachment.

358. Preoperative steps that are helpful in penetrating keratoplasty (PKP) include:
   1. pilocarpine 1% preoperatively.
   2. Honan balloon placement before surgery.
   3. intravenous mannitol preoperatively.
   4. phospholine iodide, preoperatively.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

359. T or F A scleral support ring (Flieringa’s ring) during penetrating keratoplasty (PKP) is most
helpful in eyes that are aphakic (or will become so during the procedure).

360. **T or F** The main advantage of small donor button size (7 mm or less) is decreased astigmatism, whereas the main disadvantage is increased risk of rejection.

361. **T or F** The main advantage of oversizing the donor button (0.5 mm or more larger than the host bed) is to decrease the incidence of peripheral anterior synechiae and subsequent elevated intraocular pressure (IOP).

362. The condition in which same-size or smaller-than-host-bed donor buttons are often used is:
   a. keratoconus.
   b. Fuchs’ dystrophy.
   c. herpetic keratitis.
   d. bullous keratopathy after cataract surgery.
   e. corneal stromal dystrophies.

363. Which method of closure of penetrating keratoplasty (PKP) causes the greatest amount of irregular astigmatism (before suture removal):
   a. interrupted.
   b. single running.
   c. double running.
   d. combined interrupted plus single running.
   e. combined interrupted plus double running.

364. A patient returns for follow-up 8 weeks after penetrating keratoplasty (PKP). The central cornea is clear but too irregular to permit accurate keratometry. Vision is 20/400, pinholing to 20/30. What is the most reliable and effective method of visual rehabilitation?
   a. random removal of sutures, one per week.
   b. removal of sutures that appear tightest at the slit lamp.
   c. use of keratoscope to guide suture removal.
   d. removal of all sutures simultaneously.
   e. contact lens refraction and correction.

365. **T or F** In penetrating keratoplasty (PKP) for bullous keratopathy associated with anterior chamber intraocular lenses (ACIOL), the intraocular lens (IOL) should be removed in all cases.

366. **T or F** In the setting of severe corneal disease combined with cataract, the corneal transplantation should be performed initially, with cataract extractions performed later as a secondary procedure.

367. A 62-year-old woman presents complaining of slow loss of vision in each eye. She denies any previous ocular history. Visual acuities are 20/400 in the right eye and 20/100 in the left eye. Slit-lamp examination reveals corneal guttata in both eyes with central stromal thickening more prominent on the right. The corneal epithelium is normal bilaterally. There is dense nuclear sclerosis on the right eye and moderate nuclear sclerosis on the left. The view of the fundus is consistent with the patient’s vision. The next appropriate step for visual rehabilitation in this patient might be:
   a. extracapsular cataract extraction (ECCE) only, in the right eye.
   b. ECCE with posterior chamber intraocular lens (PCiol) in the right eye.
   c. penetrating keratoplasty (PKP) alone, in the right eye.
   d. PKP with ECCE in the right eye.
   e. PKP, ECCE, and PCiol in the right eye.

368. A 65-year-old man presents to the ophthalmologist complaining of slow loss of vision in each eye. He denies any previous ocular history. Examination reveals visual acuities of 20/400 in the right eye and 20/100 in the left eye. Slit-lamp examination reveals severe corneal guttae bilaterally. The corneal epithelium and stroma are normal. There is dense nuclear sclerosis in the right eye and moderate nuclear sclerosis in the left eye. The next step for visual rehabilitation of this patient might be:
   a. extracapsular cataract extraction (ECCE) only in the right eye.
   b. ECCE with posterior chamber intraocular lens (PCiol) in the right eye.
   c. penetrating keratoplasty (PKP) alone in the right eye.
   d. PKP with ECCE in the right eye.
   e. PKP, ECCE, and PCiol in the right eye.

369. Mucous membrane grafting in anticipation of penetrating keratoplasty (PKP) is most hazardous in patients with:
   a. Stevens-Johnson syndrome.
   b. ocular cicatricial pemphigoid (OCP).
Questions

c. trachoma.
d. keratoconjunctivitis sicca.
e. herpes zoster ophthalmicus (HZO).

c. This condition is generally quite responsive to topical antibiotics.
d. Historical factors most significant in the development of the lesion include topical steroid use and keratoplasty.
e. Other organisms reported to cause a similar condition include coagulase-negative Staphylococcus and Candida species.

370. Persistent epithelial defects of the donor cornea following penetrating keratoplasty (PKP) are likely to be seen in all of the following except:
a. ocular cicatricial pemphigoid (OCP).
b. alkali burns.
c. keratoconus.
d. keratoconjunctivitis sicca.
e. herpes zoster ophthalmicus (HZO).

371. Recurrence of the original disease process has been reported following penetrating keratoplasty (PKP) for each of the following conditions except:
a. lattice dystrophy.
b. herpes simplex keratitis.
c. Reis-Bückler dystrophy.
d. Fuchs’ dystrophy.
e. macular dystrophy.

372. The most common postoperative complication seen after penetrating keratoplasty (PKP) is:
a. infectious keratitis.
b. recurrence of the original disease process.
c. acute glaucoma.
d. high astigmatism.
e. wound leak.

373. A patient presents to the ophthalmologist 12 weeks following penetrating keratoplasty (PKP). Complaints consist of increasing redness and discomfort in the operated eye. Visual acuity is the same as at the time of the previous office visit 2 weeks earlier. Examination discloses a white, crystalline infiltrate at the donor/host interface between sutures. The infiltrate has indistinct borders, and the stroma appears thickened by it. There is overlying epithelial irregularity but no confluent epithelial defect. Gram stain of a corneal scraping reveals Gram-positive cocci in chains. Cultures on blood agar grow multiple colonies with alpha hemolysis. Which one of the following statements regarding this condition is false?
a. The causative organism, as with cases originally described, is Streptococcus viridans.
b. This infection is generally slowly progressing.
following statements about this situation is probably false?

a. This is too early to represent corneal allograft rejection.
b. Elevated intraocular pressure (IOP) should be ruled out as a potential mechanism of corneal stromal edema.
c. Vitreous–endothelial touch should be ruled out as a potential cause of corneal edema.
d. An important clue for determining the etiology of the findings is the absence of similar signs in the host bed.
e. A linear array of keratic precipitates (KPs) may have been observed along the endothelium earlier in the process.

377. Which one of the following statements about corneal allograft rejection is false?

a. Epithelial rejection frequently manifests as subepithelial keratitis identical to epidemic keratoconjunctivitis (EKC).
b. Endothelial–stromal rejection is unlikely to develop in a first graft before 10 to 14 days postoperatively.
c. Endothelial–stromal rejection may be incited by suture removal or intraocular laser procedures.
d. Endothelial–stromal graft rejection should be treated as an emergency with intensive topical and systemic steroids.
e. Epithelial rejection is generally more severe and damaging to the donor button than endothelial–stromal rejection.

378. The potential advantages of lamellar keratoplasty over penetrating keratoplasty (PKP) include which of the following?

1. Restrictions on donor material are not as stringent.
2. The donor/host interface generally remains clearer.
3. There is a lower incidence of allograft rejection.
4. It is technically easier to perform.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

379. Indications for lamellar keratoplasty include all of the following except:

a. Reis-Bückler dystrophy.
b. a large corneal perforation in the bed of an infectious corneal ulcer.
c. superficial traumatic corneal scars.
d. lattice dystrophy.
e. Terrien's marginal degeneration.

380. Which one of the following statements about tissue adhesive closure of corneal defects is false?

a. Polymerization is rapid when the adhesive contacts free anions.
b. All epithelium and necrotic tissue must be cleared from around the perforation to enhance adhesion.
c. The bed to which the adhesive is to be attached must be dry at the time of treatment.
d. Polymerization releases considerable amounts of heat (exothermic).
e. Bandage contact lenses are generally needed following successful adhesive application.

381. Indications for conjunctival flap surgery include all of the following except:

a. bullous keratopathy.
b. chronic painful band keratopathy.
c. neurotrophic ulceration.
d. large perforation in the bed of an infectious corneal ulcer.
e. severe surface disruption with pain following chemical alkali burn.

382. A 30-year-old woman presents complaining of episodic bilateral eye pain, worse on the left eye and associated with blurry vision. On further questioning, she relates a history of severe red eyes approximately 3 weeks ago, which have gradually improved as her current symptoms have developed. Best corrected acuity is 20/25 in the right eye and 20/40 in the left eye. External examination is unremarkable, and there is no lymphadenopathy. Slit-lamp examination reveals bilateral corneal findings, worse on the left, as shown in the following figure. Which one of the following statements about her condition is false?

a. Three weeks earlier, she may have had a membranous conjunctivitis.
b. The corneal lesions represent active microbial replication.
c. Topical steroids could provide significant symptomatic relief.

d. Topical steroids could considerably increase the duration of the disease process.

e. Similar corneal findings can be seen in the absence of antecedent red eye.

383. A 21-year-old patient presents with a history of chronically poor vision in the right eye, and slit-lamp examination findings as shown in the figure below. Which of the following laboratory tests is useful for initial evaluation?

1. serum calcium levels.
2. chest x-ray.
3. fluorescent treponemal antibody-absorption (FTA-ABS).
4. complete human leukocyte antigen (HLA) typing.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

384. Three weeks after undergoing primary “bare-sclera” excision of a pterygium, a patient returns concerned about his eye (shown in the following figure). The surgeon should offer the patient:

a. beta-irradiation to the surgical bed.
b. mitomycin C drops immediately.
c. assurance and frequent topical prednisolone.
d. reoperation with conjunctival transplantation.
e. wide excision with map biopsies and cryotherapy.

385. Anterior stromal puncture has been most extensively used as a treatment of:

a. bullous keratopathy.
b. recurrent epithelial erosions.
c. atopic shield ulcer.
d. exposure keratopathy.
e. granular stromal dystrophy.

386. Each of the following statements about therapy for herpes simplex stromal keratitis is true except:

a. Patients treated with an appropriate regimen of topical steroid will improve more quickly than those without such treatment.
b. The total length of a steroid taper is prolonged, often exceeding 10 weeks in length.
c. Up to one in five patients who are not treated with topical steroid will recover as quickly as patients so treated.
d. Topical trifluridine appears to shorten the duration and lessen the severity of herpes simplex stromal keratitis.
e. Prompt initiation of topical steroids offers no long-term visual advantage in herpes simplex stromal keratitis.

387. Which single-dose oral agent has been shown to be as effective as 7 weeks of topical tetracycline for the treatment of active trachoma?

a. erythromycin estolate.
b. ceftazidime.
c. azithromycin.
d. norfloxacin.
e. rifampin.

388. Following incisional keratotomy, radial corneal wounds regain up to what percentage of unincised corneal tensile strength?

a. 10%.
b. 25%.
389. Which one of the following is not considered a risk factor for the development of bacterial keratitis?
   a. drug abuse.
   b. keratoconus.
   c. diabetes mellitus.
   d. dry eye syndrome.
   e. topical steroid use.

390. Which one of the following conditions is not seen more commonly in patients with keratoconus:
   a. Down’s syndrome.
   b. atopic dermatitis.
   c. Ehlers-Danlos syndrome.
   d. diabetes mellitus, type 1.
   e. vernal keratoconjunctivitis.

391. One potential complication of topical ciprofloxacin treatment of infectious corneal ulcers is:
   a. decreased corneal sensitivity.
   b. chronic corneal crystalline deposits.
   c. chronic dry eye syndrome.
   d. acute meibomitis.
   e. acute dacryocystitis.

Matching

392. Match the Gram-positive organisms numbered below with staining, culture, and clinical characteristics lettered below (note that each organism may be used more than once).

   a. grow in pairs of lancet shaped cocci with alpha hemolysis.
   b. bacterium that resembles filamentous fungi and is vitamin K– dependent.
   c. a slender, curved rod with pseudohyphae or beading.
   d. cocci in chains or rarely clusters; the most resistant Gram-positive coccus.
   e. pleomorphic bacilli resembling Chinese letters.
   f. cocci in clusters, producing coagulase.
   g. bacilli found in soil and vegetation.
   h. organism most commonly responsible for traumatic endophthalmitis.
   i. cocci in clusters; do not produce coagulase.
   j. organism frequently associated with chronic recurrent canaliculitis.
   k. alpha-hemolytic cocci associated with crystalline keratopathy.
   l. frequently associated with chronic recurrent dacryocystitis in patients with nasolacrimal stenosis.
   m. an acid-fast filamentous bacterium confused with fungus; associated with multifocal abscesses.
   n. organism most frequently associated with hypersensitivity marginal keratitis.

393. Match the Gram-negative organisms numbered below with staining, culture, and clinical characteristics lettered below.

   a. Gram-negative rod growing in gray–blue colonies with a sweet grape aroma.
   b. dumbbell- or boxcar-shaped diplobacilli, always end-to-end.
   c. associated with central nervous system (CNS) infections in adolescents and young adults.
   d. cocci in chains or rarely clusters; the most resistant Gram-positive coccus.
   e. pleomorphic bacilli resembling Chinese letters.
   f. cocci in clusters, producing coagulase.
   g. bacilli found in soil and vegetation.
   h. organism most commonly responsible for traumatic endophthalmitis.
   i. cocci in clusters; do not produce coagulase.
   j. organism frequently associated with chronic recurrent canaliculitis.
   k. alpha-hemolytic cocci associated with crystalline keratopathy.
   l. frequently associated with chronic recurrent dacryocystitis in patients with nasolacrimal stenosis.
   m. an acid-fast filamentous bacterium confused with fungus; associated with multifocal abscesses.
   n. organism most frequently associated with hypersensitivity marginal keratitis.

   1. Propionibacterium acnes.
   2. Bacillus cereus.
   3. Actinomyces.
Questions

d. associated with hyperacute purulent conjunctivitis in neonates and adults.
e. associated with a fulminant sclerokeratitis.
f. may grow as satellites of *Staphylococcus aureus* on blood agar.
g. Gram-negative rod requiring 5% carbon dioxide, fermenting glucose only.
h. pleomorphic rod requiring hemin and nicotinamide-adenine dinucleotide (NAD), associated with otitis and meningitis, and potentially fatal septicemia in children.
i. commonly associated with angular blepharitis and corneal ulcer in alcoholics.
j. enterobacterium that overgrows culture plate quickly.

394. Match the clinical manifestations lettered below with their viral etiology.

<table>
<thead>
<tr>
<th>Clinical Manifestation</th>
<th>Viral Etiology</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. pustular rash of the eyelids and conjunctiva after vaccination.</td>
<td>1. adenovirus.</td>
</tr>
<tr>
<td>b. leads to follicular conjunctivitis in poultry handlers.</td>
<td>2. molluscum contagiosum.</td>
</tr>
<tr>
<td>c. causes severe membranous conjunctivitis and punctate keratitis.</td>
<td>3. vaccinia.</td>
</tr>
<tr>
<td>d. a nonspecific conjunctivitis and benign punctate keratitis universally accompanies the exanthem.</td>
<td>4. rubeola.</td>
</tr>
<tr>
<td>e. may be associated with a secondary toxic follicular conjunctivitis, particularly inferiorly.</td>
<td>5. Newcastle’s disease.</td>
</tr>
</tbody>
</table>

396. Match the special equipment needed for corneal scraping lettered below with the proper organisms numbered below (note that there may be more than one correct answer for each lettered item).

<table>
<thead>
<tr>
<th>Special Equipment</th>
<th>Organisms</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. thioglycolate broth.</td>
<td>1. Haemophilus.</td>
</tr>
<tr>
<td>b. Giemsa stain.</td>
<td>2. anaerobic organisms.</td>
</tr>
<tr>
<td>c. blood agar with <em>Staphylococcus aureus</em> cultures.</td>
<td>3. filamentous fungi.</td>
</tr>
<tr>
<td>d. Sabouraud’s agar.</td>
<td>4. Mycobacteria.</td>
</tr>
<tr>
<td>e. Ziehl-Neelsen stain.</td>
<td>5. Acanthamoeba.</td>
</tr>
</tbody>
</table>

395. Match the clinical characteristics lettered below with the roundworms numbered below.

<table>
<thead>
<tr>
<th>Clinical Characteristic</th>
<th>Roundworms</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. generally contracted by ingestion of fecally contaminated soil or raw vegetables.</td>
<td>1. <em>Onchocerca volvulus</em>.</td>
</tr>
<tr>
<td>b. generally contracted by ingestion of raw pork.</td>
<td>2. <em>Trichinella spiralis</em>.</td>
</tr>
<tr>
<td>c. generally spread by the black fly vector, breeding in rivers.</td>
<td>3. <em>Toxocara</em>.</td>
</tr>
</tbody>
</table>
g. Löwenstein-Jensen agar.

h. nonnutrient agar with Escherichia coli overgrowth.

i. Gomori-methenamine silver stain.

397. Arrange the antivirals listed below in order of decreasing topical toxicity (1 being the most toxic and 4 being the least toxic).

a. idoxuridine.
b. acyclovir.
c. trifluridine.
d. vidarabine.

398. Arrange the various glucocorticoid agents listed below in order of decreasing systemic potency (1 being the most potent and 5 being the least potent).

a. methylprednisolone.
b. hydrocortisone.
c. cortisone.
d. prednisone.
e. dexamethasone.

399. Assign the distinguishing features lettered below with the type of papillomas numbered below.

a. usually seen in older adults.
b. usually seen in children or adolescents.
c. rarely bilateral.
d. occasionally bilateral.
e. rarely multiple.
f. occasionally multiple.
g. usually on palpebral conjunctiva.
h. generally on bulbar conjunctiva.
i. incomplete excision generally associated with solitary recurrence.
j. incomplete excision associated with multiple recurrences.
k. may be associated with fulminant conjunctivitis.
l. rarely associated with conjunctivitis.
m. may spontaneously improve.
n. rarely improve spontaneously.

400. Match the histologic features lettered below with the various layers of the epidermis.

a. flattened, elongated squamous cells with multiple keratohyaline granules.
b. cuboid or columnar cells with large nuclei aligned in a single row above the basement membrane.
c. multiple eosinophilic layers of protein with no cellular details visible.
d. polygonal cells with numerous hairlike filamentous connections between cells.

401. Match the mechanics of secretion lettered below with the glands numbered below.

a. secretion by simple exocytosis.
b. secretion by release of entire cellular contents (disruption of cell).
c. secretion by pinching or budding off from a portion of cellular cytoplasm.

402. Match the eyelid glands lettered below with the modes of secretion numbered below.

a. meibomian glands.
b. glands of Zeis.
c. Moll’s glands.
d. typical sweat glands.

403. Match the epithelial growth features lettered below with the appropriate histopathologic terminology numbered below.

a. individual cellular enlargement.

1. squamous cell carcinoma.
281

Questions

pleomorphism, prominent hyperchromasia, and abnormal mitoses.

b. loss of maturational order and normal layering of the epithelium.

c. severe atypia and abnormal polarity.

d. full-thickness dysplasia.

e. invasion of epithelial basement membrane by dysplastic squamous epithelium.

f. keratinization and formation of skinlike features by mucosal epithelium.

404. Match the cellular features lettered below with the appropriate type of nevus numbered below.

a. diffuse congenital deep dermal nevus of the periocular skin.

b. nests of nevus cells in the basal epithelial layer.

c. deeply located dermal nevus present at birth with very little elevation.

d. compound nevus of childhood with bizarre cellular components but no malignant potential.

e. deep dermal nevus present at birth with many dendritic nevus cells.

405. Match the terms for conjunctival and/or corneal intraepithelial neoplasia lettered below with the histopathologic findings numbered below.

a. mild dysplasia.

b. moderate dysplasia.

c. severe dysplasia.

d. carcinoma in situ.

e. microinvasive carcinoma.

f. squamous cell carcinoma.

1. atypia and loss of polarity involving the basal two thirds of the epithelium.

2. full-thickness atypia and loss of polarity with limited areas of basement membrane invasion.

3. atypia and loss of polarity involving the basal one third of the epithelium.

4. full-thickness atypia and loss of polarity with no remaining normal underlying architecture.

5. full-thickness atypia and loss of polarity of the epithelium, limited by intact basement membrane.

406. Match the clinical characteristics lettered below with the correct form of marginal degeneration numbered below.

a. red, inflamed eyes.

b. white, quiet eyes.

c. prominent lipid degeneration.

d. undermined edge of corneal stroma.

e. generally begins superiorly.

f. may begin anywhere peripherally.

g. spontaneous perforation possible.

h. spontaneous perforation unlikely.

i. associated with systemic collagen vascular disease.

j. may lead to disabling against-the-rule astigmatism.

k. may respond to immunosuppressives.

407. Match the clinical characteristics lettered below with the classifications of corneal disorders numbered below.

a. associated with previous or concurrent disorders, diseases, or aging.

b. generally associated with family history.

c. generally begins centrally.

1. dystrophies.

2. degenerations.

3. both.

4. neither.
d. generally begins peripherally.
e. bilaterally symmetric.
f. unilateral or asymmetric.
g. progressive over a period.
h. may be stationary.
i. may cause decreased vision.
j. may involve deposition of material not normally found in the cornea.

408. Match the clinical findings of keratoconus lettered below with the appropriate eponymic designation numbered below.

<table>
<thead>
<tr>
<th>Clinical Finding</th>
<th>Eponymic Designation</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. conical reflection of light from a temporal light source on the nasal cornea.</td>
<td>1. the Munson sign.</td>
</tr>
<tr>
<td>b. pointed deviation of the lower eyelid in downgaze.</td>
<td>2. Vogt striae.</td>
</tr>
<tr>
<td>c. iron deposition in basal epithelial cells at the inferior aspect of the cone.</td>
<td>3. the Rizzuti sign.</td>
</tr>
<tr>
<td>d. vertical oblique lines within the corneal stroma that typically disappear with digital pressure on the eye.</td>
<td>4. Fleischer’s ring.</td>
</tr>
</tbody>
</table>

409. Match the anatomic and clinical characteristics lettered below with the correct tear film layer numbered below.

<table>
<thead>
<tr>
<th>Clinical Characteristic</th>
<th>Tear Film Layer</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. the innermost layer (closest to the cornea).</td>
<td>1. aqueous phase.</td>
</tr>
<tr>
<td>b. permits diffusion of oxygen and metabolites to the cornea readily.</td>
<td>2. lipid phase.</td>
</tr>
<tr>
<td>c. retards tear evaporation.</td>
<td>3. mucus phase.</td>
</tr>
<tr>
<td>d. constitutes the bulk of tear volume (&gt;90%).</td>
<td></td>
</tr>
<tr>
<td>e. outermost layer (exposed to air).</td>
<td></td>
</tr>
<tr>
<td>f. secreted by the conjunctiva.</td>
<td></td>
</tr>
<tr>
<td>g. the middle layer.</td>
<td></td>
</tr>
<tr>
<td>h. secreted by the glands of Krause and Wolfring.</td>
<td></td>
</tr>
<tr>
<td>i. permits smooth, even distribution of tear film over corneal epithelium.</td>
<td></td>
</tr>
<tr>
<td>j. secreted by meibomian glands.</td>
<td></td>
</tr>
</tbody>
</table>

410. Match the staining characteristics lettered below with the stains numbered below.

<table>
<thead>
<tr>
<th>Staining Characteristic</th>
<th>Stain Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. adheres well to mucus.</td>
<td>1. rose bengal.</td>
</tr>
<tr>
<td>b. stains mucus less vividly.</td>
<td>2. fluorescein.</td>
</tr>
<tr>
<td>c. adheres firmly to exposed basement membrane.</td>
<td>3. both.</td>
</tr>
<tr>
<td>d. adheres to dead or devitalized cells.</td>
<td>4. neither.</td>
</tr>
<tr>
<td>e. diffuses into intercellular epithelial spaces and stroma.</td>
<td></td>
</tr>
<tr>
<td>f. does not diffuse into intercellular spaces.</td>
<td></td>
</tr>
<tr>
<td>g. stain of choice for tear deficiency states.</td>
<td></td>
</tr>
<tr>
<td>h. stain of choice for epithelial defects.</td>
<td></td>
</tr>
<tr>
<td>i. stain of choice for herpes simplex epithelial keratitis.</td>
<td></td>
</tr>
<tr>
<td>j. requires dilution in aqueous solution for visualization.</td>
<td></td>
</tr>
</tbody>
</table>

411. Match the associated clinical findings lettered below with the various disorders associated with enlarged corneal nerves numbered below.

<table>
<thead>
<tr>
<th>Clinical Finding</th>
<th>Disorder Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. pigmentary retinopathy, hearing loss, mental retardation.</td>
<td>1. old age.</td>
</tr>
<tr>
<td>b. optic nerve glioma, multiple iris nevi.</td>
<td>2. neurofibromatosis, type I.</td>
</tr>
<tr>
<td>c. sclerosing interstitial keratitis (IK), anterior uveitis, and iris granulomas.</td>
<td>3. congenital glaucoma.</td>
</tr>
<tr>
<td>d. enlarged corneal diameter, horizontal breaks in Descemet’s membrane, and dislocated lens.</td>
<td>4. multiple endocrine neoplasia (MEN), type III.</td>
</tr>
<tr>
<td>e. scaly, tight skin.</td>
<td>5. keratoconus.</td>
</tr>
<tr>
<td>f. hypertension, hypocalcemia, mucocutaneous neuromas.</td>
<td>6. Hansen’s disease.</td>
</tr>
<tr>
<td>g. myopic astigmatism, the Munson sign.</td>
<td>7. Refsum’s disease.</td>
</tr>
<tr>
<td>h. Hudson-Stähli line, cataract, Hassall-Henle warts.</td>
<td>8. ichthyosis.</td>
</tr>
</tbody>
</table>
Answers

1. b. There are five cardinal features of inflammation: rubor (redness), calor (warmth), dolor (pain), tumor (swelling), and loss of function. Many forms of inflammation (e.g., type I hypersensitivity) do not cause irreversible structural damage to involved tissues.

2. True. The final common pathway can be the same—inflammation-mediated tissue effects. An immune agent, by definition, can elicit an anamnestic response and cause inflammation.

3. False. Polymorphonuclear leukocytes (PMNs) are far more destructive. They are the “kamikaze” cells of inflammation. Macrophages are critical for the initiation of the afferent immune response, as well as “clean-up” duties.

4. b. Conjunctival papillae form whenever there is conjunctival swelling of any cause in certain areas. The tarsal and limbal conjunctiva are unique because their subepithelial substantia propria contains fibrous tissue septa that interconnect to form polygonal lobules with a central vascular bundle. Any inflammation in these regions will result in papillae. Papillae <1 mm in diameter are entirely nonspecific. When papillae are >1 mm in diameter (giant papillae), they are more specific (see answer 10). Conjunctival follicles represent focal lymphoid aggregates in the substantia propria. These are also more specific than papillae (see answers 19 and 20). Chemosis represents subepithelial edema and is nonspecific.

5. c. See answer 4.

6. b. This is essentially the morphologic definition of a conjunctival papilla.

7. a. See answer 4.

8. c. Most young people with healthy eyes will have small conjunctival papillae along the superior margin of the upper tarsus (remember that, when everted, this will be the lower edge of the tarsus).

9. False. The cutoff for giant papillae is a diameter of 1 mm.

10. b. The four disorders associated with true giant papillae are vernal and atopic keratoconjunctivitis, and contact lens–related and prosthesis-related giant papillary conjunctivitis (GPC). Trachoma has a more pronounced follicular response (although papillae may be seen as a nonspecific sign).

11. d. Horner–Trantas dots are pathognomonic for vernal keratoconjunctivitis. Herbert’s pits are punched-out limbal lesions representing necrotic limbal follicles associated with trachoma. The von Arlt line is linear subconjunctival scarring seen on the upper tarsus in “burned-out” trachoma. Fuchs’ spots are areas of punched-out chorioretinal atrophy in the macula of high myopes. Cogan’s patches are focal areas of scleral compaction (dellen) anterior to horizontal rectus insertions in the elderly patients.

12. a. See answer 11.

13. False. As a rule, the giant papillae in vernal keratoconjunctivitis are more solid appearing, with sharper margins and greater elevation. In atopic keratoconjunctivitis, the giant papillae are less well defined, less elevated, and creamier in appearance.

14. True. The papillae in contact lens–related giant papillary conjunctivitis (GPC) may last for several months or years after the symptoms are controlled.

15. d. Itching and discharge may be seen in both contact lens–related giant papillary conjunctivitis (GPC) and viral conjunctivitis. Likewise, in both cases, the bulbar conjunctiva may be mildly to severely injected. In GPC, the inferior fornix is generally the least involved, whereas in viral conjunctivitis, it tends to be the most severely involved.


17. d. Trachoma is nearly always a disease of the superior conjunctiva. This is in stark contrast to inclusion conjunctivitis, another chlamydial disease, which is more severe inferiorly. Toxic follicular conjunctivitis secondary to medications and viral keratoconjunctivitis are also more severe inferiorly.

18. b. Typically, any disease condition lasting longer than 1 month would be termed “chronic.”

19. d. Trachoma is generally insidious. By the time the patient is seen by the ophthalmologist, there is usually a history of red eyes for several months (or of several recurrences).

20. a. Although the subepithelial keratitis may last for several months, the “red eye” (conjunctivitis) of epidemic keratoconjunctivitis (EKC) is nearly always gone after 3 to 4 weeks.

21. c. Neonatal inclusion conjunctivitis may be pseudomembranous or membranous, but this is rare in adults. A pseudomembrane or membrane is a plaque of inflammatory debris—that is, mucus, fibrin, inflammatory cells, and/or hemorrhage—which can be stripped off a mucosal surface. The distinction between a true membrane and a pseudomembrane is that, when a true membrane is stripped, there is bleeding. This is because a true membrane is fused with and incorporated into the mucosal epithelium. Clinically, pseudomembranes and true membranes have similar appearances with a nearly identical differential diagnosis.

22. e. This is a feature that may help in differentiating between the two type I hypersensitivity causes of giant papillary conjunctivitis (GPC). Atopic keratoconjunctivitis rarely comes with a (pseudo)membrane, whereas vernal keratoconjunctivitis may.

23. False. Typical microcystic corneal epithelial edema is in the intercellular space and results from an imbalance between corneal turgescence (swelling due to intraocular
24. False. Two pathophysiologic factors are felt to be necessary for the development of corneal epithelial filaments—increased mucus production along with increased, deranged epithelial turnover. The filaments themselves are composed of mucus and desquamated epithelial cells.

25. d. It would be unusual for filaments to be seen in ocular cicatricial pemphigoid (OCP) because mucus production is generally diminished in this disorder.

26. c. The depositions that form the horizontal band across the exposed portions of the cornea generally start peripherally and proceed centrally, but this is not invariable. The material that forms the deposit is most often calcium salts, but it may also be composed of urate salts.

27. c. For cells to enter the corneal stroma from the aqueous humor, there must be a breach in the endothelium.

28. b. Note that “granulomatous” keratic precipitates (KPs) may have a characteristic greasy, mutton-fat appearance, without actually being granulomas themselves. Macrophages are responsible for the clinical appearance.

29. d. See answer 30—Photorefractive keratectomy (PRK) can have significant postoperative pain. PRK offers a reduced incidence of stromal haze and more rapid epithelial healing.

30. c. Endophthalmitis rates are higher with laser-assisted in situ keratomileusis (LASIK) than with photorefractive keratectomy (PRK). LASIK does offer a reduced incidence of stromal haze and more rapid epithelial perforation.

31. c. See answer 30—Photorefractive keratectomy (PRK) can have significant postoperative pain. PRK offers a reduced incidence of endophthalmitis and corneal perforation.

32. a. On Gram stain, mycobacteria may resemble corynebacteria—small pleomorphic Gram-positive rods resembling Chinese letters. Acid-fast staining will highlight the mycobacteria, not diphtheroids.

33. b. Many mycobacteria will grow well on blood agar, but the Löwenstein-Jensen medium is designed for more fastidious mycobacteria.

34. a. The standard antitymocellular agents for Mycobacterium tuberculosis are isoniazid, ethambutol, and rifampin. Aminoglycosides are also effective, particularly against some of the atypical mycobacteria.

35. True. Wet-field preparations, viewed with dark-field illumination, must be performed promptly.

36. False. The fluorescent treponemal antibody-absorption (FTA-ABS) is both more specific and more sensitive than Treponema pallidum immobilization (TPI), and it is also easier to perform.

37. True. These two, fluorescent treponemal antibody-absorption (FTA-ABS) and microhemagglutination of Treponema pallidum (MHA-TP), are the closest to a “gold standard” for syphilis testing.

38. b. The Venereal Disease Research Laboratory (VDRL) and rapid plasma reagin (RPR) tests reflect treponemal infection and revert to normal when treated. The value of these tests is primarily to monitor response to treatment because they are less sensitive and specific. The fluorescent treponemal antibody (FTA) and microhemagglutination of Treponema pallidum (MHA-TP) offer greater sensitivity and specificity and do not normalize with treatment. They are lifelong markers of previous or active treponemal infection.

39. False. The organism responsible for Lyme disease, Borrelia burgdorferi, is notoriously difficult to culture. Serology remains superior to culture techniques.

40. c. Therapy for ocular mycosis is heavily dependent on geographic locale.

41. c. Fungi may be classified into three groups:

(i) molds, or filamentous fungi (e.g., Fusarium, Penicillium); (ii) yeasts (e.g., Candida); and (iii) dimorphs (may exist as either mold or yeast, depending on environment).

42. d. Cryptococcal keratitis is distinctly uncommon.

43. False. Molds are most common, yeasts are second most common, and dimorphs (e.g., Histoplasma, Coccidiodites) are rare.

44. b. Fungi are typically hearty.

45. b. Most periocular herpes infections are type 1. Antibody titers may be helpful in cases of primary disease, but not secondary (reactivation) cases.

46. True. These are called “Lipschutz bodies.”

47. True. This is known as the “Tzanck prep” (remember: “Tzanck God, I don’t have herpes.”)

48. True. Papanicolaou (PAP) smears and Giemsa stains from the two viral infections may be indistinguishable.

49. a. There are five anti–Epstein-Barr virus (EBV) antibodies: (i) viral capsid antigen (VCA)-IgG, (ii) VCA-IgM, (iii) early antigen-diffuse (EA-D), (iv) EA-R (early antigen–restricted), and (v) Epstein-Barr nuclear antigen (EBNA). Only EBNA does not peak in the first 6 to 8 weeks of infection.

50. c. Viral capsid antigen (VCA)-IgG and Epstein-Barr nuclear antigen (EBNA) provide lifelong evidence of Epstein-Barr virus (EBV) infection.

51. d. The rates of corneal haze are similar to laser-assisted in situ keratomileusis (LASIK). All of the other answers are true. Photorefractive keratectomy (PRK) with the concomitant use of mitomycin C also appears to reduce the incidence of postoperative corneal haze.

52. d. Like bacteria (and unlike viruses), chlamydiae have both DNA and RNA, replicate by binary fission, have lipopolysaccharide cell walls, and respond to certain
Answers

antibiotics. Unlike bacteria, chlamydiae do not possess all organelles and require a host cell for replication.

55. False. Like Lyme disease, culture for chlamydial disorders is difficult. Detection of antigen in tissue scrapings using immunofluorescent techniques is the diagnostic standard.

56. (c). Treatment must consist of oral agents (erythromycin, tetracycline, or rifampin) for at least 21 days. Resistance to these agents is becoming increasingly common, and macrolides (such as azithromycin) are also clearly of benefit in patients with ocular chlamydial infection.

57. (b). Serotypes A, B, and C cause trachoma. Types D through K cause inclusion conjunctivitis, and LGV1 through 3 cause lymphogranuloma venereum.

58. (c). As with chlamydiae, culture of Acanthamoeba is difficult.

59. (a). More than 50% of flap folds occur within the first day, and >90% of flap folds occur within the first week postoperatively. Not all folds need to be repaired, but visually significant folds should be repaired within 24 hours.

60. (a). Only Phthirus pubis (crab louse) has been associated with lice infestation. Pediculus capitis (head louse) resides only in the scalp. Therefore, ocular "pediculosis" is a misnomer (the better term is "phthiriasis").

61. (a). Suffocation techniques helpful with phthiriasis are not effective for Demodex infestation.

62. (d).

63. False.

64. True. This is in distinction to adult forms of the disease.

65. (c). The contact holmium-YAG laser is no longer available because of its unpredictable effects and because of postoperative regression. All of the other surgical procedures have been U.S. Food and Drug Administration (FDA) approved to treat certain degrees of hyperopia.

66. (b). This is useful for suspected chlamydial disease.

67. (c).

68. (d). Topical acyclovir is available in Europe for herpes simplex virus (HSV) and herpes zoster virus (HZV) keratitis.

69. (b). Idoxuridine and vidarabine are available in ointment preparations, which can be useful for children.

70. (c). Idoxuridine and vidarabine inhibit DNA synthesis by acting as false nucleic acids (analogs). Trifluridine inhibits thymidylate synthetase, a herpes-specific enzyme. Acyclovir, a guanosine analog, is activated only by thymidine kinase, another virus-specific enzyme. Both thymidylate synthetase and thymidine kinase are critical for herpetic DNA synthesis.

71. True.

72. True. Antivirals have different mechanisms of action, and resistance to one does not indicate resistance to all.

73. (c). Acyclovir is activated by viral thymidine kinase and is relatively nontoxic to mammalian cells. Trifluridine (Viroptic) inhibits another virus-specific enzyme, thymidylate synthetase. At high concentrations, it may act as a false nucleoside and affect human DNA synthesis as well.

74. False. Trifluridine is more effective for herpetic ulcers.

75. (e). All of the answers are correct. Radial incisions can sometimes be used to correct postoperative astigmatism.

76. (c). The hypersensitivity reaction to idoxuridine is so strong that it may induce lymphadenopathy.

77. (d).

78. (b). Amphotericin has less predictable activity against filamentous species than natamycin.

79. (b). Fluocytosine commonly induces acquired fungal resistance.

80. (a).

81. (c). Polymers are insoluble in water and have systemic toxicity by binding to renal tubular cells and erythrocytes.

82. (e). Therefore, liver enzymes should be monitored during long-term ketoconazole therapy.

83. (c). Natamycin and amphotericin are polyenes. Fluocytosine is a pyrimidine, and miconazole is an imidazole.

84. (a). Ketoconazole, miconazole, and clotrimazole are imidazoles. Nystatin is a polyene.

85. (a). Although varied, treatment regimens for Acanthamoeba keratitis have included neomycin and propamidine. For resistant, aggressive cases, topical imidazoles also have been used. Effect of pentamidine on Acanthamoeba keratitis is unknown at present.

86. (c). Polymorphonuclear leukocytes (PMNs) release hydrolytic enzymes that denature protein and cause tissue necrosis.

87. (c). Intracorneal rings (ICR) are currently being used for the treatment of low myopia. They are also being studied as a potential treatment of keratoconus. Corneal perforation is a potential complication of the procedure but occurs less frequently than decreased corneal sensation.

88. True. Suspensions must be shaken to be effective. Acetate preparations (suspensions) are more potent than phosphate preparations.

89. (c). Although drug levels in the cornea do not necessarily correlate with antiinflammatory activity, studies involving rabbit corneas indicate that prednisolone acetate has the greatest antiinflammatory activity.

90. (e). Medrysone has weak antiinflammatory activity but the lowest risk of intraocular pressure (IOP) elevation.

91. (False). Local steroids do not reactivate latent virus, but caution should be exercised because intermittent shedding of the virus from the trigeminal ganglion (unrelated to steroid use) may coincide with steroid use and exacerbate subsequent disease.

92. (b). In active dendritic epithelial keratitis, live virus proliferates but usually does not cause permanent ocular damage, and steroids are not indicated. Also, steroids
Steroids are usually used only in acute exacerbations.

Cultures may help direct the antibiotic regimen.

Collarettes indicate contact lens-induced papillary conjunctivitis is felt.

The incidence of giant papillary conjunctivitis (GPC) is greatest with soft contact lenses (SCL), followed by rigid gas-permeable (RGP) lenses, and then polymethylmethacrylate (PMMA) lenses.

Contact lens-induced papillary conjunctivitis is felt to be secondary to immune system interactions with mucus or materials absorbed on the lens. Increased frequency of enzyme treatments may help. Heat sterilization “bakes” the irritating deposits onto the lens, so chemical methods (avoiding thimerosal) are preferred.

Collarettes indicate *Staphylococcus*; scurf indicates seborrhea. Sleeving of the lashes is a sign of *Demodex* infestation.

Staphylococcal blepharocconjunctivitis may be asymptomatic, or may present with red eyes and symptoms of ocular inflammation such as superficial punctate keratitis and subsequent epiphora and photophobia.

False. Cultures may help direct the antibiotic regimen because resistant *Staphylococcus* species are encountered in intractable cases.

Steroids are usually used only in acute exacerbations and only with concurrent antibiotic therapy.

During treatment, episodic intraocular pressure (IOP) measurement and slit-lamp examination (for cataracts) are essential.
Answers

127. b. Long-term corticosteroid use should be avoided in patients with limbal vernal keratoconjunctivitis because of possible side effects. Topical cromolyn sodium is a more useful adjunct for long-term management. Palpebral vernal is more commonly associated with shield ulcers.

128. a. Although conjunctival scraping in atopic disease usually reveals fewer eosinophils than in vernal keratoconjunctivitis, their mere presence is not a useful differentiating factor.

129. d. By definition, atopic keratoconjunctivitis is an IgE-mediated process that occurs in patients who have atopic dermatitis (eczema).

130. b. Although posterior uveitis may occur in Reiter’s syndrome, including cystoid macular edema, a severe retinal vasculitis is not a feature of this syndrome.

131. c. This history is classic for the floppy eyelid syndrome. Spontaneous eyelid eversion occurs during sleep with minimal pillow (or bedsheet) contact. The condition is treated by mechanically protecting the involved eye (taping, shield). If these conservative measures fail, horizontal lid tightening procedures may be attempted.

132. True. Noninfectious inflammatory marginal keratitis tends to remain peripheral.

133. c. Trachoma is the most common cause of irreversible blindness in the world, but not in the United States.

134. True. Genital herpes is usually type 2; however, type 1 herpes may affect the genitalia after orogenital sexual activity.

135. True. Most ocular herpetic infections are caused by type 1 herpes; however, type 2 herpes may cause ocular infection following orogenital sexual activity.

136. b. Type 2 herpes usually resides latently in spinal ganglia. The gasserian (or trigeminal) ganglion, located in Meckel’s cave, contains the cell bodies of the trigeminal nerve.

137. a. Vesicular blepharitis may occur in either primary or recurrent herpes simplex virus (HSV) infection. Dendrites are fleeting in primary inflammation. Lymphadenopathy is rare in secondary disease.

138. False. Decreased corneal sensation is sometimes difficult to detect and certainly not specific for herpetic ulcer.

139. b. Topical antiviral medication may require up to 21 days to work. Topical steroids are contraindicated in active epithelial herpetic disease.

140. a. Overtreatment with topical antivirals is common and should be avoided. Bacterial superinfection is not a complication of overtreatment.

141. a. This ulcer is typical of a “metaherpetic” lesion and does not reflect epitheliitis. Corneal epithelial healing is impaired by decreased sensation, stromal inflammation, and antiviral toxicity.

142. False. Disciform keratitis is a description only and may be seen with herpes zoster, mumps, or chemical injury (see also answer 145).

143. a. Peripheral anterior synechiae occur commonly following anterior uveitis due to herpes simplex virus (HSV) but are not usually seen with disciform keratitis.

144. a. The phenomenon is identical to the immunoprecipitate formed in the Ouchterlony gel.

145. b. The differential diagnoses for disciform keratitis and Wessely’s rings are very similar (see also answer 142).

146. False. Herpetologists feel that either (i) the visual axis must be involved or (ii) neovascularization must be progressing before steroids are indicated.

147. True. Stromal necrosis can be devastating in interstitial keratitis (IK).

148. e. All of the answers listed are justifications for acyclovir treatment.

149. False. The Hutchinson sign (vesicles at the tip of the nose) indicates a high probability of ocular involvement with herpes zoster virus (HZV).

150. False. Reinfection has been reported to lead to typical herpes zoster ophthalmicus (HZO).

151. False. Typically, the ophthalmologist is left handling ocular complications long after the rash is gone.

152. c. Some experts feel that postherpetic neuralgia is less severe if the reactivation is treated promptly with acyclovir; others disagree. Prednisone is generally delayed until virus replication has ceased. Cimetidine is controversial and has fallen from favor.

153. a. Herpes zoster “dendrites” (pseudodendrites) are typically smaller and less branching than their simplex counterparts.

154. e. Drug abuse, particularly crack cocaine smoking, is associated with contamination and damage of corneal epithelium. Diabetes mellitus is associated with defective epithelial adherence, as is aging (which also comes with relative hyposthesia).

155. c.

156. True.

157. True.

158. c. Contaminated water supplies, particularly hot tubs and homemade contact lens saline solution, are frequently implicated. Contact lens use alone is a risk factor.

159. a. See answer 158.

160. c. The organism classically causes a radial keratoneuritis, with pain that is out of proportion to the findings.

161. a. Fortunately, herpetic keratitis does not appear to increase the risk of subsequent fungal keratitis.

162. b. Leprosy is a cause of interstitial keratitis (IK) worldwide, but is uncommon in the United States.

163. True.

164. True.

165. a. Patients are also at risk of aortitis with dissecting aneurysms.

166. c. The conjunctiva is usually normal in Thygeson’s disease.

167. b. About one half of patients with superior limbic keratoconjunctivitis (SLK) have some form of thyroid
9: Cornea, External Disease, and Refractive Surgery

disease. Treatment of the thyroid disorder, however, has little effect on the SLK. A recent study suggests that orbital decompression for thyroid optic neuropathy is required more frequently in patients with thyroid-associated SLK.

168. a. Most experts now agree that steroids are not effective therapy for superior limbic keratoconjunctivitis (SLK). Surgical resection of the superior bulbar conjunctiva is definitive.

169. False. Discontinuation of contact lenses, at least temporarily, is therapeutic for contact lens-related superior limbic keratoconjunctivitis (SLK).

170. d.

171. False. This protein-centered effect is typical of acid burns. Alkaline substances saponify cell membranes.

172. True. This is in distinction to alkali burns, which propagate rapidly.

173. True. This reflects greater intraocular penetration and damage to intraocular filtration structures by alkaline substances.

174. c. The ocular surface must be irrigated until pH is normal (6.8 to 7.2). Corneal epithelial loss, clarity, and limbal ischemia (whitening) are critical early prognostic factors.

175. c. Retained foreign bodies represent a hazardous depot of alkaline material. These must be removed immediately upon detection.

176. c. Epithelial continuity is essential for the prevention of infections, inflammation, and scarring. Unfortunately, severe chemical injuries retard healing.

177. False. Avoiding drugs that are toxic to the epithelium (e.g., neomycin, tobramycin, gentamicin) is very important because these agents will inhibit corneal epithelial healing.

178. False. If corticosteroids are to be used, they should be restricted to the first 5 to 10 days. They are useful in reducing corneal and intraocular inflammation and helpful in combating the formation of symblepharon. However, corticosteroids may enhance collagenase-induced corneal melting, which often begins 1 to 2 weeks after the injury.

179. True. Repair of cicatricial entropion and autologous conjunctival transplantation or mucous membrane grafts may be helpful in late therapy for severe chemical injury. Penetrating keratoplasty (PKP) has increased success if the procedure is performed 1 to 2 years after the chemical injury.

180. d. Episcleritis is a recurrent, transient, self-limited, and usually nonspecific disease of young adults. Simple episcleritis is sectoral in 70% and generalized in 30% of cases. Pingueculae may show a distinct form of episcleritis with local superficial inflammation similar to nodular episcleritis. Episcleritis rarely progresses to scleritis; however, episcleritis nearly always accompanies scleritis. Two thirds of patients with episcleritis have recurrences, and the condition is usually self-limited without treatment. The condition can be treated with topical steroids and/or oral nonsteroidal agents.

181. a. Generally, episcleritis causes minimal pain, whereas scleritis is moderately to severely painful. Episcleritis is usually of rapid onset as opposed to scleritis, which is usually gradual, over days. In scleritis, the scleral (deep episcleral) plexus is immobile and bluish-red in color; episcleritis appears salmon pink. Scleral vessels do not blush with phenylephrine, as do those in episcleritis. Lastly, scleritis is frequently seen with systemic connective tissue disorders; this is not true of episcleritis.

182. a. Diffuse anterior scleritis is the most benign form of scleritis and is associated with the least severe systemic conditions.

183. c. Necrotizing scleritis with inflammation is the most destructive form of scleritis. Sixty percent of affected patients develop complications (in addition to scleral thinning) and 40% suffer visual loss. These patients are at risk for mortality, secondary to associated autoimmune disease.

184. d. Patients with scleromalacia perforans (necrotizing scleritis without signs of inflammation) often have long-standing rheumatoid arthritis.

185. e. Patients with posterior scleritis present with proptosis, pain, visual loss, and occasionally motility restrictions. Choroidal folds, papilledema, exudative retinal detachment, and even angle-closure glaucoma are possible. Histopathologically, posterior scleritis is a granulomatous process.

186. False. Posterior uveitis occurs in virtually all patients with posterior scleritis and is not uncommon in patients with anterior scleritis. Anterior uveitis occurs in about one third of all patients with scleritis.

187. True. Sclerokeratitis occurs when corneal changes develop in conjunction with scleritis. There are three forms: acute stromal keratitis, sclerosing keratitis, and marginal keratolysis (associated with collagenase production, seen in patients with autoimmune connective tissue disease).

188. a. Scleritis can occur in association with various systemic infectious diseases, including leprosy, tuberculosis, herpes zoster, and syphilis. Metabolic diseases such as gout also may be associated with scleritis. Approximately one half of patients with scleritis have an associated systemic disease.

189. e. Scleritis is frequently associated with autoimmune connective tissue diseases such as systemic lupus erythematosus (SLE), rheumatoid arthritis, polyarteritis nodosa, or Wegener’s granulomatosis. Inflammatory bowel disease also has been reported in conjunction with peripheral ulcerative keratitis.

190. c. Subtenon’s injections of corticosteroids are relatively contraindicated in patients with autoimmune sclerokeratitis because these drugs may result
The eyelid skin is the thinnest skin in the body.

192. a. Chronic dacryoadenitis is usually associated with systemic disease such as lymphoma, sarcoidosis, syphilis, or tuberculosis. Sarcoidosis is the most common cause of painless bilateral enlargement of the lacrimal gland.

193. b. Chronic dacryoadenitis is sometimes accompanied by inflammation and swelling of the salivary glands, which is referred to as “Mikulicz’s syndrome.” Biopsy may be required for diagnosis.

194. c. Actinomyces israelii is usually found with expression or curettage of the canaliculus. A. israelii is a Gram-positive, branching, filamentous bacterium. Canaliculitis occurs more frequently in women.

195. b. Silent dacryocystitis is usually produced by Streptococcus pneumoniae and may present with no clinical symptoms other than occasional epiphora.

196. e. A lacrimal sac abscess has formed and is not responsive to oral antibiotics. It must be drained.

197. c. Congenital obstruction of the nasolacrimal system usually produces epiphora. The lumen of the nasolacrimal duct is blocked near the lower ostium (valve of Hasner) by epithelial debris or a mucosal membrane. The ostium will open spontaneously in 90% of infants during the first 9 months of life. Gentle probing and irrigation of the nasolacrimal system are performed by 6 to 9 months of age, if the system does not open spontaneously. If probing is unsuccessful, silicone tube intubation should be considered.

198. b. Because of the patient’s age, probing and irrigation should be performed; few blocked ducts will open spontaneously after 12 months of age.

199. False. If probing of a congenitally impatent nasolacrimal system is unsuccessful, silicone tube intubation should be considered and is effective in 85% of the cases. Dacryocystorhinostomy should be delayed until 3 years of age for long-standing success.

200. a. The eyelid skin is the thinnest skin in the body.

201. d. Eyes without any astigmatism will not have any wavefront aberration. Wavefront analysis is used to analyze irregular astigmatism. An eight-leaf clover will be produced with a penetrating keratoplasty (PKP) using eight interrupted sutures (and a four-leaf clover will be produced using four interrupted sutures). Spherical aberration occurs when peripheral rays focus in front of central rays, leading to night myopia in postoperative patients who underwent laser-assisted in situ keratomileusis (LASIK) surgery.

202. False. Although it can occur, corneal haze is not a common complication of lamellar keratoplasty. It occurs much more commonly after photorefractive keratectomy (PRK).

203. e. Lamellar keratoplasty does not effectively treat conditions with damaged corneal endothelium.

204. a. Mucosal epithelia differ from epidermis in that they do not normally keratinize (absence of granular and keratin layer) and they do not have rete ridges. Mucosal epithelium also contain goblet cells.

205. d. The conjunctiva, but not the cornea, contains many mucin-secreting goblet cells and accessory lacrimal glands. They are otherwise essentially identical epithelia.

206. a. Munnerlyn’s formula estimates the ablation depth needed centrally to perform photorefractive keratectomy (PRK). It is most useful with low amounts of correction (<7 diopters). Note that the amount of ablation needed is directly proportional to the square of the optical zone, so a larger optical zone requires more ablation.

207. False. A melanophage is a macrophage that has engulfed melanin, not a melanocyte.

208. a. These benign tumors demonstrate hyperkeratosis, parakeratosis, and epidermalization, in addition to lobular acanthosis surrounding vascular cores. Papillomas of the eyelid in young people are often caused by the human papillomavirus (HPV).

209. e. Keratoacanthoma is a reactive tumor that develops rapidly over 4 to 8 weeks. It is a large, elevated, and round cutaneous tumor that contains a central core of keratin. Although acanthosis, hyperkeratosis, and dyskeratosis may be extreme, dysplasia is often absent. Inflammation is prominent because keratoacanthoma is a type of pseudoepitheliomatous hyperplasia. Keratoacanthoma is likely a low-grade squamous cell carcinoma. The other lesions listed in the question are primarily disorders of the elderly patients.

210. c. Keratoacanthoma is a reactive tumor that develops rapidly over 4 to 8 weeks. It is a large, elevated, and round cutaneous tumor that contains a central core of keratin. Although acanthosis, hyperkeratosis, and dyskeratosis may be extreme, dysplasia is often absent. Inflammation is prominent because keratoacanthoma is a type of pseudoepitheliomatous hyperplasia. Keratoacanthoma is likely a low-grade squamous cell carcinoma. The other lesions listed in the question are primarily disorders of the elderly patients.

211. False. Keratoacanthomas contain areas of acanthosis, hyperkeratosis, and dyskeratosis, but typically not severe dysplasia.

212. True. See answers 210 and 211.

213. True. Spontaneous regression is typical. However, it is relatively easy to excise these lesions without recurrence.

214. c. Seborrheic keratoses are usually verrucoid and “greasy” looking.

215. e. Keratoacanthomas show more inflammation and are generally significantly larger. (Both are nodules with central craters.)

217. c. Hyperpigmentation may be found in both lesions, as may inflammatory cells.

218. a. The fibrotic type is also known as “morpheaform.” This pattern features strands of tumor cells infiltrating out from the central, clinically apparent lesion.
Therefore, it is much more difficult to define the margins of the lesion. They are also less responsive to radiation therapy. The other forms listed tend to be more localized.

219. d. This is a slowly growing tumor of the skin—conjunctival involvement being vanishingly rare—most commonly found on the lower lid.

220. e. Tumors here tend to be more deeply invasive, and the involved structures are more difficult to be freed of tumor.

221. b. Distant metastases of any kind are very rare for this tumor. Its invasive, burrowing nature is reflected in its historical pseudonym, “rodent ulcer.”

222. c. These lesions tend to be slow-growing.

223. b. The histopathology indicates a sebaceous cell carcinoma of the eyelid, a disorder of later life. Only prompt recognition of the diagnostic possibility with prompt resection offers hope for complete cure. All too often, chalazia are repeatedly drained, with no pathologic review to exclude this crucial diagnosis. Because of this tumor’s tendency to produce “skip” lesions (discontinuous areas of diseased tissue), Mohs’ techniques may leave residual tumor behind. Map biopsies are generally necessary.

224. d. Frozen section techniques allow for preservation of tissue lipid, staining for which (oil-red O stain) plays a role in diagnosing sebaceous cell carcinoma.

225. b. This is a feature of basal cell, not sebaceous cell, carcinomas.

226. a. These are all locations of sebaceous-type glands; Moll’s glands are apocrine sweat glands.

227. a. The syringoma is derived from sweat gland tissue.

228. d. An alternate name for pilomatrixoma is the “calcifying epitheloma of Malherbe.”

229. True. The hyperpigmentation of ephelis is due to increased melanin content of normal (or decreased) numbers of melanocytes.

230. True. Melanocytes in nevi are referred to as “nevus cells” to reflect this difference. Nevus cells are rounder, with distinct margins and more abundant, eosinophilic cytoplasm.

231. b. The opposite is true. They tend to move deeper into the subepithelial space, migrating into the dermis (or conjunctival substantia propria).

232. True. The femtosecond laser is a relatively new technology that creates the flap without the use of a microtome. The technology has been relatively expensive to implement thus far.

233. c.

234. d. (This would have to be considered grossly descriptive.)

235. d. Radiation therapy is a very important and effective modality for treating these lesions.

236. a. Blockage and inflammation of a meibomian gland produces an “internal hordeolum.”

237. False. Blocked glands of Zeis, in particular, can produce an external hordeolum that is purely inflammatory in nature. (Sebum is very irritating to tissues.)

238. c. These lesions are sometimes acutely inflamed and markedly tender.

239. c. Xanthelasma tend to be flat, plaquelike lesions, as opposed to nodules.

240. False. Epibulbar dermoids are solid choristomas. Adnexal dermoids are generally cystic.

241. d. Oral mucosal leukoplakic lesions are the other common finding in benign hereditary intraepithelial dyskeratosis—an obscure, dominantly inherited condition found in a very few number of North Carolinians.

242. e. The grading of epithelial dysplasia is identical, as is the acronym (CIN: conjunctival, corneal, or cervical intraepithelial neoplasia).

243. b. Clear lens exchange is the same procedure as phacoemulsification with posterior chamber lens implantation. However, patients with high myopia are already at risk for retinal detachment, and intraocular surgery can increase this risk further. Also, if posterior capsular opacification develops, Nd:YAG capsulotomy will also increase the risk of retinal detachment. Ptosis is also a risk of cataract surgery. Flap dehiscence does not occur with clear lens exchange, because unlike laser-assisted in situ keratomileusis (LASIK), there is no stromal flap created.

244. e. The same is true for conjunctival neoplasms.

245. b. By definition, the neoplasia of the corneal epithelium is not invasive until it has begun to penetrate Bowman’s layer.

246. a. Although the conjunctiva does not have a dermal layer, subepithelial conjunctival nevi occur and are equivalent to dermal nevi of the skin. The nevus cells, with time, migrate down into the substantia propria.

247. True. The neoplasia of the corneal epithelium is not invasive until it has begun to penetrate Bowman’s layer.

248. False. Biopsies of conjunctival melanomas are not necessary.

249. False. Clear lens exchange is the same procedure as phacoemulsification with posterior chamber lens implantation. However, patients with high myopia are already at risk for retinal detachment, and intraocular surgery can increase this risk further. Also, if posterior capsular opacification develops, Nd:YAG capsulotomy will also increase the risk of retinal detachment. Ptosis is also a risk of cataract surgery. Flap dehiscence does not occur with clear lens exchange, because unlike laser-assisted in situ keratomileusis (LASIK), there is no stromal flap created.

250. False. Cryotherapy is the most important adjunctive therapy. Occasionally, enucleation or evisceration is required.
Answers

251. a. It is difficult to distinguish conjunctival lymphomas from benign reactive lymphoid hyperplasia, but a prominent vascular stroma, numerous follicles, and cellular heterogeneity are all indicative of reactive (nonneoplastic) lesions.

252. False. Over 90% of pericorneal lymphoid proliferations are B-cell dominated (all have a small subpopulation of polyclonal T cells).

253. False. Microscopically, lymphomas are characterized by sheets of relatively monotonous cells. Unlike most other malignancies, atypia is not a feature of lymphoma. In fact, mitotic figures are rare in lymphoma and common in reactive lesions.

254. c. In isolated microcornea, the cornea is flatter than normal, resulting in hyperopia.

255. c. Although the anterior segment dysgenesis anomalies were once felt to be a result of mesodermal dysgenesis, it is now believed that the affected tissues are of neural crest origin.

256. b. The Schwalbe line is the termination of the corneal endothelium.

257. a. Posterior embryotoxon is a centrally displaced Schwalbe line that is visible without gonioscopy.

258. b. Axenfeld-Rieger syndrome is a spectrum of anterior segment defects characterized by an anteriorly displaced Schwalbe line. Patients may have glaucoma, developmental defects of facial bones and teeth. It is most commonly inherited in an autosomal dominant pattern.

259. b. In the past, “Rieger’s anomaly” was defined as the components of “Axenfeld’s anomaly” plus atrophy of the iris stroma. “Rieger’s syndrome” was defined as “Rieger’s anomaly” with additional findings of skeletal, dental, or craniofacial abnormalities. However, as mentioned in answer 258, the term Axenfeld-Rieger syndrome now more appropriately given the high incidence of overlapping findings, rather than rigidly defining each syndrome/anomaly.

260. c. An anterior cataract or dislocated lens may be present but is not necessary for the diagnosis.

261. True. Peters’ anomaly is bilateral in two thirds of the cases.

262. c. Congenital glaucoma can be distinguished from birth trauma by the presence of increased intraocular pressure (IOP) and horizontal (as opposed to vertical) breaks in Descemet’s membrane. In corneal birth trauma, residual hypertrophic ridges in Descemet’s membrane are often visible even after the corneal edema clears.

263. b. The actinic damage seen in pterygium results in changes in the subepithelial collagen. Although these fibers stain with some elastin stains, the fibers are not true elastin and will not be degraded by elastase. This finding is known as “elastosis.”

264. b. Pterygia invade the cornea down to the Bowman’s layer, producing fibrovascular ingrowth at this layer.

Because excised pterygia can recur with vigor, surgery is indicated only when the visual axis has been obscured or if there is extreme irritation.

265. a. Mitomycin C, beta-irradiation with strontium 90, and conjunctival autotransplantation all have been shown to reduce the recurrence rate of pterygia.

266. b. Amyloid exhibits dichroism and birefringence when stained with Congo red.

267. e. Note that only one polarizing filter is required to elicit dichroism.

268. a. Primary localized amyloidosis consists of conjunctival amyloid plaques, which occur without systemic involvement and without a local cause.

269. b. Primary systemic amyloidosis produces ecchymotic, waxy eyelid papules. Other ocular structures also may be infiltrated, including vitreous and uveal tract.

270. a. The white limbal girdle of Vogt consists of white flecklike deposits at the nasal and temporal limbus. It consists of subepithelial elastic degeneration and is sometimes accompanied by calcium deposition. Meretoja’s syndrome is a form of systemic amyloidosis combined with corneal lattice dystrophy.

271. a. The lipid deposition in arcus senilis tends to occur at the superior and inferior poles (where the local temperature is highest) and then spreads into the palpebral fissure. In the setting of high-grade carotid stenosis, the ipsilateral eye is protected from lipid deposition.

272. True. The guttae in Fuchs’ dystrophy as well as Hassall-Henle bodies result from dysfunctional production of basement membrane by endothelial cells.

273. False. Hassall-Henle bodies are normal aging changes found in the peripheral cornea. Similar changes in the central cornea are known as “corneal guttae,” and these may be forerunners of corneal edema.

274. c. Cornea farinata is a condition that is characterized by asymptomatic tiny deep stromal opacities, which are best viewed by retroillumination. Corneal guttae are at the level of the corneal endothelium.

275. a. Although any of the causes listed may produce calcific band keratopathy, chronic ocular inflammation is the most common. Juvenile rheumatoid arthritis (JRA) is certainly the most common (sarcoidosis is probably the second most common).

276. d. Band keratopathy starts at the horizontal periphery. It may spread centrally to form a horizontal band. Occasionally, it starts paracentrally. It is always in Bowman’s zone.

277. d. The urate form is much less common and may be associated with gout or hyperuricemia.

278. True. Actinic damage, age, genetic predisposition, and other environmental factors are thought to contribute to spheroidal degeneration, pterygia, and pingueculae.

279. d. Salzmann’s nodular degeneration, spheroidal degeneration, and Coats’ white ring are corneal degenerations that typically do not involve neovascularization.
280. b. The variety of Mooren’s ulcer that occurs in young African American men is usually more aggressive and responds poorly to medical or surgical management. Perforation is more common in this group. It is felt that some patients in this group may have developed the corneal ulceration as a result of antigen–antibody reactions to helminthic toxins. The toxins may get deposited during the blood-borne phase of certain parasitic infections.

281. d. Laser-assisted in situ keratomileusis (LASIK) is contraindicated in patients with unreasonable expectations, patients with potentially impaired healing (e.g., those on long-term topical or oral corticosteroids, or those who are immunocompromised), patients with chronic ocular conditions (including keratoconus). A history of gas-permeable contact lens use is not a contraindication for LASIK per se, but patients should discontinue their lenses for at least 8 to 10 weeks and then obtain serial topographic keratometry to ensure stability before surgery.

282. c. A wide variety of systemic inflammatory diseases may be associated with peripheral ulcerative keratitis. Most have a prominent vasculitic component.

283. False. Senile furrow degeneration is not associated with sceral weakness or melting. The thinning is generally mild with intact epithelium.

284. True. Phakic intraocular lens (IOLs) have been used to treat patients who are unable to undergo refractive surgery because of high refractive error (either high hyperopia or high myopia). Their long-term safety and efficacy is still being investigated.

285. False. The widely accepted classification scheme for corneal dystrophies is based on the layer of cornea involved (i.e., epithelium, Bowman’s zone, stroma, Descemet’s membrane, endothelium).

286. b. Both congenital and acquired (degenerative) forms of map-dot-fingerprint dystrophy may be seen. The latter may develop in the setting of recurrent erosions associated with trauma, contact lens use, or chronic blepharoconjunctivitis. Reis-Bückler dystrophy involves Bowman’s zone, whereas lattice and central cloudy dystrophies are stromal dystrophies.

287. d. Dots seen in anterior membrane dystrophy are clumps of degenerated epithelial cells and basement membrane material within the epithelium. They are also known as “microcysts.” Cogan’s microcystic edema is the eponym for the pure “dot” form of this disorder.

288. c. Successful treatment is generally targeted toward stimulating production of new, healthier basement membrane material by epithelial removal or stimulation (debridement or puncture), occlusion therapy (patching, contact lenses, lubrication), and/or epithelial dehydration (for recurrent erosions).

289. b. Epithelial cysts are seen in both map-dot-fingerprint and Meesman’s dystrophies. The cysts in the former are translucent or opaque and represent degenerated epithelial cells and basement membrane. Those of Meesman’s dystrophy are transparent and are filled with periodic acid–Schiff (PAS)-positive material known as “peculiar substance.” Fibrocellular invasion of Bowman’s zone (pannus) is typical of Reis-Bückler dystrophy; hyaline stromal deposits are seen in granular dystrophy. Congophilic stromal deposits are the hallmark of lattice dystrophies.

290. b. Symptoms in Meesman’s dystrophy are usually mild and relate to blurry vision and irritation. Central cloudy dystrophy is usually silent.

291. b. Recurrent erosions and anterior corneal scarring lead to visual loss in Reis-Bückler dystrophy sooner and to a greater degree than in the others. Pre-Descemet’s dystrophy is usually clinically silent, like central cloudy dystrophy.

292. c. In granular dystrophy, recurrent erosions are seen, but much less commonly than for the anterior dystrophies or lattice dystrophy. Transmission is autosomal dominant. Visual acuity is generally affected, but not until later in life (fifth decade or later). Stromal deposits of hyaline are diagnostic and may be associated with amyloid deposits in certain subtypes (Avellino variant).

293. b. Macular dystrophy may have focal granular-like deposits but differs from granular dystrophy in several ways. First, the intervening stroma is cloudy. Second, the peripheral cornea is involved much earlier in macular dystrophy. Third, macular dystrophy is inherited recessively, so that parents or offspring are unlikely to be involved, whereas siblings are likely to be involved. Patients with macular dystrophy have a deficiency in enzymatic synthesis of keratan sulfate, and serum levels are typically depressed. The mnemonic for stromal dystrophies (dystrophy, deposit, stain) is: Marilyn Monroe Always Gets Her Man in Los Angeles County. Marilyn Monroe Always (Macular/Mucopolysaccharide/Alcian blue) Gets Her Man (Granular/Hyaline/Masson’s trichrome) in Los Angeles County (Lattice/Amyloid/Congo red).

294. d. Lattice dystrophy also may have granular deposits but also features linear, branching (lattice) deposits. Both types of deposits are amyloid and will demonstrate birefringence and dichroism when stained with Congo red. In type I, as in this patient, the deposits are randomly distributed, greater centrally. This form is localized. In type II, the deposits are along the course of the corneal nerves, so they are denser peripherally. Type II is seen only in familial amyloidotic polyneuropathy type IV (Meretoja’s syndrome) and is associated with cranial nerve palsies and dry, redundant skin. Like granular dystrophy, lattice dystrophies are inherited dominantly. This is the stromal dystrophy most likely to be associated with recurrent erosions.
295. b. Central crystalline dystrophy of Schnyder represents intrastromal accumulation of cholesterol crystals. Like xanthelasmas, a few patients have systemic hyperlipidemia.

296. c. This disorder may be highly asymmetric or unilateral and associated with a wide range of ocular or systemic disorders. Tiny flecks are visible in corneal stroma. Vision is not affected.

297. d. Central cloudy dystrophy is nearly always clinically silent.

298. True. Both corneal guttae and macular drusen represent focal accumulations of basement membrane produced by a cellular monolayer undergoing senescence, at a normal or accelerated rate.

299. b. Symptoms of Fuchs' dystrophy are typically worse in the morning, when the corneal epithelium is more hydrated. Mild corneal stromal edema may be silent in the setting of visually significant cataract, so that triple procedures (penetrating keratoplasty [PKP], cataract extraction, intraocular lens placement) may be indicated despite asymptomatic corneal changes (with preexistent corneal edema, further decompensation is almost certain). If there are no signs (as opposed to symptoms) of corneal decompensation (stromal or epithelial edema), then cataract extraction and intraocular lens implantation may be undertaken alone.

300. a. In the setting of normal corneal diameters, glaucoma is less likely. Congenital hereditary endothelial dystrophy presents like congenital glaucoma except that the corneal diameters and intraocular pressure (IOP) are normal. In congenital hereditary stromal dystrophy, the clinical appearance is similar except that the stroma is normal thickness (but diffusely hazy), and the epithelium is spared.

301. c. This bilateral disorder presenting in the young man is less likely to be iridocorneal endothelial (ICE) syndrome than posterior polymorphous dystrophy (PPMD). Transmission is autosomal dominant, but expression is highly variable and asymmetric. Clinical findings may be similar to those of ICE. Histopathologically, the disorder may simulate epithelial downgrowth, but the cell of origin is the corneal endothelial cell.

302. b. See answer 301.

303. d. The earliest stage of keratoconus is progressive myopia with high astigmatism. Exactly when an eye may be declared to have keratoconus is a matter of opinion.

304. b. Corneal thinning, myopia, and astigmatism are most likely to progress significantly during adolescence, although progression can occur at any time.

305. False. Conductive keratoplasty has been approved for the treatment of low hyperopia (<3.0D) in patients with mild astigmatism (0.75D or less).


307. c. Rigid gas-permeable (RGP) contact lenses are the optical correction of choice in keratoconus because these lenses are the most successful at eradicating irregular astigmatism.

308. False. A satisfactory optical outcome in surgery for keratoglobus is more difficult to obtain than for keratoconus.

309. c. Higher corneal protrusion above the area of corneal thinning makes fitting very difficult in pellucid degeneration. Scleral contact lenses have been tried with some success.

310. c. Diffuse lamellar keratitis, also known as “Sands of the Sahara,” typically manifests itself within the first week after laser-assisted in situ keratomileusis (LASIK) surgery. If it is diagnosed early, topical steroid treatment usually provides for an excellent prognosis. However, later stages (clumping of white blood cells [WBCs] in the central visual axis) usually require flap lifting, irrigation, intense topical steroids, and occasionally systemic steroids.

311. c. Typically, tear volume is deficient in dry eye syndrome (inadequate aqueous phase), but a large subset of patients have problems with surface wetting due to deficiencies of the other phases (mucus problems in keratinizing disorders, lipid problems in blepharitis). Schirmer testing in these cases may be normal or supranormal despite a history and examination suggestive of dry eye. Tear breakup time is frequently the important clue.

312. False. Topical anesthesia eliminates reflex tearing so that a reliable estimate of basal tear secretion may be made.

313. True. Without anesthesia, at least 15 mm of the paper strip should be moistened after 5 minutes.

314. False. The ophthalmologist’s obligation to the patient never ends (even if the operation is successful). As per the American Academy of Ophthalmology (AAO), the ophthalmologist is obligated to perform postoperative care through the patient’s “at risk” postoperative period. Furthermore, the AAO stipulates that “If an ophthalmologist does not intend to provide postoperative medical eye care, this fact is one that a reasonable patient would consider to be material in deciding whether to undergo the proposed surgery by that ophthalmologist, and should be disclosed sufficiently in advance of the surgery.” Frankly, in the authors’ opinion, it is better for both the patient and the physician, if the physician provides the patient with postoperative care as much as possible.

315. b. See answer 311.

316. c. Blepharoconjunctivitis may produce a syndrome similar to dry eye syndrome except that the symptoms
are typically worse in the morning. In dry eye syndrome, symptoms are worse in the evening. Chalazia are associated with chronic blepharoconjunctivitis, not dry eye syndrome.

317. b. The most frequent association is classic rheumatoid arthritis.

318. c. The definition of secondary Sjögren’s syndrome is sicca complex plus coexistent autoimmune disease. Autoantibodies may be more prevalent and at higher concentrations in primary disease (no systemic disorder), but this is not a reliable differentiating feature. Dry mouth is a feature of both types. Although human leukocyte antigen (HLA)-Dw3 may be seen more commonly in primary disease, this is also not foolproof.

319. a. A variety of autoimmune diseases have been reported to develop following chronic primary Sjögren’s syndrome. There is no known association with epithelial lacrimal gland neoplasms.

320. d. Punctal occlusion is a safe and effective therapy for severe dry eye syndrome. Epiphora is the most likely adverse effect, so a graded, stepwise approach is generally taken. Temporary occlusion of inferior puncta allows assessment of response prior to permanent occlusion.

321. a. In ocular cicatricial pemphigoid (OCP), a deficient mucus phase leads to tear dysfunction. In Bell’s palsy, there is exaggerated drying due to exposure and lagophthalmos. Oral contraceptives have been associated with decreased aqueous tear production. Herpes zoster renders a cornea hypesthetic with impaired reflex tearing and drying.

322. True. In both disorders, the interpalpebral cornea is the area affected earliest and most severely.

323. True. Like exposure keratopathy, neurotrophic keratopathy affects the interpalpebral zone preferentially because this is the area of greatest exposure and drying.

324. c. The combination of corneal hypesthesia and exposure/lagophthalmos is nearly always disastrous for the corneal surface. Lateral tarsorrhaphy is probably advisable before irreversible, destructive changes can commence. Remember: Five (V) + Seven (VII) = T. That is, Vth nerve (hypesthesia) plus VIIth nerve (exposure) dysfunction equals tarsorrhaphy.

325. b. Comedones are not a feature of rosacea dermatitis. This differentiates acne rosacea from acne vulgaris.

326. a. Anterior uveitis is uncommon. The other external manifestations are common, but the most common, by far, is meibomitis.

327. True. This reflects deficiency in tear function secondary to decreased lipid secretion by damaged meibomian glands.

328. c. Demodex folliculorum may play an adjuvant role in what is primarily a type IV hypersensitivity reaction to unknown stimuli.

329. c. Potent steroids may precipitate melting in eyes with rosacea keratitis. Any steroid preparation should probably be avoided in the setting of frank corneal ulceration. Once epithelial continuity has been restored and secondary infection adequately treated (hygiene, antibiotic ointments, oral tetracycline), mild steroid preparations are probably safe but must be used with adequate surveillance.

330. d. Erythema multiforme minor is strictly a skin disease. The systemic variety (erythema multiforme major) has mucous membrane ulceration, as well as dermatitis, and is life threatening (20% systemic mortality from secondary infection).

331. d. Recent sultonamide use is the clearest association. Postinfectious etiologies also have been recognized (herpes simplex virus [HSV], adenovirus, mycoplasma).

332. False. Mucous membrane lesions are bullous. Skin lesions are macular and consist of a “target” lesion—red center, inner pale annulus, outer red annulus.

333. d. Some experts feel that lysis of symblepharons may lead to secondary infection and/or worsened scarring. In addition, despite vigorous attempts, subconjunctival scarring may still occur.

334. b. Oral and conjunctival epithelia are the most frequently involved, with a slight preponderance of the latter.

335. False. Erythema multiforme is a vasculitic process. Ocular cicatricial pemphigoid (OCP) features deposition of immunoglobulin and complement in the basement membranes of skin and mucous membranes, with subsequent bulla formation.

336. e. The inferior fornix is generally the most rewarding area of the eye to examine in ocular cicatricial pemphigoid (OCP). Here, subepithelial fibrosis and fornix foreshortening may be seen to precede symblepharon formation. The superior tarsus may show subconjunctival scarring.

337. d. This is in distinction to pemphigus, in which epithelial acantholysis and intraepithelial bullae are characteristic features.

338. False. Dapsone is an acceptable first choice in mild to moderate cases in patients with no evidence of glucose-6-phosphate dehydrogenase deficiency (in whom dapsone can cause a fatal hemolytic anemia). The most reliable prognostic indicator for long-term outcome in ocular cicatricial pemphigoid (OCP) is the severity and rapidity of active inflammation. Therefore, severe cases should be treated with more potent immunosuppressive agents from the outset, such as cyclophosphamide.

339. e. Areas of metaplastic keratinization of the conjunctiva within the exposure zone are known as “Bitot’s spots.” Secondary infection by xerosis bacilli (Corynebacterium xerosis) is common. A peripheral retinopathy of minimal functional significance also has been described. Keratomalacia, diffuse corneal necrosis and melting, is the most disastrous of manifestations.
**Answers**

340. a. Any deficiency of fat absorption may lead to hypovitaminosis A. Chronic alcoholics with very poor nutrition should be suspected of relative hypovitaminosis A and undergo serum vitamin A level testing in the setting of bilateral dry eye or corneal ulceration.

341. c. The history is most compatible with recurrent erosion syndrome. Predisposing factors are numerous and include previous corneal abrasion, particularly recalcitrant ones; contact lens use; anterior corneal dystrophies (map-dot-fingerprint, Reis-Bückler); and diabetes mellitus (defective basal epithelial adherence).

342. c. Hypertonic saline may be useful in maintaining epithelial deturgescence and enhancing adherence but must be used for several (4 to 6) months if it is to have long-term effect.

343. c. Ligneous deposits nearly always recur after excision if the disease process remains active.

344. a. Type III (Sanfilippo’s syndrome) is virtually never associated with corneal clouding. Type II (Hunter’s syndrome) is rarely associated with corneal clouding (only adults with the milder variety). Type II is inherited on an X-linked–recessive basis.

345. c. The findings are indistinguishable from medication-induced vortex keratopathy.

346. b. Fabry’s disease is an X-linked–recessive disorder featuring accumulation of cerebrosides in the cardiovascular system and kidneys. Affected male patients develop renal failure, but female carriers do not.

347. c. Three forms of cystinosis are recognized. The infantile form is the most severe, followed by the juvenile or adolescent form. Renal failure is expected in both. The adult form is mild (renal failure is unusual). Monoclonal gammopathies may be associated with corneal crystals, with immunoglobulin deposits in the peripheral cornea. Peptic ulcer disease (PUD) is not associated with cystinosis or myeloma. Bietti’s crystalline dystrophy features crystalline keratopathy and a pigmentary retinopathy.

348. b. In tyrosinemia, elevated serum tyrosine levels lead to lysosomal instability with dermal and ocular inflammation, as well as mental retardation. Nonstaining pseudopods may recur and be misdiagnosed as herpes simplex virus (HSV).

349. d. Of the agents known to cause vortex keratopathy, only chlorpromazine is associated with pigmentary retinopathy. The two are generally independent. (Chloroquine can cause a bull’s-eye maculopathy as well.)

350. d. The earliest sign of copper deposition in Descemet’s membrane (Kayser-Fleischer ring) is detectable only at the far periphery with gonioscopy. Slitlamp examination alone is insufficient.

351. e. Like the sunflower cataract of Wilson’s disease, copper accumulation in Descemet’s membrane may be seen in chalcosis and other specific cholestatic disorders.

352. True. Photorefractive keratectomy (PRK) has been approved by the U.S. Food and Drug Administration (FDA) to treat hyperopia up to 5.0D, with astigmatism up to 4.0D.

353. d.

354. b.

355. d. Other favorable factors include lack of neovascularization, lack of inflammation, and no history of previous graft failure.

356. e. Other unfavorable factors include youth, larger grafts, and glaucoma.

357. c. Most intraocular lens (IOLs) capable of causing corneal decompensation also can cause chronic intraocular inflammation and secondary cystoid macular edema.

358. a. Indirect miotics such as phospholine are generally stopped several weeks before surgery to stabilize the blood-aqueous barrier and allow repletion of serum cholinesterase. Hypotensive maneuvers such as the Honan balloon and mannitol lower the incidence of vitreous loss. Pilocarpine preoperatively protects the lens from incidental damage.

359. True. Aphakic globes have lower rigidity and structural integrity, leading to collapse and vitreous presentation. A support ring helps prevent this.

360. False. The main advantage of smaller donor size is decreased risk of rejection, whereas the main disadvantage is increased central astigmatism. For larger grafts, the opposite is true.

361. True. Donor buttons that are larger than the host bed are believed to generate centrifugal radial forces that tend to keep the anterior chamber angle open. Smaller buttons are more likely to lead to collapse and peripheral anterior synechiae.

362. a. Smaller-than-host-bed donor buttons tend to be flatter and thus more hyperopic than the larger-than-host-bed donor buttons. This effect helps to neutralize part of the high myopia seen in eyes with keratoconus.

363. a.

364. c. Keratoscopy gives qualitative information regarding corneal topography when the surface is too irregular to permit quantification with keratometry.

365. False. If the anterior chamber intraocular lenses (ACIOL) is of a style known to be associated with corneal decompensation or cystoid macular edema, it should be replaced at the time of penetrating keratoplasty (PKP). If the keratopathy is primarily due to corneal failure rather than the intraocular lens (IOL), it need not be replaced.

366. False. Cataract extraction after penetrating keratoplasty (PKP) increases the risk of graft rejection or failure. If possible, these procedures are generally performed simultaneously (when indicated).

367. e. In the setting of corneal edema and cataract, the triple procedure is probably advisable. Guttae alone are not a
sufficient indication for this procedure. There should be stromal or epithelial edema before it is undertaken.

368. b. See answer 367.

369. b. Ocular surgery may precipitate increased disease activity in ocular cicatricial pemphigoid (OCP). Once the other disorders have become quiescent, they typically remain so.

370. c. Preexistent epithelial and/or surface disorders predispose the graft to chronic epithelial defects. Keratoconus has an excellent prognosis after penetrating keratoplasty (PKP).

371. d. Unless the donor endothelium is unhealthy or damaged at the time of transplantation (primary graft failure), the findings of Fuchs’ dystrophy do not recur. Primary failure is different than true recurrence, which has been reported numerous times for each of the other disorders.

372. d. This complication can frequently be controlled with selective suture removal, but in some cases refractive surgery is necessary.

373. c. Frequently, prolonged topical and systemic antibiotic therapy is necessary, with or without surgical therapy (lamellar or penetrating excision). This infectious crystalline keratopathy is closely associated with kerato- plasty and topical steroid use.

374. a. Immunologic graft failure never presents within 24 hours of transplantation.

375. d. Type II mechanisms are also important.

376. a. In first-time grafts, rejection rarely, if ever, presents within the first 10 days postoperatively. It is possible, however, with subsequent grafts in sensitized patients. Graft rejection should spare the host bed, unlike glaucoma. A linear array of keratic precipitates (KPs), the Khoudadoust line, may be seen in the earlier phases of immunologic rejection but is not universal.

377. c. Epithelial rejection is typically mild, and of no long-term significance, because host epithelium quickly populates the donor surface. Endothelial rejection is of far greater visual significance and may be precipitated by seemingly innocuous events, all of which stimulate ocular inflammation.

378. b. Actually, lamellar keratoplasty is technically more difficult. The interface frequently becomes cloudy, but rejection is less common than with penetrating surgery.

379. d. Lattice dystrophy typically involves the deeper stroma, so lamellar surgery is not definitive.

380. d. The absence of significant heat release makes adhesives safer and more reliable.

381. d. A conjunctival flap will not provide an adequate seal over an underlying fistula or perforation. The “hole” must be closed with a tectonic graft (full-thickness or lamellar, with sclera or cornea) before a flap may be advanced.

382. b. The multiple subepithelial infiltrates seen in the figure are most typical of the postinfectious stage of epidemic keratoconjunctivitis (EKC). The history and findings are most consistent with this as well. EKC can be membranous in severe cases. The infiltrates themselves are felt to represent hypersensitivity reactions to viral antigens, without active microbial replication. Topical corticosteroids are quite helpful in controlling potentially disabling photophobia and pain but can lead to a medication-dependent state requiring months or years to reverse. Subepithelial infiltrates that may be indistinguishable from those of EKC may be seen in Thygeson’s disease. In the latter, there is no conjunctivitis prior to the corneal manifestations.

383. a. This patient has band keratopathy. The most frequent etiology of findings as severe as this is chronic intraocular inflammation, particularly juvenile rheumatoid arthritis (JRA). Other uveitides implicated in band keratopathy include sarcoidosis and syphilis. Systemic metabolic disorders including hypercalcemia of any etiology and hyperuricemia, with or without gouty arthritis, are also clearly associated with band keratopathy. Uveitides related to human leukocyte antigen (HLA) type (e.g., HLA-B27 uveitis, Reiter’s syndrome, Behçet’s disease, birdshot chorioretinopathy) are less commonly associated with band keratopathy.

384. c. The time of onset (only a few weeks after excision) and appearance of the lesion are most suggestive of a pyogenic granuloma—essentially hypertrophic granulation tissue. The lesions are neither pyogenic (copious neutrophils) nor granulomatous; instead, they feature abundant immature vascular channels in a matrix of ground substance secreted by active fibroblasts. Intensive topical steroids are typically curative.

385. b. A recent study has documented its utility in bullous keratopathy as well. It reduces bullae and lessens pain and may provide an alternative for penetrating keratoplasty (PKP) in selected cases.

386. d. The Herpetic Eye Disease Study has answered several questions regarding the utility of topical steroids in herpes simplex stromal keratitis. Topical steroids reduced persistence or progression of stromal keratitis compared with placebo. Delayed initiation of steroid therapy slowed resolution but did not affect long-term (6 months) visual outcome. Recurrences following steroid taper responded well to a more gradual taper. Topical trifluridine was used in both treatment arms of the study to help lower the incidence of epithelial keratitis. A large proportion of patients treated with trifluridine and placebo had significant progression of herpetic inflammation, implying that trifluridine alone does not exert a significant beneficial effect for stromal keratitis.
1. Azithromycin and clarithromycin are among the first members of a new class of bacterial ribosomal inhibitors known as “azalides.” They are related chemically to erythromycin but are much more rapidly distributed to tissues with better absorption, bioavailability, and higher intracellular penetration. Azithromycin’s tissue half-life is also quite long, 2 to 4 days, rendering it useful as a single-dose agent. This enhances compliance relative to the traditional prolonged topical treatment, and also offers the advantage of treating extraocular reservoirs of Chlamydia.

2. a. 2, b. 3, c. 3, d. 2, e. 4.

3. a. 3, b. 3, c. 1, d. 4, e. 2.

4. a. 3, b. 2, c. 1, d. 2, e. 3, f. 1, g. 3, h. 2, i. 1, j. 3, k. 1.

5. a. 3, b. 3, c. 1, d. 1, e. 2, f. 1, g. 1, h. 2, i. 1, j. 2, k. 1.

6. a. 1, b. 1, c. 3, d. 1, e. 2, f. 1, g. 1, h. 2, i. 1, j. 2, k. 1.

7. a. 3, b. 1, c. 4, d. 2.

8. a. 3, b. 1, c. 2, d. 1, e. 2, f. 3, g. 1, h. 1, i. 3, j. 2.

9. a. 1, b. 2, c. 2, d. 1, e. 2, f. 1, g. 1, h. 2, i. 3, j. 2.

10. a. 1, b. 2, c. 6, d. 3, e. 8, f. 4, g. 5, h. 1.

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**Answers**

Anaplasia is dysplasia of such severity as to be obviously malignant. Epidermidization can be found in benign or malignant tumors of squamous tissue.

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**Suggested Readings**


Questions

1. At birth, the length of the average infant human eye is:
   a. 8 to 9 mm.
   b. 12 to 13 mm.
   c. 16 to 17 mm.
   d. 20 to 21 mm.
   e. 23 to 24 mm.

2. The factor primarily responsible for the shallow anterior chamber in the eye of a healthy infant is that:
   a. the infant’s cornea is flatter than the adult’s cornea.
   b. the infant’s iris is relatively thicker than the adult’s iris.
   c. the infant’s lens is relatively thicker than the adult’s lens.
   d. there is more positive vitreous pressure in the infant’s eye than in the adult’s eye.
   e. the anterior chamber is as deep in infants as it is in adults.

3. T or F The corneal diameter of the eye of the average healthy human infant is 10 mm (9.5 to 10.5 mm).

4. The reasons for relatively miotic pupils in infancy include:
   1. relative delay in sympathetic innervation of the eye.
   2. excessive supranuclear input to the Edinger–Westphal nucleus.
   3. increased sensitivity of the light-induced miosis reflex.
   4. immaturity of the dilator pupillae muscle.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

5. T or F The completion of optic nerve myelination and that of foveal maturation coincide postnatally.

6. Reliable methods of estimating visual acuity in the preverbal child include:
   1. optokinetic nystagmus testing (OKN).
   2. preferential looking testing (PLT).
   3. visual evoked potentials (VEPs).
   4. electroretinography.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

7. T or F Acuity estimates generated by preferential looking testing (PLT) and visual evoked potentials (VEP) generally coincide quite closely.
8. T or F In cryptophthalmos, there is a failure of development of normal lid structures over an otherwise normal globe.

9. Congenital colobomas of the eyelids are associated with which systemic syndrome?
   a. Goldenhar’s syndrome.
   c. Hallermann-Streiff syndrome.
   d. Stickler’s syndrome.
   e. trisomy 18.

10. T or F Both congenital ectropion and entropion involve the upper lid more frequently than the lower lid.

11. T or F Like congenital entropion, distichiasis generally does not cause major keratopathy.

12. T or F There are three varieties of epicanthus: palpebralis, tarsalis, and inversus.

13. T or F Telecanthus is synonymous with hypertelorism.

14. Findings seen as part of the blepharophimosis syndrome include:
   1. simple epicanthus (palpebralis).
   2. blepharoptosis.
   3. hypertelorism.
   4. blepharophimosis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

15. Which of the following statements about congenital toxoplasmosis is/are true?
   1. Fetal infection earlier in gestation generally results in more severe involvement.
   2. Seroconversion during pregnancy, marking new infection of a mother, is rarely associated with placental transfer of the organism.
   3. Most pregnant women are seronegative (i.e., susceptible to infection).
   4. When placental transfer occurs, the infant nearly always develops some obvious manifestation of the infection.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

16. T or F The incidence of congenital toxoplasmosis, both symptomatic and asymptomatic, is approximately 1 in 1,000 live births.

17. Signs and symptoms typical of congenital toxoplasmosis include:
   1. hepatosplenomegaly.
   2. seizures with intracranial calcifications.
   3. vomiting and diarrhea.
   4. diffuse pigmentary (“salt-and-pepper”) retinopathy.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

18. Which test results from an infant would support the diagnosis of congenital *Toxoplasma* infection?
   1. antitoxoplasma immunoglobulin G (IgG) antibody.
   2. antitoxoplasma immunoglobulin M (IgM) antibody.
   3. maternal antitoxoplasma IgM antibody.
   4. computed tomography (CT) scan revealing intracranial calcifications.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

19. Medications important in the control of ocular toxoplasmosis include:
   1. pyrimethamine.
   2. sulfadiazine.
   3. prednisone.
   4. folic acid.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
20. Which of the following statements about the epidemiology of congenital rubella infection is/are true?
   1. Most pregnant women are seronegative (susceptible to rubella infection).
   2. Seroconversion of a mother from negative to positive nearly guarantees infection of the fetus.
   3. Symptomatic fetal defects are uncommon, even with viremia.
   4. Maternal infection during the third trimester rarely leads to fetal infection.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

21. The most common clinical finding in infants with congenital rubella syndrome is:
   a. pigmented retinopathy.
   b. patent ductus arteriosus.
   c. sensorineural hearing loss.
   d. mental retardation.
   e. cataract.

22. Which two signs of congenital rubella infection are unlikely to be found simultaneously?
   a. microphthalmia and congenital cataract.
   b. microphthalmia and congenital glaucoma.
   c. congenital cataract and glaucoma.
   d. congenital cataract and a poorly dilating iris.
   e. pigmented retinopathy and congenital cataract.

23. Live rubella virus may be recovered from an infected infant from which of the following sources?
   1. conjunctival swabs.
   2. urine cultures.
   3. pharyngeal swabs.
   4. lens aspirates.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

24. The postoperative course following extraction of infantile cataract associated with the congenital rubella syndrome is distinguished by:
   a. a higher incidence of retinal detachment.
   b. a higher incidence of glaucoma.
   c. poor wound healing.
   d. severe inflammation.
   e. difficulty in tolerating aphakic contact lenses.

25. The most common congenital infection in humans is:
   a. toxoplasmosis.
   b. rubella.
   c. cytomegalovirus (CMV).
   d. herpes simplex virus (HSV).
   e. syphilis.

26. The most common ocular manifestation of congenital cytomegalovirus (CMV) infection is:
   a. cataract.
   b. microphthalmia.
   c. retinochoroiditis.
   d. Peters' anomaly.
   e. strabismus.

27. T or F Most cases of congenital herpes simplex infection are caused by maternal viremia during gestation.

28. T or F Most cases of congenital cytomegalovirus (CMV) infection are caused by maternal viremia during gestation.

29. T or F Like congenital cytomegalovirus (CMV) infection, congenital herpes simplex virus (HSV) infection is frequently asymptomatic.

30. T or F The ocular manifestations of congenital herpes simplex virus (HSV) infection resemble those of acquired infections in adolescence and adulthood.

31. Transplacental spread of which of the following microorganisms is important in the spread of congenital infection?
   1. *Toxoplasma*.
   2. cytomegalovirus (CMV).
   3. *Treponema pallidum*.
   4. herpes simplex virus (HSV).
Questions

32. Ocular manifestations of congenital syphilis infection frequently include:
   b. pseudoretinitis pigmentosa.
   c. interstitial keratitis.
   d. microphthalmia.
   e. 1, 2, 3, and 4.

33. Hutchinson’s triad, considered diagnostic of congenital syphilis infection, includes:
   a. peg-shaped teeth, eighth nerve deafness, and interstitial keratitis.
   b. rhagades, interstitial keratitis, and hepatosplenomegaly.
   c. pseudoretinitis pigmentosa, interstitial keratitis, and peg-shaped teeth.
   d. pseudoretinitis pigmentosa, eighth nerve deafness, and interstitial keratitis.
   e. interstitial keratitis, cataract, and pseudoretinitis pigmentosa.

34. Each of the following statements is a valid conclusion of the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity (CRYO-ROP) except:
   a. Treatment of threshold disease reduces the incidence of retinal detachment relative to no treatment.
   b. Treatment of threshold disease reduces the incidence of blindness relative to no treatment.
   c. Treatment of threshold disease results in better long-term Snellen acuity relative to no treatment.
   d. Treatment of threshold disease increases the proportion of normal-appearing posterior poles at the age of 5 years as compared to no treatment.
   e. Treatment benefit is independent of birth weight, race, and number of sectors of stage 3 involvement.

35. Which of the following are considered common etiologic agents for conjunctivitis in children?
   a. Streptococcus pneumoniae.
   b. Haemophilus influenzae.
   c. Staphylococcus aureus.
   d. Streptococcus pyogenes.
   e. 1, 2, 3, and 4.

36. Which of the following viral infections may be associated with a pronounced keratitis?
   a. A. herpes simplex.
   b. adenovirus type 8.
   c. herpes zoster.
   d. adenovirus type 3.
   e. 1, 2, 3, and 4.

37. T or F The syndrome of infectious mononucleosis is rarely accompanied by a conjunctivitis.

38. Which of the following statements about Parinaud’s oculoglandular syndrome is/are true?
   a. Common etiologic agents include the cat-scratch fever organism, Rickettsiae, Treponema pallidum, and mycobacterial species.
   b. Clinically, follicles are prominent with a moderate discharge.
   c. Historical features may include contact with animals.
   d. Histopathology reveals follicles and granulomas.
   e. 1, 2, 3, and 4.

39. The agent most commonly responsible for preseptal cellulitis in children is:
   a. Staphylococcus aureus.
   b. Pseudomonas aeruginosa.
c. *Streptococcus pyogenes*.
d. *Haemophilus influenzae*.
e. *Proteus mirabilis*.

40. The agent most frequently associated with orbital cellulitis following bacterial conjunctivitis is:
a. *Staphylococcus aureus*.
b. *Pseudomonas aeruginosa*.
c. *Streptococcus pyogenes*.
d. *Haemophilus influenzae*.
e. *Proteus mirabilis*.

41. The focus of primary infection in most cases of orbital cellulitis is:
a. maxillary sinus.
b. ethmoid sinus.
c. frontal sinus.
d. orbital foreign body.
e. meningitis with cavernous sinusitis.

42. Sudden deterioration in ocular motility without a dramatic increase in proptosis suggests which complication of orbital cellulitis?
a. panophthalmitis.
b. meningitis.
c. central retinal artery occlusion (CRAO).
d. cavernous sinus thrombosis.
e. subperiosteal abscess.

43. Which of the following statements about vernal conjunctivitis is/are true?
    1. Vernal conjunctivitis is primarily a disease of the first 2 decades of life.
    2. Vernal conjunctivitis affects girls more frequently than boys.
    3. Prominent symptoms include photophobia and itching.
    4. The palpebral form of the disease is typically more severe inferiorly.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

44. Features distinguishing vernal conjunctivitis from trachoma include:
    1. true follicles with germinal centers in vernal disease.
    2. eosinophils in trachoma.
    3. limbal nodules in vernal disease.
    4. prominent subconjunctival scarring in trachoma.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

45. T or F The limbal nodules of vernal keratoconjunctivitis are actually follicles.

46. T or F Horner-Trantas dots and Herbert’s pits are histopathologically indistinguishable.

47. T or F The presence of superior corneal pannus favors the diagnosis of trachoma over vernal conjunctivitis.

48. Corneal manifestations of vernal disease may include:
    1. superficial punctate keratitis.
    2. superior corneal pannus.
    3. transverse oval sterile ulceration in the superior cornea.
    4. deep stromal vascularization.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

49. T or F The shield ulcers of vernal keratoconjunctivitis are primarily caused by mechanical abrasion by tarsal papillae.

50. Which of the following statements about Stevens-Johnson syndrome is/are true?
    1. This is a disorder restricted to childhood.
    2. Postinfectious, postvaccine, and drug-induced autoimmune disorders have all been implicated as the cause of Stevens-Johnson syndrome.
    3. The associated vasculitis leads to a vesicular rash and mucous membrane lesions.
    4. Clinical findings include fever, pharyngitis, and headache.
Questions

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

51. Along with a fever lasting for >5 days, which of the following are diagnostic criteria for Kawasaki’s disease?
   1. bilateral conjunctival infection.
   2. mucous membrane infection with fissures.
   3. strawberry tongue.
   4. desquamating rash of the palms and/or soles.
   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

52. Systemic mortality caused by Kawasaki’s disease is most frequently due to:
a. stroke.
b. respiratory failure.
c. myocardial infarction (MI).
d. acute renal failure.
e. acute adrenal insufficiency.

53. Which one of the following statements about the anatomy of the nasolacrimal system is false?
a. The canaliculi normally run vertically for 1 or 2 mm before running medially toward the nasolacrimal sac.
b. The medial palpebral ligament straddles the lower one third of the nasolacrimal sac.
c. The nasolacrimal canal extends downward, posteriorly, and laterally through the lateral nasal wall.
d. The lining of the canaliculi is a stratified squamous epithelium, whereas that of the nasolacrimal sac and canal is a bilayered columnar epithelium.
e. The medial portion of the eyelid is more susceptible to tearing injuries because of absence of the tarsal plate.

54. Which one of the following statements about congenital impatency of nasolacrimal system is false?
a. Congenital impatency may mimic a medial canthal hemangioma.
b. Acute dacryocystitis is uncommon.
c. The defect in canalization is within the intraosseous portion of the nasolacrimal duct.
d. Common symptoms include epiphora and mucus discharge.
e. If canalization has not spontaneously occurred by the age of 12 to 15 months, it is unlikely to do so.

55. T or F Initial probing with irrigation of an impatent nasolacrimal system is successful 90% of the time.

56. T or F The neural crest migrates in toward the anterior portion of the optic cup in several waves, forming the entire cornea.

57. T or F In megalocornea, the cornea is normal except for its excessive diameter.

58. T or F Keratoglobus is frequently associated with Marfan’s syndrome.

59. Failure of proper migration and differentiation of neural crest cells may lead to all of the following disorders except:
a. internal ulcer of von Hippel.
b. Scheie’s syndrome.
c. Peters’ anomaly.
d. Rieger’s syndrome.
e. posterior embryotoxon.

60. Features that serve to distinguish congenital hereditary endothelial dystrophy (CHED) from congenital glaucoma include:
   1. intraocular pressure (IOP).
   2. epithelial edema.
   3. corneal diameter.
   4. corneal thickness.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

61. Features that serve to differentiate congenital hereditary endothelial dystrophy (CHED) from congenital hereditary stromal dystrophy include:
   1. intraocular pressure (IOP).
   2. epithelial edema.
3. corneal diameter.
4. corneal thickness.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

62. A 15-year-old boy presents with a 2-week history of malaise, low-grade fever, and headache. Four days before presentation, he developed a severe sore throat and bilateral upper eyelid swelling. Examination shows a mildly ill boy with a temperature of 100.1°F, moderately severe pharyngitis, mildly tender cervical lymphadenopathy, mildly tender enlarged lacrimal glands, and a maculopapular rash on all four extremities. A likely etiology for all the findings would be:
   a. “strep throat.”
   b. infectious mononucleosis.
   c. sarcoidosis.
   d. diphtheria.
   e. Kawasaki's disease.

63. T or F The crystalline deposits seen in the cornea in infantile cystinosis are found nowhere else in the eye.

64. Corneal ulceration and scarring seen in familial dysautonomia (Riley-Day syndrome) is secondary to:
   1. impaired epithelial-stromal adherence.
   2. impaired corneal sensation.
   3. impaired humoral immune responses.
   4. decreased tearing.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

65. Which of the following statements about the epidemiology of infantile glaucoma is/are true?
   1. Most cases of infantile glaucoma are bilateral.
   2. Subsequent offspring of parents with an affected child have approximately a 5% chance of manifesting the condition.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

66. T or F The basic pathophysiologic mechanism in all cases of infantile glaucoma involves an imperforate membrane covering the angle.

67. Symptoms and/or signs of infantile glaucoma include:
   1. tearing.
   2. enlargement of corneal diameter.
   3. loss of corneal clarity.
   4. photophobia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

68. T or F Acute ruptures in Descemet's membrane associated with infantile glaucoma are typically vertical, whereas those associated with birth trauma are typically horizontal.

69. T or F Corneal clouding or tearing is more likely to be the presenting symptom in patients with glaucoma in whom the onset is before the age of 3 months, whereas corneal enlargement will probably be the presenting finding in older infants.

70. T or F Like optic nerve cupping, an afferent pupillary defect carries little prognostic significance in infantile glaucoma.

71. Recognized methods for long-term management of infantile glaucoma include:
   1. trabeculotomy.
   2. trabeculectomy.
   3. goniotomy.
   4. oral carbonic anhydrase inhibitors.
Questions

72. Which of the following is/are considered a negative prognostic factor(s) in infantile glaucoma?
   1. presence of an afferent pupillary defect.
   2. corneal diameter >14 mm at the time of diagnosis.
   3. onset at <3 months of age.
   4. optic nerve cupping.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

73. Systemic evaluation of the neonate with glaucoma should include:
   1. serum galactose levels.
   2. serum antirubella immunoglobulin G (IgG) levels.
   3. serum phytanic acid levels.
   4. urinalysis for proteinuria and aminoaciduria.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

74. Which of the following agents used in general anesthesia tend to lower intraocular pressure (IOP)?
   1. halothane.
   2. ketamine.
   3. thiopental.
   4. succinylcholine.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

75. T or F The upper end of normal intraocular pressure (IOP) for infants and children is felt to be the same as that for adults—22 mm Hg.

76. Which of the following statements about simple ectopia lentis is/are true?
   1. The dominant variety is more common than the recessive variety.
   2. The recessive variety is bilateral, whereas the dominant variety is unilateral.
   3. The recessive variety may be accompanied by a displaced and abnormally shaped pupil.
   4. All dominant forms of the disorder develop by age 20 years.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

77. Which of the following statements about Marfan’s syndrome is/are true?
   1. Most cases are sporadic.
   2. Most patients with the full syndrome develop ectopia lentis.
   3. The average refraction on patients with the syndrome reveals moderate hyperopia.
   4. Systemic treatment of the condition may include propranolol and antibiotic prophylaxis before dental procedures.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

78. Which of the following statements about homocystinuria is/are true?
   1. This disorder is inherited on an autosomal-dominant basis.
   2. Most patients will develop ectopia lentis.
   3. Systemic mortality is caused by coagulopathy with clotting deficiency and bleeding diatheses.
   4. The primary defect leading to lens dislocation is a structural deficiency in the zonules.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

79. Ectopia lentis associated with mental retardation may be seen in which of the following disorders:
   2. hyperlysinemia.
   3. Down’s syndrome.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

80. Which of the following types of congenital cataract do not require systemic laboratory evaluation?
   1. bilateral cataract with no family history.
   2. monocular cataract with no family history.
   3. cataract associated with retinal pigment epithelial abnormalities.
   4. posterior lenticous.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

81. In which of the following maternal–fetal infections is congenital cataract virtually unheard of?
   a. toxoplasmosis.
   b. rubella.
   c. cytomegalic inclusion disease.
   d. herpes simplex virus (HSV) infection.
   e. syphilis.

82. Which of the following congenital cataract scenarios mandates the most urgent surgical intervention?
   a. monocular anterior polar cataract.
   b. binocular posterior lenticous.
   c. monocular lamellar cataract.
   d. monocular nuclear cataract.
   e. binocular nuclear cataract.

83. Which one of the following statements about alterations in surgical strategy for pediatric cataract extraction (relative to adults) is false?
   a. Intracapsular surgery is generally avoided in younger patients because of more prominent hyaloideocapsular attachments.
   b. Extracapsular nuclear expression is generally avoided because of the small, relatively soft nucleus of the juvenile cataract.
   c. Pediatric cataracts are usually soft and can be aspirated entirely.
   d. Primary posterior capsulectomy is generally not undertaken because of more prominent hyaloideocapsular attachments in the young eye.
   e. One or more peripheral iridectomies are generally indicated by the higher risk of exaggerated inflammation and by secondary pupillary block.

84. The systemic evaluation of a patient with bilateral congenital cataracts might include which of the following?
   a. bilateral audiograms.
   b. serum calcium level.
   c. urinalysis.
   d. karyotyping.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

85. Which of the following disorders should be considered in the differential diagnosis of congenital nystagmus associated with photophobia?
   a. oculocutaneous albinism.
   b. Leber’s congenital amaurosis.
   c. achromatopsia.
   d. cystinosis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
Questions

86. Which of the following statements about juvenile rheumatoid arthritis (JRA) is/are true?
   1. It is the most common etiology of anterior uveitis in the pediatric population.
   2. Although it may have onset at any age, it is extraordinarily rare in children younger than 2 years.
   3. To conclude that uveitis is associated with JRA, there must be an antecedent history of joint symptoms.
   4. The uveitis of JRA is nearly always entirely anterior.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

87. Which of the following statements about the five subtypes of juvenile rheumatoid arthritis (JRA) is/are true?
   1. With the exception of the pauciarticular, late-onset type (HLA-B27 positive), all of the subtypes of JRA are considerably more common in girls.
   2. The group at highest risk of developing anterior uveitis is the pauciarticular early-onset group with rheumatoid factor negative and antinuclear antibody negative.
   3. Iridocyclitis is frequently seen as part of the syndrome of systemic JRA (Still’s disease).
   4. The joints involved in patients with iridocyclitis are typically the large joints (e.g., knee, ankle, and elbow).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

88. Which of the following statements about the clinical manifestations of juvenile rheumatoid arthritis (JRA) is/are true?
   1. The findings of chronic uveitis may be discovered incidentally.
   2. Visual loss caused by amblyopia is rarely a problem.
   3. Surgery for band keratopathy may be indicated for photophobia and discomfort, as well as for visual loss.
   4. Cataract surgery with intraocular lens implantation may be indicated in cases of visually significant cataract.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

89. Other etiologies of uveitis and joint complaints in children that must be considered in the differential diagnosis of juvenile rheumatoid arthritis (JRA) include:
   1. inflammatory bowel disease.
   2. sarcoidosis.
   3. Lyme disease.
   4. herpes simplex virus (HSV) infection.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

90. Which one of the following statements about the pediatric uveitis associated with herpes zoster virus (HZV) is false?
   a. Typically, the uveitis develops during convalescence from acute varicella infection.
   b. The uveitis in (reactivated) zoster ophthalmicus may have both anterior and posterior components.
   c. Reactivation disease also may be accompanied by a keratitis, either epithelial or stromal.
   d. In the setting of immunosuppression, reactivation disease should be treated with systemic as well as topical corticosteroids.
   e. Those at risk for reactivation disease include patients with acute leukemia and patients who have undergone post renal transplantation.

91. T or F Pediatric uveitis caused by herpes simplex virus (HSV) infection is usually accompanied by keratitis.

92. T or F More than 50% of uveitis in the pediatric population has a posterior component.
93. The most common etiology of posterior uveitis in the pediatric population is:
   a. toxocarasis.
   b. toxoplasmosis.
   c. syphilis.
   d. sarcoidosis.
   e. idiopathic.

94. Tissues in which the *Toxoplasma* parasite survives the best include:
   1. hepatic cells.
   2. cerebral neurons.
   3. red blood cells.
   4. ganglion cells.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

95. Manifestations of acquired systemic toxoplasmosis include all the following except:
   a. acute arthritis.
   b. rash.
   c. meningocencephalitis.
   d. influenzalike syndrome.
   e. retinitis.

96. T or F Recurrent toxoplasmosis frequently presents as a granulomatous anterior uveitis.

97. Which one of the following statements about ocular histoplasmosis is false?
   a. Symptoms consistent with histoplasmosis include a flulike syndrome and malaise.
   b. There is a geographic, but not a seasonal, predilection for the development of systemic or ocular histoplasmosis.
   c. The vitritis that may accompany the ocular infection may lead to decreased visual acuity.
   d. The initial infection consists of a choroidal granuloma.
   e. Although skin testing may support the diagnosis, it may lead to worsening of the macular disease.

98. Which of the following statements about toxocarasis is/are true?
   1. The infectious cycle in humans generally starts with the consumption of fecally contaminated soil.
   2. The condition may present as a peripheral granuloma in an otherwise quiet eye.
   3. There may be an associated peripheral eosinophilia.
   4. The associated uveitis is caused by hypersensitivity reaction to a living organism.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

99. Which of the following statements about idiopathic pars planitis is/are true?
   1. It may have a mild course, with floaters as the only symptom.
   2. Peripheral retinal periphlebitis is frequently associated.
   3. Infectious etiologies are usually not found.
   4. It is usually unilateral.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

100. Findings in a patient with known juvenile rheumatoid arthritis (JRA) and uveitis that should prompt an increase in topical steroid administration include:
    1. worsening cataract.
    2. flare.
    3. band keratopathy.
    4. aqueous cells.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
101. Which one of the following statements about persistent hyperplastic primary vitreous (PHPV) is false?
   a. Mittendorf’s dots and Bergmeister’s papillae may be considered mild variants of the disorder.
   b. In severe cases, fibrovascular overgrowth within the primary vitreous may invade the lens substance itself.
   c. A common complication is glaucoma, secondary to either vitreous hemorrhage or angle closure.
   d. The presence of dense leukocoria in an eye that is abnormally small suggests the diagnosis of retinoblastoma rather than PHPV.
   e. The condition is typically unilateral.

d. 4 only.

e. 1, 2, 3, and 4.

106. T or F Zone 1 is considered to be a circular area with a diameter of 30 degrees centered around the optic nerve.

107. The CRYO-ROP (Cryotherapy for Retinopathy of Prematurity) study established that cryotherapy will reduce the incidence of unfavorable outcomes (i.e., posterior retinal detachment, fixed macular folds, or retrolental fibroplasia) in the eyes with which of the following criteria?

   1. stage 3 disease or worse.
   2. “plus” disease.
   3. involvement of at least five contiguous or eight interrupted clock hours.
   4. disease involving only zone I or zone II.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

108. Sequelae of retinopathy of prematurity (ROP) may include:

   1. angle-closure glaucoma.
   2. cataract.
   3. pseudoexotropia.
   4. pseudoesotropia.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

109. Which of the following statements about screening protocols for premature infants is/are true?

   1. Only infants of birth weight <1,500 g require screening.
   2. Initial examination should take place 4 to 6 weeks after birth or at 30 weeks’ gestational age, whichever is later.
   3. In patients with mild or no retinopathy of prematurity (ROP), examinations may be conducted every 2 weeks.
   4. In patients with threshold ROP, cryotherapy should be delivered within 72 hours.
10. Which one of the following statements about Coats’ disease is false?
   a. True Coats’ disease is a disorder of childhood, more often affecting boys.
   b. The condition is more commonly bilateral than unilateral.
   c. Diagnosis of Coats’ disease may not be made in the setting of subretinal exudate without obvious abnormal retinal vessels.
   d. Full treatment involves obliteration of the abnormal vessels and subsequent treatment of associated retinal detachment.
   e. In up to one half of untreated cases, the condition may be nonprogressive.

110. Which of the following statements about Coats’ disease is false?
   a. False Coats’ disease is a disorder of childhood, more often affecting boys.
   b. The condition is more commonly bilateral than unilateral.
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   d. Full treatment involves obliteration of the abnormal vessels and subsequent treatment of associated retinal detachment.
   e. In up to one half of untreated cases, the condition may be nonprogressive.

111. T or F Juvenile cataracts associated with type 1 diabetes mellitus generally develop independent of the level of blood sugar control.

112. Children of mothers with diabetes are at increased risk for the development of:
   a. Coats’ disease.
   b. Pigmentary glaucoma.
   c. Aniridia.
   d. Optic nerve hypoplasia.
   e. Pseudotumor cerebri.

113. The most common fundus finding in a patient with acute leukemic oculopathy is:
   a. Choroidal infiltration (creamy elevated subretinal patches).
   b. Nerve fiber layer hemorrhages.
   c. Cotton-wool spots.
   d. Roth’s spots.
   e. Optic disc edema.

114. T or F A patient with known acute leukemia who presents with disc edema and loss of vision should be evaluated for radiotherapy within the next 2 weeks.

115. T or F Patients with leukemic optic neuropathy may be more sensitive to radiation neuropathy than other patients.

116. Other ocular manifestations common in leukemic oculopathy include:
   1. Uveitic glaucoma.
   2. Spontaneous hyphemas.
   3. Iris heterochromia.
   4. Cataract.

117. Which of the following statements about the gangliosidoses is/are true:
   1. The most common gangliosidose is GM2, type I (Tay-Sachs disease).
   2. Inheritance is generally on an X-linked recessive basis.
   3. Prominent cherry-red spots are typically seen in Tay-Sachs and Sandhoff’s diseases.
   4. Patients generally succumb to neurologic deterioration in their late teens or early twenties.

118. Which one of the following statements about the oculorenal syndromes is false?
   a. Lowe’s syndrome is inherited on an X-linked recessive basis.
   b. Female carriers of Lowe’s syndrome may be detected by punctate cortical opacities of the lens.
   c. The most common ocular disorder in Lowe’s syndrome is glaucoma.
   d. The most common ocular finding in Alport’s syndrome is anterior lenticonus and/or anterior polar cataract.
   e. Unlike Alport’s syndrome, Senior-Loken syndrome features a progressive retinal degeneration with profound visual symptoms.

119. Ocular findings consistent with renal failure from any etiology include:
   1. Exudative retinal detachment.
   2. Calcium crystals in the conjunctiva.
3. macular edema.
4. diffuse arteriolar attenuation.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

120. T or F In albinism, more ganglion cell fibers decussate at the chiasm than in normal visual pathways.

121. T or F The tyrosinase-negative type of albinism generally has more severe clinical findings than the tyrosinase-positive type.

122. T or F The vitreous is normal in juvenile retinoschisis.

123. Reliable methods of distinguishing juvenile retinoschisis from Goldmann-Favre dystrophy include:

1. macular examination.
2. electrooculography (EOG).
3. electroretinogram (ERG).
4. careful family history.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

124. Fundus findings that may be seen in association with tapetoretinal degeneration include:

1. cystoid macular edema.
2. vitreous cells.
3. posterior subcapsular cataract.
4. subretinal exudation.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

125. T or F Most cases of sector retinitis pigmentosa ultimately progress to macular involvement with a poor visual prognosis.

126. The most common underlying disorder in a patient with a “bull’s-eye” maculopathy is:

a. Stargardt’s disease.
b. cone dystrophy.
c. chloroquine retinopathy.
d. hydroxychloroquine retinopathy.
e. Best’s disease.

127. Which one of the following statements about the various forms of congenital stationary night blindness (CSNB) is false?

a. Retinitis punctata albescens is associated with dots deep in the retina.
b. Fundus albipunctatus reveals normalization of the scotopic electrotretinography after prolonged dark adaptation (after 3 to 12 hours).
c. One recessive form is frequently associated with high myopia.
d. One variety may have a normal scotopic a-wave with no apparent b-wave.
e. Oguchi’s disease displays the Mizuo phenomenon: a golden sheen of the retina returning to normal after several hours of dark adaptation.

128. T or F Visual acuity, although usually better in blue cone monochromatism than rod monochromatism, is not reliable for distinguishing between the two.

129. Clinical findings associated with Leber’s congenital amaurosis include:

1. oculodigital sign.
2. keratoconus.
3. high hyperopia.
4. sensorineural hearing loss.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

130. T or F In most cases designated as Stargardt’s disease, the presenting symptom is night blindness.

131. T or F Visual function in the pattern dystrophies of the retinal pigment epithelium (RPE) is usually good.
132. All of the following are features of Aicardi’s syndrome except:
   a. X-linked recessive inheritance.
   b. agenesis of the corpus callosum.
   c. infantile spasms.
   d. lacunar chorioretinal degeneration.
   e. severe mental retardation.

133. T or F Morning glory disc anomaly is generally an incidental finding with little functional significance.

134. T or F Colobomata involving the optic nerve may be associated with nonrhegmatogenous retinal detachment.

135. T or F Medullation of optic nerve fibers generally begins at the optic chiasm and is completed by the end of the first month of life.

136. T or F Myelinated nerve fibers are more common in boys than in girls.

137. Findings consistent with the tilted disc syndrome include:
   1. prominence of the superior portion of the disc.
   2. an inferior or inferonasal scleral crescent.
   3. situs inversus.
   4. binasal field defects.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

138. Which of the following statements about optic nerve hypoplasia is/are true?
   1. The condition may be unilateral or bilateral.
   2. Visual acuity may vary from normal to no light perception.
   3. A classic finding is the double ring sign.
   4. The association of optic nerve hypoplasia, the absence of the septum pellucidum, and the presence of midline central nervous system anomalies and hypothalamic–pituitary abnormalities are stronger for unilateral than for bilateral optic nerve hypoplasia.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

139. The most common location for optic disc pits is:
   a. superonasal.
   b. superotemporal.
   c. inferotemporal.
   d. inferonasal.
   e. central.

140. Which of the following techniques aid(s) in distinguishing pseudopapilledema with buried drusen from true papilledema?
   1. red free photographs.
   2. computed tomography (CT) scanning.
   3. visual fields.
   4. ultrasonography.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

141. T or F The prognosis for visual recovery is better for the optic neuritis associated with Devic’s disease than for idiopathic optic neuritis.

142. Which one of the following statements about histiocytosis X is false?
   a. These disorders reflect abnormal proliferation of dendritic histiocytes.
   b. Radiographically, these tumors produce osteosclerosis.
   c. Conventional radiography will frequently disclose a greater number of bony lesions than are apparent clinically.
   d. The classic triad in Hans-Schüller-Christian disease is diabetes insipidus, lytic skull lesions, and proptosis.
   e. Patients with Letterer-Siwe disease are frequently very ill.

143. Options for the treatment of the histiocytoses include:
   1. curettage.
   2. intralesional steroid injections.
   3. low-dose radiation.
   4. systemic steroid therapy.
144. Prognosis for histiocytoses is unfavorable in children with:
   1. age of disease onset younger than 2 years.
   2. multiple lesions of the skull.
   3. hepatic or bone marrow involvement.
   4. bilateral proptosis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

145. Which one of the following statements about the fibroosseous disorders of the orbit is false?
   a. The distinction between fibrous dysplasia and ossifying fibroma is generally made radiologically.
   b. Fibrous dysplasia may radiologically be either sclerotic or lytic.
   c. Generally, fibrous dysplasia stabilizes after skeletal maturity is attained.
   d. The polyostotic variety of fibrous dysplasia may be accompanied by sexual precocity and hyperpigmented skin macules.
   e. The most significant visual implication of fibroosseous orbital lesions is optic nerve compression.

146. Which one of the following statements about capillary hemangiomas is false?
   a. Systemic interferon may lead to involution.
   b. They are more common in girls than in boys.
   c. They characteristically blanch with pressure.
   d. Phlebolith formation is common.
   e. Indications for treatment include occlusion amblyopia and/or significant astigmatism.

147. **T or F** The pathology of the nevus flammeus (port-wine stain) is consistent with cavernous dilation of dermal blood vessels and cellular proliferation.

148. Which of the following statements about lymphangiomas is/are true?
   1. Lymphangiomas are primarily a disorder of the pediatric age range.
   2. Superficial lesions may have a bluish or violaceous hue.
   3. Classic presenting symptoms include spontaneous ecchymosis, and proptosis with crying or following upper respiratory infections.
   4. Surgical intervention is indicated early in the course of the disorder in order to remove the tumor when it is small.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

149. Which lesion(s) may be associated with proptosis with the Valsalva maneuver or crying?
   1. orbital lymphangioma.
   2. capillary hemangioma.
   3. orbital varix.
   4. orbital cavernous hemangioma.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

150. Which of the following statements about the epidemiology of rhabdomyosarcoma is/are true?
   1. It is one of the most common soft-tissue malignancies in children.
   2. It is the most common solid malignant tumor of the orbit in children.
   3. A common presentation is an orbital cellulitis-like picture.
   4. The average age at diagnosis is 7 years.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

151. **T or F** Any child with unexplained acquired ptosis should have radiographic imaging of the orbit to rule out rhabdomyosarcoma.
152. Which of the following statements about the histopathology of rhabdomyosarcoma is/are true?

1. The embryonal type is the most common.
2. The embryonal type has the best prognosis.
3. The alveolar type has the worst prognosis.
4. The differentiated (pleomorphic) type is the second most common.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

153. The proper intervention for newly diagnosed rhabdomyosarcoma is:

a. exenteration.
b. radiotherapy.
c. chemotherapy.
d. a., b., and c.
e. b. and c.

154. Which of the following statements about the neurilemmoma (schwannoma) is/are true?

1. Most patients with neurofibromatosis will develop at least one lesion.
2. The lesion can be exquisitely tender or painful.
3. Malignant degeneration is common.
4. There are two classic histopathologic patterns.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

155. Which of the following statements about neurofibroma is/are true?

1. The nodular neurofibroma is the most specific for neurofibromatosis.
2. Like schwannomas, neurofibromas grow in close relation to peripheral nerves.
3. Neurofibromas are generally osteosclerotic.
4. The association of neurofibroma with congenital glaucoma is strongest with lesions of the upper eyelid.

a. 1, 2, and 3.
b. 1 and 3.

156. Which of the following features might be used to differentiate between neurofibroma and neurilemmoma?

1. the presence of axons and perineural cells in the neurofibroma.
2. the presence of Schwann’s cells in the neurilemmoma.
3. the presence of a true capsule around a neurilemmoma.
4. positive S-100 staining for neurofibroma.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

157. Which of the following statements about optic nerve glioma is/are true?

1. The age range with the highest incidence is 2 to 6 years.
2. Two common means of presentation include visual loss and proptosis.
3. The classic radiographic appearance of the lesion on computed tomography (CT) scanning is a fusiform enlargement of the optic nerve.
4. A syndrome strikingly similar to spasmus nutans may be seen in gliomas involving the hypothalamus or optic-chiasm.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

158. Which of the following statements about the treatment of optic nerve gliomas is/are true?

1. Biopsy findings are important in guiding appropriate therapy.
2. Extent of spread is important in guiding appropriate therapy.
3. Tumors arising from the optic nerve have a poorer prognosis than those arising from the optic chiasm.

4. Increased intracranial pressure and involvement of contiguous central nervous system (CNS) structures confer a poorer prognosis.

159. T or F Meningiomas are more likely to appear osteolytic than osteoblastic on radiographic studies.

160. Which of the following statements about the epidemiology of neuroblastoma is/are true?

1. In some pediatric series, the incidence of neuroblastoma is greater than that of rhabdomyosarcoma.
2. The second most common site of origin is the retroperitoneal sympathetic chain.
3. The site of origin is the adrenal gland in at least half of the cases.
4. This tumor presents as metastases in over half of the cases.

162. All of the following are features of the histopathology of metastatic neuroblastoma except:

a. sheets of indistinct round cells with scanty cytoplasm.

b. copious mitotic figures.

c. areas of tumor necrosis.

d. Homer-Wright rosettes.

e. bony invasion.

163. Which of the following is/are considered ominous prognostic factors for metastatic neuroblastoma?

1. liver metastases.
2. age <1 year.
3. bone marrow metastases.
4. bone metastases.

164. The paraneoplastic syndrome most commonly associated with metastatic neuroblastoma is:

a. photoreceptor degeneration.

b. optic neuropathy.

c. opsoclonus.

d. facial myokymia.

e. cerebellar degeneration.

165. Which of the following statements about Ewing's sarcoma is/are true?

1. Like neuroblastoma, this tumor may present with an orbital cellulitislike picture.
2. Invasion of the globe is common.
3. The age at onset is older than that for neuroblastoma.
4. Unlike neuroblastoma, there is no role for radiotherapy.
10. Pediatric Ophthalmology and Strabismus

166. The histopathologic “starry sky” (i.e., histiocytes scattered amidst a monotonous background of lymphocytes) is the classic appearance of:
   a. chloroma.
   b. Burkitt’s lymphoma.
   c. Wilms’ tumor.
   d. metastatic neuroblastoma.
   e. Ewing’s sarcoma.

167. Which one of the following statements about ocular adnexal dermoid cysts is false?
   a. They are choristomatous arrests of epithelial tissue.
   b. The most common location is the superonasal orbital rim.
   c. Generally, they do not enlarge after the first year of life.
   d. Rupture may lead to an orbital cellulitis like picture.
   e. Radiography of orbital lesions generally demonstrates bony excavation.

168. Which of the following may be considered features that distinguish pediatric orbital pseudotumor from that seen in adults?
   2. More frequent constitutional symptoms such as headache, fever, vomiting, and lethargy in children.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

169. The epibulbar lesion most commonly seen in children younger than 15 years is:
   a. dermoid.
   b. pyogenic granuloma.
   c. dermolipoma.
   d. nevus.
   e. epithelial inclusion cyst.

170. Findings consistent with the Goldenhar–Gorlin syndrome include:
   1. vertebral anomalies.
   2. eyelid colobomas.
   3. limbal dermoids.
   4. aniridia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

171. **T or F** Other than those seen in Goldenhar’s syndrome, limbal dermoids are rarely associated with other abnormalities.

172. **T or F** Limbal dermoids are generally more difficult to excise completely than dermolipomas are.

173. Which of the following statements about the incidence of retinoblastoma is/are true?
   1. The most frequent age at diagnosis is 18 months.
   2. Ninety percent of cases are diagnosed by the age of 3 years.
   3. Almost 95% of newly diagnosed cases will have no family history of retinoblastoma.
   4. The most reliable clue to the presence of a new germline mutation is unilateral involvement.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

174. **T or F** Germline mutations leading to retinoblastoma are inherited on an autosomal-dominant basis with 100% penetrance.

175. Which of the following statements about the genetics of inherited retinoblastoma is/are true?
   1. The locus is on the long arm of chromosome 13 (13q).
   2. The gene product of interest induces malignant transformation if produced in sufficient concentration.
3. The chromosomal defect is expressed on an autosomal-recessive basis.
4. The effects of the retinoblastoma gene are expressed only within the eye.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

176. A couple gives birth to a child who, at the age of 9 months, is diagnosed with bilateral retinoblastoma. There is no previous family history of the disorder. Which one of the following statements about this situation is incorrect?

a. The child most likely carries one abnormal copy of chromosome 13 in each of his cells.
b. The chance of this child having an affected brother or sister is approximately 6%.
c. Either the mother or the father must carry an abnormal copy of chromosome 13 in their germ cells.
d. The child’s life expectancy is less than normal.
e. Chromosomal analysis on the parents and the affected child could elucidate the origin of the genetic defect.

177. Two years later, the same couple gives birth to another child who, at the age of 15 months, is diagnosed with bilateral retinoblastoma. When the parents inquire about the probability of their next child developing retinoblastoma, they should be told that the probability is approximately:

a. <1%.
b. 6%.
c. 25%.
d. 40%.
e. 80%.

178. The probability of a patient who survives bilateral retinoblastoma giving birth to a child with retinoblastoma is:

a. <1%.
b. 6%.
c. 25%.
d. 40%.
e. 80%.

179. Patients who have received radiation therapy for bilateral retinoblastoma are at increased risk for the development of:

1. osteogenic sarcoma of the long bones.
2. osteogenic sarcoma of the orbital bones.
3. malignant melanoma of the eyelids.
4. leiomyosarcomas of the eye or orbit.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

180. T or F The prognosis for survival in patients who develop a second malignancy after treatment for retinoblastoma is poor.

181. Which of the following is the least common presentation for retinoblastoma?

a. decreased vision.
b. strabismus.
c. orbital cellulitis.
d. incidental finding.
e. leukocoria.

182. Which of the following is the most common presentation for retinoblastoma?

a. decreased vision.
b. strabismus.
c. orbital cellulitis.
d. incidental finding.
e. leukocoria.

183. Clinical features that are particularly helpful in the diagnosis of retinoblastoma include:

1. spontaneous hyphema.
2. ophthalmoscopically evident calcification.
3. heterochromia iridis.
4. vitreous seeding.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

184. T or F Retinoblastoma that grows into the vitreous humor in a mushroom or spherical shape is termed “exophytic.”
185. T or F If a child’s nystagmus has a rotary component, congenital nystagmus may be eliminated from the differential diagnosis.

186. Blood vessels in a retinoblastoma may absorb released nucleic acids from the necrotic cells, and consequently take on what appearance microscopically?
   a. eosinophilia.
   b. basophilia.
   c. fibrinoid necrosis.
   d. xanthomatization.
   e. congophilia.

187. T or F An interesting light microscopic characteristic of retinoblastoma is zonal necrosis of tumor surrounding blood vessels.

188. T or F The genetic implications of retinoma (retinocytoma) are identical to those of retinoblastoma.

189. Which of the following is the most common site of retinoblastoma spread outside the eye?
   a. skull bones.
   b. liver.
   c. lymph nodes.
   d. central nervous system (CNS).
   e. distal bones.

190. T or F Case reports of pineal gland neoplasms associated with retinoblastoma probably represent central nervous system (CNS) metastasis.

191. Which of the following might be considered part of a workup for a patient with suspected retinoblastoma?
   1. aqueous humor paracentesis.
   2. computed tomography (CT) scanning.
   3. serum levels of carcinoembryonic antigen.
   4. cranial magnetic resonance imaging (MRI).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

192. Metastatic workup for a patient with established retinoblastoma should include:
   1. bone marrow biopsy.
   2. bone scan.
   3. lumbar puncture.
   4. liver-spleen scan.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

193. Which one of the following statements about the treatment of retinoblastoma is false?
   a. For large tumors, treatment generally includes enucleation.
   b. In advanced or metastatic cases, chemotherapy is used.
   c. Photocoagulation may be used for small, obviously localized tumors.
   d. Cryotherapy is avoided because it typically leads to dissemination of viable tumor cells within the eye.
   e. Cobalt plaque therapy has been used in eyes that have incompletely responded to external beam irradiation.

194. Ocular disorders associated with optic nerve hypoplasia include:
   1. albinism.
   2. aniridia.
   3. coloboma.
   4. Duane’s syndrome.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

195. The typical color of a juvenile xanthogranuloma (JXG) is:
   a. black.
   b. pearly.
   c. orange.
   d. blue.
   e. salmon.

196. T or F The most common location of extraocular juvenile xanthogranuloma (JXG) is skeletal muscle.
197. **T or F** The skin lesions of juvenile xanthogranuloma (JXG) are generally left untreated, whereas iris lesions generally mandate intervention.

198. Which one of the following statements about medulloepithelioma (diktyoma) is false?
   - a. The cell of origin is probably nonpigmented ciliary epithelium.
   - b. Like hemangiopericytoma, tumors with benign histopathologic features hide a significant metastatic potential.
   - c. A teratoïd variant exists that may contain cartilage, muscle, or neural tissue.
   - d. Leukocoria may be the presenting finding.
   - e. Enucleation before transscleral extension is frequently curative.

199. A 17-year-old girl undergoes dilated fundusscopic examination after being fitted for contact lenses. A creamy orange, geographic placoid elevation deep to the retina is noted to extend superotemporally from the disc margin. There is subretinal fluid in the inferior periphery. Ultrasonography reveals a very highly reflective thickening of choroid in the same region. Computed tomography (CT) scan reveals calcification. The most likely diagnosis is:
   - a. amelanotic melanoma.
   - b. choroidal hemangioma.
   - c. choroidal osteoma.
   - d. choroidal metastasis from an ovarian primary.
   - e. regressed retinoblastoma.

200. Which of the following neoplastic disorders may originate within intraocular structures?
   1. leukemia.
   2. rhabdomyosarcoma.
   3. Letterer-Siwe syndrome.
   4. neuroblastoma.
   - a. 1, 2, and 3.
   - b. 1 and 3.
   - c. 2 and 4.
   - d. 4 only.
   - e. 1, 2, 3, and 4.

201. **T or F** If indicated, prophylactic irradiation following the diagnosis of acute leukemia in childhood should involve both the eyes.

202. Which of the following are found in >90% of adolescents or adults with neurofibromatosis?
   1. *café au lait* spots.
   2. neurofibromas.
   3. Lisch nodules.
   4. gliomas.
   - a. 1, 2, and 3.
   - b. 1 and 3.
   - c. 2 and 4.
   - d. 4 only.
   - e. 1, 2, 3, and 4.

203. Each of the following is seen in increased frequency in patients with neurofibromatosis except:
   - a. malignant melanoma of the uveal tract.
   - b. Wilms’ tumor.
   - c. rhabdomyosarcoma.
   - d. choroidal osteoma.
   - e. schwannomas.

204. **T or F** The light microscopic findings of Lisch nodules are indistinguishable from those of iris nevi.

205. **T or F** The optic nerve gliomas associated with neurofibromatosis generally have a poorer prognosis than isolated lesions.

206. Which of the following tests will always be normal in severe unilateral optic nerve hypoplasia?
   - a. visual acuity test.
   - b. swinging flashlight test.
   - c. electroretinography.
   - d. visual evoked responses (VERs).
   - e. fluorescein angiography.

207. Seizures, mental retardation, and facial angiofibroma form the classic triad for:
   - a. neurofibromatosis.
   - b. tuberous sclerosis.
   - c. von Hippel-Lindau disease.
   - d. ataxia–telangiectasia.

208. **T or F** Adenoma sebaceum is common in tuberous sclerosis and may be confused with acne vulgaris.
209. **T or F** The complete Sturge-Weber syndrome includes facial hemangioma, ipsilateral glaucoma, and ipsilateral epilepsy.

c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

210. **T or F** The classic fundus finding in a patient with Sturge-Weber syndrome is the focal choroidal hemangioma.

211. **T or F** The glaucoma seen ipsilateral to facial hemangioma in Sturge-Weber syndrome is due entirely to elevated episcleral venous pressure (EVP).

212. What percentage of patients with capillary hemangiomas of the retina will develop hemangioblastomas of the cerebellum?

a. 5%.
b. 20%.
c. 50%.
d. 75%.
e. 100%.

213. **T or F** Like Sturge-Weber syndrome, iris neovascularization may complicate the course of retinal angiomatosis.

214. **T or F** The Wyburn-Mason syndrome is always unilateral.

215. Anatomic sites to look for the vascular changes associated with ataxia-telangiectasia include:

1. retroauricular skin.
2. conjunctiva.
3. popliteal fossae.
4. malar skin.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

216. Which of the following immune deviations is/are associated with ataxia-telangiectasia?

1. hypogammaglobulinemia.
2. thymus hypoplasia.
3. monoclonal gammopathies.
4. immunoglobulin A (IgA) deficiency.

a. 1, 2, and 3.
b. 1 and 3.

217. **T or F** The life expectancy of patients with ataxia-telangiectasia is normal.

218. Which of the following disorders is inherited on an autosomal-recessive basis?

a. neurofibromatosis.
b. tuberous sclerosis.
c. Sturge-Weber syndrome.
d. von Hippel-Lindau disease.
e. ataxia-telangiectasia.

219. Which of the following disorders has no clear inheritance pattern?

a. neurofibromatosis.
b. tuberous sclerosis.
c. Sturge-Weber syndrome.
d. von Hippel-Lindau disease.
e. ataxia-telangiectasia.

220. Potential ocular manifestations of the craniosynostoses include:

1. papilledema.
2. exposure keratitis.
3. tortuous retinal vasculature.
4. optic nerve hypoplasia.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

221. The strabismus most frequently associated with the craniosynostoses is:

a. double elevator palsy.
b. Duane's syndrome.
c. V-pattern exotropia.
d. A-pattern esotropia.
e. infantile esotropia.

222. **T or F** In a patient with strabismus associated with craniosynostosis, it is best to correct the strabismus as early as possible to enhance binocular function.
Questions

223. Features of Pierre Robin’s sequence/syndrome include:
   1. cleft palate.
   2. glossoptosis.
   3. micrognathia.
   4. bird face.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

224. Lower lid colobomas, pronounced antimongoloid slant (downward displacement of the lateral canthus), and orbital rim defects are typical of:
   a. Hallermann-Streiff syndrome.
   b. Treacher Collins’ syndrome.
   c. Goldenhar’s syndrome.
   d. Waardenburg’s syndrome.
   e. Pierre Robin’s sequence/syndrome.

225. T or F The difference between primary telecanthus and secondary telecanthus is the presence of true hypertelorism in the former.

226. Ocular findings in fetal alcohol syndrome include all of the following except:
   a. a blepharophimosislike picture.
   b. hypertelorism.
   c. anterior segment dysgeneses.
   d. tortuous retinal vessels.
   e. optic nerve hypoplasia.

227. In which of the following inborn errors of metabolism are corneal findings seen?
   1. mucopolysaccharidosis (MPS) I-H (Hurler’s syndrome).
   2. MPS III (Sanfilippo’s syndrome).
   3. Fabry’s disease.
   4. Tay-Sachs disease.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

228. The pupillary light reflex can be shown to develop at approximately what age?
   a. 20 weeks’ gestation.
   b. 30 weeks’ gestation.
   c. 40 weeks’ gestation.
   d. 4 weeks postnatally (44 weeks postconception).
   e. 8 weeks postnatally (48 weeks postconception).

229. Signs of poor vision in a 1-month-old infant might include:
   1. paradoxical pupillary response to light.
   2. oculodigital sign.
   3. fixation only on extremely bright lights.
   4. skew deviation.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

230. An infant boy is born with a disorder not previously seen in his family except in an elder sister. Five other siblings are not affected. This is most consistent with what type of inheritance?
   a. autosomal dominant.
   b. autosomal recessive.
   c. X-linked dominant.
   d. X-linked recessive.
   e. mitochondrial.

231. Disorders to be considered in a neonate with very poor vision and a normal ocular examination include:
   1. Lowe’s syndrome.
   2. achromatopsia.
   3. optic nerve hypoplasia.
   4. Leber’s congenital amaurosis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

232. Which of the following does not arise from the annulus of Zinn?
   1. superior oblique.
   2. levator palpebrae superioris.
3. inferior oblique.
4. superior rectus.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

233. The action(s) of the medial rectus muscle with the eye in primary position is/are:
   a. adduction, elevation, intorsion.
   b. adduction, depression, intorsion.
   c. adduction and intorsion.
   d. adduction and extorsion.
   e. adduction.

234. To maximize the elevation generated by the superior rectus, how must the eye be rotated from primary position?
   a. adducted 51 degrees.
   b. abducted 51 degrees.
   c. adducted 23 degrees.
   d. abducted 23 degrees.
   e. adducted 67 degrees.

235. To maximize the depression generated by the superior oblique, how must the eye be rotated (see the figure shown below)?
   a. adducted 51 degrees.
   b. abducted 51 degrees.
   c. adducted 23 degrees.
   d. abducted 23 degrees.
   e. adducted 67 degrees.

236. T or F Both oblique muscles are characterized by a physical distinction between the anatomic origin and the mechanical origin.

237. T or F The superior oblique tendon passes between the superior rectus muscle and the globe on the way to its insertion.

238. T or F The inferior oblique muscle passes between the inferior rectus muscle and the globe on the way to its insertion.

239. T or F The superior oblique muscle becomes tendinous after turning through the trochlea.

240. The primary elevator of the eye in primary position is the:
   a. superior oblique.
   b. superior rectus.
   c. inferior oblique.
   d. inferior rectus.
   e. levator muscle.

241. The primary intorter of the globe in primary position is the:
   a. superior oblique.
   b. superior rectus.
   c. inferior oblique.
   d. inferior rectus.
   e. levator muscle.

242. The extraocular muscle with the shortest length of active muscle belly is the:
   a. superior rectus.
   b. inferior rectus.
   c. superior oblique.
   d. inferior oblique.
   e. levator palpebrae superioris.

243. The muscle with the shortest length of tendon is the:
   a. superior rectus.
   b. inferior rectus.
   c. superior oblique.
   d. inferior oblique.
   e. levator palpebrae superioris.
Questions

244. Which one of the following statements about the physiology and anatomy of the extraocular muscles is false?
   a. Each of the rectus muscles receives two muscular arteries, which continue on as anterior ciliary arteries.
   b. Each muscle contains both fast-type (twitch) and slow-type (tonic) muscle fibers.
   c. The slow, tonic-type muscles rely on aerobic metabolism and are innervated by en grappe endings.
   d. The fast, twitch-type muscle fibers rely on glycolysis and are innervated by en plaque nerve endings.
   e. The motor units in extraocular muscle are among the smallest in the human body.

245. T or F Of the extraocular muscles, only the inferior oblique fails to penetrate Tenon’s capsule.

246. T or F Extraconal orbital fat may be encountered as far as 10 mm anterior from the limbus.

247. T or F The average point of entry for a motor nerve into its muscle is approximately the junction of the posterior two thirds and the anterior one third of the active muscle belly.

248. Which of the following changes in lid position is/are consistent with the muscle surgery described?
   1. narrowing of palpebral fissure with superior rectus resection.
   2. narrowing of palpebral fissure with inferior rectus recessed.
   3. narrowing of the palpebral fissure with inferior rectus resection.
   4. narrowing of the palpebral fissure with superior oblique tenotomy.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

249. In young, healthy eyes, anterior segment ischemia becomes a concern after surgery on how many rectus muscles?
   a. 1.
   b. 2.
   c. 3.
   d. 4.
   e. Anterior segment ischemia may follow unpredictably after any muscle surgery.

250. Each of the following is a correct match of muscular synergist and antagonist except:
   a. medial rectus: synergist, superior rectus; antagonist, lateral rectus.
   b. lateral rectus: synergist, superior oblique; antagonist, medial rectus.
   c. superior rectus: synergist, superior oblique; antagonist, inferior rectus.
   d. inferior rectus: synergist, superior oblique; antagonist, superior rectus.
   e. inferior oblique: synergist, superior rectus; antagonist, superior oblique.

251. T or F Hering’s law governs binocular motor function, whereas Sherrington’s law governs monocular motor function.

252. Which one of the following constitutes a violation of Hering’s law?
   a. inhibitional palsy of the contralateral antagonist.
   b. dissociated vertical deviation (DVD).
   c. Brown’s syndrome.
   d. cyclic esotropia.
   e. Duane’s syndrome.

253. Which one of the following constitutes a violation of Sherrington’s law?
   a. inhibitional palsy of the contralateral antagonist.
   b. disassociated vertical divergence.
   c. Brown’s syndrome.
   d. cyclic esotropia.
   e. Duane’s syndrome.

254. T or F By definition, versions are conjugate binocular movements, whereas vergences are disconjugate binocular movements.

255. T or F In both fusional convergence and fusional divergence, there is an obligatory change in the refractive status of the eye.
256. The site of origin of neural impulses leading to a rightward saccade is the:
   a. right frontal lobe.
   b. left frontal lobe.
   c. right parietooccipital lobe.
   d. left parietooccipital lobe.
   e. the site of origin has not been determined.

d. A normal divergence amplitude is 14 prism diopters at distance and is 16 prism diopters at near.
e. Normal vertical fusional amplitude varies from 2 to 4 prism diopters and is independent of fixation distance.

257. The site of origin of neural impulses leading to a leftward pursuit movement is the:
   a. right frontal lobe.
   b. left frontal lobe.
   c. right parietooccipital lobe.
   d. left parietooccipital lobe.
   e. site of origin has not been determined.

258. T or F Physiologically, any point not lying on the empirical horopter will be perceived doubly by the human visual system.

259. T or F Panum’s area of single binocular vision is broader for points in space clustered around the central portion of the empirical horopter and narrower in the periphery.

260. T or F If simultaneous stimulation of retinal areas in two eyes leads to the perception of one image, normal retinal correspondence (NRC) is said to exist.

261. T or F Stereopsis only exists for points in space that lie on the empirical horopter.

262. T or F For fusion to exist, there must be simultaneous stimulation of corresponding retinal areas with normal retinal correspondence.

263. T or F For fusion to exist, the two retinal images must be similar in size and shape.

264. Which one of the following statements about motor fusion is false?
   a. Motor fusion is a means of avoiding diplopia.
   b. Motor fusion is the process by which similar retinal images are made to fall on corresponding retinal areas.
   c. A normal convergence amplitude is 14 prism diopters at distance and is 38 prism diopters at near.

265. T or F The most important visual clues for depth perception require binocular vision.

266. A vertical slit pattern is projected onto the fovea of the right eye, whereas a horizontal slit pattern is projected onto the fovea of the left eye. The subject perceives rapidly alternating images of each pattern—first one and then the other, never simultaneously. This perception is an example of:
   a. fusion.
   b. stereopsis.
   c. suppression.
   d. retinal rivalry.
   e. normal retinal correspondence (NRC).

267. The first cell type in the visual pathway to have receptive fields consisting of circular centers and annular surrounds is the:
   a. photoreceptor.
   b. bipolar cell.
   c. ganglion cell.
   d. lateral geniculate neuron.
   e. occipital cortical neuron.

268. T or F The “critical period” for proper development of the visual pathways is longer in humans than in most experimental animals.

269. T or F Diplopia occurs when each of the two foveae of a single patient contains a distinct retinal image.

270. T or F If a patient with manifest strabismus does not complain of diplopia, then suppression must be active.

271. Which of the following statements about amblyopia is/are true?
   1. The incidence in the general population is approximately 2% to 3%.
   2. The presence of an afferent pupillary defect clearly establishes an organic etiology for visual loss, rather than amblyopia.
3. Patients with amblyopia will frequently perform better with single-symbol acuity test targets than with line targets (crowded stimuli).

4. A neutral density filter placed over an amblyopic eye will generally cause a greater decrement in visual acuity than the same filter placed over an eye with maculopathy.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

272. In which of the following types of strabismus is amblyopia least frequently seen?
   a. infantile esotropia.
   b. esotropia with high accommodative convergence to accommodation ratio (AC/A).
   c. alternating esotropia.
   d. esotropia associated with Duane’s syndrome.
   e. esotropia associated with craniosynostosis.

273. T or F The proper guideline for intervals between examinations for a child undergoing full-time occlusion therapy is 1 week for every month of age.

274. A 7-year-old patient presents to a pediatric ophthalmologist after failing his school vision test. Visual acuity is tested with patching and Snellen targets and is found to be 20/20 in the right eye and 20/50 in the left eye. Motility is full, and there is no apparent tropia on cover–uncover testing. The child has stereoacuity with targets disparate by no less than 60 seconds of arc. Distance Worth four-dot testing reveals fusion. Convergence and divergence amplitudes are normal at distance. The most likely diagnosis is:
   a. cyclic esotropia.
   b. monofixation syndrome.
   c. central fixation with anomalous retinal correspondence.
   d. Duane’s syndrome.
   e. factitious visual loss.

275. The patient in question 274 has no history of previous eye surgery. The remainder of his examination is most likely to disclose:
   a. high axial myopia bilaterally.
   b. retraction of the globe on adduction.
   c. anisometropia >2D.
   d. macular edema.
   e. esotropia developing sometime within the next 24 hours.

276. The most practical and valuable test to perform next on this patient would be:
   a. Lancaster red-green test.
   b. afterimage testing.
   c. Bagolini glass testing.
   d. four-prism–diopter base-out test.
   e. fogging refraction.

277. T or F The patient in questions 274 through 276 must have anomalous retinal correspondence (ARC).

278. A 42-year-old patient presents to the emergency room with a manifest right esotropia. A red glass is placed over the left eye and the patient is asked to fixate at a distant point-light target. In the absence of suppression, and with normal retinal correspondence (NRC), the patient should perceive the red light:
   a. above the white light.
   b. below the white light.
   c. to the right of the white light.
   d. to the left of the white light.
   e. the lights will appear to be superimposed.

279. The patient in question 278 reports that he sees only the red light. The examiner could conclude that:
   1. the patient must have anomalous retinal correspondence.
   2. the motility defect is not recent in origin.
   3. the patient will have normal stereoacuity with his distance correction in place.
   4. the patient may be malingering.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
280. On further questioning, the patient reports that he actually perceives the white light to be to the right of the red light. These images are superimposed with a 10-prism-diopter prism placed base-out over the left eye. Simultaneous prism cover test with a distance target and no red glass reveals a 20-prism-diopter right esotropia. These results indicate:

a. normal retinal correspondence (NRC).
b. harmonious anomalous retinal correspondence (ARC).
c. unharmonious ARC.
d. visual confusion on the patient’s part.
e. total confusion on the examiner’s part—the measurements must be wrong.

281. Which of the following statements about the afterimage test for retinal correspondence is/are true?

1. It is best to flash the vertical line into the fixating eye.
2. Regardless of fixation behavior, in the setting of normal retinal correspondence (NRC), the patient will perceive a cross with a single central gap.
3. A patient with a right exotropia, central fixation, and harmonious anomalous retinal correspondence (ARC) will perceive the vertical line flashed into his right eye as being to the left of the horizontal image placed into his fixating left eye (crossed diplopia).
4. To appropriately interpret this test, the patient’s fixation behavior must be determined.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

282. T or F Testing with Bagolini striated glasses for retinal correspondence requires preparation with cover-uncover testing and assessment of fixation behavior.

283. During routine examination, an alternate-cover test reveals outward fixation shifts of each eye as the cover is moved. The cover-uncover test reveals no shift of either eye as the cover is placed over either eye. The correct description of the patient’s motility status would be:

a. orthophoric, orthotropic.
b. orthophoric, esotropic.
c. orthotropic, esophoric.
d. orthotropic, exophoric.
e. this set of findings is not possible.

284. During a routine examination, the cover-uncover test reveals an outward fixation shift of either eye as the cover is placed over the contralateral eye. The alternate-cover test reveals no shift as the cover is moved back and forth. The correct description of this patient’s motility status would be:

a. orthophoric, orthotropic.
b. orthophoric, esotropic.
c. orthotropic, esophoric.
d. orthotropic, exophoric.
e. this set of findings is not possible.

285. T or F Most of the normal adult population is orthophoric and orthotropic.

286. For a strabismus to be appropriately termed congenital, the disturbance must be documented:

a. at birth.
b. within the first 4 weeks of life.
c. within the first 3 months of life.
d. within the first 6 months of life.
e. within the first 12 months of life.

287. The design of effective occlusion therapy for a toddler with strabismus requires careful evaluation of which of the following factors?

1. age at which the deviation was initially noted.
2. amount of the day during which the child’s eyes are straight.
3. which eye, if either, the patient prefers fixing with.
4. a family history of strabismus.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
Questions

288. Of the following acuity tests, which is least likely to lead to an overestimation of actual recognition acuity?
   a. single Snellen letters.
   b. illiterate E.
   c. optotype cards.
   d. Allen cards.
   e. Landolt’s rings.

289. Which of the following is useful primarily as a screening test for the presence of visual behavior?
   a. Snellen letters.
   b. visual evoked responses (VERs).
   c. preferential looking tests.
   d. illiterate E.
   e. optokinetic nystagmus.

290. Which of the following ocular alignment tests require foveal fixation in the deviated eye for quantification of the angle of strabismus?
   1. the cover–uncover test with prisms.
   2. the alternate-cover test with prisms.
   3. the simultaneous prism-cover test.
   4. the Krimsky test.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

291. A patient with strabismus is asked to fixate on a penlight held by the examiner. The examiner notes that the corneal reflex in the right eye is central, whereas that in the left eye is displaced approximately 3 mm temporal to the center of the pupil. Using Hirschberg’s method for estimating the angle of strabismus, the examiner concludes that the patient has a:
   a. 45-degree esotropia.
   b. 45-prism-diopter esotropia.
   c. 45-degree exotropia.
   d. 45-prism-diopter exotropia.
   e. There is not sufficient information to use Hirschberg’s method.

292. To estimate the deviation for the patient in question 291, the examiner chooses to use the Krimsky method. To do this he should:
   a. perform simultaneous prism-cover testing until there is no net movement of either eye.
   b. place prisms over the nonfixing eye until its light reflex appears centered.
   c. place prisms over the fixing eye until the visuscope target is foveal in the nonfixing eye.
   d. place prisms over the fixing eye until the corneal light reflex from the nonfixing eye appears central.
   e. multiply the distance of the decentered light reflex from the center of the pupil (in millimeters) by 15 to estimate the deviation (in prism diopters).

293. When used with prisms, which of the following is best suited for quantification of a tropia only, with no contribution from a phoria?
   a. cover-uncover test.
   b. alternate-cover test.
   c. Maddox rod testing.
   d. simultaneous prism-cover test.
   e. double Maddox rod testing.

294. T or F The image a subject perceives from a Maddox rod is a real image of a line perpendicular to the orientation of the cylinders in the Maddox rod.

295. T or F To test for a vertical deviation, the Maddox rod should be aligned over the eye with its cylinders running vertically.

296. When tested with a Maddox rod held over the affected eye with its cylinders running horizontally, a patient with new-onset exyclotropia will perceive:
   a. a horizontal line.
   b. a vertical line.
   c. an oblique line running superotemporal to inferonasal.
   d. an oblique line running superonasal to inferotemporal.
   e. a curved line concave toward the nose.

297. An adult with a right esotropia caused by an acquired right abducens paresis is tested with the Lancaster red-green test. He wears the goggles
with the red glass over his right eye and the green glass over his left. An examiner holds the green light central on the chart and gives the patient the red light. The patient is then instructed to superimpose his red light on the examiner’s green light. To the examiner:

a. the red light will appear to the left of the green light.
b. the red light will appear above the green light.
c. the red light will appear to the right of the green light.
d. the red light will appear below the green light.
e. the lights will be superimposed.

298. In the test mentioned in question 297, to the patient:

a. the red light will appear to the left of the green light.
b. the red light will appear above the green light.
c. the red light will appear to the right of the green light.
d. the red light will appear below the green light.
e. the lights will be superimposed.

299. The same patient described in question 297 is retested with the goggles reversed—that is, the green lens over the right eye and the red lens over the left eye. The examiner holds the green light as a fixation target centrally, and the patient moves the red light. This time, the examiner will observe:

a. the red light to the left of the green light at the same distance between the two as before.
b. the red light to the left of the green light at a larger distance between the two than before.
c. the red light to the right of the green light with the same distance between the two as before.
d. the red light to the right of the green light at a larger distance than before.
e. the red light to the right of the green light at a smaller distance than before.

300. If a patient with untreated congenital esotropia is tested with the Lancaster red-green test, when the glasses are reversed and the test is repeated, which one of the following statements is true?

a. The position of the lights on the screen will reverse, and the distance will increase.
b. The position of the lights on the chart will not reverse, and the distance will increase.
c. The position of the lights on the chart will reverse, and the distance will remain the same.
d. The position of the lights on the chart will remain the same.
e. None of the above.

301. Broad nasal bridges with abnormally large angle kappa may lead to an error in the diagnosis of strabismus with which of the following methods?

1. alternate-cover tests.
2. Maddox rod testing.
3. cover-uncover testing.
4. Hirschberg testing.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

302. T or F Temporal displacement of the macula will lead to positive angle kappa.

303. T or F Negative angle kappa simulates esotropia.

304. To accurately quantify an esodeviation, a prism is most appropriately placed over either eye:

a. base-up.
b. base-out.
c. base-down.
d. base-in.
e. apex-out.

305. To quantify a hyperdeviation accurately, a prism is most appropriately placed in front of the deviated eye:

a. base-up.
b. base-out.
c. base-down.
d. base-in.
e. apex-down.

306. T or F A patient whose distance esotropia increases by >10 prism diopters at near is said to have a clinically high accommodative convergence to accommodation ratio (AC/A).

307. A 31-year-old man with moderate hyperopia presents for routine examination. There is a 10-prism-diopter alternating esotropia at distance. While reading through his distance correction at
20 cm, there is a 35-prism-diopter esotropia. Eye movements are full, and he denies any history of prior surgery. It can be concluded that:

a. he has the sequelae of pure, classic infantile esotropia.
b. he must have amblyopia in one eye.
c. he probably has restrictive strabismus.
d. he probably will note double vision if questioned appropriately.
e. he has a high accommodative convergence to accommodation ratio (AC/A).

308. Continuing with the example in question 307, the examiner elects to calculate the patient’s accommodative convergence to accommodation ratio (AC/A). His interpupillary distance is 60 mm, and his near deviation increases to 50 prism-diopters when he views an acuity target through a +1.00D sphere over each eye. By the gradient method, his AC/A measures:

a. 5:1.
b. 11:1.
c. 15:1.
d. 25:1.
e. 50:1.

309. By the heterophoria method, the accommodative convergence to accommodation ratio (AC/A) measures:

a. 5:1.
b. 11:1.
c. 15:1.
d. 25:1.
e. 50:1.

310. In the setting of a heterophoria, fusional vergence amplitudes may be diminished by which of the following?

1. intercurrent illness.
2. fatigue.
3. alcohol consumption.
4. improvement in visual acuity.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

311. T or F Suppression with the distance Worth four-dot test but fusion on near Worth four-dot testing indicates peripheral fusion with no central fusion.

312. A 27-year-old man presents to an ophthalmologist complaining of double vision and difficulty descending stairs since an automobile accident 1 week earlier. The examiner notes that the patient has a left head tilt and concludes that he must have a right superior oblique paresis. Which one of the following findings could not possibly be present if the examiner is correct?

a. right hypertropia in primary position.
b. left hypertropia in right gaze.
c. right hypertropia aggravated by left gaze.
d. right hypertropia aggravated by right gaze.
e. V-pattern esotropia.

313. The muscle that is most likely responsible for aggravation of the right hypertropia seen upon right head tilt with a right superior oblique paresis is the:

a. right superior rectus.
b. right inferior rectus.
c. right inferior oblique.
d. right superior oblique.
e. left inferior rectus.

314. A patient undergoes left orbital exploration for biopsy of a suspicious infiltrate on computed tomography (CT) scanning. Postoperatively, the patient is noted to have a widely dilated pupil and poor vision at near in the left eye. He also complains of binocular diplopia. You note an inability to elevate the eye when it is adducted. What findings would you expect on the three-step test?

a. a right hypertropia worse in right gaze and left head tilt.
b. a left hypertropia worse in left gaze and right head tilt.
c. a right hypertropia worse in right gaze and right head tilt.
d. a left hypertropia worse in right gaze and left head tilt.
e. a right hypertropia worse in left gaze and right head tilt.

315. Atropine is relatively contraindicated as a cycloplegic agent in:

1. albinos.
2. neonates.
3. patients with Down’s syndrome.
4. patients with heart block.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

316. Systemic manifestations of cycloplegic intoxication include:

1. flushing.
2. agitation.
3. tachycardia.
4. somnolence.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

317. T or F The purpose of the prism adaptation test is to demonstrate whether the patient’s deviation is stable over time.

318. Which of the following statements about infantile esotropia is/are true?

1. Although it is typically seen in isolation, it may be associated with neurologic abnormalities.
2. Amblyopia develops as the child cross-fixates.
3. Characteristically, the esotropia is large (>30 prism diopters).
4. There is never an accommodative component.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

319. Findings commonly associated with infantile esotropia on examination include:

1. latent nystagmus.
2. overaction of the inferior obliques.
3. disassociated vertical divergence (DVD).
4. high accommodative convergence to accommodation ratio (AC/A).
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

320. Appropriate options for initial surgical intervention in infantile esotropia include:

1. bimedial recession.
2. bilateral resection.
3. ipsilateral medial rectus recession and lateral rectus resection.
4. bimedial resection.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

321. T or F A monofixation syndrome with peripheral fusion, good cosmesis, and limited stereopsis is a fairly common outcome of successful management of infantile esotropia.

322. T or F Nystagmus blockage syndrome develops purposefully as the patient seeks his or her null point.

323. Parents bring their 3-year-old boy for examination after they note the development of “cross-eyes.” Upon closer review, the parents report that they originally noted the deviation to be present throughout the day at age 2 years. A brief “glance” at the child makes a constant moderate-angle esotropia obvious. Which of the following is/are true?

1. Amblyopia is highly unlikely.
2. With further careful questioning, it may be possible to document that the deviation was originally intermittent.
3. The child should be able to perceive the wings of the Titmus fly in three dimensions.
4. It would not be surprising to find a family history of a similar disorder.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
Questions

d. 4 only.
e. 1, 2, 3, and 4.

324. As part of the comprehensive examination of the patient in question 323, a cycloplegic refraction is performed and reveals +8.50D in both eyes. Which of the following is/are true?

1. There may be bilateral amblyopia.
2. The deviation will certainly be greater at near than at distance.
3. +3.00D lenses are likely to have little effect on the distance deviation.
4. The deviation at distance is likely to measure >50 prism diopters.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

325. The initial step in management of the patient in questions 323–324 must be:

a. penalization with atropine bilaterally.
b. alternate occlusion therapy if the visual acuity is normal bilaterally.
c. full correction of the cycloplegic refractive error.
d. bifocals.
e. bimedial recession.

326. Parents bring their child to the ophthalmologist after noting “cross-eyes.” On further questioning, they report that they noted the deviation to be present throughout the day since the child has been approximately 2 years old. A quick glance at the child reveals an obvious intermittent moderate-angle esotropia. It seems to be larger when the child plays with an object in his hands. Cycloplegic refraction reveals +1.50D in both eyes. Which of the following is/are true?

1. The deviation at distance is not likely to be large.
2. The deviation at near is likely to be moderate (20 to 30 prism diopters).
3. The deviation at near is likely to be lessened with +3.00D lenses over each eye.
4. The accommodative convergence to accommodation ratio (AC/A) is likely to be <5.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

327. Which of the following statements about the treatment of esotropia is/are true?

1. Accommodative esotropia is more likely to require surgical intervention than infantile esotropia.
2. Bifocals are most helpful in the management of patients with refractive accommodative esotropia.
3. If refractive correction fails to solve the problem, the only solution is surgical.
4. Accommodative esotropia may progress over the first 5 to 7 years of life and should be monitored carefully.

a. 1, 2, and 3.
b. 2 and 4.
c. 1 and 3.
d. 4 only.
e. 1, 2, 3, and 4.

328. Clinical features of esotropia that are predictive of the need for future surgical intervention include all of the following except:

a. presence of overaction of the inferior obliques.
b. equal vision.
c. large-angle esotropia (>50 prism diopters).
d. age of onset between 2 and 3 years.
e. low hyperopia or myopia.

329. Which of the following statements about the treatment of accommodative esotropia is/are true?

1. No improvement of esotropia with miotic therapy rules out the possibility of an accommodative component.
2. Delay in refractive correction of an accommodative esotropia increases the probability of a permanent residual esotropia after full correction is given.
3. It is important to attempt surgical realignment before prolonged occlusion therapy.
4. Surgical realignment resulting in a residual esotropia of <10 prism diopters may permit the development of peripheral fusion.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
a. 1, 2, and 3.  
b. 1 and 3.  
c. 2 and 4.  
d. 4 only.  
e. 1, 2, 3, and 4.

330. T or F In accommodative esotropia, patients with fully corrected esotropia due to high hyperopia are more likely to develop stress-induced decompensation in their esotropia than patients with a high accommodative convergence to accommodation ratio (AC/A).

331. In intermittent accommodative esotropia, some ophthalmologists do not prescribe full hyperopic correction because:
   a. distance vision will be blurred.  
   b. the greater deviation at near will not be fully compensated.  
   c. the patient may be converted to a constant esotropia without glasses.  
   d. the patient may become exotropic with full hyperopic correction.  
   e. the problem never becomes constant.

332. Which one of the following statements about cyclic esotropia is false?
   a. The cycle consists of 1 week of orthotropia and 1 week of esotropia.  
   b. The angle of deviation is generally moderate.  
   c. The time of onset is similar to accommodative esotropia.  
   d. Full hyperopic correction may correct the esotropia.  
   e. Amblyopia is possible but relatively uncommon.

333. Divergence insufficiency resembles lateral rectus palsy except that:
   a. there is typically an esodeviation.  
   b. the deviation is generally worse at distance.  
   c. there are commonly no associated neurologic abnormalities.  
   d. the deviation is comitant.  
   e. therapy for nonresolving cases includes surgical intervention.

334. Findings that favor the diagnosis of spasm of the near reflex rather than accommodative esotropia include:
   1. new myopia.  
   2. esotropia worse at near than at distance.  
   3. miosis on attempted lateral gaze.  
   4. no vertical component.

335. T or F Isolated abducens paresis in a child is a neurologic emergency demanding immediate neuroimaging and lumbar puncture.

336. The features least consistent with acquired abducens paresis include all of the following except:
   a. esotropia.  
   b. deviation greater at distance than near.  
   c. amblyopia.  
   d. head turn toward the side of the paretic muscle.  
   e. noncomitant deviation.

337. Treatment alternatives in acquired abducens paresis include:
   1. patching.  
   2. base-in Fresnel prisms.  
   3. botulinum toxin injections.  
   4. ipsilateral medial rectus resection.

338. T or F Consecutive exotropia should be repaired immediately because spontaneous recovery is highly unlikely.

339. T or F Sensory deprivation in a child younger than 6 years is more likely to lead to esotropia, whereas that occurring in adults is more likely to lead to exotropia.

340. An exodeviation that is greater at distance than at near is known as:
   a. basic exotropia.  
   b. divergence excess exotropia.  
   c. true divergence excess exotropia.  
   d. simulated divergence excess exotropia.  
   e. convergence insufficiency exotropia.
Questions

341. To distinguish true divergence excess exotropia from “simulated divergence excess” exotropia:
   a. the deviations are remeasured after cycloplegia.
   b. the accommodative convergence to accommodation ratio (AC/A) is calculated by the heterophoria method.
   c. the AC/A ratio is calculated by the gradient method.
   d. the deviations are remeasured after prolonged (30 to 45 minutes) monocular occlusion.
   e. the deviations are remeasured with +1.00D lenses over the patient’s distance correction.

342. A patient with constant exotropia has a deviation of 35 prism diopters at distance and 15 prism diopters at near. After wearing lenses over his distance correction for 1 hour, the deviations are remeasured and found to be 35 prism diopters at distance and 30 prism diopters at near. The classification of his strabismus is:
   a. basic exotropia.
   b. divergence excess exotropia.
   c. true divergence excess exotropia.
   d. simulated divergence excess exotropia.
   e. convergence insufficiency exotropia.

343. A patient presents for ophthalmic examination that discloses a comitant exotropia. At distance, his deviation measures 5 prism diopters of exotropia and at near 15 prism diopters of exotropia. His strabismus is best classified as a:
   a. basic exotropia.
   b. divergence excess exotropia.
   c. true divergence excess exotropia.
   d. simulated divergence excess exotropia.
   e. convergence insufficiency exotropia.

344. T or F Congenital exotropia is equally as common as congenital esotropia.

345. The most common etiology for constant exotropia is:
   a. decompensated intermittent exotropia.
   b. sensory exotropia.
   c. third nerve palsy.
   d. Duane’s syndrome type 2.
   e. exotropia associated with craniofacial anomalies.

346. T or F Intermittent exotropia, like accommodative esotropia, usually fades away as the child progresses through early adolescence.

347. Clinical features frequently associated with intermittent exotropia include:
   1. variable angle of deviation.
   2. high accommodative convergence to accommodation ratio (AC/A).
   3. reflex closure of one eye in bright light.
   4. amblyopia.

348. T or F Patients with intermittent exotropia generally have reduced stereoaucity.

349. Useful treatment modalities for intermittent exotropia include:
   1. minus lenses.
   2. phospholine iodide.
   3. base-in prism.
   4. bilateral rectus resections.

350. T or F Virtually all cases of intermittent exotropia will progress to constant exotropia over time.

351. T or F The exotropia associated with Duane’s syndrome type 2 is frequently noncomitant.

352. Clinical features of convergence insufficiency include all of the following except:
   a. asthenopia.
   b. blurry reading vision.
   c. diplopia while reading.
   d. exophoria at near.
   e. brow-ache.

353. T or F Treatment of convergence insufficiency may include the use of either base-out or base-in prisms.

354. T or F Convergence insufficiency may occur with normal accommodation.
355. T or F Innervational vertical deviations are as common as innervational horizontal deviations.

356. A patient presents for evaluation of “wandering eyes.” On alternate-cover testing, with the left eye covered, the right eye fixes on a distance target. As the cover is shifted to the right eye, the left eye moves down to pick up fixation. As the cover is moved back over the left eye, the right eye moves upward to reassume fixation. This set of findings is consistent with:
   a. right hyperdeviation.
   b. left hyperdeviation.
   c. overaction of the inferior obliques.
   d. dissociated vertical deviation.
   e. overaction of the superior obliques.

357. A patient presents for evaluation of “wandering eyes.” On alternate-cover testing, with the left eye covered, the right eye fixes on a distance target. As the cover is shifted to the right eye, the left eye moves down to pick up fixation. As the cover is shifted back over the left eye, the right eye does not move in order to reassume fixation. This set of findings is most consistent with:
   a. right hyperdeviation.
   b. left hyperdeviation.
   c. overaction of the inferior obliques.
   d. dissociated vertical deviation.
   e. overaction of the superior obliques.

358. Which of the following statements about dissociated vertical deviation is/are true?
   1. It is frequently associated with congenital esotropia.
   2. It is often bilateral.
   3. It can be made manifest by monocular visual loss of organic etiology.
   4. The deviation is typically reproducible and measurable.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

359. A 4-year-old child with a moderate-angle esotropia is noted to have a left hypertropia on right gaze and a right hypertropia on left gaze. When fixing with the left eye in right gaze, there is a right hypotropia, and when fixing with the right eye in left gaze, there is a left hypotropia. The most likely clinical diagnosis is:
   a. right hypotropia.
   b. left hypotropia.
   c. esotropia associated with overaction of the inferior oblique muscles.
   d. bilateral trochlear palsy.
   e. esotropia with dissociated vertical deviation.

360. Which one of the following statements about overaction of the superior oblique muscle(s) is false?
   a. Some cases are secondary to weakness of the ipsilateral inferior oblique muscle.
   b. It may be associated with an exotropia in primary gaze.
   c. It is frequently associated with exotropia in downgaze.
   d. Unilateral cases may develop an ipsilateral hypertropia in primary gaze.
   e. The hallmark is depression on attempted adduction.

361. Which of the following features argue(s) for a bilateral rather than a unilateral superior oblique paresis?
   1. head tilt.
   2. symptomatic excyclotorsion.
   3. A-pattern esotropia.
   4. aggravation of diplopia with right or left head tilt.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

362. Indications for surgical treatment of superior oblique paresis include:
   1. significant head tilt.
   2. large hypertropia in primary position.
   3. symptomatic diplopia.
   4. V-pattern esotropia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
Questions

363. T or F In long-standing superior oblique paresis, the hypertropia may be greatest in all directions of downgaze.

364. T or F Each millimeter of recession of a vertical rectus muscle will result in approximately 8 prism diopters of vertical correction.

365. Surgical strategies for the management of a right superior oblique paresis with symptomatic diplopia include all of the following except:
   a. right inferior oblique myectomy.
   b. right superior oblique tuck.
   c. right inferior rectus recession.
   d. right superior rectus recession.
   e. left superior oblique tenectomy.

366. The procedure of choice in acquired bilateral superior oblique pareses with symptomatic diplopia is the:
   a. bilateral inferior oblique myectomy.
   b. bilateral superior oblique tuck.
   c. bilateral inferior rectus recession.
   d. right superior rectus recession and right inferior rectus resection.
   e. left superior oblique tenectomy.

367. The surgical procedure of choice in a superior oblique paresis with excyclotorsion only (no vertical diplopia) is the:
   a. ipsilateral superior oblique tuck.
   b. ipsilateral inferior oblique myectomy.
   c. recession of the ipsilateral superior rectus muscle.
   d. lateral transposition of the superior oblique tendon.
   e. recession of the contralateral inferior rectus.

368. All of the following are features consistent with double elevator palsy except:
   a. ipsilateral hypotropia with large secondary deviation.
   b. ptosis.
   c. forced ductions indicating inferior rectus restriction.
   d. chin-down head position.
   e. poor Bell phenomenon on the side of the palsy.

369. Which one of the following statements about Brown’s syndrome is false?
   a. Both congenital and acquired forms exist.
   b. A common manifestation is hypotropia of the involved eye in adduction.
   c. Duction and version testing mimic weakness of the ipsilateral superior oblique muscle.
   d. Forced duction testing is necessary to confirm the diagnosis.
   e. Acquired cases should be observed for a considerable period because many spontaneously improve.

370. Late clinical findings consistent with an inferior blowout fracture of the orbit include all of the following except:
   a. proptosis.
   b. paresthesia or hypesthesia of the infraorbital region.
   c. ipsilateral hypotropia on upgaze.
   d. ipsilateral hypertropia on downgaze.
   e. positive forced ductions.

371. Which of the following statements about A and V patterns of horizontal strabismus is/are true?
   1. A patterns must measure at least a 15-prism-diopter difference between upgaze and downgaze to be considered significant.
   2. V patterns must measure at least 10 prism diopters between upgaze and downgaze to be considered significant.
   3. These forms of noncomitance are seen in <5% of horizontal strabismus.
   4. All the extraocular muscles (in varying combinations) have been implicated as responsible for these patterns.

372. T or F Oblique muscle dysfunction is uncommon in AH1- or VH1-pattern strabismus.
373. Factors that are crucial in the selection of appropriate surgical therapy for A- or V-pattern strabismus include:

1. measurements and type of deviation in primary position.
2. presence or absence of peripheral fusion.
3. presence and type of oblique muscle dysfunction.
4. presence or absence of head tilt.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

374. A patient presents with an exotropia measuring 15 prism diopters in primary position. In downgaze it diminishes to <5 prism diopters, and in upgaze it increases to >30 prism diopters. There is no significant oblique muscle dysfunction noted. Appropriate surgical intervention might include:

1. recession of the ipsilateral lateral rectus.
2. resection of the ipsilateral medial rectus.
3. upward transposition of the lateral rectus and downward transposition of the medial rectus.
4. inferior oblique myectomy.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

375. A patient presents with a 15 prism diopter esotropia in primary gaze that increases to <5 prism diopters in downgaze and increases to >30 prism diopters in upgaze. There is overaction of the inferior obliques bilaterally. Appropriate surgical intervention might include:

1. recession of the ipsilateral medial rectus muscle.
2. resection of the ipsilateral lateral rectus muscle.
3. bilateral inferior oblique myectomies.
4. upward transposition of the lateral rectus and downward transposition of the medial rectus muscles ipsilaterally.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

376. A patient presents with a 20 prism diopter esotropia in primary gaze that increases to 35 prism diopters in downgaze and diminishes to 15 prism diopters in upgaze. There is bilateral depression with attempted adduction. Choices for surgical intervention should include:

1. recession of the ipsilateral medial rectus muscle.
2. recession of the contralateral medial rectus muscle.
3. tenotomy of both superior oblique muscles.
4. upward transposition of the lateral rectus muscle and downward transposition of the medial rectus muscle.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

377. A patient presents with a 25 prism diopter esotropia in primary position. The esotropia increases to 45 prism diopters in upgaze and diminishes to 5 prism diopters in downgaze. There is significant elevation of each eye with adduction bilaterally. Appropriate surgical intervention might include:

1. recession of the ipsilateral medial rectus muscles.
2. recession of the contralateral lateral rectus muscle.
3. bilateral inferior oblique myectomies.
4. upward transposition of the ipsilateral medial rectus muscle and downward transposition of the ipsilateral lateral rectus muscle.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
378. A patient presents with a 20 prism diopter esotropia in primary position. The esotropia increases to 35 prism dipters in upgaze and diminishes to 15 prism dipters in downgaze. Appropriate choices for surgical intervention might include:

1. recession of the contralateral medial rectus muscle.
2. recession of the ipsilateral medial rectus muscle.
3. upward transposition of the ipsilateral medial rectus muscle and downward transposition of the ipsilateral lateral rectus muscle.
4. tenotomy of both superior oblique muscles.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

379. T or F Adjustable suture techniques are useful for nearly every type of strabismus surgery except superior oblique procedures.

380. Superior oblique overaction is most likely to be encountered in:

a. A-pattern esotropia.
b. A-pattern exotropia.
c. V-pattern esotropia.
d. V-pattern exotropia.
e. Y-pattern exotropia.

381. Systemic associations with Duane’s syndrome include:

1. sensorineural hearing loss.
2. Goldenhar’s syndrome.
3. spinal column anomalies.
4. congenital absence of the pectoral muscle.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

382. T or F Like sixth nerve palsy, abduction deficit associated with Duane’s syndrome leads to progressive contracture of the medial rectus muscle.

383. Indications for surgical intervention in Duane’s syndrome include:

1. amblyopia.
2. significant head turn.
3. significant globe retraction.
4. significant deviation in primary position.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

384. T or F An adduction deficit coincident with abducens and facial palsies makes the diagnosis of Möbius’ syndrome very unlikely.

385. The most common cause of third nerve palsy in the pediatric population is:

a. congenital.
b. traumatic.
c. inflammatory.
d. migrainous.
e. tumor.

386. In adults, the most common cause of third nerve palsy is:

a. microvascular.
b. traumatic.
c. inflammatory.
d. migrainous.
e. tumor.

387. Strabismus surgery for patients with Graves’ ophthalmopathy is generally performed:

a. before orbital decompression.
b. before orbital radiation.
c. before tarsorrhaphy.
d. before yield surgery.
e. none of the above.

388. T or F Prisms are frequently a permanent solution for strabismus associated with Graves’ disease.

389. T or F Helpful techniques in the surgical management of patients with Graves’ disease and diplopia include muscle resections with adjustable sutures.
390. T or F Surgical realignment of the eyes plays a role in the management of strabismus associated with medication-resistant myasthenia gravis.

c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

391. Which of the following conditions is more commonly seen in boys than in girls?
a. congenital third nerve palsy.
b. Duane’s syndrome.
c. congenital sixth nerve palsy.
d. congenital oculomotor apraxia.
e. Möbius’ syndrome.

392. Which of the following conditions is seen more commonly in girls than in boys?
a. congenital third nerve palsy.
b. Duane’s syndrome.
c. congenital sixth nerve palsy.
d. congenital oculomotor apraxia.
e. Möbius’ syndrome.

393. A child’s nystagmus is noted to have equal velocity in all directions and to be symmetric in direction, amplitude, and frequency in each eye. The nystagmus would most appropriately be described as:
a. uniplanar.
b. pendular, conjugate.
c. jerk, conjugate.
d. pendular, disconjugate.
e. jerk, disconjugate.

394. T or F In a neonate or infant, pendular nystagmus is more likely to be sensory than motor nystagmus.

395. T or F The hallmark of congenital nystagmus is an exponentially increasing velocity of the slow phase on electronystagmography.

396. T or F In congenital motor nystagmus, near visual acuity may be better than distance visual acuity.

397. Sensory nystagmus in an infant may be seen in association with:
1. aniridia.
2. bilateral macular colobomata.
3. bilateral optic nerve hypoplasia.
4. rod monochromatism.
a. 1, 2, and 3.
b. 1 and 3.

398. Which of the following statements about latent nystagmus is/are true?
1. Its fast phase is toward the uncovered eye.
2. It may become manifest in the setting of monocular visual loss like amblyopia.
3. It is associated with congenital nystagmus and infantile esotropia.
4. It may be the cause of unexpectedly low monocular visual acuity testing.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

399. Characteristics considered classic for spasmus nutans include:
1. head nodding.
2. a rapid, small-amplitude, conjugate nystagmus.
3. torticollis.
4. hypertonia.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

400. T or F Spasmus nutans generally has its onset within the first year of life and resolves within 2 years.

401. The entity in the differential diagnosis with spasmus nutans that must be ruled out is:
a. optic nerve meningioma.
b. parasellar glioma.
c. pontine glioma.
d. cerebellar astrocytoma.
e. syringomyelia.

402. Treatment for a patient with congenital nystagmus whose null zone is in right gaze and who has adopted an extreme left head turn might include:
a. prism base down in both eyes.
b. prism base in, in both eyes.
Questions

c. prism base in, in the right eye and base out, in the left eye.
d. prism base out, in the right eye and base in, in the left eye.
e. prism base up in both eyes.

403. T or F The goal of muscle surgery in congenital nystagmus is to move the null zone to primary position.

404. Surgical intervention for the patient with a null zone in right gaze and severe left head turn might include:

1. recession of the right lateral rectus muscle.
2. resection of the right medial rectus muscle.
3. recession of the left medial rectus muscle.
4. resection of the left lateral rectus muscle.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

405. For which of the following procedures might a two-stage adjustable suture technique be advisable?

1. disassociated vertical divergence (DVD).
2. strabismus associated with Graves’ disease.
3. muscle transposition surgery in A- or V-pattern horizontal strabismus.
4. infantile esotropia.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

406. For which of the following procedures would a rectus muscle transposition procedure be particularly appropriate?

1. nonresolving sixth nerve palsy.
2. double elevator palsy.
3. nonresolving third nerve palsy.
4. dissociated vertical deviation (DVD).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

407. A 3-year-old child presents to the ophthalmologist with parents complaining of “cross-eyes” of approximately 1 year duration. Examination discloses visual acuity of 20/30 in the right eye and 20/100 in the left eye with Allen cards. There is a 35 prism diopter esotropia at distance increasing to 45 prism diopters at near. Refraction reveals +3.50D in both eyes. Initial steps in managing this patient should include:

a. bimedial recessions.
b. bilateral resections.
c. prescription of +3.50D in both eyes with add +3.50 in both eyes (bifocals).
d. prescription of +3.50D in both eyes and patching of the right eye.
e. a Kestenbaum procedure (bilateral recess/resect procedure).

408. A 7-year-old boy presents with an exotropia. His deviation measures 30 prism diopters in primary position, 20 prism diopters in right gaze, and 40 prism diopters in left gaze. Near deviation is 15 prism diopters in all directions. Fixation appears to alternate, and visual acuity is 20/20 in both eyes. The patient’s parents strongly desire some form of correction. You recommend:

a. addition of +2.00D to his current distance refraction.
b. base-in prism.
c. bilateral rectus recessions, equal on each side.
d. bilateral rectus recessions with a greater distance of recession on the left.
e. bilateral rectus recessions with a greater distance of recession on the right.

409. A patient presents with symptomatic vertical diplopia from a right hypertropia that is greatest in left eye: left upgaze and left downgaze. Appropriate surgical intervention could include all of the following except:

a. right superior oblique tuck.
b. left superior oblique tenotomy.
c. right inferior oblique myectomy.
d. right superior rectus resection.
e. left inferior rectus recession.

410. A 1-year-old child is brought to the ophthalmologist by his parents who have noted that “cross-eyes” developed over the previous 6 months. Your examination reveals an approximately 40 prism diopter esotropia at near that does not seem to
diminish significantly when the child fixes at longer distances. The child can maintain fixation with either eye easily. Cycloplegic refraction reveals +1.50D in both eyes. An appropriate next step in the management of this patient might be:

a. prescription of +1.50D glasses in both eyes.

b. prescription of +1.50D in both eyes with +3.50D add in both eyes.

c. alternate patching throughout the day.

d. bilateral rectus recession of 8 mm in both eyes.

e. bimedial rectus recession of 5.5 mm in both eyes.

411. After treating a child with mixed mechanism esotropia for 8 months with full hyperopic correction and occlusion therapy, surgery is undertaken to realign his eyes. Visual acuity measures 20/50 in the right eye and 20/25 in the left eye with HOTV letter cards. There is an esotropia of 30 prism diopters in all directions of gaze. Appropriate surgical intervention would consist of:

a. bimedial rectus recession of 4.5 mm in both eyes.

b. bilateral rectus resection of 7 mm in both eyes.

c. bimedial rectus recession of 7 mm in both eyes.

d. recession of the right medial rectus 4.5 mm and resection of the right lateral rectus 7 mm.

e. recession of the left medial rectus 4.5 mm and resection of the left lateral rectus 7 mm.

412. T or F It is probably better initially to overcorrect an exodeviation than to undercorrect it.

413. A 6-year-old patient presents to an ophthalmologist for the first time after failing a school eye examination. Complete ophthalmic examination discloses a visual acuity of 20/20 in the right eye and 20/100 in the left eye. There is a comitant left exotropia measuring 30 prism diopters. Cycloplegic refraction reveals +0.50D in both eyes. A year of occlusion therapy of the right eye is undertaken, with little improvement in acuity in the left eye. The next step in the management of this patient might be:

a. prescription of +1.50D in both eyes.

b. base-in prism.

c. bilateral rectus recession of 7 mm.

d. recession of the left lateral rectus 6 mm and resection of the left medial rectus 7 mm.

e. recession of the left lateral rectus 7 mm and resection of the left medial rectus 6 mm.

414. The maximal advisable recession of the medial rectus muscle in the initial surgical management for esotropia is:

a. 3 mm.

b. 4 mm.

c. 5 mm.

d. 5.5 mm.

e. 6 mm.

415. The maximal advisable resection of the lateral rectus muscle in the initial surgical management of esotropia is:

a. 6 mm.

b. 7 mm.

c. 8 mm.

d. 9 mm.

e. 10 mm.

416. The maximal advisable resection of the lateral rectus muscle in the initial surgical management of exotropia is:

a. 5 mm.

b. 6 mm.

c. 7 mm.

d. 10 mm.

e. 12 mm.

417. The maximal advisable recession of the medial rectus muscle in the initial surgical management of exotropia is:

a. 6 mm.

b. 8 mm.

c. 10 mm.

d. 12 mm.

e. 15 mm.

418. Inferior oblique myectomy, performed to correct either primary overaction of the inferior obliques or to correct a V-pattern exotropia, can be expected to yield how much correction of hyperdeviation in adduction or how much esodeviation in upgaze?

a. 5 prism diopters.

b. 10 prism diopters.

c. 25 prism diopters.

d. 40 prism diopters.

e. 50 prism diopters.

419. Bilateral superior oblique tenotomies, as part of the treatment for A-pattern exotropia, can be
expected to cause how much of an esodeviation in
downgaze?
a. 5 prism diopters.
b. 15 prism diopters.
c. 25 prism diopters.
d. 40 prism diopters.
e. 50 prism diopters.

420. Manifestations of ischemia following excessive
rectus muscle surgery include:
   1. corneal edema.
   2. anterior chamber reaction.
   3. hypotony.
   4. retinal neovascularization.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

421. T or F Surgery on the superior rectus muscle is
more likely to lead to lid malposition postopera-
tively than surgery on the inferior rectus muscle.

422. Which of the following statements about diplopia
after surgery for esotropia is/are true?
   1. Adults with acquired strabismus are more
      likely to suffer from symptoms than
      children.
   2. Postoperative diplopia is most likely to de-
      velop in undercorrection of intermittent
      exotropia.
   3. A trial of preoperative prisms may be help-
      ful in predicting who is likely to suffer from
      this complication.
   4. The complication, if persistent, must be
      managed with a second surgical procedure.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

423. A 2-year-old child undergoes bimedial recession
for infantile esotropia. On the first postoperative
day, the deviation is measured as <10 prism
diopters of residual esotropia, with fairly good
versions. At the 1-week visit, there is a prominent
right exotropia, which increases in left gaze.
Duction testing reveals an inability to adduct the
right eye past the midline. The most likely diag-
nosis is:
a. surgical undercorrection.
b. consecutive exotropia caused by surgical over-
correction.
c. postoperative third nerve palsy.
d. lost or slipped right medial rectus.
e. restriction of the right medial rectus caused by
   orbital fat prolapse.

424. Early signs in the development of malignant
hyperthermia include all of the following
except:
a. tachycardia.
b. arrhythmia.
c. elevated body temperature.
d. darkening of the blood in the operative
   field.
e. trismus.

425. The mechanism by which botulinum toxin
(Oculinum) works is:
a. sarcoplasmic capture of extracellular calcium.
b. direct blockage of postsynaptic acetylcholine
   receptors.
c. inhibition of the formation of actin-myosin
   complexes.
d. inhibition of release of acetylcholine from
   presynaptic nerve terminals.
e. direct blockage of muscle cell membrane cal-
   cium channels.

426. Botulinum toxin has been shown to be effective
for which of the following conditions?
   1. congenital nystagmus.
   2. acute lateral rectus palsy.
   3. dissociated vertical deviation.
   4. essential blepharospasm.
a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

427. The most common complication of botulinum
injections is:
a. vertical strabismus.
b. Adie’s pupil.
c. ptosis.
d. perforation of the globe.
e. retrobulbar hemorrhage.
428. Which of the following have been established as risk factors for esotropia?
   1. white race.
   2. maternal cigarette smoking.
   3. increasing maternal age.
   4. low birth weight.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

429. Which of the following have been established as risk factors for exotropia?
   1. white race.
   2. maternal cigarette smoking.
   3. increasing maternal age.
   4. low birth weight.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

431. Match the causes of congenital/neonatal corneal clouding with their tendency to be unilateral or bilateral.
   a. sclerocornea. 1. unilateral.
   b. forceps injury. 2. bilateral.
   c. mucopolysaccharidoses. 3. either.
   d. anterior segment dysgenesis (Peters’ anomaly).
   e. congenital hereditary endothelial dystrophy.
   f. congenital hereditary stromal dystrophy.
   g. corneal dermoids.
   h. congenital glaucoma.

432. Match each of the systemic disorders lettered below with its most likely manifestations.
   a. Fabry’s disease. 1. hematuria and hearing loss.
   b. homocystinuria. 2. flaking dermopathy and enlarged corneal nerves.
   c. Refsum’s disease. 3. conjunctival and retinal telangiectasis.
   d. Wilson’s disease. 4. ectopia lentis, tall stature, and mental retardation.
   e. Lowe’s syndrome. 5. frontal bossing, baldness, and testicular atrophy.
   f. Alport’s syndrome.
   g. myotonic dystrophy.
   h. incontinentia pigmenti.
   i. ichthyosis.
   j. Hallermann-Streiff syndrome.
6. alopecia and birdlike facies.
7. progressive tapetoretinal degeneration and hearing loss.
8. peripheral retinal neovascularization.
9. hepatic failure, dementia, and peripheral pigmentation of Descemet's membrane.
10. aminoaciduria, hypotonia, and renal rickets.

433. Match the name of the histopathologic feature numbered below with the light microscopic characteristics.
   a. a cluster of nearly mature-appearing photoreceptor elements.
   b. a ring of nuclei surrounding parallel arranged fibrils with a central clear lumen.
   c. a ring of nuclei and cell processes surrounding a blood vessel.
   d. a ring of nuclei surrounding a mass of haphazardly arranged cell processes with no central lumen.

435. Match the diagnostic criteria lettered below with the type of neurofibromatosis numbered below.
   a. plexiform neurofibroma.
   b. intertriginous freckling.
   c. bilateral acoustic neuromas.
   d. unilateral optic glioma.
   e. first-degree relative with the same disorder plus two neurofibromas, meningiomas, gliomas, or schwannomas.
   f. two or more Lisch's nodules.
   g. posterior subcapsular cataract.
   h. sphenoidal dysplasia.

434. Match each of the fundus tumors of childhood with its systemic disorder.
   a. capillary hemangioma of the retina.
   b. astrocytic hamartoma of the optic nerve.
   c. astrocytic hamartoma of the retina.
   d. diffuse choroidal hemangioma.
   e. peripheral acquired hemangioma of the retina.

436. Select the appropriate distance from limbus to muscle insertion for each of the muscles listed below.
   a. lateral rectus.
   b. medial rectus.
   c. inferior rectus.
   d. superior rectus.
1. **c.** Axial length increases most rapidly over the first 4 years of life and more slowly thereafter until age 10 to 12 years.

2. **c.** The lens is proportionately the largest part of the infant eye.

3. **True.** Corneal diameters >13 mm are termed megalocornea.

4. **d.**

5. **False.** The optic nerve is generally myelinated to the lamina cribrosa 4 to 6 months before foveal maturation.

6. **a.** Acuity estimates are highest with visual evoked potentials (VEPs). Optokinetic nystagmus testing (OKN) and preferential looking testing (PLT) are fairly close. OKN is more cumbersome and difficult to quantify accurately than PLT.

7. **False.** Visual evoked potentials (VEPs) document 20/20 acuity by the age of 6 to 8 months. Preferential looking testing (PLT) cannot document 20/20 until 24 to 30 months.

8. **False.** The globe obscured by the fused lids is typically disorganized.

9. **a.** The upper lids are more frequently involved. Other features include limbal dermoids, branchial cleft abnormalities, and vertebral anomalies.

10. **False.** The lower lids are more frequently involved by congenital ectropion and congenital entropion.

11. **True.**

12. **False.** There are four varieties of epicanthus: palpebralis (simple), broader above; inversus, broader below; tarsalis, equally broad above and below; and supraciliaris, origin from eyebrow.

13. **False.** Hypertelorism implies an abnormally wide distance between medial orbital walls. Telecanthus is an abnormally wide distance between medial canthi. Telecanthus may be isolated (primary) or secondary to hypertelorism.

14. **c.** Epicanthus inversus and telecanthus are the other components of this tetrad. Ectropion or euryblepharon is seen in some cases as well.

15. **b.** Seventy percent of pregnant women are seronegative. Placental transfer of organism is common in the newly infected mother; however, many infected infants will not have systemic manifestations.

16. **True.**

17. **a.** Microphthalmos and cataracts are rare. Focal chorioretinal lesions are common. Diffuse retinopathy is not seen.

18. **c.** Immunoglobulin G (IgG) against *Toxoplasma* found in the infant’s serum is probably maternal and may reflect old maternal infection. Immunoglobulin M (IgM), on the other hand, does not cross the placenta and probably represents a response to fetal infection. Computed tomography (CT) scan evidence is very strong.

19. **c.** Pyrimethamine and sulfadiazine are antimicrobial agents effective against *Toxoplasma*. Prednisone reduces inflammatory tissue destruction. Pyrimethamine blocks synthesis of folinic acid, so it must be given as well. Note that folinic acid is synthesized from dietary folic acid, so folic acid administration is not sufficient.

20. **d.** The later in the course of the pregnancy a mother is infected, the lower the chance of fetal infection, and if infection occurs, the lower the likelihood of symptomatic defects. Infection early in pregnancy leads to a high rate of abnormalities. Thanks to vaccinations, most mothers in developed countries are seropositive (protected).

21. **c.** Many cases of “oculounditory syndrome” are caused by rubella.

22. **c.** Although children with congenital rubella syndrome may develop either cataract or glaucoma, their coincidence in an infant is rare.

23. **d.** Excessive rebound uveitis is caused by live virus retained within the residual lens cortex.

24. **d.** Virus in lens material can lead to rebound uveitis after cataract extraction.

25. **c.** Approximately 2% of human infants are infected. Most cases are silent.

26. **c.**

27. **False.** Most neonatal herpes is contracted at the time of delivery caused by passage through an infected birth canal.

28. **True.**

29. **False.** Most cases of congenital herpes declare themselves with vesicular dermatitis, keratoconjunctivitis, pneumonitis, or meningoencephalitis.

30. **False.** A posterior inflammatory component is far more common in congenital disease, with severe retinitis and vitritis. This is unusual in acquired disease but may be the cause of some cases of acute retinal necrosis.

31. **a.** Herpes simplex is generally transmitted at the time of birth as the child passes through an infected birth canal.

32. **a.** Microphthalmia is an unusual manifestation of congenital syphilis.

33. **a.** Rhagades, linear scars around the mouth, are characteristic of congenital lues, but they are not a part of Hutchinson’s triad.

34. **c.** Treatment clearly reduces the likelihood of a no light perception (NLP) outcome, but the long-term Snellen acuity levels are disappointing. In fact, treated eyes were slightly, albeit not significantly, less likely to retain 20/40 or better than eyes randomized to no treatment.

35. **a.** *Staphylococcus aureus*, *Streptococcus pneumoniae*, and *Haemophilus influenzae* are far more common.
36. **a.** Pharyngoconjunctival fever (adenovirus type 3) generally spares the cornea, although subepithelial infiltrates or superficial punctate keratitis may rarely be seen. **False.** A self-limited conjunctivitis is very common but frequently obscured by other, more prominent symptoms and signs. **c.** The inflammation in Parinaud’s syndrome is always granulomatous. **False.** The inflammation in Parinaud’s syndrome is always granulomatous. **e.** The inflammation in Parinaud’s syndrome is always granulomatous. **b.** Boys are more frequently involved than girls, and the upper tarsal conjunctiva is more commonly and more severely involved than the lower. **False.** The limbal lesions in vernal keratoconjunctivitis are more papilliform because they have a central vascular core. When a central pit full of degenerated eosinophils forms, the term Horner-Trantas dot is applied. **d.** True follicles are rare in vernal disease, whereas eosinophils are numerous. Both conditions may have prominent limbal nodules (limbal papillae and Horner-Trantas dots in vernal, limbal follicles in trachoma). **False.** The limbal lesions in vernal keratoconjunctivitis are more papilliform because they have a central vascular core. When a central pit full of degenerated eosinophils forms, the term Horner-Trantas dot is applied. **d.** Horner-Trantas dots are clumps of degenerated eosinophils, whereas Herbert’s pits are depressed, necrotic follicles at the limbus. **True.** Many experts have maintained that corneal vascularization is unlikely in vernal disease. So-called shield ulcers—large oval, central epithelial defects—are classic corneal manifestations of the disease. **False.** Debridement of giant palpebral papillae has been tried therapeutically in an attempt to speed reepithelialization of shield ulcers. Typically, this fails, indicating that some factor other than mechanical irritation is at play. Some experts have suggested that inflammatory factors released by diseased conjunctiva may be responsible. **b.** Stevens-Johnson syndrome can affect patients of all ages and typically causes target-shaped macular, rather than vesicular, dermatitis. The mucous membrane lesions are vesicular. **c.** Other diagnostic criteria include injected pharynx, edema of upper or lower extremities, nonspecific rash, and lymphadenopathy. Fever plus four of the other criteria must be present to make the diagnosis. **c.** The vasculitis may involve the coronary arteries and lead to coronary artery aneurysm or occlusion. **b.** The medial canthal tendon straddles the superior one third of the nasolacrimal sac. Therefore, distention of the sac caused by obstruction presents as a mass below the tendon. **c.** Dacryocele, or congenital mucocele of the nasolacrimal sac, typically has a bluish-red hue, similar to a hemangioma, and it results from chronic obstructive tear stasis. Recurrent conjunctivitis is far more common than dacroyocystitis. Obstruction is nearly always at the inferior extreme of the nasolacrimal duct—the valve of Hasner (within the inferior nasal meatus). If it persists beyond 1 year of age, surgical intervention is generally required. **True.** Failure is generally followed by a repeat probing and irrigation. If this fails a second time, then silastic intubation is tried. If this also fails, then dacryocystorhinostomy is the next step. **b.** Sacue’s syndrome, or mucopolysaccharidosis I-S, is (MPS), an inborn error of metabolism. Each of the other disorders is a “neurocristopathy.” **b.** Congenital hereditary endothelial dystrophy (CHED) occurs secondary to a defect of the corneal endothelium and Descemet’s membrane. Although the corneal edema may resemble that seen in congenital glaucoma, there is no elevated intraocular pressure (IOP) or increased corneal diameter. **c.** In congenital hereditary stromal dystrophy, the cloudy stroma is of normal thickness, and the epithelium is normal. In congenital hereditary endothelial dystrophy (CHED), there is epithelial edema and a thickened stroma. In both conditions, intraocular pressure (IOP) is usually normal. **b.** Lacrimal gland involvement can often occur in acute Epstein-Barr virus infection. The authors suggest it may be far more common than the 20% reported previously. Neither streptococcal nor diphtheritic infections tend to cause dacryoadenitis. Sarcoidosis never causes acute pharyngitis, and the dacryoadenitis is typically painless. **c.** Cystine crystals may be found in the cornea, conjunctiva, and uvea. **b.** Corneal ulceration and scarring in familial dysautonomia (Riley-Day syndrome) is secondary to decreased or absent corneal sensation and decreased lacrimation.
65. a. Approximately two thirds of the cases of infantile glaucoma are bilateral, and approximately two thirds of the patients are boys. An affected parent has about a 5% chance of having a child with infantile glaucoma, and the next subsequent sibling has approximately a 5% chance of being affected. The incidence of open-angle glaucoma in grandparents is no different from age-matched controls.

66. False. There is controversy about the basic pathophysiological mechanism underlying the cause of infantile glaucoma. Many anomalies in and around the chamber angle have been described.

67. c.

68. False. Ruptures in Descemet’s membrane associated with infantile glaucoma are usually horizontal, and those associated with birth trauma are usually vertical.

69. True. This is probably true because younger infants spend most of the day with their eyes closed, making corneal enlargement difficult to detect.

70. False. Although optic nerve cupping in infants may be reversible, the presence of an afferent pupillary defect carries a poor visual prognosis.

71. a. Medication is used only for preoperative control of intraocular pressure (IOP) in children. Definitive management consists of surgery.

72. a. Optic cupping is reversible in children. Afferent pupillary defects generally indicate irreversible optic nerve damage.

73. d. Rubella immunoglobulin M (IgM) titers might be of value in a neonate, but IgG titers are not diagnostic because they are of maternal origin during the first 3 to 6 months of life. Glaucoma is not a feature of galactosemia or Refsum’s disease. Lowe’s syndrome must be considered.

74. b. Ketamine and succinylcholine elevate intraocular pressure. Ketamine only does so at high doses. Succinylcholine does so by triggering undirected extraocular muscle contractions (fasciculations).

75. False. Unlike adults, pressures in children >20 mm Hg must raise a suspicion of glaucoma.

76. b. All inherited forms are typically bilateral, but recessive forms may be asymmetric. Dominant varieties may not present until the third, fourth, fifth, or sixth decades.

77. c. Autosomal-dominant patterns are most frequently reported, but 15% have no family history. Aortic valvular insufficiency and aneurysm may be life threatening and should be treated with cardiac depressants and/or surgery. Mitral valve prolapse also may be found. Myopia, high astigmatism, or frank keratoconus is seen in association with ectopia lentis in many cases. Up to 60% of patients with rigorously diagnosed Marfan’s syndrome will develop ectopia lentis.

78. c. Homocystinuria is an elastosis and leads to hypercoagulability with thrombotic vascular occlusions. The elastosis also causes defective zonular function and ectopia lentis in 90% of patients. It is inherited on an autosomal-recessive basis.

79. c. Mentation is typically normal in Sturge-Weber syndrome, whereas ectopia lentis is unusual in Down’s syndrome (except after trauma).

80. c. Bilaterality makes a systemic condition more likely. Monocular cataracts rarely herald significant systemic derangements. Anterior lenticonus may be part of Lowe’s syndrome, but posterior lenticonus is usually isolated. Pigmentary retinopathy raises the possibility of congenital infections.

81. d. Congenital cataract formation secondary to a maternal herpes simplex virus (HSV) infection would be extremely unusual because the infection is typically acquired at birth.

82. d. Monocular visual deprivation, most likely with nuclear cataract, is the most “amblyogenic.” Every visually significant infantile cataract should be operated on as soon as it is feasible.

83. d. Because of the high incidence of capsular opacification, many surgeons advocate primary posterior capsulotomy (at the time of cataract extraction).

84. e. A complete systemic evaluation of a patient with bilateral congenital cataracts should rule out congenital rubella infection, hypocalcemia, Lowe’s syndrome, and trisomies.

85. b. Nystagmus is prominent in Leber’s congenital amaurosis, but photophobia would be extremely unusual. Photophobia is prominent in cystinosis, but unless there are other complicating features, nystagmus would not be expected.

86. c. There is a pediatric uveitis identical to juvenile rheumatoid arthritis (JRA) with no previous history of joint disease, but this is generally considered a separate disorder.

87. d. Pauciarticular, early-onset patients are most likely to develop uveitis, but they are generally antinuclear antibody (ANA) positive, rheumatoid factor negative. Uveitis is rare in Still’s disease, which is more common in boys.

88. b. The symptoms of chronic uveitis in juvenile rheumatoid arthritis (JRA) may be minimal. As a result, routine examinations should be frequent to avoid amblyopia from undetected sequelae (cataract, keratopathy). Placement of an intraocular lens (IOL) is absolutely contraindicated following cataract surgery in these patients because of the aggressive postoperative inflammation aggravated by an IOL. Secondary inflammatory membranes are stimulated by implants and will contract, leading to intractable, blinding hypotony.

89. a.

90. d. In immunocompromised patients, a high risk of systemic spread with severe complications exists and may be more likely if systemic steroids are used. Topical steroids may be used in immunocompromised patients.

91. True. Although keratitis is not universal in patients with herpes simplex virus (HSV) uveitis, it is usually present in affected children.
Answers

92. True.
93. b. Up to 50% of pediatric patients with posterior uveitis have ocular toxoplasmosis.
94. c. The brain and retina (a part of the brain) are the sites where Toxoplasma organisms survive best.
95. a. Acute arthritis is not a typical finding in acquired systemic toxoplasmosis.
96. True. The anterior flare-up may occur without obvious reactivation of retinal disease.
97. c. In ocular histoplasmosis, there is no vitreous inflammation.
98. a. The infectious cycle generally starts with a child consuming sand or dirt contaminated by the feces of a house pet that ingested the organism (dogs more often than cats). A peripheral granuloma with overlying vitreous opacification and chronic uveitis are typical findings in toxocarasis. The uveitis may die down and leave a quiescent peripheral granuloma. The inflammation in the eye often flares after the larva dies, and corticosteroids are the drug of choice in the treatment of this condition.
99. a. Pars planitis is bilateral in about 75% of cases. Tuberculosis, Lyme's disease, or syphilis may be the underlying cause, but the workup is usually negative.
100. d. The number of aqueous cells in the anterior chamber, and not the amount of flare, should be the basis for treatment with topical steroids. Topical steroids may worsen the cataract and will not reverse band keratopathy.
101. d. Persistent hyperplastic primary vitreous (PHPV) occurs frequently in small eyes, whereas retinoblastoma rarely occurs in association with microphthalmia.
102. d. Studies of the natural history of retinopathy of prematurity (ROP) show that 65% of premature infants with birth weight <1,250 g will develop some stage of the disease.
103. False. Vascularization of the nasal retina is complete by (approximately) the eighth gestational month. The temporal retina is completely vascularized 1 to 2 months later.
104. False. The risk of retinopathy of prematurity (ROP) does not correlate with the postdelivery age but with gestational age (postconception age).
105. b. Although increased oxygen tension has been implicated in the pathogenesis of retinopathy of prematurity (ROP), the correlation with disease severity is not direct. Although necrotizing enterocolitis (NEC) shares similar risk factors with ROP (low birth weight), no known pathologic connection exists between the two conditions.
106. False. Zone 1 is a circle of radius 30 degrees (twice the distance from optic disc to macula) centered on the optic disc. Zone 2 extends to the nasal ora and the temporal equator (radius 60 degrees). Zone 3 is the remaining crescent-shaped region anterior to zone 2 in the temporal retina.
107. e. All of these criteria together define subjects shown to benefit from cryotherapy.
108. e. All of these complications can occur from retinopathy of prematurity (ROP). Pseudopanretinal vitreous hemorrhages (Roth's spots), cotton-wool spots, optic disc swelling, and perivascular infiltration also may be seen but are less common. Choroidal infiltration is usually not ophthalmoscopically apparent.
109. False. The incidence of juvenile cataracts appears to be higher in patients with poorly controlled diabetes.
110. d. Optic nerve hypoplasia, particularly segmental, is more common in children of mothers with diabetes. Central vision is usually good, but sector or altitudinal field defects are frequently present.
111. False. The administration of chemotherapeutic agents to patients with leukemia makes the optic nerve more susceptible to radiation optic neuropathy than other eyes. Radiation dosage may need to be adjusted accordingly.
112. e. Glaucome may occur because of posterior synechiae formation with increased relative pupillary block or pupillary seclusion, from tumor cells clogging the trabecular meshwork or from spontaneous hyphema. Cataract may occur secondary to inflammation or radiation therapy.
113. b. Flame-shaped nerve fiber hemorrhages are the most common funduscopy finding. White-centered hemorrhages (Roth's spots), cotton-wool spots, optic disc swelling, and perivascular infiltration also may be seen but are less common. Choroidal infiltration is usually not ophthalmoscopically apparent.
114. False. Permanent loss of central acuity may occur rapidly with leukemic involvement of the optic nerve. Radiation therapy should be undertaken (within 24 to 48 hours) on an emergent basis in such cases.
115. True. The administration of chemotherapeutic agents to patients with leukemia makes the optic nerve more susceptible to radiation optic neuropathy than other eyes. Radiation dosage may need to be adjusted accordingly.
116. e. Glaucome may occur because of posterior synechiae formation with increased relative pupillary block or pupillary seclusion, from tumor cells clogging the trabecular meshwork or from spontaneous hyphema. Cataract may occur secondary to inflammation or radiation therapy.
117. b. All of the gangliosidoses are autosomal-recessive diseases. Most patients die in the first decade of life.
118. c. The most common ocular disorder associated with Lowe's syndrome is a characteristic small, thick, opaque cataract.
119. a. Diffuse arteriolar attenuation is not a characteristic finding in renal failure, although it may occur if there is severe associated hypertension.
120. False. It is felt that fewer ganglion cells decussate at the chiasm than in healthy subjects.

121. True. Patients with albinism of the tyrosinase-negative variety usually have severe photophobia and nystagmus. Visual acuity is usually <20/200. As the name implies, tyrosinase-negative albinos have no tyrosinase enzyme in their hair shafts or skin.

122. False. Extensive vitreous syneresis, vitreous veils, and strands are common features of X-linked juvenile retinoschisis.

123. c. The electrooculography (EOG) is abnormal in Goldmann-Favre disease and normal in X-linked juvenile retinoschisis. Goldmann-Favre disease is autosomal recessive. Both diseases have abnormal electoretinograms (ERGs). In mild forms of juvenile retinoschisis, the scotopic b-wave is lost, with preservation of the a-wave. In severe cases, the ERG is extinguished, as it is in most cases of Goldmann-Favre. Both diseases are characterized by foveal and peripheral retinoschisis, as well as retinal pigment epithelial disturbances, although the latter are more severe in Goldmann-Favre.

124. c. Some forms of tapetoretinal degeneration can have prominent subretinal exudation and mimic Coats’ disease.

125. False. Although electrophysiologic testing may show diffuse retinal involvement in sector retinitis pigmentosa, the condition rarely progresses, and the prognosis is good. This is reflected in normal b-wave implicit times.

126. b. Cone dystrophy is the most common “bull’s-eye” maculopathy.

127. a. Retinitis punctata albescens is not a form of congenital stationary night blindness (CSNB) but is progressive (although more slowly than typical retinitis pigmentosa).

128. True. With complete rod monochromatism, acuity is typically 20/200. With blue cone monochromatism, acuities are 20/40 to 20/200.

129. e. The ocular digital sign refers to a tendency to gouge the eyes with a finger or fist. It is felt that afflicted patients do this to provide entopic stimulation of the neurosensory retina. Five percent to 10% of patients with Leber’s congenital amaurosis have associated hearing loss.

130. False. The presenting symptom is usually decreased central visual acuity.

131. True. Central vision is usually preserved in the pattern dystrophies.

132. a. Aicardi’s syndrome is limited to the female sex because it is X-linked dominant. It is lethal in male infants or fetuses.

133. False. Most cases of morning glory disc have poor vision. The condition is usually unilateral and may be associated with cranial vault abnormalities. It is more common in girls.

134. True. Nonrhegmatogenous retinal detachments occur with optic nerve pits, primarily involving the macula.

135. False. Myelination of the optic nerve begins at the lateral geniculate ganglion, not at the optic chiasm. Myelination of the nerve is completed within a month of birth. Some studies claim that it is complete up to the lamina cribrosa by 1 month before birth; others claim that the process is complete 1 month after.

136. True. This condition occurs bilaterally in 20% of cases.

137. a. The visual field defects usually correspond to the area of fundus ectasia (inferior or inferonasal) and are consequently in the upper temporal field. They often cross the midline, unlike chiasmal defects.

138. a. De Morsier’s syndrome (septo-optic dysplasia) is far more common in bilateral optic nerve hypoplasia. Most authorities feel that neuroimaging should be conducted in both unilateral and bilateral optic nerve hypoplasia.

139. c. Optic nerve pits are usually in the inferotemporal quadrant or central part of the disc.

140. c. Both computed tomography (CT) scanning and ultrasonography will reveal the presence of disc drusen, if calcified. Red-free photographs are of no value. However, disc drusen do autofluoresce on fluorescein angiography. To elicit this, fluorescein barrier filters must be used with no injection. The visual fields of chronic papilledema and disc drusen may appear similar and are of less value.

141. False. Devic’s disease, or neuromyelitis optica, is considered by most experts as an entity separate from multiple sclerosis (MS). It is characterized by acute optic neuritis (usually bilateral) and transverse myelitis. Visual prognosis is much poorer for the optic neuritis of Devic’s disease than for typical optic neuritis.

142. b. The tumors produce osteolysis, not osteosclerosis.

143. e. Depending on the extent of the disease, any or all of these treatments may be useful.

144. b. Children with disease onset younger than 2 years with multifocal disease (especially those with compromised liver or lung function, or impaired hematopoiesis) have a poor prognosis.

145. a. The distinction between fibrous dysplasia and ossifying fibroma is generally made histologically. If osteoblasts are present, then the lesion is called an ossifying fibroma; if absent, the term fibrous dysplasia is applied.

146. d. Blood stagnation and phleboliths are characteristic of cavernous, not capillary, hemangiomas.

147. False. The pathology reveals dilated capillaries without the endothelial cell proliferation characterizing capillary hemangioma. This accounts for their flat clinical appearance.

148. a. Surgery is not prudent unless absolutely necessary because these tumors are difficult to remove completely and have a propensity to hemorrhage.

149. a.

150. c. Wilms’ tumor, neuroblastoma, and rhabdomyosarcoma are the three most common solid pediatric
malignancies. Leukemia is the most common of all pediatric malignancies.

151. True. An unexplained mass in the lids or acquired ptosis in a child should prompt radiology to rule out rhabdomyosarcoma.

152. b. Differentiated rhabdomyosarcoma is the least common type but has the best prognosis.

153. e. Exenteration is reserved for treatment failures and recurrences.

154. c. Few patients with neurofibromatosis (1.5%) will develop a schwannoma. It rarely undergoes malignant degeneration. The two classic histologic appearances are Antoni A (regular arrangement of cosinophilic spindle cells with palisading nuclei) and Antoni B (haphazardly arranged stellate cells in a myxomatous matrix). Perineural spread and compression account for pain.

155. d. Plexiform neurofibroma is most specific for neurofibromatosis. Unlike schwannomas, neurofibromas grow independent of peripheral nerves. In addition, they are generally osteolytic.

156. b. Schwann’s cells (neural crest derivatives) may be seen in both neurilemomas and neurofibromas. Positive staining for S-100 is characteristic of any cell derived from the neural crest. Thus, this will be seen in both lesions.

157. e. Pathologically, these are pilocytic astrocytomas of the juvenile type.

158. c. Optic gliomas are usually benign in children. If the tumor is located within the orbit, observation may be appropriate. On the other hand, if the tumor is posterior (i.e., involving the chiasm and optic tract), radiation therapy may be appropriate to avoid hydrocephalus and damage to contiguous central nervous system structures.

159. False. Radiographically, meningiomas produce hyperplastic bone changes (although bone destruction can be seen).

160. a. The tumor presents with metastases in about one third of patients.

161. a. Enophthalmos is characteristically seen with metastatic scirrhouous carcinoma of the breast in adult women.

162. d. Homer-Wright rosettes are usually not seen in orbital metastases, being limited to the better differentiated primary tumor.

163. c. Even with metastases to the liver, bone marrow, and spleen, survival may be as high as 84%. Bone metastasis is a poor prognostic factor. Age at onset is the most powerful predictor of survival.

164. c. Opsoclonus ("dancing eyes") is the paraneoplastic syndrome most commonly associated with metastatic neuroblastoma.

165. b. Ewing’s sarcoma is a primary intramedullary malignancy of bone, and periocular metastasis usually presents with proptosis, hemorrhage, and inflammation from tumor necrosis. It occurs in individuals aged 10 to 25 years, older than the population with neuroblastoma. Usually, there is no globe involvement, and treatment is principally chemotherapy, although radiation to remote sites has been used with some success.

166. b. The most common location is the superotemporal orbital rim, in association with the zygomaticofrontal suture line.

167. c. In children, one third of pseudotumor cases is bilateral but is rarely associated with systemic disease. Half of the patients have headache, abdominal pain, or lethargy. In addition, peripheral blood and local tissue eosinophilia are more common in children.

168. d. One study in which 302 epibulbar lesions were removed from children younger than 15 years reported the following frequencies:

- nevus: 29%
- dermoid: 19%
- epithelial inclusion cyst: 11%
- dermolipoma: 10%
- pyogenic granuloma: 6%

170. a. Goldenhar-Gorlin syndrome features limbal and conjunctival dermoids with upper lid colobomas, preauricular appendages, aural fistulas, and vertebral deformities. It is a syndromic abnormality of branchial arch development.

171. False. Approximately 30% of epibulbar dermoids have associated systemic and ocular findings.

172. False. Dermolipomas are often closely associated with extraocular muscles or the lacrimal gland, and complete surgical excision is difficult.

173. a. Retinoblastoma occurs in approximately 1 in 20,000 births. Only 6% have a positive family history. The familial tumors appear to be inherited in an autosomal-dominant pattern with 80% penetrance. Genetic counseling is complex, but bilateral retinoblastoma probably represents a genetic mutation.

174. False. As in question 173, the inheritance pattern appears to be autosomal dominant with 80% to 90% penetrance. Therefore, the offspring of a patient with bilateral retinoblastoma has approximately a 40% chance of developing a tumor (80% of 50%).

175. b. The locus of the responsible gene is located on 13q14, and the defect must occur on both chromosomes 13 (recessive), consistent with Knudson’s hypothesis. The absence of the growth regulatory gene results in the expression of the retinoblastoma. The retinoblastoma gene is likely expressed in other tissues as well. In particular, the pineal gland tumors accompanying some bilateral retinoblastomas are felt to be primary tumors and not metastasis. Musculoskeletal and integumentary systems are at increased risk of malignancy.

176. c. With no family history of bilateral retinoblastoma and healthy parents, this child probably had a germline mutation of chromosome 13. The copies of the child’s
parents are probably normal. Again, genetic counseling is complex, but the following generalizations serve as guidelines:
1. Healthy parents with one affected child have a 6% chance of producing more affected children.
2. If two or more siblings are affected, this implies a chromosomal defect in one parent. Therefore, each additional child has a 40% chance of inheriting the tumor.
3. A parent with bilateral retinoblastoma has a 50% chance of transmitting the affected chromosome to his or her children. With 80% penetrance, 40% of offspring will be affected phenotypically.

177. d. See answer 176.
178. d. See answer 176.
179. e. Patients who are post–radiation therapy for retinoblastoma are susceptible to many malignancies, including all of the listed tumors, but most notably osteogenic sarcoma.
180. True. Life expectancy is markedly diminished.
181. c. Although the presenting symptoms and signs depend on the extent of the tumor, the frequency of some of the presenting signs and symptoms are summarized below, with leukocoria and strabismus presenting in most of the instances:
- leukocoria: 60%
- strabismus: 22%
- decreased vision: 5%
- incidental finding: 4%
- orbital cellulitis: 2%
182. e.
183. c. Two diagnostic features of retinoblastoma are chalky white deposits on the tumor (which represent calcifications) and multiple clumps of tumor cells floating in the vitreous (vitreous seeding).
184. False. Growth into the vitreous is termed endophytic (endo-, toward the inner retina). Growth in the direction of the sclera is exophytic (exo-, toward the outer retina).
185. False. A rotary component is a common finding in congenital nystagmus.
186. b. Given the acidic nature of nucleic acids, they will be basophilic.
187. False. The rapid tumor growth exceeds the blood supply. Necrosis occurs with preservation of cells around blood vessels.
188. True. Although the course of retinoma is benign, the fellow eye may show a retinoblastoma, and the genetic implications are identical.
189. d. Direct spread into the optic nerve and central nervous system (CNS) is most common. Bone metastasis is seen in over half of metastatic cases. Lymph node metastases are the second most common type.
190. False. This relationship is common enough to be labeled “trilateral” retinoblastoma and likely represents a primary malignancy of the pineal gland.

191. c. Computed tomography (CT) scanning and ultrasonography are standard because they can demonstrate otherwise undetectable calcifications. Magnetic resonance imaging (MRI) is helpful in determining the extent of tumor spread. Serum levels of carcinomaembryonic antigen may be elevated in large tumors. Paracentesis is not a typical part of the workup but may be used in difficult cases to show an elevated level of lactate dehydrogenase in aqueous humor (relative to serum levels).
192. a.
193. d. Cryotherapy is as effective as photocoagulation for small tumors that are difficult to manage with photocoagulation.
194. e. All the disorders listed can be associated with optic nerve hypoplasia.
195. c. The combination of high lipid content (yellow) and vascularity (red) leads to the color of these benign growths. They can also be yellow-brown, especially when located on the iris.
196. False. Skin lesions are more common than those in skeletal muscle.
197. True. The risk of spontaneous hyphema from iris lesions is high enough to prompt treatment.
198. b. The only medulloepitheliomas associated with significant metastatic risk are those that extend into the orbit.
199. c. This is a classic description of a choroidal osteoma. These tumors are composed of mature bone; they commonly become yellow-white by the late teenage years (secondary to overlying retinal pigment epithelium [RPE] atrophy).
200. b. Both of these neoplasms can involve the choroid. The other two are primarily extraocular/orbital neoplasms.
201. True. The highly vascular uveal tissues are common sites for leukemic infiltration.
202. a. Gliomas and other central nervous system (CNS) neoplasms (aside from neurofibromas) are found in <10% of patients with this disorder.
203. d. Cutaneous melanoma does not appear to be more frequent.
204. True. Lisch’s nodules, like nevi, are composed of groups of benign, nevus-type melanocytes.
205. False. These gliomas are more “hamartomatous” and less neoplastic than isolated glioma.
206. c. The photoreceptors should not be affected, and the electroretinogram will be normal.
207. b. Adenoma sebaceum consists microscopically of an angioblastoma.
208. True.
209. False. The seizures are contralateral (secondary to the presence of leptomeningeal hemangioma overlying the ipsilateral cerebral convexity).
210. False. Diffuse choroidal hemangioma is classic, leading to the so-called tomato ketchup fundus. Focal
choroidal hemangioma is also seen in this disorder but is more frequently isolated.

211. False. This is a leading candidate for the etiology, but it has not been shown to be the sole cause (or even a definitive primary cause). Some patients have less elevated episcleral venous pressure (EVP) with an angle anomaly. Rubeosis may occur later in the disease course, as well.

212. b.

213. True. In Sturge-Weber syndrome, the retina overlying a diffuse hemangioma may develop serous detachment or ischemia and provoke neovascularization. In retinal angiomatosis, the cause tends to be serous detachment of the retina secondary to leakage at the capillary hemangioma.

214. False. Rare bilateral cases have been reported, but this is quite rare.

215. c.

216. c.

217. False. Patients with ataxia-telangiectasia tend to die from recurrent respiratory infections or leukemia/lymphoma before reaching middle age.

218. e. The others are autosomal dominant except for Sturge-Weber syndrome, which is sporadic.

219. c. See answer 218.

220. a. Optic nerve atrophy—from chronically high intracranial pressure or abnormalities of the optic canal—has been described, but hypoplasia is not a common finding.

221. c. V-pattern exotropia is common among the craniosynostoses.

222. False. Abnormalities in the development and origins of the extraocular muscles make correction of the strabismus very difficult; furthermore, should reconstructive surgery for the orbits be necessary, as is often the case, any good results obtained from muscle surgery would almost certainly be ruined.

223. a. The findings of this deformity/anomaly are present in a number of specific syndromes.

224. b.

225. False. Primary telecanthus is an increased distance between the medial canthi that is the result of soft tissue abnormality alone; secondary telecanthus is, as the name implies, caused by an underlying process such as hypertelorism (in which the medial walls of the bony orbits are actually further apart).

226. b. Primary telecanthus may be seen, but not hypertelorism (see answer 225).

227. b. Marked corneal clouding is seen with Hurler’s syndrome, and a spiral or whorl pattern (cornea verticillata) is associated with Fabry’s disease.

228. b.

229. a. Skew deviation may occur without reason for alarm during the early perinatal period. The other signs are of great concern.

230. b. The disorder could not be dominant, or at least one of the parents would have had it. X-linked recessive is unlikely because the children’s father would have had to have the phenotype for the daughter to inherit it (she would need two affected X chromosomes). Some previous history, in the maternal lineage, would be necessary to support a mitochondrial mode of inheritance; also, one would expect more affected siblings.

231. c. Achromatopsia, with few or no functioning cones, and Leber’s congenital amaurosis, with few or no functioning photoreceptors, cause severe visual compromise. Hypoplastic optic nerves usually appear small on examination, although this may often appear subtle on examination. Lowe’s syndrome should feature an abnormal examination (cataracts or glaucoma).

232. a. The superior oblique and the levator muscles each arise posteriorly, above the annulus. The inferior oblique arises from the anteromedial orbital floor. The annulus gives rise to the four rectus muscles.

233. e. The horizontal rectus muscles have no torsional or vertical action in primary gaze. In upgaze, they are elevators (weak); in downgaze, they are depressors (weak).

234. d. With the eye abducted 23 degrees, the superior rectus is parallel to the visual axis, and its contraction will result in maximal elevation. For the same reason, in this position the inferior rectus is maximized as a depressor. In primary gaze, contraction of the superior rectus not only elevates the eye but also intorts and adducts the eye. (The inferior rectus will extort and adduct the eye, as well as depress it, in primary gaze.)

235. a. With the eye adducted 51 degrees, the tendon of the superior oblique is parallel to the visual axis, and its contraction will result in maximal depression. For the same reason, in this position the inferior oblique will be maximized as an elevator. In primary gaze, contraction of the superior oblique not only depresses the eye but also intorts and adducts the eye. (The inferior oblique will extort and abduct the eye, as well as elevate it, in primary gaze.)

236. False. The inferior oblique’s anatomic and mechanical origins are the same—from the anteromedial orbital floor. The superior oblique has its anatomic origin at the orbital apex. Its mechanical origin is at the trochlea because this is where its effective force is generated.

237. True. The oblique muscles always run below the corresponding rectus muscle.

238. False. The inferior rectus lies between the globe and the inferior oblique (the obliques pass below the recti).

239. False. By the time it reaches the trochlea, the superior oblique is already tendinous.

240. b. The inferior oblique also contributes to elevation in primary gaze.

241. a. The superior muscles act as intorters, whereas the inferior muscles act as extorters. The oblique muscles are more important for torsion than the vertical recti.
Therefore, the primary intorter of the globe in primary gaze is the superior oblique.

242. c. The belly of the superior oblique is approximately 32 mm (the others are 37 to 40 mm).

243. d. The inferior oblique is muscle virtually all the way to its insertion over the posterior sclera.

244. a. The lateral rectus muscle contributes only one anterior ciliary artery to the major arterial circle. Therefore, there are a total of seven anterior ciliary arteries.

245. False. All six extraocular muscles that attach to the globe must penetrate Tenon’s capsule. The levator muscle does not.

246. True.

247. False. Typically, the motor nerve penetrates each muscle at the junction of the posterior one third and anterior two thirds of the belly.

248. b. Because the superior rectus is loosely attached to the levator complex, recession will pull the levator back and widen the fissure, whereas resection will pull the levator forward and narrow the fissure. The same relationships apply for the inferior rectus because its sheath gives rise to the capsulopalpebral fascia. Operation on the superior oblique should not affect the palpebral fissure.

249. c. Two muscles may nearly always be safely removed. Removal of four muscles is certain to cause ischemia. Removal of three muscles is likely to lead to some degree of ischemia.

250. c. The main synergist of the superior rectus is the inferior oblique (both elevate the globe).

251. True. Hering’s law of yoke muscles states that “yoked” (contralateral) muscles must always receive equal innervational input. Sherrington’s law states that innervation to an (ipsilateral) antagonist decreases as innervation to the agonist increases.

252. b. In dissociated vertical deviation (DVD), the affected eye drifts up under cover. When the cover is shifted to the other eye, if Hering’s law is followed, yoke innervation should generate a contralateral hypotropia. This is not seen in DVD for reasons that are unclear.

253. c. In Duane’s syndrome, innervational impulses to the medial rectus are not associated with decreased innervation of the ipsilateral lateral rectus muscles, as Sherrington’s law demands. This is because of anomalous innervation of the lateral rectus, in this case from the oculomotor (III) nerve, which leads to c innervation of the horizontal rectus muscles, with globe retraction on adduction.

254. True. Conjugate binocular movements imply both eyes move a similar amount in a similar direction (i.e., right, left, up, and down) at the same time.

255. False. Fusional convergence is always accompanied by accommodation, but divergence is not always accompanied by relaxation of accommodation.

256. b.

257. d. Note that saccades are generated contralaterally, whereas pursuits are generated ipsilaterally.

258. False. Within a certain range around the empirical horopter—that is, Panum’s space—objects will be perceived as one, with stereopsis. Any point on the horopter must be perceived singly.

259. False. Panum’s space is broader peripherally and narrower centrally. Again, this range describes the area of object space where an object off the horopter will still be perceived singly and with stereopsis.

260. False. If simultaneous stimulation of two retinal areas leads to perception of one image, then sensory fusion is said to exist. If the two retinal areas have identical topographic locations relative to the fovea, then normal retinal correspondence (NRC) exists.

261. False. Stereopsis can be generated for points off the horopter, if they lie within Panum’s space. Some discrepancy in perception between the two eyes must exist for stereopsis to follow. There is no discrepancy for objects located on the horopter.

262. False. In abnormal or anomalous retinal correspondence (ARC), simultaneous stimulation of two areas of retina that do not have the same topographic relation to the fovea (noncorresponding) still leads to perception of one image (sensory fusion).

263. True. If the two images are sufficiently different, visual confusion will be created.

264. d. Normal divergence amplitudes at distance is 6 prism diopters. At near, this increases to 16 prism-diopters.

265. False. Monocular clues such as image size, motion, and parallax are probably more important than stereopsis (a higher binocular function) for satisfactory depth perception.

266. d.

267. c.

268. True. In kittens, the critical or sensitive period is probably 6 to 12 weeks. In children, the first 3 to 6 months are particularly important, but the visual system is susceptible to developmental arrest (amblyopia) for the first 6 years of life, perhaps longer.

269. False. Diplopia, or double images of a single object, exists when the two images of the object fall on non-corresponding areas of the retina. When two different objects are simultaneously imaged on the foveas and perceived, visual confusion exists.

270. False. Although suppression is the most common explanation for absence of diplopia with manifest strabismus, marked organic visual loss in the deviating eye will prevent diplopia, requiring no central suppression. In large-angle esotropia, the nasal bridge may “occlude” the deviating eye and prevent diplopia. Anomalous retinal correspondence also may prevent diplopia despite an obvious tropia.
271. b. Approximately one in 50 people suffer from amblyopia. The incidence of strabismus, with or without amblyopia, is slightly higher, at one in 25 to 30 people. Afferent pupillary defects may be seen in amblyopia of any cause but should heighten suspicions about an organic lesion. “Crowding” refers to poorer recognition acuities in amblyopia when targets are presented multiply rather than singly. Neutral density filters will not significantly affect vision in amblyopes, unlike patients with maculopathy.

272. c. Amblyopia in Duane’s syndrome is uncommon but is more likely than in alternating esotropia, where each eye shares the visual workload.

273. False. One week for every year of age is the typical pattern for follow-up of the patient undergoing treatment for amblyopia.

274. b. Cyclic esotropia is rarely accompanied by amblyopia. With full eye movements and no globe retraction, there can be no Duane’s syndrome, particularly given the amblyopia. Parafoveal fixation is difficult to assign to factitious etiologies. All of the findings described (mild amblyopia, mildly impaired stereopsis, peripheral fusion, eccentric fixation with a central suppression scotoma) are typical of a microtropia (the tropia is either too small to detect or does not exist because of parafoveal fixation and anomalous retinal correspondence).

275. c. Microtropia is most commonly seen after successful (or nearly so) strabismus surgery, as well as in the setting of anisometropia.

276. d. A weak base-out prism will shift a fixation target nasally. If placed over the microtropic eye, the shift will not move the target out of the suppression scotoma, and no refixation movement is generated. When placed contralaterally, both eyes will shift away from the base of the prism. No fusional convergence will be seen contralaterally because of the central scotoma. In normal eyes, the contralateral eye converges to fuse the fixation images.

277. False. A small central scotoma may permit fusion with normal retinal correspondence (NRC) and without visual confusion.

278. d. Esotropia gives uncrossed diplopia on red glass testing. Thus, the left light (red) should appear to the left of the right light (white).

279. c. The right image is probably being suppressed. There may be either normal retinal correspondence (NRC) or anomalous retinal correspondence (ARC). To determine which, the right image is moved out of the central suppression scotoma with a vertical prism. In ARC, the right white light will appear directly below the left red light. On the other hand, it is possible that the patient is malingering (voluntary convergence spasm) and being less than truthful about his perceptions. Findings supportive of this conclusion include a variable and unpredictable amount of esotropia in various gaze positions, as well as noticeable miosis with lateral gaze, particularly to the right.

280. c. When identical images are projected into each eye, if the angle of the tropia and the angle of subjective image separation are equal, there is normal retinal correspondence (NRC). If images are superimposed by a prism whose power is different than that required to neutralize the tropia, there is unharmonious anomalous retinal correspondence (ARC). If ARC is harmonious, there will be fusion with no prism. An amblyoscope is used to project similar images onto each fovea. The angle of deviation between the two targets matches the tropia angle in NRC. If the angle of deviation is less than the tropia, ARC is harmonious. If the angle is 0 in the setting of a tropia, ARC is harmonious.

281. d. It is better to flash the vertical line into the deviated eye because displacement of a horizontal line may be missed with a suppression scotoma. If there is eccentric fixation, the test is more difficult to interpret. In right exotropia and anomalous retinal correspondence (ARC), the image the patients see with the right eye (vertical) will be displaced to the right (because it lies on the anatomic fovea, nasal to the "pseudofovea" of ARC).

282. True. Bagolini-glass testing cannot be interpreted without knowledge of the strabismus findings and fixation behavior.

283. c. There is no manifest deviation (tropia, cover–uncover test) but there is a latent esodeviation (phoria, alternate-cover test).

284. e. In the setting of an esotropia (detected by cover-uncover test), the alternate-cover test must disclose some strabismus because it detects both active/accommodative components, alternating versus nonalternating) as well as which eye is more at risk. Family history is not critical in determining the actual strategy as a rule.

285. False. Most of the normal adult population has a small phoria, more often exophoria.

286. d.

287. a. Decisions about occlusion therapy require an assessment of the risk of amblyopia (age of onset, refractive/accommodative components, alternating versus nonalternating) as well as which eye is more at risk. Family history is not critical in determining the actual strategy as a rule.

288. a. Snellen figures, particularly presented in rows, are the most challenging, and perhaps most relevant, type of acuity test.

289. e. Optokinetic nystagmus has been used to quantify acuity, but the procedure is more cumbersome and difficult to score accurately than preferential looking.

290. a. The Krimsky test uses prisms over the fixating eye to center the light reflex over the pupil in the deviating eye. Foveal fixation is not required.
291. b. Each millimeter of decenteration is 7 degrees of deviation. Each degree is approximately 2 prism diopters. Thus, each millimeter is approximately 15 prism diopters. Three millimeters is 21-plus degrees, or 45 prism diopters. Because the reflex is displaced temporally, the eye must be deviated inward (esotropia).

292. d. See answer 291.

293. d. As the fixing eye is covered, a prism is simultaneously placed over the deviated eye. When there is no refixation shift, the prism has neutralized the tropia. The alternate-cover test with prisms will neutralize total phoria plus tropia. The cover-uncover test with prisms will neutralize tropia plus part of the phoria.

294. False. The real image of a Maddox rod is a line parallel to the axes of the rod, immediately in front of the rod. It requires too much accommodation to be seen. The image that is perceived is a virtual image of a line perpendicular to the rod axes, behind the rod.

295. True. The image formed is a horizontal line that permits perception of vertical image disparity.

296. d. In excyclotropia, the superonasal retinal quadrant is rotated vertically toward the 12-o’clock position, and the inferotemporal quadrant is rotated toward the 6-o’clock position. The vertical line from the Maddox rod will run superonasal to inferotemporal on the retina and be perceived the same way.

297. a. In the Lancaster red-green test, the fovea of each eye is isolated with duochrome glasses. The eye under green glass (left) will not see the red light and the eye under the red glass (right) will not see the green light. By holding the green light centrally, the examiner holds the patient’s left fovea centrally. Assuming normal retinal correspondence (NRC), the patient will direct the red light in space along his right visual axis to place the red light on his right fovea and superimpose the foveal images. The patient’s right visual axis (in esotropia) crosses his left visual axis, so the right foveal image will fall to the left of the left foveal image, as seen by the examiner.

298. e. This is true because the strabismus is acquired. In that case, we can assume normal retinal correspondence (NRC) and central fixation.

299. d. In this case, the deviated right eye is fixing centrally (green glass). The larger secondary deviation, a left esotropia of greater magnitude, will cause the red light (left foveal image) to appear to the right of the green light (right foveal image).

300. e. Because of suppression, the Lancaster test is not useful for congenital strabismus, but only for acquired cases. In this case, the patient will see only one light.

301. d. The first three tests utilize actual fixation behavior and will not fall victim to large angle kappa or facial anomalies. The Krimsky and Hirschberg methods use apparent eccentricity of pupillary reflexes and may be misguided by these factors.

302. True. Nasal displacement of the light reflex corresponds to positive angle kappa and pseudoesotropia. This occurs with temporal macular ectopia. Temporal displacement of light reflex corresponds to negative angle kappa (pseudoesotropia). This occurs with nasal macular ectopia.

303. True.

304. b. The base of the prism is always placed opposite to the direction of the deviation.

305. c. See answer 304.

306. True.

307. c. A 25 prism diopter increase in esotropia from distance to near is almost certainly a high accommodative convergence to accommodation ratio (AC/A).

308. c. By the gradient method, accommodative convergence to accommodation ratio (AC/A) equals the difference in the deviation induced by a lens divided by the specific accommodative gradient (of an extra lens over the distance correction). Minus lenses stimulate accommodation, whereas plus lenses blunt it. In this example: \( \frac{35 - 10}{50} = 5 \), plus 6 (PD in cm) = 11:1.

Heterophoria method:

\[ \frac{\text{AC/A}}{\text{deviation at near} - \text{deviation at distance}} = \text{accommodation (D)} \]

Gradient method:

\[ \frac{\text{AC/A}}{\text{deviation with lens} - \text{deviation without lens}} = \text{lens power (D)} \]

310. a. Factors known to be associated with decompensated latent strabismus include alcohol, fatigue, and illness. Decreased visual acuity (e.g., cataract) also may lead to fusional breakdown, with resultant manifest strabismus.

311. True. When performed at distance, central retinal elements are stimulated, and central fusion is evaluated. When performed at near, more peripheral fusion processes are tested. The test is not capable of detecting small suppression scotomata (i.e., results of near Worth four-dot testing may be normal in the setting of a central scotoma).

312. d. The finding of a left hypertropia indicates that more than a simple right trochlear palsy is present but does not eliminate this possibility because the left hypertropia is seen only in right gaze (indicating...
probable left superior oblique palsy). A right hypertropia in primary gaze that is worsened with left gaze is classic for right trochlear palsy. The V-pattern esotropia is convincing evidence of a bilateral superior oblique palsy, but a right hypertropia worse in right gaze by the three-step test, maps to the right inferior rectus or the left inferior oblique and is therefore inconsistent with the diagnosis.

313. a. Right head tilt stimulates intorsion of the right eye to keep the visual field at the proper angle. The intortors are the superior rectus and the superior oblique. In the setting of a superior oblique palsy, the vertical, elevating force generated by the superior rectus is not effectively balanced by the depressing force of the superior oblique, so the eye goes up.

314. c. The clinical findings of mydriasis and accommodative paresis indicate damage to the parasympathetic supply to the globe. These nerves travel with the nerve to the inferior oblique before forming the short root of the ciliary ganglion. The motility findings in this case indicate a probable inferior oblique palsy (inability to elevate the adducted eye). The three-step test should show a left hypotropia (right hypertropia) worsened in right gaze and right head tilt.

315. a. Exaggerated sensitivity to cholinergic blockade has been reported in albinos and patients with Down’s syndrome. Infants are also particularly sensitive. Atropine is a treatment for heart block (acute) and should not cause arrhythmias in patients with this disorder.

316. c. The central nervous system (CNS) side effects of atropine include both alerting/agitation and somnolence. Flushing and tachycardia are particularly common in infants.

317. False. The prism adaptation test involves prescription of corrective prisms in anticipation of muscle surgery to determine if the patient can achieve sensory fusion with alignment. This test may be associated with an increase in the angle of strabismus because the patient becomes accustomed to the prisms.

318. b. With cross-fixation, amblyopia is less likely to develop because the younger will use each eye at different times. An accommodative component may be discovered in many cases of infantile esotropia.

319. a. Latent nystagmus, overacting inferior obliques, and dissociated vertical deviation are such common concomitants that they should be specifically sought in the examination of a child with infantile esotropia. A high accommodative convergence to accommodation ratio (AC/AS) may be seen (see answer 318) but is not typical.

320. a. Surgery for esotropia must provide either weakening of the medial rectus muscles (recession) or strengthening of the lateral rectus muscles (resection). Recession always has a greater effect than a resection of the same amount. Thus, bimedial recession is generally performed before bilateral resection. In some cases, however, bilateral resection may be the first procedure (for instance, in esotropia that is greater at distance). Combined medial and lateral resection is an equally effective alternative.

321. True. Success in surgical management of infantile esotropia is multifaceted, but with reference to the strabismus, the residual turn should be <10 prism diopters for the procedure to be termed successful. This amount of residual tropia is frequently cosmetically undetectable. If the residual turn is too small to be detected with typical cover-uncover testing (<5 to 8 prism diopters), a microtropia exists. This situation is usually stable over time and is associated with peripheral fusion and limited stereopsis. If there is eccentric fixation near the fovea, visual acuity will be mildly depressed with no tropia evident on cover testing (but no central fusion).

322. True. Congenital motor nystagmus frequently has a point where the amplitude and frequency are dampened (null point). Some experts believe that some children are esotropic in order to hold their null point in eye position. This contention is not universally accepted, however.

323. c. Amblyopia is certainly possible given the constant esotropia. A large-angle turn might provide cross-fixation, but amblyopia must be ruled out. Given the age of onset, an accommodative component is likely, so an originally intermittent turn is also possible. No patient with manifest strabismus has any stereopsis whatsoever. Certainly, a family history of esotropia of any mechanism is possible.

324. b. This patient probably has refractive accommodative esotropia. Amblyopia is possible bilaterally, given the high ametropia. The deviation may be greater at near than distance, but this is not highly likely because patients with refractive accommodative esotropia typically have normal accommodative convergence to accommodation ratios (AC/AS). In this type of esotropia, the turn is usually moderate (20 to 40 prism diopters), although it may be greater. Lenses of +3.00D may lessen nonrefractive accommodative esotropia but typically have little effect on this type (with +3.00D lenses, there would still be a residual SD of hyperopia).

325. c. In all cases of accommodative esotropia, full hyperopic correction is warranted immediately. Penalization with atropine may be useful in cases of noncompliance with spectacles and/or patching therapy. Bifocals may be of value with high accommodative convergence to accommodation ratio (AC/A) with a residual turn at near after full correction.

326. a. This is probably a case of nonrefractive accommodative amblyopia (intermittent turn, worse at near, with normal refractive error and high accommodative convergence to accommodation ratio (AC/A)). Lenses of +3.00D for near work will relieve the accommodative demand and prevent accommodative convergence from causing an esotropia.
327. d. Surgery is necessary in virtually all cases of infantile esotropia, whereas many cases of pure accommodative esotropia will resolve with time and refractive correction. Bilifocals are generally most helpful in nonrefractive accommodative esotropia, where high accommodative convergence to accommodation ratios (AC/A) make the esotropia worse at near. If full refractive correction is not the solution, some experts advocate atropine penalization with full correction or pilocarpine treatment to provide accommodation without convergence. These steps frequently fail, however, and are controversial.

328. d. Age of onset between 2 and 3 years makes an accommodative mechanism more likely, with better prognosis for refractive correction. The other findings are consistent with a large-angle, congenital esotropia.

329. c. Persistent esotropia on pilocarpine treatment may be seen in accommodative esotropia, so spectacle correction must always be attempted. Surgical results are much more stable and predictable in the setting of maximal visual acuity (after occlusion therapy).

330. False. The converse is true. That is, with illness or stress, nonrefractive accommodative esotropia (high accommodative convergence to accommodation ratio [AC/A]) is more likely to decompensate than the refractive variety.

331. c. Correction with full hyperopic prescription may weaken the patient’s fusional divergence, which is the force keeping accommodative esotropia intermittent at its outset. Then, the esotropia may become constant without the “crutch” of the spectacles.

332. a. The cycle is typically 48 hours long: 1 day of esotropia, and 1 day of orthotropia. Many experts believe that this is a variety of accommodative esotropia, with many features similar to it.

333. d. Divergence insufficiency is indistinguishable from sixth nerve palsy except it is typically comitant. Sixth nerve palsy is more likely to have an esotropia at near as well.

334. b. The myopia may be hard to establish if the spasm is intermittent, but the miosis is detectable and is the diagnostic “clincher.” Also, the angle of turn is highly variable and unpredictable.

335. False. Although many experts will obtain timely neuroimaging tests (computed tomography [CT]/magnetic resonance imaging [MRI]), abducens palsy in a child is a common postviral syndrome and typically resolves uneventfully. The second most common etiology is increased intracranial pressure (central nervous system [CNS] mass lesions, pseudotumor cerebri).

336. c. Unless the palsy is acquired early in childhood, goes untreated (with occlusion), and/or does not resolve, amblyopia is highly unlikely. Some children will develop a permanent esotropia after abducens palsy, probably representing a decompensated esophoria. Thus, all cases must be followed to resolution.

337. b. Medial rectus resection would aggravate the esotropia. The most widely accepted approach in sixth nerve palsy is a muscle-splitting operation utilizing half of the superior and inferior rectus muscles in transposition laterally (Jensen’s procedure). Prisms may be of temporary assistance, but should be base-out to correct the esotropia.

338. False. Consecutive exotropia of moderate degree after surgery for esotropia is not uncommon. It commonly resolves within several weeks or months of surgery with no therapy, other than continued occlusion.

339. True. This is, in general, a useful guideline. It is by no means foolproof, particularly in adults with longstanding esophoria, who may decompensate into esotropia with unilateral visual loss.

340. b. Exotropia that is equal at distance and near is basic. If the deviation is greater at near, then it is termed convergence insufficiency exotropia. If it is greater at distance than near, it is termed divergence excess exotropia.

341. d. Divergence excess exotropia may be divided into two subtypes. In some cases of divergence excess exotropia, the turn appears larger at a distance because of enhanced fusional convergence at near related to accommodation. Interrupting fusion with prolonged occlusion (>30 minutes) or relaxing accommodation with plus lenses may cause the deviation at near to increase to a measurement similar to the original distance measurement. This is simulated divergence excess. If these manipulations have no effect (i.e., the distance measurement is still larger), true divergence excess is said to exist.

342. d. See answer 341.

343. c. See answer 341.

344. False. Congenital exotropia is relatively uncommon. Most exotropias develop during childhood as intermittent deviations.

345. a. Most constant exotropia is intermittent originally.

346. False. Some cases of exotropia may resolve entirely, but this is much less common than with accommodative esotropia. Most cases remain intermittent, becoming manifest with fatigue, stress, illness, or alcohol consumption. Some cases become constant and require surgery.

347. a. High accommodative convergence to accommodation ratios (AC/A) develop as a fusional mechanism for near work. The turn in intermittent exotropia is highly variable and sensitive to external stimuli (see answer 346). Bright light typically causes reflex closure of the deviating eye. Amblyopia is highly unlikely if the turn is truly intermittent.

348. False. Although this may be occasionally the case, most patients with truly intermittent exotropia have excellent stereoacuity.

349. b. Minus lenses stimulate accommodation and may provide the extra convergence needed for fusion. Base-in prisms may permit fusion but are not advocated by all investigators because they may weaken
Many cases remain intermittent, with decreasing frequency and amount of manifest exotropia. False. It is often larger with attempted adduction. It will also be marked by globe retraction on adduction.

By definition, convergence insufficiency is a latent deviation, not a manifest one. (This is not the same entity as convergence insufficiency exotropia, which is a manifest deviation.) True. Base-out prisms may be used to stimulate fusional convergence, whereas base-in prisms may be used to lessen the convergence demand. True.

False. Innerveational (i.e., idiopathic) vertical strabismus is far less common than the horizontal varieties.

The left eye deviates upward under cover, and the right eye deviates downward under cover. This is a left hyper–or a right hypodeviation.

Dissociated vertical deviation simulates a hyperphoria but violates Hering’s law. When covered, the left eye drifts up. When uncovered, the left eye moves down to assume fixation. By Hering’s law, the yoke muscles of the right eye, depressors, should receive equal innervation as the left eye moves down. The right eye should be deviated downward under cover (right “hypo”) and move up to reassume fixation when the cover is shifted back to the left eye. This does not occur, consistent with dissociated vertical divergence. (The dissociation refers to the violation of Hering’s law with dissociation of yoke muscles.)

Dissociated vertical deviation is a frequent concomitant of infantile esotropia and frequently is made manifest only when visual input to the affected eye is interrupted (occlusion, amblyopia, other organic disease). The deviation is highly variable and difficult to measure, making surgery difficult to quantify accurately.

This set of findings is not particularly unusual in infantile esotropia. There are two potential explanations for the vertical deviation in lateral gaze. The first is overaction of the inferior obliques. In this condition, on lateral gaze, there is hypotropia of the abducted, nonfixing eye. In dissociated vertical deviation (DVD), in lateral gaze there is no associated hypotropia, in violation of Hering’s law. In fact, if the DVD is bilateral, there may be hyperdeviation of the abducted, nonfixing eye. The distinction between the two is important for surgical planning.

Overaction of the inferior oblique may be seen as a primary disorder or secondary to underaction of the ipsilateral, antagonist superior oblique. This duality of causes for superior oblique overaction is not seen. Because isolated underaction of the inferior obliques is quite uncommon, secondary overaction of the superior oblique is also uncommon. Only primary mechanisms are recognized.

Head tilt and excyclotorsion are not uncommonly seen in unilateral cases and are not useful. V-pattern, not A-pattern, esotropia results from underaction in downgaze and subsequent unopposed adduction in downgaze by the inferior rectus. In unilateral superior oblique palsies, lateral gaze toward the involved side generally relieves the diplopia. Aggravation of diplopia in both directions argues for bilateral involvement, as does aggravation with head tilt in either direction.

If the V-pattern esotropia does not cause troublesome diplopia, then surgery is not indicated.

Superior oblique pareses are particularly prone to develop “spread of comitance,” so that the deviation becomes more difficult to localize. Surgical therapy must also address this.

Each millimeter of vertical muscle recession will correct approximately 3 prism diopters of vertical strabismus.

Surgical management of superior oblique underaction is generally aimed at weakening the ipsilateral inferior oblique (myectomy) and/or strengthening the ipsilateral superior oblique. If the hyperdeviation is >35 prism diopters in primary gaze, then strengthening the ipsilateral inferior rectus or weakening the ipsilateral superior rectus may be of value. Contralateral superior oblique weakening also may be useful in certain cases.

This procedure, called the Harada-Ito procedure, increases the force vector for incyclotorsion by moving the lateral half of the insertion of the paretic muscle. It has no effect on vertical eye movement or fusion.

Double elevator palsy may be primarily caused by elevator weakness or restriction of the depressors. Ptosis, hypotropia, and poor elevation in any direction are characteristic. A subset will have positive forced ductions for the inferior rectus. Head position is generally an automatic compensation for the hypotropia, with the chin up.

In Brown’s syndrome, elevation is limited in adduction, but not in abduction. Duction testing mimics paresis of the ipsilateral inferior oblique.

Enophthalmos is frequently cosmetically unacceptable. In the acute setting, there may be proptosis, but this usually gives way to enophthalmos as swelling subsides.

For an A deviation to be significant, the difference in measurements between upgaze and downgaze must exceed 10 prism diopters. For V patterns, the difference must measure 15 prism diopters. Approximately 15% of horizontal strabismus cases have a significant A or V component.

Oblique underaction or overaction is frequently implicated in the pathogenesis of A and V patterns.
The basic procedure must address the deviation in primary gaze. If there is no significant oblique dysfunction, then transposition of the rectus insertions may be performed to correct the A or V patterns. If there is oblique dysfunction, then the type and amount must be determined.

In V-pattern exotropia, one should search for overaction of the inferior oblique or underaction of the superior obliques. If neither is present, then lateral rectus recession and medial rectus resection should be performed with downward transposition (toward the apex of the V) of the medial rectus insertion, by one-half the insertion width, and upward transposition (toward the empty space of the V) of the lateral rectus insertion, again by half the insertion width. The mnemonic for this is “MALE”: Medial rectus toward the Apex and Lateral rectus toward the Empty space. This holds for both A patterns and V patterns.

In this case of V-pattern exotropia, there is significant overaction of the inferior obliques. Bilateral inferior oblique myectomies will lessen the V pattern by 15 to 25 prism diopters but leave a residual exotropia in primary gaze, necessitating a recess–resect procedure. When oblique muscle surgery has been performed, rectus transpositions are not necessary.

In V-pattern exotropia with overacting inferior obliques, inferior oblique myectomy will cause 15 to 25 prism diopters of esohist in upgaze. This only makes the deviation more comitant. Thus, the deviation in primary gaze must be addressed with appropriate medial rectus recession and lateral rectus resection. Because oblique surgery is indicated, rectus transpositions are not.

Here, because the A-pattern esotropia is >35 prism diopters in upgaze, overaction of the superior obliques may be addressed with superior oblique tenotomies bilaterally. This will create an esohist of up to 40 prism diopters in downgaze (only). As with V-pattern esotropia and inferior oblique myectomy, this only makes the deviation more comitant. Recess–resect (or bimedial recession) procedures must be included.

In this case, the A pattern is <35 prism diopters, so bilateral superior oblique surgery might leave a residual esotropia in downgaze after horizontal muscle surgery. In this case, the A pattern can be addressed through transpositions of the rectus insertions (medial rectus toward the apex—up; lateral rectus toward the empty space—down).

False. Quantifying the effect of superior oblique tenotomy is particularly difficult, making adjustable sutures valuable.

b. Remember that the superior oblique muscles are abductors, particularly in downgaze. Thus, overaction will result in overabduction (exotropia) in downgaze (A pattern).

a. Klippel-Feil syndrome is one spinal anomaly associated with Duane’s syndrome. (Goldenhar’s syndrome features cervical vertebral anomalies as well.)

False. This is an important differentiating feature between Duane’s syndrome and sixth nerve palsy. The medial rectus muscle nearly always undergoes progressive contracture in the setting of a nonresolving sixth nerve palsy. This virtually never occurs in Duane’s syndrome. Forced ductions may help differentiate between the two in puzzling cases of a long-standing abduction deficit.

Amblyopia in Duane’s syndrome is generally caused by anisometropia rather than strabismus. In any case, it must be treated before attempting surgical realignment. Globe retraction may be lessened by lateral rectus recessions but is rarely if ever a solitary indication for surgery.

False. The brainstem nuclei most likely to be involved by Möbius’ syndrome are the sixth, seventh, ninth, and twelfth. The third nerve nucleus also may be involved, however, and oculomotor weakness does not make the diagnosis less likely.

The etiologies for pediatric third nerve palsy, in descending frequency, are congenital, traumatic, inflammatory, migrainous, and neoplastic.

The etiologies for adult third nerve palsy, in descending frequency, are microvascular, aneurysmal, traumatic, and neoplastic.

Because muscle surgery, particularly the vertical muscles, often indicated in Graves’ disease, can affect the position of the eyelids, it is generally wise to perform any strabismus surgery before eyelid reposi-
tioning. For instance, some of the lid retraction seen in affected patients may be directly attributable to superior rectus overaction (with subsequent lid retraction) attempting to counter inferior rectus contracture.

False. The deviations of Graves’ disease are classically noncomitant and not easily addressed with prism.

Adjustable sutures are very helpful in this condition (which may respond unpredictably to muscle surgery). Resection techniques, however, are generally avoided because Graves’ disease generally causes considerable restriction. Resection would tend to exacerbate this and leave an eye nearly frozen if overdone.

Because the deviations in myasthenia gravis are particularly variable and noncomitant, surgery may cause more difficulties than it relieves.

duane’s syndrome is more common in girls, whereas congenital apraxia is more common in boys.

b. See answer 391.

Jerk nystagmus has clearly biphasic velocities—fast in one direction and slow in the other. Disconjugate nystagmus has different amplitude and/or frequency in one eye relative to the other. Uniplanar nystagmus...
is, as its name implies, present in one plane only, usually horizontal.

394. True. Motor nystagmus is virtually always of the jerk type. In fact, some “nystagmologists” are reluctant to term any pendular variety as true nystagmus, maintaining that biphasic velocity is part of the definition. Many “pendular” cases of motor nystagmus are actually jerk, with very similar velocities requiring electrooculography (EOG) for differentiation.

395. True. This is in distinction to latent nystagmus, in which the slow phase shows exponentially decreasing velocity.

396. True. The hallmarks of congenital motor nystagmus include dampening with convergence and aggravation by fixation. Near work may be associated with convergence and dampening with subsequent improvement in acuity.

397. c. Latent nystagmus becomes manifest with monocular occlusion or visual loss and is probably underdiagnosed. It may be an unsuspected cause of “amblyopia” if acuity testing is performed with total occlusion. Using fogging plus lenses that provide blurred images to the “occluded” eye may circumvent this problem (decreased acuity caused by a moving, distorted image).

398. b. The nystagmus of spasmus nutans is typically small in amplitude but characterized by its disconjugate nature. It may appear to be entirely monocular and raise the specter of chiasmal or hypothalamic glioma. The third part of the classic triad is head bobbing.

400. True. The disorder is rarely discovered after the age of 3 years.

401. b. Many pediatric ophthalmologists will obtain neuroimaging tests of children with monocular nystagmus, unless combined with obvious head bobbing and torticollis. Cases of anterior visual pathway glioma associated with head bobbing have been reported.

402. c. This combination of prisms will force the child to gaze to the right in order to see objects straight ahead. This will obviate a head turn, which is typically the end point of treatment.

403. True. This is done by shifting the resting position out of primary position toward the direction opposite to the null zone. Then, to obtain primary gaze, the child will need to innervate ocular muscles as if gaze were toward the null zone.

404. c. This combination of procedures will shift both eyes to the left in resting position. In order to look straight ahead, the patient will be forced to innervate muscles in a pattern identical to right gaze (null zone) before surgery.

405. a. In these disorders, the angle of deviation is difficult to quantify or the effects of muscle surgery are hard to predict.

406. a. For cases in which the force-generating capability of a muscle is permanently and significantly depressed, resection techniques will offer little. Here, transposition of neighboring, healthy rectus muscles (Jensen’s procedure) is often helpful.

407. d. This may be a case of partially or totally accommodative esotropia, which will respond nicely to hyperopic correction. Bifocals might be added later if a significant residual esodeviation remains at near with distance correction. Any amblyopia must be treated aggressively with careful follow-up of the treatment effect on each eye.

408. d. Stimulation of accommodative convergence is unlikely to resolve the problem. Because the parents are determined to correct the cosmetic problem, and because the deviation is greater in left gaze, weakening of both lateral recti, more so on the left, should be undertaken.

409. d. Strengthening the ipsilateral superior rectus will aggravate the right hypertropia. A common approach might be superior oblique tuck alone (if the deviation is <25 prism diopters) or combined with inferior oblique tenotomy (if >25 prism diopters).

410. a. Although this probably represents a case of basic esotropia, there may be an accommodative component. There is no way to rule this out without a trial of hyperopic correction. Bifocals might be added later if a significant residual esodeviation remains at near with distance correction. Early surgery might be advocated to maximize the retention of fusion, but only the angle of deviation that remains after refractive correction should be addressed. Close follow-up, as always, is critical.

411. d. Because there is asymmetric vision caused by amblyopia, a right-eyed procedure should be chosen (to minimize operative risks on the better-seeing left eye). The guidelines for a 30-prism-diopter esotropia are 4.5 mm medial rectus recession and 7 mm lateral rectus resection.

412. True. Undercorrected exodeviations nearly always increase in size until there is total recurrence.

413. c. Monocular surgery is preferable. The guidelines for a 30 prism diopter exotropia are 6 mm of medial rectus resection and 7 mm of lateral rectus recession.

414. c. If this results in undercorrection, then additional surgery is necessary.

415. d.

416. d.

417. c.

418. c. The range is 15 to 25 prism diopters.

419. d. This procedure has little eso effect in primary gaze and none in upgaze.

420. a. The ischemia induced by surgery on three or more muscles is entirely anterior.

421. False. The converse is true—inferior rectus surgery nearly always involves manipulation of the lid retractors.
10: Pediatric Ophthalmology and Strabismus

422. b. Acquired forms of strabismus may be associated with troublesome postoperative strabismus as images are moved closer but not close enough. In congenital cases, suppression will prevent the complication. The prism adaptation test is indeed valuable for predicting postoperative behavior. Undercorrected intermittent exotropia will be associated with suppression and a quick recurrence of large-angle turns. The most annoying diplopia is frequently caused by small-angle residual turns, which are manageable with prisms. Many cases will resolve spontaneously over months.

423. d. Consecutive exotropia caused by overcorrection alone should not be associated with striking impairment in adduction.

424. c. Elevated body temperature is a relatively late sign. Careful surveillance for the earlier signs is critical.

425. d.

426. c.

427. c. Ptosis is the most common and is seen slightly more frequently in children. Secondary vertical strabismus is the second most common complication.

428. c.

429. c.

430. a. 4, b. 3, c. 1, 2, d. 2, e. 3, f. 1, g. 3, h. 2, i. 3, j. 2, k. 4. The time of onset of neonatal conjunctivitides after birth is important. Silver nitrate prophylaxis (medicamentosa) tends to present earliest, followed by gonococcus, herpes simplex virus (HSV), and chlamydiaceae. Gonorrhea must be treated with intravenous antibiotics. Intravenous acyclovir is now used for all forms of neonatal (HSV) infections. Treatment with mild topical antibiotics, or no treatment at all, is sufficient for chemical conjunctivitis.

431. a. 3, b. 1, c. 2, d. 3, e. 2, f. 2, g. 1, h. 3.

432. a. 3, b. 4, c. 7, d. 9, e. 10, f. 1, g. 5, h. 8, i. 2, j. 6.

433. a. 4, b. 1, c. 3, d. 2. Note that Flexner-Wintersteiner rosettes are more specific for retinoblastoma than Homer-Wright rosettes.

434. a. 3, b. 1, c. 1, d. 2, e. 5. Capillary hemangioma of the retina may be accompanied by intracranial hemangioblastomas and other systemic tumors in von Hippel-Lindau disease. Glial tumors of the retina and optic disc (such as astrocytic hamartoma of the optic disc and retina) are seen with tuberous sclerosis but also may be associated with neurofibromatosis. Diffuse choroidal hemangioma can be associated with Sturge-Weber syndrome.

435. a. 1, b. 1, c. 2, d. 1, e. 3, f. 1, g. 2, h. 1.

436. a. 3, b. 1, c. 2, d. 4. The spiral of Tillaux describes the ever-increasing distance from limbus to muscle insertion with rotation from the medial rectus inferiorly around to the superior rectus muscle (clockwise in the right eye, counterclockwise in the left eye).

### Suggested Readings


Retina and Vitreous

Questions

1. Effects of acute or chronic systemic hypertension on the retinal vascular system include all of the following except:
   a. focal or generalized vasoconstriction.
   b. breakdown of the blood–retinal barrier with subsequent hemorrhage and exudate.
   c. thickening of venous walls with secondary nicking of arterioles.
   d. histopathologic evidence of endothelial hyperplasia.
   e. development of micro- and macroaneurysms.

2. Which of the following statements about blood pressure–induced choroidal disease is false?
   a. Crucial pathophysiologic events lead to occlusion of the choriocapillaris.
   b. Elschning’s spots are characteristic.
   c. Exudative retinal detachment (RD) may develop as a secondary manifestation.
   d. Hypertensive choroidopathy may be associated with chronic elevation in systemic blood pressure.
   e. Hypertensive choroidopathy may be associated with acute elevations in intraocular pressure (IOP).

3. What is the approximate prevalence of endophthalmitis after an intravitreal triamcinolone injection?
   a. 0%.
   b. 0.01% to 0.05%.
   c. 0.1% to 0.3%.
   d. 0.5% to 1.5%.
   e. 2% to 3%.

4. Abnormalities in retinal physiology that are felt to be important in the early stages of diabetic retinopathy include:
   1. impaired photopigment recycling and metabolism.
   2. impaired autoregulation of retinal blood flow.
   3. impaired retrograde axoplasmic transport.
   4. breakdown in the blood–retinal barrier.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

5. T or F Evidence of breakdown in the blood–retinal barrier in patients with diabetes may be detected before the onset of clinical retinopathy.

6. T or F The abnormalities in the blood–retinal barrier in patients with diabetes are believed to be caused by loss of tight junctions between retinal pigment epithelium (RPE) cells.

7. Histopathologic features seen in the retinal vasculature of patients with early diabetic retinopathy include:
   1. loss of arteriolar pericytes.
   2. thickening of endothelial basement membranes.
3. capillary closure and/or nonperfusion.
4. medial hyperplasia.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

8. **T or F** In diabetes, the visual prognosis for diffuse macular edema is better than that for focal macular edema.

9. According to the Early Treatment Diabetic Retinopathy Study (ETDRS), which of the following is defined as clinically significant macular edema?
   1. retinal thickening within the temporal arcades.
   2. hard exudate within 500 microns of the center of the macula with adjacent thickening of the retina.
   3. extensive foveal and parafoveal nonperfusion on fluorescein angiography (FA).
   4. an area of retinal thickening ≥1 disc area, any part of which lies within 1 disc diameter of the center of the macula.

   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

10. Which of the following is a poor prognostic sign in a patient with nonproliferative diabetic retinopathy?
   a. numerous cotton-wool spots.
   b. numerous microaneurysms.
   c. extensive intraretinal microvascular abnormalities.
   d. extensive exudate.
   e. neovascularization (NV) of the disc.

11. In untreated eyes with preproliferative disease, the probability of progression to proliferative retinopathy over 2 years is approximately:
   a. 5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

12. Which factor is most strongly correlated with the development of choroidal effusion following panretinal photocoagulation?
   a. systemic hypertension.
   b. increasing age.
   c. male gender.
   d. total retinal surface area treated.
   e. short axial length (<23 microns).

13. **T or F** Rhegmatogenous retinal detachment (RRD) (of any type) is uncommon in proliferative diabetic retinopathy.

14. Each of the following statements are valid conclusions of the Diabetes Control and Complications Trial (DCCT) except:
   a. Among patients with type 1 diabetes with no retinopathy, intensive treatment can lower the incidence of progressive retinopathy by a factor of five compared to conventional treatment.
   b. Patients with macular edema realize the benefits of intensive control sooner than patients with proliferative retinopathy.
   c. The early worsening seen in patients initiating intensive control has no long-term effect on the severity of retinopathy.
   d. The study only tested patients with type 1 diabetes and the conclusions reached may not necessarily apply to patients with type 2 diabetes.
   e. The benefit of intensive control is not seen for the first 3 to 5 years of treatment.

15. Which of the following are the favorable clinical prognostic features for visual stabilization following laser treatment of diabetic macular edema?
   1. macular nonperfusion.
   2. cystoid macular edema (CME).
   3. extensive hard exudate within the fovea.
   4. focal leakage and thickening.

   a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
16. Which one of the following characteristics is felt to confer the greatest protection from the development of proliferative diabetic retinopathy?
   a. complete posterior vitreous separation (PVS).
   b. partial PVS.
   c. younger age (<30 years).
   d. ipsilateral carotid artery stenosis.
   e. no history of hypertension.

17. T or F The Early Treatment for Diabetic Retinopathy Study (EDTRS) showed that laser treatment of clinically significant diabetic macular edema leads to an improvement in vision in twice as many treated patients as untreated patients.

18. T or F The Diabetic Retinopathy Study (DRS) showed that panretinal photocoagulation could reduce the incidence of severe visual loss in certain patients by >50%.

19. High-risk characteristics of proliferative retinopathy mandating immediate prethreshold retinopathy of prematurity (ROP) include:
   1. neovascularization (NV) of the disc covering more than half of its area, only if associated with vitreous hemorrhage.
   2. any neovascularization elsewhere (NVE).
   3. severe NVE.
   4. moderate to severe NVE only if associated with vitreous hemorrhage.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

20. Adverse effects of prethreshold retinopathy of prematurity (ROP) may include which of the following?
   1. decreased night vision.
   2. angle-closure glaucoma.
   3. retinal detachment (RD).
   4. decreased reading acuity.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

21. Which one of the following statements about hemoglobinopathy and retinopathy is false?
   a. The incidence of proliferative retinopathy is higher in patients with sickle cell thalassemia (Hb SThal) than in patients with sickle cell SS disease (Hb SS) Br.
   b. The incidence of sickle cell trait (Hb AS) in the African American population is approximately 8%.
   c. The incidence of sickle cell SC disease (Hb SC) in the African American population is <0.5%.
   d. Fortunately, retinopathy has never been reported in patients with Hb AS.
   e. The ocular findings of Hb SC are not limited to the retina.

22. T or F Like diabetic retinopathy, the earliest pathophysiologic changes in proliferative sickle cell (SC) retinopathy include capillary closure and drop out.

23. T or F Like diabetic retinopathy, sickle cell (SC) retinopathy may have both nonproliferative and proliferative forms.

24. Which of the following is/are common manifestations of nonproliferative sickle cell (SC) retinopathy?
   1. salmon patches.
   2. iridescent deposits.
   3. black sunbursts.
   4. preretinal hemorrhage.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

25. Which of the following conditions may be causes of visual loss in sickle cell (SC) disease?
   1. vitreous hemorrhage.
   2. parafocal capillary nonperfusion.
   3. retinal detachment (RD).
   4. choroidal neovascularization (CNV).

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.
26. T or F Angioid streaks have been associated with both sickle cell (SC) disease and SC trait.

27. T or F Patients with sickle cell (SC) hemoglobinopathy are more prone to develop retinal arterial occlusions, both central and branch.

28. T or F The retinopathy of sickle cell SC disease is generally worse than that of sickle cell SS disease or sickle cell thalassemia (Hb SThal) disease.

29. Modalities useful in the treatment of proliferative sickle cell (SC) retinopathy include:
   1. pars plana vitrectomy with endolaser.
   2. scatter photocoagulation in the region of a neovascular frond.
   3. feeder-vessel photocoagulation.
   4. macular grid photocoagulation.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

30. Which of the following statements about ranibizumab is/are true?
   1. It is a humanized mRNA aptamer.
   2. It is derived from a monoclonal antibody to vascular endothelial growth factor (VEGF).
   3. It is administered intravenously.
   4. Phase III studies have shown significant improvement of visual acuity in patients with wet age-related macular degeneration (AMD).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

31. T or F The temporal retina is vascularized at 36 weeks’ gestation, approximately 4 weeks before the nasal retina.

32. Spontaneous regression of retinopathy of prematurity (ROP) occurs in approximately what percentage of the eyes of infants?
   a. 5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. >75%.

33. Which of the following descriptions of the severity of retinopathy of prematurity (ROP) is correct?
   a. stage 1: presence of a demarcation line having width and height (protruding into the vitreous humor).
   b. stage 2: presence of an elevated demarcation line with preretinal neovascularization (NV).
   c. stage 3: presence of a demarcation line having width and height (protruding into the vitreous).
   d. stage 4: total retinal detachment (RD).
   e. stage 5: total RD.

34. T or F The development of retinopathy of prematurity (ROP) requires the exposure of an immature retinal vascular system to some supplemental oxygen.

35. Which of the following criteria are part of the definition of prethreshold retinopathy of prematurity (ROP) according to the Early Treatment for Retinopathy of Prematurity (ETROP) study?
   1. zone I: any stage retinopathy of prematurity (ROP) less than threshold.
   2. zone II: stage 2 ROP with plus disease.
   3. zone II: stage 3 ROP without plus disease.
   4. zone II: <5 contiguous clock hours stage 3 ROP with plus disease.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

36. Which of the following statements about laser treatment of retinopathy of prematurity (ROP) is false?
   a. The technology for laser treatment is portable, making it possible for use in the neonatal intensive care unit (NICU).
   b. Laser treatment has not yet been shown to be as effective as cryotherapy, which was revealed in a randomized study.
   c. Laser treatment causes significantly less soft-tissue inflammation than cryotherapy.
d. Significant vitreous hemorrhage may require the use of cryotherapy in addition to or instead of laser treatment.

e. Like cryotherapy, laser photocoagulation is directed at the peripheral, avascular retina.

37. Which of the following statements about branch retinal vein occlusion (BRVO) is/are true?

1. The superotemporal quadrant is the most commonly affected.
2. The site of focal occlusion is usually an arterial-venous crossing.
3. A history of glaucoma is a risk factor for BRVO development.
4. A history of systemic hypertension is a risk factor for BRVO development.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

38. Common complications of branch retinal vein occlusion (BRVO) include:

1. macular edema.
2. macular nonperfusion.
3. neovascularization elsewhere (NVE).
4. rubeotic glaucoma.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

39. T or F The Branch Vein Occlusion Study (BVOS) documented the recovery of a final visual acuity of 20/40 or better in nearly twice as many patients treated with argon macular grid laser (compared with those who were untreated).

40. Other conclusions of the Branch Vein Occlusion Study (BVOS) include which of the following?

1. Quadrantic scatter photocoagulation reduces the risk of vitreous hemorrhage in the eyes with established neovascularization (NV).
2. Quadrantic scatter photocoagulation reduces the risk of developing NV if the area of retina affected by the vein occlusion is at least 5 disc areas in size.
3. Large areas of nonperfusion were a significant risk factor for the development of NV.
4. Quadrantic scatter photocoagulation significantly reduces the risk of severe visual loss in patients with at least 5 disc areas of nonperfusion.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

41. T or F Nonischemic and ischemic central retinal vein occlusions (CRVOs) should probably be considered as separate pathophysiologic disorders.

42. Which of the following conditions would define a central retinal vein occlusion (CRVO) as nonischemic?

a. mild vessel dilatation.
b. mild disc edema.
c. lack of significant parafoveal leakage on fluorescein angiography (FA).
d. lack of significant capillary nonperfusion on FA.
e. visual acuity better than 20/60.

43. T or F Nearly half the cases of nonischemic central retinal vein occlusion (CRVO) resolve entirely without treatment.

44. Which of the following systemic disorders may be associated with central retinal vein occlusion (CRVO)?

1. diabetes.
2. multiple myeloma.
3. hypertension.
4. syphilis.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

45. T or F Central retinal vein occlusion (CRVO) may be induced by acute angle-closure glaucoma.

46. T or F Central retinal vein occlusion (CRVO) may induce acute angle-closure glaucoma.
47. Important markers of ischemic central retinal vein occlusion (CRVO), compared to nonischemic CRVO, include all of the following except:
   a. numerous cotton-wool spots.
   b. extreme venous dilation.
   c. retinal hemorrhages in all four quadrants.
   d. extensive nonperfusion on fluorescein angiography (FA).
   e. b-wave/a-wave ratio of <1.0 on dark-adapted electroretinography (ERG).

48. Venous stasis retinopathy (carotid occlusive retinopathy) and central retinal vein occlusion (CRVO) routinely share which of the following features?
   1. venous dilation.
   2. cotton-wool spots.
   3. midperipheral blot retinal hemorrhages.
   4. diminished central retinal arterial pressure.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

49. T or F Regardless of etiology, the final clinical manifestation of precapillary arteriolar occlusion is the cherry-red spot.

50. Branch retinal arterial occlusion (BRAO) may be associated with which of the following conditions?
   1. mitral valve prolapse.
   2. cardiac myxoma.
   3. systemic lupus erythematosus (SLE).
   4. intravenous drug abuse.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

51. T or F Compared with ischemic central retinal vein occlusion (CRVO), central retinal arterial occlusion (CRAO) has a better visual prognosis (i.e., a larger percentage of patients with final visual acuity of 20/400 or better).

52. Irreversible structural damage occurs in the retina after what duration of total ischemia?
   a. 15 to 20 minutes.
   b. 30 to 40 minutes.
   c. 45 to 60 minutes.
   d. 90 to 100 minutes.
   e. 120 minutes.

53. Which of the following statements about idiopathic juxtafoveal telangiectasis is/are true?
   1. Of the subtypes, only one seems to fall within the spectrum of Coats’ disease.
   2. The histopathology of the adult forms of the disorder resembles diabetic retinopathy.
   3. Visual loss is generally caused by macular edema and/or exudate.
   4. The risk of retinal neovascularization (NV), as with diabetic retinopathy, may be lessened with scatter photocoagulation.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

54. Which of the following statements about idiopathic primary retinal vasculitis (Eales’ disease) is/are true?
   1. It is primarily a disease of childhood and young adulthood, more commonly affecting girls.
   2. It may be associated with tuberculous infection.
   3. It is generally unilateral.
   4. It may be associated with epistaxis and a cerebral vasculitis.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

55. Retinal vasculitis may be associated with which of the following conditions?
   1. systemic lupus erythematosus (SLE).
   2. multiple sclerosis (MS).
3. polyarteritis nodosa.
4. intravenous drug abuse.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

56. Which of the following statements about arterial macroaneurysms of the retina is/are true?

1. They are generally associated with hypertension and are more frequent in older women.
2. Visual loss is always caused by associated macular edema.
3. Laser treatment may be applied in the region of the macroaneurysm to reduce macular edema.
4. Direct treatment of the macroaneurysm is most effective for reducing associated macular edema.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

57. Which one of the following statements about aphakic or pseudophakic cystoid macular edema (CME) is false?

a. The incidence is lower with extracapsular surgery compared with intracapsular surgery.
b. Intraocular lens implantation decreases the incidence of CME following cataract surgery.
c. The presence of an intact posterior capsule is associated with a lower incidence of CME.
d. More than 75% of mild cases partially regress spontaneously within 6 months.
e. Topical nonsteroidal antiinflammatory medications have been shown to reduce the incidence as well as improve vision in cases of CME following cataract surgery.

58. Which one of the following statements about Coats’ disease (retinal telangiectasis) is false?

a. The male-to-female ratio is approximately 7:1.
b. It is primarily a disease of childhood, although adults may be affected.
c. Inheritance is autosomal dominant with incomplete penetrance.
d. In patients who have an onset of the disease before the age of 4 years, the disease follows a more fulminant course.
e. Treatment alternatives include photocoagulation and cryotherapy.

59. T or F Pathologically, a von Hippel's lesion is a cavernous hemangioma.

60. T or F Rhegmatogenous retinal detachment (RD) is uncommon in retinitis pigmentosa.

61. Findings in a patient with von Hippel-Lindau disease may include all of the following except:

a. café-au-lait spots.
b. pancreatic and renal cysts.
c. hemangioblastomas of the brainstem.
d. renal cell carcinoma.
e. pheochromocytoma.

62. T or F An ophthalmologist treating a retinal hemangioma may expect temporary worsening of exudation following successful treatment.

63. T or F Both congenital arteriovenous malformations of the retina (racemose angioma) and cavernous hemangiomas of the retina are distinguished from the vascular malformations of von Hippel's disease by the lack of exudation and subretinal fluid.

64. T or F The most common complication of retinal cavernous hemangioma is vitreous hemorrhage.

65. Which of the following constitutes the histologic definition of the macula?

a. the area of the retina with increased xanthophyll pigment concentration.
b. the area of the retina whose ganglion cell layer is more than one cell layer thick.
c. the area of the retina whose innermost layer is the outer plexiform layer.
d. the area of the retina with the tallest retinal pigment epithelium (RPE) cells.
e. the area of the retina within the temporal arcades.
66. Which is the outermost layer supplied by the central retinal circulation?
   a. ganglion cell layer.
   b. inner plexiform layer.
   c. inner nuclear layer.
   d. outer plexiform layer.
   e. outer nuclear layer.

67. Which is the correct order of the five layers of Bruch’s membrane (from the retina toward the sclera)?
   a. retinal pigment epithelium (RPE) basement membrane, inner collagenous zone, elastic layer, outer collagenous zone, and choriocapillaris basement membrane.
   b. RPE basement membrane, elastic layer, inner collagenous zone, outer collagenous zone, and choriocapillaris basement membrane.
   c. elastic layer, RPE basement membrane, inner collagenous zone, outer collagenous zone, and choriocapillaris basement membrane.
   d. inner collagenous zone, elastic layer, outer collagenous zone, RPE basement membrane, and choriocapillaris basement membrane.
   e. RPE basement membrane, inner collagenous zone, elastic layer, choriocapillaris basement membrane, and outer collagenous zone.

68. T or F Changes in the retinal pigment epithelium (RPE) and Bruch’s membrane, seen with aging, may be caused by an accumulation of metabolic breakdown products from the photoreceptor outer segments.

69. T or F The retinal pigment epithelium (RPE) acts as a depot for dietary and excess vitamin A (retinol).

70. T or F Hypertrophy of the retinal pigment epithelium (RPE) generally leads to a flat, jet-black subretinal lesion, whereas hyperplasia most frequently leads to intraretinal bone spicule pigment deposition.

71. Which one of the following statements about fluorescein angiography (FA) and the blood–ocular barriers is true?
   a. Fluorescein is a high-molecular weight compound normally confined to the intravascular space.
   b. Fluorescein absorbs light in the yellow–green range—530 nm.
   c. After stimulation, fluorescein emits light in the blue range—490 nm.
   d. The “red-free” filter is the initial filter through which white light passes before entering the eye.
   e. The most dreaded complication of FA is mortality from anaphylactic shock.

72. Which of the following statements about hyperfluorescence patterns on fluorescein angiography (FA) is/are true?
   1. Staining generally refers to the uptake of fluorescein by solid collagenous tissue.
   2. Transmitted fluorescence, or a window defect, generally implies a focal defect in the retinal pigment epithelium (RPE).
   3. Pooling implies collections of fluorescein within fluid-filled spaces.
   4. True leakage consists of early hyperfluorescence that diminishes in late views.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

73. Which of the following features are considered necessary for the development of retinal neovascularization (NV)?
   1. production of a local angiogenic factor.
   2. some source of endothelial cells capable of replication and migration.
   3. a structural scaffold for vascular growth, presumably posterior cortical vitreous.
   4. a focal defect in the internal limiting membrane.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

74. Which of the following may lead to visual loss in choroidal neovascularization (CNV)?
   1. focal subretinal exudate.
   2. subretinal hemorrhage.
3. sensory retinal detachment (RD).
4. photoreceptor atrophy.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

75. Which of the following statements about central serous retinopathy (CSR) is/are true?

1. The classic fundus finding is a small serous detachment of the macula, although serous detachment of the pigment epithelium also may be seen.
2. Fluorescein angiography (FA) performed at presentation will show a “smokestack” pattern of leakage 90% of the time.
3. Affected women tend to present at later ages than affected men.
4. Laser photocoagulation of fluorescein leakage sites leads to improved final Snellen visual acuity.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

76. T or F Eighty percent to 90% of the cases of central serous retinopathy (CSR) will spontaneously resolve, but 40% to 50% will recur.

77. Patients who should be considered for laser treatment of central serous retinopathy (CSR) include:

1. a patient whose sensory retinal detachment (RD) has persisted for >6 weeks.
2. a monocular patient with occupational visual needs.
3. a patient with pigment epithelial detachment and surrounding sensory (RD).
4. a patient whose prior episodes of CSR have been associated with permanently decreased acuity.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.

78. As of 2005, what types of functional visual prostheses have been successfully implanted in humans?

a. optic nerve stimulators.
b. epiretinal implants.
c. passive subretinal implants.
d. occipital cortex implants.
e. all of the above.

79. T or F The feature necessary and sufficient for the diagnosis of age-related macular degeneration (AMD) is the presence of at least one type of drusen.

80. Risk factors for progressive visual loss in age-related macular degeneration (AMD) include all of the following except:

a. hyperopia.
b. smoking.
c. light iris color.
d. hypertension.
e. family history of visual loss caused by AMD.

81. Which of the following statements about the classification of age-related macular degeneration (AMD) is/are true?

1. “Dry” AMD accounts for 90% of all patients affected by this disorder.
2. “Wet” AMD accounts for 90% of all patients with severe visual loss (worse than 20/200) who are affected by this disorder.
3. Patients with either pigment epithelial detachment or choroidal neovascularization (CNV) should be considered as part of those affected by the wet variety of AMD.
4. In patients affected with dry AMD, those with central geographic atrophy generally preserve the best visual acuity.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 1 and 3.
e. 1, 2, 3, and 4.

82. T or F Pigment epithelial detachment in a patient younger than 50 years generally has a better prognosis than in a patient older than 50 years.
83. Argon laser photocoagulation for exudative age-related macular degeneration (AMD) has been shown to be effective in reducing the rate of severe visual loss or preserving visual function among which categories of the disease?
   1. extrafoveal (>200 microns from the center of the foveal avascular zone) choroidal neovascularization (CNV).
   2. juxtafoveal CNV (1 to 199 microns from the center of the foveal avascular zone).
   3. subfoveal CNV.
   4. juxtafoveal pigment epithelial detachment.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

84. Which of the following factor(s) support(s) an etiologic connection between ocular Histoplasma infection and the presumed ocular histoplasmosis syndrome (POHS)?
   1. More than 90% of patients with POHS will have a positive histoplasmin skin reaction.
   2. The highest prevalence of POHS is among the populations of the southwest states such as New Mexico, Arizona, and California.
   3. Histoplasma organisms have been recovered from the human choroid.
   4. Systemic treatment with antifungal agents leads to resolution of the ocular findings.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

85. Accepted methods of treatment of choroidal neovascularization (CNV) secondary to presumed ocular histoplasmosis syndrome (POHS) include:
   1. systemic antifungal medications.
   2. photodynamic therapy.
   3. cryotherapy.
   4. argon laser photocoagulation.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.

86. The best predictor of future contralateral visual loss in a patient with a disciform macular scar from presumed ocular histoplasmosis syndrome (POHS) is the presence or absence of:
   a. a focal macular scar in the better eye.
   b. peripapillary scarring in the better eye.
   c. active vitritis in the better eye.
   d. symmetric peripheral punched-out lesions of each eye.
   e. anti-Histoplasma antibodies.

87. Which of the following statements about angioid streaks is/are true?
   1. They always extend in continuity from the optic nerve head.
   2. They appear as window defects on fluorescein angiography (FA).
   3. The typical pattern forms concentric circles around the optic nerve head.
   4. Histopathologically, they represent discontinuities in a thickened, abnormal Bruch’s membrane.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

88. Systemic disorders associated with angioid streaks include:
   2. pseudoxanthoma elasticum.
   3. Ehlers-Danlos syndrome.
   4. high myopia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

89. Idiopathic epiretinal membranes are bilateral in what proportion of affected patients?
   a. 5%.
   b. 20%.
   c. 50%.
Questions

d. 75%.
e. 100%.

90. T or F Most patients with idiopathic epiretinal membranes maintain vision better than 20/50.

91. A randomized clinical trial (RCT) has documented that vitreous surgery with gas–fluid exchange and prone positioning offers no long-term benefit relative to observation for which of the following conditions?
   a. idiopathic macular hole, stage 1.
   b. idiopathic macular hole, stages 1 and 2.
   c. idiopathic macular hole, stages 1, 2, and 3.
   d. idiopathic macular hole, regardless of stage.
   e. As of 2004, no RCT of vitreous surgery for idiopathic macular hole has been completed.

92. The prevalence of contralateral full-thickness macular hole in a patient with an established full-thickness macular hole is approximately:
   a. 5%.
   b. 25%.
   c. 50%.
   d. 75%.
   e. 100%.

93. The lesion that is felt to be the immediate precursor to a full-thickness macular hole is:
   a. a partial posterior vitreous detachment.
   b. a macular cyst.
   c. a sensory retinal detachment (RD) involving the fovea.
   d. a pigment epithelial detachment under the fovea.
   e. the Watzke sign.

94. Which of the following statements about ocular toxicity of chloroquine and hydroxychloroquine is/are true?
   1. Only chloroquine has been clearly associated with retinal toxicity.
   2. Chloroquine may be associated with a vortex keratopathy.
   3. The most useful and most sensitive parameter for detecting chloroquine retinopathy is the electrooculogram (EOG).
   4. Important tests in the evaluation for subclinical chloroquine retinopathy include color vision testing and threshold central visual field testing.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

95. T or F Cessation of chloroquine treatment in a patient with established chloroquine retinopathy should lead to rapid reversal of the retinal findings.

96. T or F Both antimalarial agents (e.g., chloroquine) and antipsychotics (e.g., thioridazine) may cause peripheral pigmentary retinopathy with electoretinographic abnormalities.

97. T or F Both thioridazine and chlorpromazine may lead to abnormal pigment deposition in eyelids, cornea, lens, and retina.

98. T or F All of the mucopolysaccharidoses (MPSs) are autosomal-dominant disorders except type II, which is X-linked recessive.

99. Corneal clouding is a common manifestation of which of the following mucopolysaccharidoses (MPS)?
   1. Type I-H (Hurler’s syndrome).
   2. Type II (Hunter’s syndrome).
   3. Type I-S (Scheie’s syndrome).
   4. Type III (Sanfilippo’s syndrome).
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

100. A pigmentary retinopathy indistinguishable from typical retinitis pigmentosa (ERP) may be seen in which of the mucopolysaccharidoses (MPS)?
   1. Type I-H (Hurler’s syndrome).
   2. Type II (Hunter’s syndrome).
3. Type III (Sanfilippo’s syndrome).
4. Type IV (Morquio’s syndrome).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

101. Optic atrophy may be seen in which of the following mucopolysaccharidoses (MPS)?

1. Type II (Hunter’s syndrome).
2. Type III (Sanfilippo’s syndrome).
3. Type IV (Morquio’s syndrome).
4. Type VII (Sly’s syndrome).

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

102. T or F Gangliosides are sphingolipids found only in the central nervous system (CNS), whereas cerebrosides are sphingolipids found throughout the body.

103. Which of the following disorders may be associated with a cherry-red spot?

1. Tay-Sachs disease.
2. Sandhoff’s disease.
3. Niemann-Pick disease.
4. Fabry’s disease.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

104. T or F In Fabry’s disease, the leading cause of death is myocardial infarction (MI).

105. Ocular manifestations of Fabry’s disease include:

1. cornea verticillata.
2. spoke-like posterior subcapsular cataract.
3. conjunctival and retinal telangiectases.
4. pigmentary retinopathy.
109. Which of the following statements about the distribution of photoreceptors in the normal human retina is true?

1. The ratio of rods to cones is approximately 4:1.
2. The numbers of rods and cones in the macula (central 18 degrees) is equal.
3. Cone density is maximal in a ring 20 to 40 degrees eccentric to the foveola.
4. Forty percent of cones lie outside the macula.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

110. T o F A giant retinal tear is a circumferential retinal break of 90 degrees or greater (3 clock hours or more).

111. What diagnosis should be considered in a 30-year-old man with a history of bilateral giant retinal tears, cleft palate, severe myopia, severe arthritis requiring a total hip replacement, and a family history of severe arthritis and blindness?

a. Ehlers-Danlos syndrome.
b. Marfan’s syndrome.
c. Stickler’s syndrome.
d. Wagner’s syndrome.
e. Weill-Marchesani syndrome.

112. T o F In a dark-adapted patient, a blue flash of light will generate an electroretinogram (ERG) with rod input only.

113. T o F In a light-adapted patient, a blue flash of light will generate an electroretinogram (ERG) with cone input only.

114. In order to truly isolate cone function, it is necessary to present a light stimulus as flicker-flash at what minimum frequency?

a. 5 Hz.
b. 10 Hz.
c. 15 Hz.
d. 25 Hz.
e. 40 Hz.

115. The key feature on electroretinography (ERG) distinguishing focal or nonprogressive retinal disease from a diffuse progressive degeneration is an abnormality in the:

a. early receptor potential.
b. a-wave amplitude.
c. b-wave implicit time.
d. c-wave amplitude.
e. d-wave amplitude.

116. T o F Increasing the intensity of the stimulus flash in a scotopic electroretinogram (ERG) will result in a decrease in both implicit time and amplitude of the b-wave.

117. Which of the following disorders may be considered forms of congenital stationary night blindness?

1. Oguchi’s disease.
2. choroideremia.
3. fundus albipunctatus.
4. fundus flavimaculatus.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

118. T o F Eyes with retinal dialyses usually have posterior vitreous detachments.

119. Which of the following statements about ocular antiangiogenesis is false?

a. Ocular antiangiogenesis therapy may be beneficial in the treatment of age-related macular degeneration (AMD) and diabetic retinopathy.
b. Pegaptanib sodium (Macugen) is a monoclonal antibody that has been approved by the U.S. Food and Drug Administration (FDA) for treatment of certain forms of wet AMD.
c. Ranibizumab (Lucentis) is a monoclonal antibody fragment that binds to vascular endothelial growth factor (VEGF), preventing VEGF from binding to VEGF receptors.
d. Both Ranibizumab and Pegaptanib sodium are administered by intravitreal injection.
e. VEGF is a potent cell mitogen that induces angiogenesis and increases vascular permeability, and it is elevated in various retinal disorders involving hypoxic conditions and ischemia.

120. Which of the following statements about electrooculography (EOG) is/are true?

1. The corneal surface or vitreal space is positive relative to sclera.
2. Amplitudes generally diminish with light adaptation and increase with dark adaptation.
3. Amplitudes are typically measured by alternating lateral gaze from left to right.
4. An EOG is generally considered abnormal if the dark-peak to light-trough ratio is <1.75.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

121. Electrooculography (EOG) and electroretinography (ERG) are similarly depressed for all of the following except:

a. choroideremia.
b. gyrate atrophy.
c. Oguchi’s disease.
d. Best’s disease.
e. X-linked–recessive retinitis pigmentosa.

122. The electrooculogram (EOG) may be valuable in evaluating patients with potential retinal toxicity from:

a. ultraviolet light.
b. chloroquine.
c. phenothiazine.
d. isoniazid.
e. amiodarone.

123. After dark adaptation, how much more sensitive are rods than cones?

a. 10 times (1 log unit).
b. 100 times (2 log units).
c. 1,000 times (3 log units).
d. 10,000 times (4 log units).
e. 100,000 times (5 log units).

124. T or F Anaphylaxis has been reported as a potential complication of intravitreal injection.

125. T or F A ganglion cell whose receptive field consists of an off-center on-surround will increase its firing rate if stimulation to its central photoreceptors is diminished.

126. T or F The Purkinje shift refers to the enhancement in light sensitivity during dark adaptation (the shift from cone threshold sensitivity to rod threshold sensitivity).

127. T or F A red cone attached directly to a ganglion cell could induce a high rate of ganglion cell firing if appropriately stimulated with monochromatic green light.

128. Which of the following statements is true regarding Refsum’s disease?

a. Both infantile and adult forms exist.
b. Night blindness can be an early symptom in patients.
c. Phytanic acid levels are typically elevated.
d. Dietary restriction may slow down the disease.
e. All of the above are true.

129. The most common pattern of congenital dyschromatopsia is:

a. deuteranomaly.
b. protanomaly.
c. protanopia.
d. deuteranopia.
e. tritanopia.

130. Which forms of congenital dyschromatopsia are accompanied by abnormally low visual acuity?

1. protanopia.
2. blue cone monochromatism.
3. red cone monochromatism.
4. rod monochromatism.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
Questions

131. Which of the following help to distinguish between rod monochromats and blue cone monochromats?
   a. specific vision color testing.
   b. the presence or absence of nystagmus.
   c. careful family history.
   d. electroretinographic (ERG) waveforms.
   e. 1, 2, and 3.

132. Which of the following color vision tests detect both red–green and blue–yellow defects?
   a. Hardy-Rand-Rittler plates.
   b. Farnsworth-Munsell 100 test.
   c. Farnsworth panel D-15 test.
   d. Ishihara plates.
   e. 1, 2, 3, and 4.

133. What proportion of patients with intracranial hemorrhage have Terson’s syndrome?
   a. 5%.
   b. 10%.
   c. 33%.
   d. 50%.
   e. 90%.

134. Which of the following conditions is generally not associated with Purtscher’s or Purtscherlike retinopathy?
   a. systemic lupus erythematosus (SLE).
   b. thrombotic thrombocytopenic purpura (TTP).
   c. long-bone fractures.
   d. chronic renal failure.
   e. hepatic encephalopathy.

135. T or F The vast majority of severe hearing loss associated with retinitis pigmentosa (RP) is congenital.

136. Which one of the following statements about retinitis pigmentosa (RP) is false?
   a. In Leber’s congenital amaurosis, the infant is typically blind at birth.
   b. In Leber’s congenital amaurosis, the electroretinogram (ERG) is typically nonrecordable at birth.
   c. In Leber’s congenital amaurosis, the fundus examination is typically normal at birth.
   d. A variety of congenital RP is associated with macular coloboma and skeletal abnormalities.
   e. The most common pattern of inheritance in congenital RP is autosomal dominant.

137. T or F Usher’s syndrome describes any combination of pigmentary retinopathy and partial or complete deafness.

138. T or F Many patients with Usher’s syndrome may have cerebellar and/or vestibular abnormalities.

139. Preservation of central acuity past the age of 45 in a patient with a retinal degeneration and an X-linked inheritance pattern suggests the diagnosis of:
   a. recessive cone–rod degeneration.
   b. gyrate atrophy.
   c. Refsum’s disease.
   d. choroideremia.
   e. Usher’s syndrome type I.

140. T or F Bone spicule pigmentation is unusual in choroideremia.

141. Which of the following statements about gyrate atrophy is/are true?
   a. It is inherited on an X-linked–recessive basis.
   b. There is a systemic deficiency in ornithine aminotransferase activity.
   c. Serum abnormalities include hyperornithinemia and hyperlysinemia.
   d. Life span is normal in this disorder.
   e. 1, 2, 3, and 4.
11: Retina and Vitreous

142. Which of the following statements about fundus flavimaculatus is/are true?

1. The pisciform lesions seen in the posterior fundus represent lipofuscin deposits within hypertrophied retinal pigment epithelium (RPE) cells.
2. The most common mode of inheritance is autosomal recessive.
3. Visual acuity loss may be mild or severe depending on the extent of macular involvement.
4. The degree of electroretinographic (ERG) abnormality parallels the amount of fundus involvement.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

143. Which of the following statements about familial drusen (Doyne’s honeycomb dystrophy) is/are true?

1. The initial fundus manifestations generally appear in the third decade of life.
2. Most cases are diagnosed in the third decade of life.
3. There are both atrophic (dry) and exudative (wet) forms of the disease.
4. The electroretinogram (ERG) is generally markedly depressed.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

144. Which of the following is considered the hallmark of fundus albipunctatus?

a. loss of dark adaptation with relative preservation of color vision and visual field.
b. loss of the scotopic electroretinogram (ERG) with preservation of cone responses.
c. normalization of scotopic ERG after 4 to 8 hours of dark adaptation.
d. punctate yellow–white deposits scattered throughout the equatorial and peripheral fundus.
e. the Mizuo phenomenon.

145. T or F In Best’s disease, the macular appearance is considerably worse than the visual acuity.

146. T or F In Stargardt’s disease, the macular appearance is considerably worse than the visual acuity.

147. The Mizuo phenomenon is a feature of which of the following disorders?

1. Oguchi’s disease.
2. fundus albipunctatus.
3. X-linked cone dystrophy.
4. fundus flavimaculatus.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

148. Symptoms of cone degeneration include which of the following?

1. loss of visual acuity.
2. photophobia.
3. progressive dyschromatopsia.
4. difficulty driving at night.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

149. T or F The risk of anaphylaxis caused by fluorescein angiography (FA) is significantly <1 in 10,000 patients.

150. The differential diagnosis of peripheral pigmentary retinopathy includes:

1. syphilitic chorioretinitis.
2. prior exudative retinal detachment (RD).
3. phenothiazine toxicity.
4. ophthalmic artery occlusion.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.
151. Fundus abnormalities associated with congenital stationary night blindness (CSNB) may be seen in:

1. autosomal-dominant CSNB.
2. fundus albipunctatus.
3. autosomal-recessive CSNB.
4. Oguchi’s disease.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

152. Foveal hypoplasia may be associated with which of the following disorders?

1. albinism.
2. congenital cytomegalovirus (CMV) disease.
3. aniridia.
4. Leber’s congenital amaurosis.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

153. Which one of the following statements about Stargardt’s disease is false?

a. It is generally inherited on an autosomal-dominant basis.
b. It may be associated with peripheral flecks indistinguishable from fundus flavimaculatus.
c. In its early stages, the maculopathy has a beaten metal appearance, similar to cone–rod dystrophy.
d. In fundus flavimaculatus, the fluorescein angiogram (FA) may reveal a “dark choroid.”
e. The terminal stages of the maculopathy resemble central areolar choroidal dystrophy.

154. T or F In typical vitelliform dystrophy, it is unusual for visual acuity to deteriorate beyond 20/200.

155. Which of the following statements about Best’s vitelliform dystrophy is/are true?

a. The general mode of inheritance is autosomal dominant.
b. Because of its profound effect on visual acuity, the “egg yolk” stage of the disease is frequently found at presentation.

b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

156. T or F The adult type of vitelliform macular dystrophy features “egg yolk” lesions that are generally smaller than those of classic Best’s disease.

157. T or F Adult vitelliform macular dystrophy (foveomacular dystrophy) may involve regions other than the fovea but never simultaneously.

158. Which of the following statements about pattern dystrophies of the retinal pigment epithelium (RPE) is/are true?

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

159. Albinism and albinoidism share which of the following features?

a. iris transillumination.
b. markedly decreased vision.
c. blond fundi.
d. nystagmus.

e. 1, 2, 3, and 4.
160. Abnormalities in the visual pathways of patients with true albinism include:
   1. a reduced number of ganglion cells.
   2. inappropriate decussation of temporal retinal nerve fibers.
   3. grossly hypoplastic lateral geniculate nuclei.
   4. foveal hypoplasia.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

161. T or F Albinism and albinoidism may be distinguished on the basis of inheritance.

162. The only form of true albinism that is not inherited on an autosomal-recessive basis is:
   a. Hermansky-Pudlak syndrome.
   b. tyrosinase-positive oculocutaneous albinism.
   c. Nettleship-Falls ocular albinism.
   d. Chédiak-Higashi syndrome.
   e. yellow-mutant variety of oculocutaneous albinism.

163. Which form(s) of true albinism is/are potentially lethal?
   1. tyrosinase-positive.
   2. Chédiak-Higashi syndrome.
   3. yellow-mutant variety.
   4. Hermansky-Pudlak syndrome.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

164. Disadvantages of xenon arc photocoagulation include:
   1. expensive, sophisticated machinery.
   2. unfocused white light emission.
   3. inability to generate large amounts of power.
   4. considerable amount of emitted blue light.
   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

165. Which laser is least absorbed by hemoglobin or red blood cells?
   a. xenon arc.
   b. argon blue-green.
   c. argon green.
   d. krypton red.
   e. dye yellow.

166. Which laser is best for retinal photocoagulation in the setting of dense cataract or vitreous hemorrhage?
   a. xenon arc.
   b. argon blue-green.
   c. argon green.
   d. krypton red.
   e. dye yellow.

167. T or F Decreasing the spot size of an argon laser burn increases the energy delivered per unit area.

168. Regarding the Subfoveal New Choroidal Neovascularization (CNV) Study, coordinated by the Macular Photocoagulation Study, each of the following is true except:
   a. With lesions <3.5 disc areas, eyes assigned to laser treatment lost more acuity over the short term (3 to 6 months) than untreated eyes.
   b. With lesions <3.5 disc areas, treated eyes lost less acuity over the long term (1 to 3 years).
   c. With lesions <3.5 disc areas, reading speed results paralleled those for visual acuity.
   d. With lesions <3.5 disc areas, contrast sensitivity results paralleled those for visual acuity.
   e. At the outset of the study, patients with lesions >3.5 disc areas or initial acuity better than 20/80 were excluded.

169. Which of the following is/are potential complications of fundus photocoagulation?
   1. choroidal neovascularization (CNV).
   2. traction retinal detachment (RD).
Questions

3. epiretinal membrane.
4. angle-closure glaucoma.

a. 1, 2, and 3.
b. 1 and 3.
c. 2 and 4.
d. 4 only.
e. 1, 2, 3, and 4.

176. The prevalence of lattice retinal degeneration in the adult population is approximately:
   a. 5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

177. The prevalence of lattice retinal degeneration in patients with rhegmatogenous retinal detachment (RRD) is approximately:
   a. 5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

178. T or F In lattice retinal degeneration, histopathology reveals abnormally strong vitreoretinal attachments over the affected retina.

179. Which of the following conditions is/are felt to increase the risk of rhegmatogenous retinal detachment (RRD)?
   1. lattice degeneration.
   2. ora pearls.
   3. meridional complexes.
   4. cobblestone degeneration.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

180. The incidence of cobblestone degeneration in the adult population is approximately:
   a. 5%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

181. In what proportion of patients with rhegmatogenous retinal detachment (RRD) can a retinal break not be found?
   a. 1%.
   b. 3%.
   c. 5%.

170. Points of firm vitreoretinal attachment include:
   1. the vitreous base.
   2. the edge of retinal scars.
   3. the edge of lattice retinal degeneration.
   4. the vortex veins.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

171. T or F The prevalence of posterior vitreous detachment is higher after intracapsular cataract surgery than after extracapsular cataract surgery.

172. What proportion of patients with an acute symptomatic posterior vitreous detachment have a retinal tear?
   a. 0.05%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

173. What proportion of patients with acute symptomatic posterior vitreous detachment and associated vitreous hemorrhage have a retinal tear?
   a. 1%.
   b. 10%.
   c. 25%.
   d. 50%.
   e. 75%.

174. T or F Most retinal dialyses occur in the inferotemporal quadrant.

175. T or F Most superonasal retinal dialyses will be associated with a clear history of head or eye trauma.
182. Findings consistent with rhegmatogenous retinal detachment (RRD) rather than exudative or tractional detachment include:
   1. shifting fluid.
   2. “tobacco dust.”
   3. smooth, domed appearance of the retina.
   4. undulation of the retina with eye movements.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

183. The most common cause of redetachment following initially successful surgical repair of rhegmatogenous retinal detachment (RRD) is:
   a. occult retinal breaks.
   b. new retinal breaks.
   c. inadequate retinopexy.
   d. proliferative vitreoretinopathy (PVR).
   e. failure to relieve vitreoretinal traction adequately.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

184. Prognostic factors favoring an excellent outcome after surgery for rhegmatogenous retinal detachment (RRD) include:
   1. aphakic detachment.
   2. retinal detachment (RD) associated with choroidal detachment.
   3. RDs with breaks >180 degrees.
   4. retinal dialyses.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

185. The key prognostic factor in predicting postoperative visual acuity following surgical repair of rhegmatogenous retinal detachment (RRD) is:
   a. the size of the largest retinal break.
   b. the number of retinal breaks.
   c. the presence and duration of macular detachment.

   d. the presence or absence of lattice degeneration.
   e. the presence or absence of myopia.

186. The finding most frequently associated with degenerative retinoschisis is:
   a. peripheral cystoid degeneration.
   b. bullous retinoschisis.
   c. reticular retinoschisis.
   d. rhegmatogenous retinal detachment (RRD).
   e. retinal dialysis.

187. Which of the following statements about retinoschisis is/are true?
   1. Typical degenerative retinoschisis is associated with an increased risk of retinal detachment (RD).
   2. Most patients with retinoschisis are hyperopic.
   3. The absolute scotoma caused by posterior extension of retinoschisis is highly symptomatic.
   4. Retinoschisis is bilateral in most patients.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

188. T or F Simple retinoschisis no retinal detachment (RD) is generally associated with a demarcation line.

189. T or F Retinal detachment (RD) associated with retinoschisis requires holes in both the inner and outer walls of the schisis cavity.

190. T or F Fluorescein angiography (FA) is contraindicated in individuals with an allergy to shellfish or iodine.

191. Regarding the surgical treatment of proliferative vitreoretinopathy (PVR), which one of the following is false?
   a. Silicone oil and SF₆ gas are equally effective tamponade agents.
   b. Silicone oil and perfluoropropane (C₃F₈) gas are equally effective tamponade agents.
Questions

c. When silicone oil is used as tamponade in PVR, there is no significant difference in outcomes between eyes undergoing primary (i.e., their first) vitrectomy and previously vitrectomized eyes.
d. When C3F8 is used as tamponade in PVR, there is no significant difference in outcomes between eyes undergoing primary vitrectomy and previously vitrectomized eyes.
e. Postoperative macular pucker is equally common following tamponade with gas or oil.

192. Features differentiating persistent fetal vasculature/persistent hyperplastic primary vitreous (PHPV) from retinoblastoma include:
   1. age of onset of leukocoria.
   2. presence of microphthalmos.
   3. calcification.
   4. presence of cataract.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

193. Features differentiating Wagner’s disease from Stickler’s syndrome include:
   1. cataract.
   2. midfacial flattening.
   3. high myopia.
   4. high incidence of retinal detachment (RD).

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

194. T or F The best way of differentiating familial exudative vitreoretinopathy from retinopathy of prematurity (ROP) is careful examination of family members.

195. Which one of the following statements about asteroid hyalosis is false?
   a. It is more common with aging.
   b. It is more commonly bilateral.
   c. It is generally associated with no decrease in visual acuity.
   d. The vitreous is otherwise normal.
   e. The particulate matter seen clinically consists of calcium soaps.

196. T or F The deposits of synchysis scintillans are more likely to settle inferiorly than those of asteroid hyalosis.

197. The most common cause of vitreous hemorrhage in the American adult population is:
   a. trauma.
   b. retinal tear.
   c. posterior vitreous detachment.
   d. diabetic retinopathy.
   e. retinal neovascularization (NV) secondary to retinal vein occlusion.

198. What condition(s) mandate(s) repair with pars plana vitrectomy rather than conventional scleral buckling +/− pneumatic retinopexy?
   1. retinal dialysis.
   2. retinal detachment (RD) with accompanying vitreous hemorrhage following penetrating trauma.
   3. aphakic RD.
   4. RD associated with marked proliferative vitreoretinopathy (PVR).

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

199. Advantages of early vitrectomy surgery in a patient who is diabetic and has vitreous hemorrhage include:
   1. an opportunity to remove the vitreous scaffold that fosters neovascularization (NV).
   2. a lower incidence of complete blindness following vitrectomy surgery in patients who are diabetic compared with those who are nondiabetic.
   3. an opportunity to treat retinal ischemia intensively with endolaser.
   4. an opportunity to remove the lens surgically and improve visualization of the retina for subsequent treatment.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
11: Retina and Vitreous

200. Indications for emergent pars plana vitrectomy surgery on an eye with an acute (i.e., unrepaired) rupture or laceration include:
   1. vitreous hemorrhage and associated retinal detachment (RD).
   2. intraocular foreign body.
   3. traumatic endophthalmitis.
   4. vitreous hemorrhage.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

201. Which of the following conditions is/are likely to lead to permanently decreased visual acuity following trauma?
   1. commotio retinae.
   2. subretinal hemorrhage under the fovea.
   3. Valsalva retinopathy involving the fovea.
   4. traumatic macular hole.

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

202. Purtscher's retinopathy has characteristic fundus findings that may be seen in association with:
   1. vigorous chest compression.
   2. acute pancreatitis.
   3. long-bone fracture.
   4. systemic lupus erythematosus (SLE).

   a. 1, 2, and 3.
   b. 1 and 3.
   c. 2 and 4.
   d. 4 only.
   e. 1, 2, 3, and 4.

203. A 58-year-old man presents for a routine examination and refraction. Best acuities are 20/400 in the right eye and 20/30 in the left eye. The results of his examination are normal except for the findings shown in figures a., b., c., and d., below. Each of the following statements about this case is false except:
   a. Each of the patient's children has a 50% chance of being similarly affected.
   b. This disorder is strikingly unusual in women.
   c. Histopathology of the left retina would show dilated, irregular retinal capillaries in the temporal macula.
   d. The patient should undergo some form of glucose tolerance testing.
   e. This condition is a forme fruste of Coats' disease.
Questions

204. A 17-year-old boy presents with complaints of difficulty reading his schoolwork. He is completely healthy. Visual acuities measure 20/80 in the right eye and 20/100 in the left eye. His fundi are shown in figures a., b., c., and d., below. Which one of the following statements about his case is false?
   a. One of his parents almost certainly has similar findings.
   b. His electroretinogram (ERG) may be normal.
   c. Examination of his peripheral retina may show white flecks.
   d. His visual fields may be normal.
   e. He can be counseled to expect further visual loss.

205. A 58-year-old man with essential hypertension is referred to a retinal specialist to “rule out cystoid macular edema” 4 weeks after uncomplicated phacoemulsification on his right eye. His best corrected acuity is 20/40 in the right eye and 20/200 in the left eye. Examination of the right eye shows a quiet pseudophakic anterior segment. The vitreous is clear bilaterally. There is a posterior subcapsular cataract in the left eye, which is felt to be sufficient to account for his acuity. His fundi are shown in the figures below (left and middle). Peripherally, he has bilateral inferior retinal detachments (RDs) that shift with head position. No retinal breaks are seen. A fluorescein angiogram (FA) for the right eye is shown in the figure below (right) (angiographic quality for the left eye was poor due to the cataract). Which one of the following statements about this man is true?
   a. Therapy should include retrobulbar steroid and a topical nonsteroidal agent.
   b. Therapy should include laser photocoagulation of the right macula.
   c. Therapy should include oral steroids.
   d. Proper serologic testing will confirm the diagnosis.
   e. The patient should be promptly referred to his internist.
206. Results of the Endophthalmitis–Vitrectomy Study (EVS) are applicable to which group of patients?
   a. any patient with presumed exogenous bacterial endophthalmitis.
   b. any patient with presumed bacterial endophthalmitis within 1 year of intraocular surgery.
   c. any patient with presumed bacterial endophthalmitis within 6 weeks of cataract surgery.
   d. any patient with presumed bacterial endophthalmitis within 6 weeks of cataract or filtration surgery.
   e. any patient with presumed bacterial endophthalmitis within 6 weeks of cataract surgery or trauma.

207. The Endophthalmitis–Vitrectomy Study (EVS) has documented which one of the following?
   a. Intravenous ceftazidime reduces the duration of severe visual loss in acute postoperative bacterial endophthalmitis but does not improve long-term visual outcome.
   b. Intravenous ceftazidime reduces the duration of severe visual loss and improves long-term visual outcome in patients with acute postoperative bacterial endophthalmitis.
   c. Intravenous amikacin reduces the duration of severe visual loss in acute postoperative bacterial endophthalmitis but does not improve long-term visual outcome.
   d. Intravenous amikacin reduces the duration of severe visual loss and improves long-term visual outcome in patients with acute postoperative bacterial endophthalmitis.
   e. Neither intravenous ceftazidime nor amikacin offers any therapeutic advantage in acute postoperative bacterial endophthalmitis.

208. What single factor from the presenting history or physical examination most strongly dictates appropriate surgical management of acute postoperative bacterial endophthalmitis?
   a. visual acuity at presentation.
   b. type of cataract extraction (extracapsular versus phacoemulsification).
   c. age of patient (younger than 60 years versus older than 60 years).
   d. duration of symptoms.
   e. none of the above.

209. Among men, each of the following constitutes a significant risk factor for the development of a central retinal vein occlusion (CRVO) except:
   a. systemic hypertension.
   b. diabetes mellitus.
   c. elevated erythrocyte sedimentation rate.
   d. open-angle glaucoma.
   e. decreased physical activity.

210. Among women, each of the following constitutes a significant risk factor for central retinal vein occlusion (CRVO) except:
   a. systemic hypertension.
   b. diabetes mellitus.
   c. elevated erythrocyte sedimentation rate.
   d. open-angle glaucoma.
   e. moderate alcohol consumption.

211. Among patients with initially nonperfused (ischemic) central retinal vein occlusion (CRVO), which one of the following is not a risk factor for the development of at least 2 hours of iris or angle neovascularization (NV)?
   a. >30 disc areas of nonperfusion.
   b. male gender.
   c. duration <1 month.
   d. severe intraretinal hemorrhage.
   e. cigarette smoking.

212. What percentage of patients diagnosed with a nonischemic central retinal vein occlusion (CRVO) of <1 year’s duration will develop iris or angle neovascularization (NV) within 4 months?
   a. 0% to 1%.
   b. 5% to 6%.
   c. 10% to 15%.
   d. 20% to 30%.
   e. 50% to 60%.

213. What percentage of patients diagnosed with an indeterminate central retinal vein occlusion (CRVO) of <1 year’s duration will develop iris or angle neovascularization (NV) within 4 months?
   a. 0% to 1%.
   b. 5% to 6%.
   c. 10% to 15%.
   d. 20% to 30%.
   e. 50% to 60%.
214. Findings of the Central Vein Occlusion Study (CVOS) include each of the following except:
   a. Grid pattern photocoagulation does not significantly reduce cystoid macular edema (CME) associated with central retinal vein occlusion (CRVO).
   b. Grid pattern photocoagulation does not significantly alter visual outcome in patients with CME associated with CRVO.
   c. Roughly 25% to 30% of patients with at least 10 disc areas of nonperfusion will develop iris or angle neovascularization (NV) within 3 years.
   d. Prophylactic scatter laser showed a trend toward reducing the incidence of iris or angle NV, but baseline differences in the treatment versus no treatment groups eliminated any statistical significance.
   e. Prophylactic scatter laser appears to reduce the efficacy of additional scatter laser treatment of iris or angle NV that develops subsequently.

215. According to results of the Subfoveal New Choroidal Neovascularization (CNV) Study, by the Macular Photocoagulation Study, which of the following would be the best candidate for immediate argon laser treatment?
   a. a 67-year-old woman with presenting acuity of 20/200 and 1 disc area of subfoveal CNV.
   b. an 82-year-old man with presenting acuity of 20/100 and 3 disc areas of subfoveal CNV.
   c. a 67-year-old man with presenting acuity of 20/400 and 3 disc areas of subfoveal CNV.
   d. a 67-year-old woman with presenting acuity of 20/400 and 3 disc areas of subfoveal CNV.
   e. an 82-year-old man with presenting acuity of 20/400 and 3 disc areas of subfoveal CNV.

216. According to results of the Subfoveal New Choroidal Neovascularization (CNV) Study, by the Macular Photocoagulation Study, which of the following would be the worst candidate for immediate argon laser treatment?
   a. a 67-year-old woman with presenting acuity of 20/200 and 1 disc area of subfoveal CNV.
   b. an 82-year-old man with presenting acuity of 20/100 and 3 disc areas of subfoveal CNV.
   c. a 67-year-old man with presenting acuity of 20/400 and 3 disc areas of subfoveal CNV.
   d. a 67-year-old woman with presenting acuity of 20/400 and 3 disc areas of subfoveal CNV.
   e. an 82-year-old man with presenting acuity of 20/400 and 3 disc areas of subfoveal CNV.

217. What is the approximate prevalence of idiopathic macular hole in a population of healthy Americans older than 55 years?
   a. 1 in 10,000.
   b. 1 in 1,000.
   c. 3 in 1,000.
   d. 12 in 1,000.
   e. 20 in 1,000.

218. What is the approximate prevalence of age-related macular degeneration (AMD) in a population of healthy Americans older than 55 years?
   a. 1 in 10,000.
   b. 1 in 1,000.
   c. 3 in 1,000.
   d. 12 in 1,000.
   e. 20 in 1,000.

219. T or F Foveal (focal) electroretinographic (ERG) testing can be used to provide useful information with regard to the presence or absence of organic disease within the macula.

220. Which of the following statements about optical coherence tomography (OCT) is false?
   a. OCT uses a combination of infrared light and sound waves to create images.
   b. Images created with OCT have much better resolution than standard 10-MHz B-scan ultrasound.
   c. OCT image quality will decrease as the amount of vitreous hemorrhage increases.
   d. Ultra-high resolution OCT can create images with resolutions of 2 to 3 microns.
   e. OCT also provides quantitative information with regard to macular thickness and retinal nerve fiber layer thickness.

221. For a patient with relapsing cytomegalovirus (CMV) retinitis originally controlled with maintenance doses of either foscarnet or ganciclovir,
which one of the following regimens has been shown to be most effective?

a. reinduction with the original agent.

b. stopping the first agent and inducing with the second.

c. inducing with the second agent and continuing the first at maintenance levels.

d. Options a., b., and c., are equally effective.

e. There is no clear answer because relapses of CMV retinitis are extremely rare.

222. The single most important physical property of perfluoropropane ($C_3F_8$) gas rendering it useful as a vitreous substitute in vitreoretinal surgery is its:

a. high interfacial surface tension.

b. high vapor pressure.

c. high specific gravity.

d. high index of refraction.

e. high viscosity.

223. The single most important physical property of silicone oil rendering it useful as a vitreous substitute in vitreoretinal surgery is its:

a. high interfacial surface tension.

b. high vapor pressure.

c. high specific gravity.

d. high index of refraction.

e. high viscosity.

224. The single most important physical property of perfluorocarbon liquids rendering them useful as vitreous substitutes in vitreoretinal surgery is their:

a. high interfacial surface tension.

b. high vapor pressure.

c. high specific gravity.

d. high index of refraction.

e. high viscosity.

225. Which of the following should probably be avoided in a patient with active central serous retinopathy (CSR)?

a. topical beta-blockers.

b. topical steroids.

c. oral beta-blockers.

d. oral steroids.

e. fava beans.

226. The annual incidence of development of choroidal neovascularization (CNV) in the fellow eyes of patients with ocular histoplasmosis syndrome and initially unilateral CNV is approximately:

a. 1%.

b. 2%.

c. 5%.

d. 8%.

e. 10%.

227. In a patient with unilateral choroidal neovascularization (CNV) secondary to ocular histoplasmosis syndrome, the presence of contralateral macular histo spots confers what relative risk for bilateral CNV?

a. $1.5 \times$.

b. $2 \times$.

c. $3 \times$.

d. $8 \times$.

e. $10 \times$.

228. Approximately what percentage of patients with ocular histoplasmosis syndrome will develop new atrophic scars over a period of 5 to 15 years?

a. <1%.

b. 1% to 5%.

c. 5% to 10%.

d. 10% to 15%.

e. 15% to 20%.

229. A 75-year-old woman presents to you complaining of painless decreased vision in her left eye, worsening over the past 6 hours. She denies any other systemic problems and has no other medical history. Blood pressure, measured in your clinic, is 145/90. Ophthalmic examination reveals a visual acuity of 20/20 oculus dexter (OD) and counting fingers (CF) at 6 feet oculus sinister (OS). Ophthalmic examination reveals a normal right eye, a relative afferent pupillary defect OS, and the left fundus as shown in the following figure. Which of the following would be the most appropriate next step?

a. Obtain an erythrocyte sedimentation rate (ESR).

b. Obtain a serum ESR and C-reactive protein.

c. If the patient does not have a history of jaw claudication, initiate aspirin (acetylsalicylic acid [ASA]) 325 mg sublingually.
Questions

d. Initiate ASA 325 mg orally.
e. Admit the patient to the hospital for a computed tomography (CT) scan.

d. Foveal changes as shown in this figure are almost always seen.
e. A late petalloid leakage pattern seen with fluorescein angiography (FA) is pathognomonic.

230. A 50-year-old woman presents to you complaining of gradual visual loss in her left eye over the past year. Ophthalmic examination is normal except for her left fundus, which is shown in the figure below. Which of the following statements is true?

a. The chance that she has the same condition in the right eye is 5%.
b. Most patients with her condition have visual acuities worse than 20/60.
c. A single intravitreal injection of perfluoropropane (C3F8) gas can be used to improve visual acuity.
d. Metamorphopsia is rare in these patients.
e. Without vitrectomy, the chance of spontaneous improvement in visual acuity is <10%.

232. A 45-year-old man complains of blurry vision in his right eye, which has gradually worsened over the past two years. His right fundus is shown in the figure below. There are no other abnormalities that you detect in his right eye. Which of the following statements about the condition shown in this figure is true?

a. The condition is typically bilateral.
b. Patients with this condition generally have a good visual prognosis.
c. Amazingly, the lesions shown do not create any visual field deficits.
d. The condition generally is not chronic.
e. The condition generally does not recur.

233. Which of the following statements about the condition shown in the following figure is true?

a. Most cases of this condition are bilateral.
b. Visual acuity in patients with this condition is generally <20/200.
c. The prevalence of this condition is increased in patients with diabetes.
d. The condition is common in young Asian men.
e. There is no role for surgery in patients with this condition.
234. A 20-year-old Jewish man complains of blurry vision bilaterally and difficulty driving at night. The fundus of his right eye (shown in the figure below) is very similar to the fundus of his left eye. Which of the following statements about the condition shown in this figure is false?

a. The condition can be inherited through several inheritance patterns.

b. Night blindness is one of the earliest symptoms of the disorder.

c. An electroretinogram (ERG) would typically show a reduction of both a-wave and b-wave amplitudes.

d. Visual acuity can remain excellent, despite a severely constricted visual field.

e. The clinical presentation of bone spiculation and chorioretinal atrophy is virtually pathognomonic for the condition.

d. Although it can occur, neovascularization (NV) is not common.

e. The yellow material shown in this figure is xanthophyll.

235. An 8-year-old boy is having difficulty seeing with his left eye. His ophthalmic (including funduscopy) examination is normal except for his left fundus. A peripheral photograph of his left fundus is shown in the following figure. Which of the following statements is true regarding the patient's condition?

a. Although not so in this specific case, most cases of this condition are bilateral.

b. The condition is typically inherited in an X-linked–recessive pattern and has a high male preponderance.

c. The condition can generally be managed with a single session of laser photocoagulation or cryotherapy.

d. To date, the gene responsible for this condition has not been identified.

e. All of the above statements are true.

236. The eye of a 10-year-old boy is found to have the fundus appearance shown in the figure below. Which of the following statements about the patient's condition is true?

a. The condition is typically unilateral and idiopathic.

b. The condition typically leads to rhegmatogenous retinal detachments (RRDs).

c. The condition has been associated with nystagmus and amblyopia in the past.

d. The condition requires cryotherapy or focal surrounding photocoagulation in symptomatic patients in order to prevent retinal detachments (RDs).

e. All of the above statements are true.

237. Which of the following statements about the condition shown in the following figure is true?

a. The condition can be a precursor for malignant transformation.

b. The condition typically leads to rhegmatogenous retinal detachments (RRDs).

c. The condition has been associated with nystagmus and amblyopia in the past.

d. The condition requires cryotherapy or focal surrounding photocoagulation in symptomatic patients in order to prevent retinal detachments (RDs).

e. All of the above statements are true.
Questions

238. Which of the following statements about the condition shown in the figure below is true?

a. The most likely diagnosis is an ophthalmic artery obstruction.
b. An erythrocyte sedimentation rate (ESR) and congenital retinitis pigmentosa (CRP) should be drawn to rule out giant cell arteritis (GCA) in patients who present with this condition.
c. The most likely diagnosis is iatrogenic subretinal silicone oil status post–pars plana vitrectomy.
d. Sphingolipidoses are usually the cause of this condition.
e. None of the above.

239. Which of the following statements about the condition shown in the figure below is true?

a. A defect in the gene for fibrillin is the most common systemic disease associated with the condition.
b. Photodynamic therapy currently has no known role in the treatment of this condition.
c. Fluorescein angiography (FA) of the eye shown in this figure will most likely demonstrate early hypofluorescence followed by late staining.
d. Up to half of all patients with this condition will have no systemic medical condition.
e. The lesions shown generally represent sclerosed retinal vessels.

240. A 25-year-old woman complains of blurry vision in her right eye over the past three days. She denies any past medical or ocular history. Anterior segment exam is normal bilaterally. Her right fundus is shown in the figure below (left). Unsure of your diagnosis, you decide to obtain a fluorescein angiogram (FA), part of which is depicted in the figures below (middle and right). Which of the following statements is true?

a. The condition shown is generally unilateral.
b. Visual prognosis is generally poor.
c. The condition can be associated with a fatal vasculitis.
d. Most patients have a viral prodrome before visual loss occurs.
e. None of the above is true.
241. Which of the following statements about the pathology shown in the figure below is true?
   a. Visual acuity in this patient is probably normal.
   b. If this eye is from a patient with type 2 diabetes, early vitrectomy may be indicated.
   c. If this eye is from a patient with type 1 diabetes with severe proliferative diabetic retinopathy, early vitrectomy may be beneficial.
   d. Hypertension is the most frequent cause of this condition in adults.
   e. B-scan ultrasound examination is always necessary in patients with this condition, regardless of whether the underlying cause is known.

242. Which of the following statements about the condition shown in the figure below is true?
   a. The condition shown is a posterior staphyloma.
   b. Patients with acquired immunodeficiency syndrome (AIDS) presenting with this condition must obtain neuroimaging.
   c. This particular patient will likely need a prolonged course of steroids to help preserve visual acuity.
   d. Folic acid is used in patients being treated for this condition to protect against thrombocytopenia and leukopenia.
   e. None of the above.

243. A 35-year-old retinal surgeon complains of the onset of blurry vision in his left eye over the past week. Ophthalmic examination is normal bilaterally, except for the left fundus, which is shown in the figure below (left). To confirm your diagnosis, you decide to obtain a fluorescein angiogram (FA), part of which is depicted in the figures below (middle and right). Which of the following statements is true?
   a. The angiographic presentation shown is the most common manifestation of the condition.
   b. More than half of all eyes with this condition eventually develop permanently reduced visual acuity.
   c. Early focal photocoagulation in this patient would be the treatment of choice, given his occupation.
   d. Oral steroids may be of moderate benefit in controlling this condition.
   e. None of the above.
244. Which of the following most likely represents the condition shown in the figure below?
   a. retinitis punctate albescens.
   b. fundus albipunctatus.
   c. Oguchi’s disease.
   d. enhanced S-cone syndrome.
   e. familial drusen.

245. A 28-year-old Asian woman complains of blurry vision bilaterally, which has dramatically worsened over the past two days. She denies any past medical or ocular history and any recent history of trauma. Her left fundus is shown in the figure below (left). Two frames from a fluorescein angiogram (FA) are shown in the figures below (middle and right). Which of the following statements is true?
   a. The patient most likely has sympathetic ophthalmia.
   b. The patient most likely has endophthalmitis.
   c. Most patients with this condition develop the Sugiura sign.
   d. The prognosis of this condition is generally poor.
   e. None of the above.

246. A 50-year-old man complains of fever, malaise, and visual loss in his right eye from two weeks ago. He recently got a new cat, which scratched him. The right eye fundus is shown in the figure to the right. Which of the following statements is false?
   a. Given his history, the most likely etiologic agent for his condition is *Bartonella henselae*.
   b. Oral ciprofloxacin is an effective therapy for this patient.
   c. The patient may have a painful lymphadenopathy as well.
   d. This patient will likely have a chronic recurrent course with poor visual prognosis.
   e. Several other etiologic agents can cause the condition that is shown in this figure.
247. Which of the following conditions in a patient with a history of limbal opacities is most likely to represent the fundus shown in the figure below?
   a. hydroxychloroquine retinopathy (retinal crystals and cornea verticillata).
   b. Bietti crystalline corneoretinal dystrophy.
   c. cystinosis.
   d. synchysis scintillans.
   e. fleck retina of Kandori.

248. A 65-year-old man undergoes uncomplicated phacoemulsification and posterior chamber intraocular lens implantation in his right eye. Two months later, he complains of blurry vision in his right eye, but his vision remains 20/20 in that eye. A fluorescein angiogram (FA) is performed and is depicted in the figure below. Which of the following statements about the patient’s condition is false?
   a. The patient has Irvine-Gass syndrome.
   b. The patient's condition will most likely spontaneously resolve.
   c. Angiographic evidence of this condition is more common than clinical evidence.
   d. The incidence of this condition is generally higher with complicated cataract surgery.
   e. The peak incidence of this condition is generally within two weeks after surgery.
Answers

1. c. There is thickening of arteriolar walls, which leads to nicking of venules, not vice versa.
2. d. Acute hypertensive episodes (rather than chronic hypertension) can lead to fibrinoid necrosis of choroidal arterioles. The choroid responds to chronic systemic hypertension in a complex fashion that is usually silent clinically.
3. d. The prevalence of endophthalmitis is 1.4% after intravitreal triamcinolone injection. However, this also includes cases of pseudoendophthalmitis. Excluding pseudoendophthalmitis, the prevalence is approximately 0.6%.
4. c. The early stages of diabetic retinopathy primarily involve vascular changes. Trouble with photopigment recycling and catabolism is not a factor in the pathogenesis of diabetic retinal disease.
5. True. Vitreous fluorophotometry may reveal leakage of fluorescein into the vitreous from abnormally permeable retinal vessels before any clinically detectable changes have occurred.
6. False. Loss of retinal vascular endothelial tight junctions have been implicated, but the barrier function of the retinal pigment epithelium (RPE) is generally intact.
7. a. Pericyte loss occurs early in diabetic retinopathy. Capillary closure also may be seen. Basement membrane thickening is found not just in the eye but systematically in diabetics. Medial hyperplasia is a feature of hypertensive vasculopathy.
8. False. The diffuse pattern is associated with more widespread vascular insult, often ischemic, and is more difficult to treat than most cases of focal edema.
9. c. Retinal thickening within the temporal arcades would encompass all macular edema, much of which does not imminently threaten vision. Extensive foveal and parafoveal nonperfusion would imply that the likelihood of improving vision would be small, even with treatment of any associated edema. The third definition of clinically significant macular edema is any thickening within 500 microns of the center of the fovea.
10. c. Formation of extensive intraretinal microvascular abnormalities implies widespread severe injury to small arterioles, with the resulting ischemic state commonly being the immediate predecessor or neovascularization (NV). The other options are features of lower-risk nonproliferative retinopathy, with the exception of disc neovascularization (NVD) (which is a form of proliferative diabetic retinopathy).
11. d.
12. e. Shorter eyes have greater resistance to fluid efflux from the suprachoroidal space. The percentage of retinal surface area treated is a very important factor and represents the combined effect of total treatment area and the size of the eye.
13. False. Retinal tears can easily occur as a result of vitreoretinal adhesions formed at areas of neovascularization (NV). Tractional-rhegmatogenous retinal detachment (RRD) is very common in patients with proliferative diabetic retinopathy.
14. b. The early worsening seen in the Diabetes Control and Complications Trial (DCCT) was temporary and had no long-term implications regarding the progression of retinopathy. Patients who had early worsening were more likely to recover, and actually improve, than patients in the conventional treatment group. The benefit of intensive control was seen in all subgroups of patients stratified by severity of retinopathy at the study’s outset. The DCCT only evaluated patients with type 1 diabetes. The benefit of intensive treatment took up to 3 years to be realized for patients with proliferative retinopathy and 5 years for patients with macular edema. Apparently, diabetic retinopathy builds “momentum” that requires years of intensive control to arrest.
15. d. All of the others are either difficult to treat successfully (cystoid macular edema [CME]) or associated with considerable retinal damage from previous insult (foveal nonperfusion and extensive foveal hard exudate).
16. a. The vitreous acts as a scaffold for the growth of neovascularization (NV) and will exert traction once it has developed, leading to hemorrhage and retinal detachment (RD).
17. False. The Early Treatment for Diabetic Retinopathy Study (ETDRS) found that twice as many untreated patients lost vision. It is not accurate to conclude that laser treatment is likely to improve vision in diabetic macular edema.
18. True. A 60% reduction in progression to severe visual loss in patients with so-called high-risk characteristics was reported.
19. d. Disc neovascularization (NVD) greater than one fourth to one third of a disc area is considered high risk, whether or not it is associated with vitreous hemorrhage. This is in distinction to neovascularization elsewhere (NVE), which must be associated with bleeding to qualify as high risk. Furthermore, to meet the Diabetic Retinopathy Study (DRS) criteria for high-risk disease, NVE also must comprise at least half the disc area. Any NVD associated with hemorrhage is high risk.
20. c. Decreased night vision results from destruction of extramacular rods. Angle-closure glaucoma may occur after particularly heavy treatment associated with choroidal effusions. Regression of neovascular fronds may be associated with contracture and secondary rhegmatogenous retinal detachment (RRD) or traction
11: Retina and Vitreous

retinal detachment (RD). Laser can cause temporary or permanent dysfunction of the long ciliary nerves passing through the outer choroid and cause difficulties with corneal sensation and accommodation. The latter probably occurs far more often than suspected. Prethreshold retinopathy of prematurity (ROP) also can aggravate macular edema, further compromising near acuity.

21. d. Although rare, cases of retinopathy have been reported in association with sickle cell trait (Hb AS) only. In addition, sickle cell SC disease can be associated with angioid streaks and comma-shaped conjunctival capillaries. The incidence of proliferative retinopathy is as follows: sickle cell SC disease (Hb SC), 33%; sickle cell thalassemia (Hb SThal), 14%; sickle cell SS disease (Hb SS), 3%.

22. True. Both are felt to be caused by a similar mechanism: capillary nonperfusion with subsequent retinal ischemia, leading to the production of a vasogenic, diffusible substance. In sickle cell (SC) retinopathy, elevated blood viscosity may occlude venules and arterioles, as well as capillaries.

23. True.

24. a. Preretinal hemorrhage may rarely be seen in nonproliferative disease, but it typically heralds neovascularization (NV).

25. e. All can occur. With sickle cell (SC) disease, obstruction of the central macular blood supply may occur due to thrombosis of a cilioretinal or macular branch artery. This complication is very unusual in other forms of sickle hemoglobinopathies. Conversely, neovascularization (NV) is a relatively uncommon feature of SC retinopathy. If choroidal neovascularization (CNV) occurs, it is usually related to angioid streaks. As in diabetes, contracture of neovascular fronds may lead to retinal detachment (RD).

26. True. A number of systemic disorders are associated with angioid streaks, including pachychromatoma elasticum (“P”), Ehlers-Danlos syndrome (“E”), Paget’s disease of bone (“P”), sickle cell (SC) disease and SC trait (“S”), and idiopathic (“I”) causes—remember these by the acronym “PEPSI.” Approximately 50% of cases of angioid streaks are idiopathic. Photodynamic therapy may be of some benefit in the treatment of choroidal neovascularization (CNV) secondary to angioid streaks.

27. True.

28. False. Retinal neovascularization (NV), vitreous hemorrhage, and retinal detachments (RDs) are more characteristic of sickle cell SC disease and sickle cell thalassemia (Hb SThal) than of sickle cell SS disease (Hb SS).

29. a. Photocoagulation to close the feeder vessels of neovascular fronds can cause regression of the neovascularization (NV) but is associated with a relatively high rate of complications (hemorrhage). Scatter photocoagulation to the region involved with the NV can effectively control it, with fewer complications. With dense vitreous hemorrhage, vitrectomy may be necessary to permit laser treatment.

30. c. Ranibizumab (Lucentis) is a potent antiangiogenic agent administered through intravitreal injection and is derived from bevacizumab (Avastin), a monoclonal antibody to VEGF. Phase III data from Genentech’s MA-RINA study demonstrated visual acuity improvement in approximately one third of patients.

31. False. The nasal retina is usually vascularized prior to the temporal side, usually at 36 weeks’ gestation. Vascularization of the temporal retina is generally complete by term.

32. e. Natural history studies estimate an 85% rate of spontaneous regression.

33. e. Stage 5 retinopathy of prematurity (ROP) is defined as a total retinal detachment (RD). Stage 1 is a flat demarcation line. The demarcation line of stage 2 has three dimensions—that is, it protrudes into the vitreous. Stage 3 features extra retinal fibrovascular proliferation. Stage 4 is a subtotal RD (stage 4A is with the macula attached and stage 4B is with the macula detached).

34. False. It is hypothesized that immature vascular precursor tissue is susceptible to oxygen-induced cytotoxicity. Environmental oxygen may be sufficient to induce retinopathy of prematurity (ROP) in some cases.

35. e. All of the following definitions are part of prethreshold disease. The Early Treatment for Retinopathy of Prematurity (ETROP) study found that early treatment of type I prethreshold retinopathy of prematurity (ROP) using retinal ablative therapy reduced unfavorable outcomes to a clinically important degree. Type 1 ROP was defined as (i) zone I, any stage ROP with plus disease; (ii) zone I, stage 3 ROP without plus disease; or (iii) zone II, stage 2 or 3 ROP with plus disease.

36. b. Several retrospective studies have suggested that laser therapy is at least as effective as cryotherapy, with fewer complications and less stress on the child. A few randomized prospective studies have confirmed this.

37. e. The supertemporal quadrant is most commonly affected in branch retinal vein occlusion (BRVO) (almost two thirds of cases). The most common systemic disease association with BRVO is systemic hypertension. A clear association with primary open-angle glaucoma (POAG), as shown for central retinal vein occlusion (CRVO), has not been established, although several small studies claim this is true.

38. a. Complications of branch retinal vein occlusion (BRVO) can be divided into acute and chronic categories. Macular edema, macular nonperfusion, and hemorrhage may occur acutely. Macular edema, subretinal fibrosis, and posterior neovascularization (NV) can be delayed causes of visual loss following
Answers

BRVO. Rubeosis iridis occurs in <1% of cases with BRVO.
39. True. The Branch Vein Occlusion Study (BVOS) showed that eyes treated for macular edema with photocoagulation had a 60% chance of recovering 20/40 or better vision, compared with 34% for controls.
40. a. Treatment is generally not undertaken until signs of neovascularization (NV) are evident, because the long-term incidence of severe visual loss in patients with extensive nonperfusion is low (only a subset of these actually develop NV). This risk increases once NV occurs. Greater than 5 disc diameters of ischemia was found to be associated with a 31% risk of developing NV.
41. False. All forms of central retinal vein occlusion (CRVO) are caused by thrombosis of the central retinal vein at the level of the lamina cribrosa.
42. d. Fluorescein angiography (FA) of a nonischemic central retinal vein occlusion (CRVO) should show minimal nonperfusion in contrast to an ischemic CRVO, which will show extensive capillary nonperfusion.
43. True. According to one study, approximately 48% of nonischemic central retinal vein occlusion (CRVO) will resolve completely.
44. e. The most common systemic disease associations for central retinal vein occlusion (CRVO) include atherosclerotic heart disease, hypertension, and diabetes. Less common associations include paraproteinemias, syphilis, sarcoidosis, increased intraorbital pressure, and hypersensitivity and other vasculitides.
45. True. Extremely high intraocular pressure (IOP) may occlude a healthy central retinal vein.
46. True. In central retinal vein occlusion (CRVO), the vitreous swells as transudate and seeps out of the congested retinal veins. This may cause narrowing of the angle and precipitate angle closure.
47. c. Any central retinal vein occlusion (CRVO) will have hemorrhage in all quadrants, although asymmetrically.
48. b. The retinopathy of carotid artery occlusive (CRVO) disease may feature retinal hemorrhages and venous dilatation such as central retinal vein occlusion (CRVO), but the central retinal artery pressure is low. In CRVO, the retinal arteriolar pressure should be normal or elevated. Cotton-wool spots should not be seen in simple carotid occlusion.
49. False. Occlusion of precapillary arterioles results in cotton-wool spots. Cherry-red spots may arise because of axonal swelling in central retinal arterial occlusion (CRAO) or because of axonal accumulation of metabolites in storage diseases such as Tay-Sachs disease.
50. c. Retinal artery occlusion is caused by embolization or thrombosis of the involved vessel. Uncommon causes of emboli include fat emboli, cardiac myxoma, and talc emboli in intravenous drug abusers. Mitral valve prolapse, vasculitides, and connective tissue disorders also have been associated with branch retinal arterial occlusion (BRAO).
51. True. The visual prognosis for ischemic central retinal vein occlusion (CRVO) is poor, with only 10% of affected eyes obtaining better than 20/400 vision. Central retinal arterial occlusion (CRAO) has a better visual prognosis than ischemic CRVO, although it is still devastating. In one study, 66% of eyes with CRAO had final vision of <20/400, whereas 18% of eyes with CRVO recovered vision of at least 20/40.
52. d. Studies in monkeys whose central retinal arteries were ligated established that irreversible damage to the retina occurs after 97 minutes of total ischemia.
53. a. Type I is probably a forme fruste of Coats’ disease, but types II and III are bilateral and occur in both sexes. Microscopically, the structural abnormalities are similar to diabetic microangiopathy, rather than a true telangiectasis. Unlike diabetes, there is no stimulus for retinal neovascularization (NV). However, exudation in type I may respond to laser photocoagulation. Choroidal neovascularization (CNV) is another cause of visual loss.
54. c. Eales’ disease is an idiopathic retinal vasculitis in young boys or men (most commonly from India) and is generally bilateral. The original syndrome was defined as retinal vasculitis in a young man with associated epistaxis, constipation, and positive reaction to dermal purified protein derivative. A potentially lethal cerebral vasculitis also has been recognized as an occasion finding. Neovascularization (NV) can be treated with scatter photocoagulation and visual prognosis is generally good with prompt appropriate therapy.
55. e. Additional causes of retinal vasculitis include Behçet’s disease, temporal arteritis, Wegener’s granulomatosis, sarcoidosis, herpes zoster, syphilis, and toxoplasmosis.
56. b. Potential causes of visual loss include subinternal limiting membrane hemorrhage, vitreous hemorrhage, subretinal hemorrhage, and macular edema. Photocoagulation around the macroaneurysm may reduce edema and improve acuity. Direct focal treatment of the aneurysm is not recommended because hemorrhage is likely.
57. b. Intraocular lens (IOL) implantation at the time of extracapsular cataract extraction does not affect the incidence of cystoid macular edema (CME).
58. c. Retinal telangiectasia (Coats’ disease, Leber’s military aneurysms) is defined by the presence of an exudative retinal detachment (RD) with associated vascular anomalies. This condition is not hereditary and is not associated with systemic vascular abnormalities. Usually, only one eye is involved, and men are predominantly affected (85%).
59. False. The vascular von Hippel’s lesion is a capillary hemangioma of the retina or optic nerve. It also may be
11: Retina and Vitreous

termed a “retinal hemangioblastoma.” If both retinal angiomas and central nervous system (CNS)/visceral involvement are present, the term “von Hippel-Lindau” disease is used.

60. True. Despite retinal atrophy (or perhaps because of it), breaks and detachments rarely occur spontaneously.

61. a. Café-au-lait spots are characteristic of neurofibromatosis and are not seen as part of von Hippel-Lindau disease.

62. True. In fact, treatment may be associated with a total exudative detachment (which typically resolves).

63. False. Both congenital retinal arteriovenous malformations and retinal capillary hemangiomas (von Hippel-Lindau disease) are associated with subretinal fluid and exudate. Cavernous hemangiomas bleed but usually do not leak.

64. True. Retinal cavernous hemangioma can cause vitreous hemorrhage, which is presumed to be secondary to traction.

65. b. Clinically, the macula is 5 to 6 mm in diameter, centered between the temporal vascular arcades. In this region, ganglion cells form two or three sublayers within the ganglion cell layer.

66. c. The retinal vessels supply the nerve fiber layer, the ganglion cell layer, the inner plexiform layer, and the inner third of the inner nuclear layer. The choriocapillaris supplies the outer two thirds of the inner nuclear layer, the outer plexiform layer, the outer nuclear layer, the photoreceptors, and the retinal pigment epithelium (RPE).

67. a. The membrane may be thought of as an elastin sandwich—the bread is composed of collagenous zones and basement membrane on either side.

68. True. The retinal pigment epithelium (RPE) continually ingests membranes shed by the outer segments of the photoreceptor cells. RPE phagosomes containing ingested outer segment debris are discharged into Bruch’s membrane after being processed. This is seen most prominently in the macula, the most metabolically active portion of the retina.

69. True. Vitamin A required for vision is delivered to and stored in the retinal pigment epithelium (RPE) (of course, most vitamin A stores are in the liver).

70. True. Metaplasia of the retinal pigment epithelium (RPE) can result in both preretinal and subretinal membranes.

71. c. In fluorescein angiography (FA), white light from the camera first passes through a blue filter. The blue light (wavelength of 490 nm) is absorbed by the fluorescein molecules in the retinal and chorial vascular network, stimulating them to emit yellow–green light (530 nm). A yellow–green filter is placed to block the blue light reflected from the eye, allowing the yellow–green light into the camera. Fluorescein molecules (not bound to albumin), which have a molecular weight <600 daltons, can easily pass through the spaces between endothelial cells of the choriodicapillaris but normally cannot leak through the tight junctions of the retinal pigment epithelium (RPE) or retinal vascular endothelium (blood–retinal barrier). Anaphylactic shock is thought to occur at a rate of 1 in 100,000 procedures.

72. a. Leakage appears as an area of early hyperfluorescence that gradually increases in size and intensity throughout the angiogram.

73. a. A focal defect in the internal limiting membrane is not necessary for retinal neovascularization (NV) to progress.

74. c. Photoreceptor atrophy is unavoidable in the setting of marked retinal pigment epithelium (RPE) dysfunction. This is probably the mechanism of visual loss in severe atrophic age-related macular degeneration (AMD).

75. b. Central serous retinopathy (CSR) presents in men most commonly in their 30s. Women tend to present with CSR later, in their 40s. The classic “smokestack” pattern is uncommon (10% of cases). It is much more common to see a small focal hyperfluorescent leak from the retinal pigment epithelium (RPE) that appears early in the angiogram and increases in size and intensity with time (inkblot). Laser treatment shortens the course of each attack, but final visual acuity and rate of recurrence are not affected.

76. True.

77. c. Treatment is generally reserved for (i) occupational or other demands for rapid recovery of binocular function, (ii) persistent serous detachment (>4 months), (iii) prior episodes of central serous retinopathy (CSR) that have been associated with permanently decreased visual acuity, and (iv) permanent visual loss caused by CSR in the contralateral eye.

78. c. All of the above types of visual prostheses have been successfully implanted in humans. Visual restoration, to date, has been limited.

79. False. The diagnosis of age-related macular degeneration (AMD) is made when eyes with drusen or their associated complications develop a decrease in visual acuity not attributable to other ocular conditions.

80. d. Possible risk factors for progressive visual loss in age-related macular degeneration (AMD) include a positive family history, hyperopia, cigarette smoking, and light iris color. Hypertension has not been directly related.

81. a. Findings in the nonexudative form of age-related macular degeneration (AMD) include pigmentary changes, drusen, and areas of geographic atrophy. Patients with central geographic atrophy generally have a guarded visual prognosis.

82. True. A pigment epithelial detachment in a patient younger than 50 years is generally thought to be a variant
of central serous retinopathy (CSR) and has a better prognosis. Visual acuity often returns to 20/30.

83. a. The Macular Photocoagulation Study has documented that argon laser photocoagulation is effective in reducing the rate of severe visual loss for extrafoveal choroidal neovascularization (CNV) (>200 microns from the center of the foveal avascular zone) and juxtafoveal CNV (1 to 199 microns). For subfoveal CNV, laser treatment may preserve contrast sensitivity and reading speed, but visual acuity is generally worse immediately after laser treatment.

84. b. Endemic areas include Ohio and the states of the Mississippi River valleys. Systemic antifungal treatment does not lead to resolution of the ocular findings.

85. c. At present, effective methods of treatment of choroidal neovascularization (CNV) secondary to presumed ocular histoplasmosis syndrome (POHS) are photodynamic therapy and argon laser photocoagulation.

86. a. The likelihood of contralateral choroidal neovascularization (CNV) in a patient with a disciform macular scar from presumed ocular histoplasmosis syndrome (POHS) is increased if there are focal macular scars in the better eye.

87. c. Angioid streaks represent discontinuities in abnormally thickened and calcified Bruch's membrane. They do not always extend in continuity from the optic nerve head and appear to radiate from the optic nerve head, rather than forming concentric circles around it. Because the overlying retinal pigment epithelium (RPE) is often atrophic, angioid streaks may appear as window defects on fluorescein angiography (FA).

88. a. Sickle cell (SC) anemia is another potential systemic disease associated with angioid streaks. Up to 50% of patients with angioid streaks have no identifiable systemic illness. High myopia is not associated with angioid streaks but may feature lacquer cracks, which are similar histopathologically.

89. b.

90. True. Approximately 75% of eyes with idiopathic epiretinal membranes maintain a visual acuity of 20/50 or better.

91. a. The first study of surgery for macular holes evaluated patients with the earliest stage, stage 1. The natural history of the disorder, with up to 50% enjoying spontaneous improvement, is at least as favorable as, if not superior to, surgical intervention.

92. a. The prevalence of bilaterality of idiopathic macular holes, including earlier stages before a full-thickness dehiscence develops, will be higher, approaching 20% to 25%. Because a large proportion of early holes spontaneously resolve, only a small fraction (3%) develop bilateral full-thickness defects.

93. c. Idiopathic macular holes are believed to arise because of both tangential and anteroposterior vitreous traction. The earliest change is loss of the normal foveal depression due to elevation of the fovea itself off the retinal pigment epithelium (RPE), thereby constituting a tiny sensory retinal detachment (RD). This appears clinically as a yellow dot or ring (stages IA and IB, respectively).

94. c. Although hydroxychloroquine is thought to be less toxic, both hydroxychloroquine and chloroquine have a significant risk of retinal toxicity. Color vision testing and threshold central visual field testing are thought to be important in elevating subclinical retinopathy. The electrooculogram (EOG) is not the most sensitive parameter in detecting chloroquine retinopathy, although it does reflect pathophysiology of retinal pigment epithelium (RPE) damage when abnormal.

95. False. Because of their slow excretion, toxic effects of chloroquine and hydroxychloroquine may progress despite cessation of the drug. Any abnormalities caused by these medications are probably permanent, although mild deficits may be reversible.

96. True. Both the phenothiazines and the antimalarials (chloroquine and hydroxychloroquine) may cause electroretinographic abnormalities and peripheral pigmentary retinopathy. Thioridazine is probably the most likely phenothiazine to cause retinal toxicity.

97. True. Chlorpromazine (Thorazine) may lead to abnormal pigmentation of the conjunctiva, cornea, eyelids, anterior lens capsule, and retina. Thoridizine (Mellaril) is more likely to cause retinal pigment stippling or widespread atrophy of the pigment epithelium and choriocapillaris, but it is less likely to affect other ocular structures.

98. False. Type II (Hunter's) is indeed X-linked recessive. However, the rest of the mucopolysaccharidoses (MPS) are autosomal recessive (not autosomal dominant).

99. b. Corneal clouding is common in mucopolysaccharidoses (MPS) types I-H (Hurler’s), I-S (Scheie’s), IV (Morquio’s), and VI (Maroteaux-Lamy). It may rarely be seen in type II (Hunter’s) but is never seen in type III (Sanfilippo’s).

100. a. Retinal pigmentary degeneration resembling typical retinitis pigmentosa (RP) is seen in types I-H (Hurler’s), I-S (Scheie’s), II (Hunter’s), and III (Sanfilippo’s).

101. a. Optic atrophy can be found in all of the types in the Macular Photocoagulation Study except type VI (mild phenotypic) and type VII.

102. True. Gangliosides are found in cerebral gray matter, whereas cerebrosides are found in cell membranes throughout the body.

103. a. In the sphingolipidoses, a cherry-red spot is caused by accumulation of sphingolipids in the retinal ganglion cell layer. The orange-red color of the retinal pigment epithelium (RPE) and choroid stands out in
the central fovea, where the ganglion cell layer is very thin or absent. There is no gross accumulation of lipid in Fabry’s or Krabbe’s disease; therefore, no cherry-red spot is present. 104. False. Ceramide accumulation in blood vessels leads to diffuse angiopathy, with myocardial infarction (MI) or stroke quite common. Still, renal failure is the leading cause of death.

105. a. Pigmentary retinal changes are not characteristic of Fabry’s disease.

106. b. Deposition of cystine crystals in the conjunctiva and cornea with resultant photophobia occurs in all three types of cystinosis (nephropathic, late onset, and benign). A pigmentary retinopathy with a “salt-and-pepper” appearance occurs only in the nephropathic type. However, no significant visual disturbance occurs. In addition to its benefits as a topical medication, systemic cysteamine can also be used to help prevent renal failure.

107. c. Radiation retinopathy may occur with doses of approximately 3,000 rad (30 Gy) or more, although rare cases have been reported after as little as 2,400 rad (24 Gy).

108. c. Visual acuity in solar retinopathy is generally not reduced below 20/200, and is frequently only minimally reduced. Recovery is good.

109. c. The ratio of rods to cones ranges between 12:1 and 15:1. Some investigators claim a ratio as high as 20:1. Rod density is maximal in a ring that is 20 to 40 degrees eccentric to the fovea, whereas cone density is greatest in the fovea. Although the cone density is greater in the macula than in the peripheral retina, the number of rods and cones in the macula is roughly equal, and nearly half of all cones lie outside the macula.

110. True. This is the definition of a giant retinal tear.

111. c. The description is classic for Stickler’s syndrome, or hereditary arthroophthalmopathy. It is inherited in an autosomal-dominant fashion and is characterized by progressive myopia with a high incidence of retinal detachment (RD) and abnormal epiphyseal development with premature degenerative changes to various joints. Most cases are secondary to premature termination codons in COL2A1, the gene for type II collagen, which is a major constituent in both cartilage and vitreous.

112. True. The scotopic electroretinogram (ERG) represents the ERG with rod input only. It is generated with a dim white or blue flash below the cone threshold in a dark-adapted state.

113. False. Some rod function is represented even in a light-adapted electroretinogram (ERG). Rod input can be minimized or eliminated by using only the longest wavelengths (orange–red), or more practically, by using a flicker stimulus.

114. d. Rods can respond to flickering stimuli of rates up to 20 Hz. Only cones can respond at higher frequencies, with a maximum of approximately 70 Hz.

115. c. Although the b-wave amplitude can be increased in focal or stationary retinal disease, the b-wave implicit time is increased only in diffuse, progressive retinal disease.

116. False. With increasing stimulus intensity, the b-wave amplitude increases and the implicit time (the time from stimulus to b-wave peak) decreases.

117. b. Oguchi’s disease and fundus albipunctatus are forms of congenital stationary night blindness. Choroideremia is an X-linked recessive form of rod–cone degeneration. Fundus flavimaculatus (Stargardt’s disease) is a condition with abnormal lipofuscin deposition in the retinal pigment epithelium (RPE). Functional problems in this syndrome are not limited to night blindness.

118. False. Posterior vitreous detachments are generally absent in eyes with retinal dialyses. The converse is true in eyes with giant retinal tears.

119. b. Pegaptanib sodium (Macugen) is a mRNA aptamer (not a monoclonal antibody) that has been approved by the U.S. Food and Drug Administration (FDA) to treat wet age-related macular degeneration (AMD). All of the other statements are true.

120. b. In electrooculogram (EOG), amplitudes generally increase with light adaptation and diminish with dark adaptation. The EOG is considered abnormal if the light-peak to dark-trough ratio (the Arden ratio) is <1.75.

121. d. In Best’s disease, the electroretinogram (ERG) is generally normal, whereas the electrooculogram (EOG) is abnormal.

122. b. An abnormally low light-peak to dark-trough ratio on electrooculogram (EOG) has been found to occur in retinal toxicity from hydroxychloroquine and chloroquine.

123. c. Rods are approximately 1,000 times more sensitive than cones during dark adaptation.

124. True. Anaphylaxis, although extremely rare, is a potential complication of intravitreal injection.

125. True. An off-center-on-surround receptive field implies that stimulation of the central photoreceptors is inhibitory (ganglion cell impulses diminish in frequency), whereas stimulation of surrounding photoreceptors is excitatory (ganglion cell impulses increase in frequency).

126. False. The Purkinje shift refers to a shift in peak spectral sensitivity, from 555 nm to 505 nm, with dark adaptation. Dark adaptation does increase light sensitivity by a factor of 1,000, but this is not the Purkinje shift.

127. True. Each cone pigment is capable of absorbing a broad range of wavelengths. Thus, although a cone with a red-sensitive pigment absorbs red light most efficiently, it could produce the same firing rate if stimulated with a much higher intensity of green light.
128. **c.** All of the answers are true. Refsum’s disease is characterized by ataxia, pigmentary retinopathy, deafness, and cardiac myopathy.

129. **a.** Deuteranomalous trichromats are the most common. They are X-linked recessive and found in 5% of the male population.

130. **c.** Rod monochromats typically have nyctalopia and acuities in the 20/200 range. Blue monochromats have variable nyctalopia and acuities in the 20/40 to 20/200 range.

131. **b.** Decreased acuity and nyctalopia are seen in both disorders. The electroretinograms (ERGs) in both disorders are similar in that they show cone dysfunction with relatively normal rod function. Rod monochromatism is an autosomal-recessive disorder, whereas blue cone monochromatism is X-linked recessive.

132. **a.** Ishihara plates detect only red-green defects.

133. **c.** Up to one third of patients with intracranial hemorrhage will have intraocular hemorrhage (the presence of both simultaneously is called “Terson’s syndrome”).

134. **e.** All of the other answers have been associated. Hepatic encephalopathy is generally not associated with Purtscher’s.

135. **True.** Some patients with retinitis pigmentosa (RP) will develop moderate hearing loss with age, but most cases of severe hearing loss are congenital.

136. **e.** Most forms of Leber’s congenital amaurosis are autosomal recessive. There is an autosomal-dominant infantile-onset retinitis pigmentosa (RP) that behaves like a progressive cone–rod degeneration of very early onset.

137. **False.** Usher’s syndrome only refers to the association of pigmentary retinopathy and congenital deafness.

138. **True.** Types I and III feature vestibular dysfunction. Patients with profound deafness are more likely to have cerebellar atrophy.

139. **d.** The other syndromes are autosomal recessive. With choroideremia, central acuity is spared until later in life relative to X-linked retinitis pigmentosa (RP).

140. **True.** The retinal findings in choroideremia include scalloped retinal pigment epithelium (RPE) atrophy with no or scanty hyperpigmentation.

141. **c.** Gyrate atrophy is an autosomal-recessive deficiency in ornithine aminotransferase activity. This deficiency causes an increase in serum ornithine levels and a decrease in serum lysine levels.

142. **e.** Although the inheritance in fundus flavimaculatus is usually autosomal recessive, autosomal-dominant pedigrees have also been documented.

143. **b.** Familial drusen are an autosomal-dominant disorder that begins with asymptomatic retinal changes in the third decade of life and does not become symptomatic until the fourth or fifth decade of life. The electroretinogram (ERG) is usually normal or mildly decreased.

144. **c.** This scotopic electroretinographic (ERG) pattern helps to distinguish fundus albipunctatus from retinitis punctata albescens and other forms of congenital stationary night blindness. Normalization of the scotopic ERG after dark adaptation has been shown in two pedigrees.

145. **True.** Vision may be 20/30 to 20/60 in the early stages of the disease, despite the prominent central “egg yolk” lesion.

146. **False.** On the contrary, patients with Stargardt’s disease may have poor vision with barely detectable macular changes.

147. **b.** The Mizuo-Nakamura effect occurs in Oguchi’s disease and X-linked cone dystrophy. The eponym refers to a relative lightening of the retinal pigment epithelium (RPE) and fundus after 3 to 4 hours of dark adaptation.

148. **e.** Although difficulty in driving at night would seem to be more likely with rod degenerations, urban night driving is generally performed with background illumination at low photopic intensities. Therefore, patients with cone degeneration may complain of this.

149. **True.** The risk of anaphylaxis is at least an order of magnitude lower (<1 in 100,000 patients).

150. **e.** All may mimic a generalized tapetoretinal degeneration. Harada’s disease and toxemia of pregnancy are exudative disorders that may resolve and lead to pseudo–retinitis pigmentosa (RP).

151. **c.** Fundus albipunctatus features multiple, tiny white flecks in the deep retina. Oguchi’s disease features the Mizuo phenomenon, a gold-colored fundus sheen that normalizes after several hours of dark adaptation.

152. **b.** Persistent hyperplastic primary vitreous (PHPV) also has been associated with foveal hypoplasia.

153. **a.** Stargardt’s disease is generally inherited on an autosomal-recessive basis, but dominant pedigrees also have been described.

154. **True.**

155. **b.** Despite the striking fundus appearance of the “egg yolk” lesion, visual acuity is usually quite good (20/30 to 20/50) at this stage. The amount of lipofuscin cannot be correlated with the amount of electrooculogram (EOG) depression.

156. **True.**

157. **False.** So-called multifocal Best’s disease features more than one lesion simultaneously.

158. **b.** Pattern dystrophies of the retinal pigment epithelium (RPE) are better seen on fluorescein angiography (FA) than with ophthalmoscopy. As in Best’s disease, the electrooculogram (EOG) is usually abnormal despite a normal electroretinogram (ERG).

159. **b.** Albinoidism differs from albinism in that the former does not have severe visual consequences. Both share certain clinical features, including photophobia, iris translumination defects, and fundus hypopigmentation.
11. Retina and Vitreous

160. c. True albinos have abnormal retinogeniculostriate projections as well as foveal hypoplasia. Temporal hemiretinal fibers that would normally project to the ipsilateral geniculate nucleus decussate abnormally. The lateral geniculate nuclei are grossly normal.

161. True. In general, albinism is inherited as an autosomal-dominant trait with incomplete penetrance, whereas true albinoism is inherited as either an autosomal-recessive or X-linked–recessive trait.

162. c. Nettleship-Falls albinism is inherited in an X-linked–recessive pattern.

163. c. Hermansky-Pudlak syndrome is characterized by bleeding diatheses, whereas Chédiak-Higashi syndrome is characterized by an immune deficiency and abnormal susceptibility to childhood infections.

164. c. Xenon arc emits polychromatic white light that cannot be as precisely focused as monochromatic light. In addition, blue light (harmful to the retina and lens) emission is significant. Other disadvantages include more pain and a greater rate of breaks in Bruch’s membrane (leading to choroidal effusions and/or choroidal neovascularization [CNV]) after xenon prethreshold retinopathy of prematurity (ROP) compared to argon laser photocoagulation.

165. d. Krypton red is useful for laser therapy in the presence of vitreous hemorrhage. In addition, because it is poorly absorbed by xanthophyll, it is useful for macular laser treatment.

166. d. Red light penetrates cataract and vitreous hemorrhage better than lights of other wavelengths.

167. True. This is in contrast to the xenon arc, in which the decreasing spot size decreases the total energy delivered.

168. d. Contrast sensitivity was better preserved over both the short term and the long term among treated patients, relative to untreated patients. This is in distinction to acuity and reading speed, both of which were worse among the treated patients, over the short term and the long term among treated patients.

169. e. Other potential complications include vitreous hemorrhage, nerve fiber bundle defects, incidental foveal burns, and retinal vascular occlusions. Angle-closure glaucoma, after extensive scatter treatment, is caused by choroidal effusion.

170. a. Other areas of firm vitreoretinal attachment include the optic nerve and major blood vessels. The vortex veins represent a firm point of attachment between the choroid and sclera.

171. True. Removal of the lens and posterior capsule allows the hyaluronic acid in the vitreous to diffuse into the anterior chamber and out of the eye.

172. b.

173. e. Hemorrhage or vitreous cells (“tobacco dust”) are suggestive of retinal breaks.

174. True. Spontaneous and familial dialyses occur inferotemporally. Traumatic dialyses also may occur superonasally.

175. True. See answer 174. This location is probably reflective of contrecoup injury to the ora from blunt forces delivered inferotemporally, where the globe is most exposed.

176. b. Although 8% to 10% of the general population has lattice degeneration, only a small subset will develop a retinal detachment (RD).

177. c. Estimates range from 20% to 40%.

178. False. Important histologic features of lattice degeneration include discontinuity of the internal limiting membrane and a pocket of liquefied vitreous overlying the degeneration. Vitreoretinal condensation and adherence occur at the margin of the lesion. Other features include sclerosis of the vessels and variable degrees of retinal atrophy.

179. b. Lattice degeneration and meridional complexes (redundant retinal folds in the same meridian as a ciliary process, usually occurring superonasally) increase the risk of rhegmatogenous retinal detachment (RRD). Cobblestone degeneration has occasionally been observed to limit the spread of retinal detachment (RD) and does not predispose to RRD. It also may be the site of secondary retinal breaks as the advancing subretinal fluid reaches the edge of a cobblestone, where choriretinal adherence is enhanced.

180. c. Approximately 22% of patients older than 20 years have cobblestone degeneration (RD), usually in the inferior quadrants.

181. b. In 97% of cases of rhegmatogenous retinal detachment (RRD), a definite break can be found. In the other 3%, one break is presumed to be present. This most frequently occurs in the setting of aphakic or pseudophakic retinal detachment, in which the breaks are frequently tiny.

182. c. Signs suggestive of rhegmatogenous retinal detachment (RRD) include (i) lower intraocular pressure (IOP) compared with the other eye, (ii) “tobacco dust” (small clumps of retinal pigment epithelium [RPE] cells floating in the vitreous), and (iii) corrugated appearance of the retina that undulates with eye movements. A smooth, domed appearance and shifting fluid are more suggestive of exudative detachment.

183. d. Proliferative vitreoretinopathy (PVR) is the most common cause of redetachment after successful repair. Retinal pigment epithelium (RPE) and glial cells proliferate and subsequently contract, causing fixed folds, traction, and/or detachment.

184. d. Approximately 100% reattachment can be achieved with rhegmatogenous retinal detachment (RRD) secondary to dialysis or small round holes, detachments with demarcation lines (chronic), or those with minimal subretinal fluid. Giant tears and combined
retinal–choroidal detachments have a poorer prognosis, whereas aphakic detachments have intermediate prognosis.

185. c. Degeneration of photoreceptors limits recovery of vision. Seventy-five percent of patients with macular detachment for a duration <1 week will recover vision >20/70. In contrast, those patients with macular detachment for a duration of 8 days or more have only a 50% chance of regaining acuity of at least 20/70.

186. a. In virtually all cases of typical degenerative retinoschisis, peripheral cystoid degeneration can be found, usually adjacent to the schisis cavity.

187. c. Greater than 70% of patients with retinoschisis are hyperopic, and this condition is bilateral in 50% to 80% of cases. However, in typical retinoschisis, complications such as hole formation and retinal detachment (RD) are rare. With retinoschisis posterior to the equator, the scotoma, although absolute and readily identifiable on perimetry, is rarely noted by the patient.

188. False. Demarcation lines with retinoschisis indicate current or previous full-thickness retinal detachment (RD).

189. False. A hole in the outer wall alone is sufficient for formation of RD. Indeed, holes in both inner and outer walls may lead the cavity to collapse, making the origin of detachment difficult to locate.

190. False. Indocyanine green (ICG) (not fluorescein) should be used with caution in patients with a history of shellfish or iodine allergy.

191. a. The Silicone Oil Study Group found that reattachment rates and visual outcomes were both worse when using SF6 as the tamponade agent, compared with either perfluoropropane (C3F8) or oil.

192. c. Persistent fetal vasculature or persistent hyperplastic primary vitreous (PHPV) should be included in the differential diagnosis of leukocoria and differentiated primary vitreous (PHPV) is unilateral in >90% of cases and is associated with microphthalmia. Retinoblastoma is often bilateral and has no significant associations with microphthalmia or cataracts.

193. c. Classification of hereditary hyaloideoretinopathies into two categories has been suggested: those with ocular manifestations only (Wagner’s disease) and those with systemic symptoms (Stickler’s syndrome). Wagner’s disease consists of high myopia, posterior subcapsular cataract, and an optically empty vitreous. Stickler’s syndrome is Wagner’s disease plus increased incidence of (i) retinal detachment (RD), (ii) facial anomalies, and (iii) musculoskeletal anomalies.

194. True. Familial exudative vitreoretinopathy is autosomal dominant and occurs in full-term infants with normal respiratory status.

195. b. Astroid hyalosis is monocular in 75% of cases.

196. True. Astroid hyalosis usually does not settle, suggesting some collagenous support of asteroid bodies within formed vitreous. Synchysis scintillans, the crystalline lipid breakdown products of previous hemorrhage, usually settle inferiorly in liquefied vitreous (or in an eye that has undergone vitrectomy).

197. d. Estimates of the proportion of vitreous hemorrhage caused by diabetic retinopathy have ranged from 39% to 54%.

198. c. Aphakic retinal detachment (RD) and retinal dialysis may be repaired with scleral buckling +/- pneumatic retinopexy alone. However, traumatic RD with vitreous hemorrhage or with severe proliferative vitreoretinopathy (PVR), may require vitrectomy to remove vitreous scaffolds that can promote future neovascularization (NV) and contraction.

199. b. Severe visual loss after vitrectomy seems more common in patients who are diabetic. Lens removal may increase the risk of rubeosis, although a concomitant retinal detachment (RD) may be the key factor prompting lensectomy.

200. a. Indications for emergent pars plana vitrectomy in the setting of globe penetration include retinal detachment (RD), intraocular foreign body, and endophthalmitis. Vitreous hemorrhage without RD may be observed or addressed 7 to 10 days after the initial repair of globe rupture.

201. c. Traumatic macular hole and subfoveal hemorrhage may lead to a permanent decrease in visual acuity. Commotio retinae and Valsalva retinopathy generally have a good visual prognosis.

202. e. Severe compressive injuries of the head and trunk or fractures may lead to patches of retinal whitening and hemorrhages known as “Purtscher’s retinopathy.” The disorder is thought to be mediated by air or fat embolism and consequent acute endothelial damage. Virtually identical fundus findings can result in patients with acute pancreatitis, systemic lupus erythematosus (SLE), dermatomyositis, or scleroderma.

203. d. This patient has a classic case of type II idiopathic juxtapfoveal telangiectasis (IJT). The right eye has developed the most serious complication of the disorder, subfoveal choroidal neovascularization (CNV). The changes in the left eye are more typical for the condition. Men and women are nearly equally affected. Type I IJT is unilateral and virtually always affects men. This form of the disease is probably a limited form of Coats’ disease, unlike type II. Although the angiogram suggests telangiectasis, or vascular dilation, histopathologic review of a clinical case showed marked thickening of the capillary walls, with narrowing of vascular lumina. Thus, the term “telangiectasis” is not accurate histologically. Type II IJT is seen more frequently among patients with mild type 2 diabetes mellitus. Interestingly, the vascular changes are strikingly similar to those of diabetic retinopathy but are limited to the parafoveal circulation. Furthermore, IJT features...
significant subretinal changes, including retinal pigment epithelium (RPE) hyperplasia and, as in this case, choroidal neovascularization (CNV). These are uncommon in uncomplicated diabetic retinopathy.

204. a. Figures a., b., c., and d., show a symmetric pigmented maculopathy with an evolving “bull’s-eye” appearance. In a young man with no history of treatment with hydroxychloroquine (Plaquenil) or chloroquine (Aralen), the differential diagnosis is limited to a few macular dystrophies. The two most commonly associated with “bull’s-eye” maculopathy are cone dystrophy and Stargardt’s disease (juvenile macular degeneration). The fluorescein shows the “dark choroid” nearly pathognomonic for Stargardt’s disease. This is usually inherited on an autosomal-recessive basis, meaning his parents are likely carriers with no detectable ophthalmoscopic changes. Electoretinogram (ERG) and visual fields are typically preserved or minimally affected. The white flecks seen in the periphery of some affected patients lead most experts to believe the disorder is on a pathophysiologic or genetic spectrum with fundus flavomaculatus. Typically, both eyes of affected patients stabilize at approximately 20/200 visual acuity.

205. e. The patient’s history and clinical findings are classic for an unusual paraneoplastic syndrome, bilateral diffuse uveal melanocytic proliferation. The syndrome is seen in patients with cancer, often undiagnosed. In men, lung and colon primaries predominate. In women, neoplasms of the reproductive system (e.g., ovarian and uterine) have been reported. The hallmarks of the syndrome, as defined by Gass, include focal red patches at the level of the retinal pigment epithelium (RPE) in the posterior pole, which hyperfluoresce; multiple pigmented and nonpigmented melanocytic “tumors” as well as diffuse proliferation of choroidal melanocytes; exudative retinal detachment (RD); and rapidly developing cataract. Visual loss is initially secondary to cataract and may precede the diagnosis of the cancer. Subsequent photoreceptor loss leads to inexorable, severe visual loss. This is reflected in the electoretinogram (ERG) as a rod degeneration more than a cone degeneration (scotopic greater than photopic loss). The underlying cause is not known, but is felt to be caused by hormonal effects of the primary carcinoma on preexistent uveal nevus cells. The angiogram shows multiple window defects, reflecting widespread RPE damage corresponding to the red patches and some of the melanocytic deposits. Unfortunately, no treatment is effective at stopping the visual decline. Because there is no serologic test, the diagnosis is made on a completely clinical basis.

206. c. Only patients meeting these criteria were entered into the study. Therefore, the results of the study are strictly applicable only to patients meeting similar criteria.

207. e. The choice of parenteral antibiotics was controversial. Many surgeons outside the study group felt that intravenous vancomycin would have been a better choice for Gram-positive coverage than amikacin, which leaves some staphylococcal species poorly covered. Thus, some retina specialists continue to advocate the use of intravenous vancomycin. The study clearly demonstrated that neither amikacin nor cefazidime offer any therapeutic advantage.

208. e. If you answered “a,” give yourself a half credit. One of the little details about the Endophthalmitis-Vitreectomy Study (EVS), which is usually overlooked, is that randomization typically took place in the operating room. Patients randomized to “tap” actually underwent a limited vitrectomy. That is, the vitreous sample was collected with a typical guillotine cutter run by a posterior segment vitrectomy machine, at parameters identical to standard vitrectomy (400 to 600 cuts per minute, suction 100 to 200 mm Hg). A true vitreous tap, typically performed in an office setting, involves no mechanized cutting; rather, a vitreous biopsy sample is obtained with a larger bore needle and active suction without any cutting. The risk of peripheral retinal breaks with the latter technique is higher, but the EVS did not specifically evaluate this method of obtaining a vitreous sample. Thereby, the EVS did document that for patients with acuity of hand motions or who are better at presentation, an extensive vitrectomy is no better than a limited vitrectomy (versus a true vitreous tap, with a needle only).

209. c. According to the Eye Disease Case-Control Study, elevated erythrocyte sedimentation rate is a risk for central retinal vein occlusion (CRVO) only among women.

210. e. Moderate alcohol consumption reduces the risk of central retinal vein occlusion (CRVO) for both women and men. Exogenous estrogen use also reduces risk.

211. c.

212. b.

213. d.

214. a. Clinical and angiographic edema did respond to grid laser treatment. There was no significant effect on long-term visual acuity, so the treatment is no longer recommended.

215. a. Subgroup analysis of outcomes by the Macular Photocoagulation Study evaluated outcome based on lesion size and presenting acuity. Age and gender were not analyzed. The ideal candidate has poor acuity (<20/125 for small lesions and <20/200 for medium lesions) and a small (<1 disc area) or a medium (1 to 2 disc areas) lesion.

216. b. The candidate who enjoys no treatment benefit whatsoever has a large lesion (>2 disc areas) with acuity better than 20/200.

217. c.
219. **True.** Foveal (focal) electroretinographic (ERG) testing can be used to test patients with physical findings that do not correspond to the degree of visual loss.

220. **a.** Optical coherence tomography (OCT) only uses light waves to create images. Ultra-high resolution OCT uses a different laser to create images with much greater resolution.

221. **c.** Relapses of cytomegalovirus (CMV) are far too common. Unfortunately, none of the treatment options are viricidal for CMV, only virostatic.

222. **a.** The surface tension at a gas–aqueous interface is very high and responsible for the ability of a gas-fluid exchange to flatten a detached retina. The interfacial tension prevents the aqueous phase from reentering the subretinal space, unless the retinal breaks are very large or under severe traction. These factors can outweigh the beneficial effects of surface tension.

223. **c.** Interfacial surface tension between silicone oil and aqueous is very low. The high viscosity of oil makes it more difficult for the aqueous phase remaining in an eye to gain access to retinal breaks. The oil is also less likely to enter the subretinal space, once again because of its viscosity. Once aqueous reaches a break, however, the oil cannot prevent it from entering the subretinal space because there is no significant interfacial surface tension. Silicone oil also has lower solubility for various inflammatory and vasogenic mediators, which may beneficially “compartmentalize” the eye following complicated retinal surgery.

224. **c.** The high specific gravity of perfluorocarbon liquids renders them heavier than water and makes them a revolutionary addition to the vitreoretinal armamentarium. They can “steamroll” giant retinal tears flat, act as “third hands” in complicated surgery, and effect drainage of subretinal fluid through breaks in the far periphery, obviating the need for drainage retinotomies. They also have been used for repositioning dislocated lens implants and draining choroidal hemorrhage. As time passes, their utility is bound to increase. They are not tolerated by the retina for >24 to 48 hours (histopathology shows compression of the retina). They are not tolerated by anterior chamber structures at all. Liquids with higher vapor pressures (e.g., perfluoro-octane) may be safer to use because they will readily evaporate in the gas-filled eye.

225. **d.** Several case reports have documented significant deterioration in patients with central serous retinopathy (CSR) started or maintained on oral corticosteroids. The most common finding is dramatically increased exudation with the development of bilateral bullous retinal detachments (RDs). Fava beans should be avoided in patients with glucose-6-phosphate dehydrogenase (G6PD) deficiency because they can precipitate acute hemolysis.

226. **b.** The only factor independently predictive of bilateral choroidal neovascularization (CNV) is the presence of histo spots in the contralateral eye at the time of diagnosis of unilateral CNV.

227. **c.**

228. **e.**

229. **b.** The history and the fundus is consistent with a diagnosis of anterior ischemic optic neuropathy (AION), a condition presenting with acute painless visual loss and typically with optic disc edema and pallor. Anyone with AION should be ruled out for arteritic AION (AAION) secondary to giant cell arteritis (GCA), as this is a potentially fatal condition. The best initial way to rule out GCA is with an erythrocyte sedimentation rate (ESR) and congenital retinitis pigmentosa (CRP), as up to 20% of patients with AAION can present with no systemic symptoms (jaw claudication, scalp pain, fever, malaise, etc.). ESR and CRP (when both are abnormal) have a 97% specificity for AAION. The gold standard for diagnosing GCA is a temporal artery biopsy. Steroids can be initiated before the biopsy (which should be performed within 1 week).

230. **c.** The history and the fundus is consistent with a diagnosis of a macular epiretinal membrane. The chance of this condition being bilateral is approximately 15% to 20%. Most patients have visual acuities that are 20/50 or better, although they often complain of metamorphopsia. Intravitreal perfluoropropane (C₃F₇) will not help improve visual acuity. Instead, pars plana membranectomy is required to improve visual acuity.

231. **d.** The condition shown is X-linked juvenile retinoschisis, a condition that is inherited on an X-linked–recessive basis. Splitting of the retina occurs at the nerve fiber layer. Foveal schisis is shown in the figure is almost always observed. There is no leakage observed with fluorescein angiography (FA).

232. **a.** The condition shown is serpiginous chorioretinopathy, a bilateral condition that usually spreads outward from the optic nerve and/or macula in a serpentine fashion. Serpiginous chorioretinopathy is chronic and recurrent with poor visual prognosis and scotomata affecting the areas of involvement. Treatment with potent immunosuppressives may slow down the disease in some cases, but generally the visual prognosis is poor, especially with macular involvement.

233. **c.** The condition shown is asteroid hyalosis, a benign condition that is usually unilateral (three quarters of cases), with patients having good visual acuity. There is a clear association of this condition in patients with diabetes. It usually occurs in patients older than 50 years, and vitrectomy may sometimes be used to help visualize other retinal conditions (e.g., diabetic retinopathy), which are difficult to assess because of the asteroid hyalosis.
234. **c.** The condition shown is retinitis pigmentosa (RP). Although it can be inherited through several inheritance patterns, X-linked inheritance usually has the worst prognosis. Electroretinogram (ERG) should be performed on all suspected patients to confirm the diagnosis because the presence of bone spiculation and chorioretinal atrophy can occur in a variety of conditions, including severe uveitis, syphilis, and chloroquine toxicity.

235. **d.** The condition shown is Coats’ disease. Coats’ disease is not inherited and is generally unilateral. The diagnosis of Coats’ disease requires the presence of retinal telangiectasia (small anomalous vessels) as shown in the figure in the question. It can recur and must be watched carefully, with difficult cases requiring multiple photocoagulation or cryotherapy treatments. Retinal detachments (RDs) can be a late sequela of the disease. The yellow material is lipid exudate. Posterior neovascularization (NV) is not common. It can recur and must be watched carefully, with difficult cases requiring multiple photocoagulation or cryotherapy treatments. Retinal detachments (RDs) can be a late sequela of the disease. The yellow material is lipid exudate. Posterior neovascularization (NV) is not common.

236. **b.** The condition shown is Best’s disease (or vitelliform dystrophy). It is inherited in an autosomal-dominant fashion and is secondary to a mutation in the vitelliform macular dystrophy gene (VMD2) (bestrophin) on chromosome 11. Visual acuity in the “sunny side up” stage shown in the figure is generally good. The electroretinogram (ERG) is usually normal whereas the electrooculogram (EOG) is generally abnormal in Best’s disease.

237. **c.** The condition shown is a myelinated nerve fiber layer (MNFL), a benign condition. It has been associated with nystagmus and amblyopia, but retinal detachments (RDs) are not associated with MNFL.

238. **b.** The condition shown is a central retinal (or hemiretinal) artery occlusion, manifested by nerve fiber layer edema. Atherosclerotic emboli are often the cause, and giant cell arteritis (GCA) is discovered in approximately 5% of cases.

239. **d.** The figure demonstrates angiod streaks that can be associated with several diseases (think “PEPSI”—see answer 26). Up to half of all cases are idiopathic, however. Fluorescein angiogram (FA) generally shows a window defect representing the breaks in Bruch’s membrane, through which choroidal neovascularization (CNV) can migrate. Photodynamic therapy (PDT) has been recently shown to be of benefit in subfoveal CNV secondary to angiod streaks.

240. **c.** The condition shown is acute posterior multifocal placoid epitheliopathy (AMPEP). It is usually bilateral and has a good visual prognosis. Up to one half of patients have a viral prodrome. Fluorescein angiography (FA) often demonstrates early hypofluorescence (indicated blockage of the choroidal pattern) with late hyperfluorescence (as shown in the middle figure and in the right figure, respectively). Although AMPEP is generally benign, it can be rarely associated with a fatal cerebral vasculitis.

241. **c.** The figure shows vitreous hemorrhage overlying the fovea, with a likelihood of significantly decreased visual acuity. The most common cause of vitreous hemorrhage is diabetic retinopathy in adults. The Diabetic Retinopathy Vitrectomy Study (DRVS) demonstrated the benefit of early vitrectomy (within 1 to 6 months after vitreous hemorrhage) in patients with type 1 diabetes or those patients with vitreous hemorrhage and severe proliferative diabetic retinopathy. B-scan ultrasound is not necessary in patients for whom the cause of the vitreous hemorrhage is known.

242. **b.** The condition shown is toxoplasmic retinochoroiditis (a posterior staphyloma generally would not have satellite lesions). The lesions appear old in this patient and antibiotic therapy and steroids would be of no use (unless the condition recurs). Patients with acquired immunodeficiency syndrome (AIDS) presenting with toxoplasmic retinochoroiditis must obtain neuroimaging to rule out cerebral lesions. Folinic acid (not folic acid) is used in combination with pyrimethamine to protect against iatrogenic thrombocytopenia and leukopenia.

243. **c.** The condition shown is central serous chorioretinopathy (CSCR), a condition usually affecting men between the ages of 30 and 50, typically with type A personalities (e.g., the author’s). The “smokestack” presentation shown in these figures only occurs in approximately 10% of cases of CSCR. Most eyes with this condition recover visual acuity within 6 months. Early focal photocoagulation in this particular patient (a retinal surgeon) would be contraindicated, given the lesion’s proximity to the center of the macula and the potential for a permanent visual scotoma with focal photocoagulation. Waiting for spontaneous remission would be the most appropriate treatment option.

244. **b.** The condition shown is fundus albipunctatus. It can be differentiated from retinitis punctata albescens because of the normal vasculature (typically attenuated in retinitis punctata albescens, a retinitis pigmentosa [RP] variant). Familial drusen would appear similar, although the lesions would not be as uniform in size and would form grapelike clusters. Patients with this condition may have good visual acuity, and night blindness may be their only symptom.

245. **c.** On the basis of the history, the fundus appearance, and the fluorescein angiogram (FA), the patient most likely has Vogt-Koyanagi-Harada syndrome (VKH), which is a bilateral condition characterized by granulomatous panuveitis. VKH is a systemic condition with both neurologic and dermatologic manifestations. The FA clearly demonstrates multiple areas of hyperfluorescence with late leakage into the subretinal space. Sugiura sign, or perilimbal vitiligo, occurs in 75% of
cases. Given the lack of trauma and the overall health of the patient before the examination, both sympathetic ophtalmia and endophthalmitis are unlikely. VKH generally has a good prognosis when effectively and rapidly treated with steroids and immunosuppressives.

246. d. The condition shown is neuroretinitis, which, along with a painful lymphadenopathy, can be a manifestation of cat-scratch disease caused by *Bartonella henselae*. Neuroretinitis, however, can have a variety of different infectious and immunologic causes, and, consequently, a thorough history should be obtained from the patient. Both erythromycin and ciprofloxacin can be used to treat catscratch disease. The visual prognosis is generally good with the disease being self-limited.

247. b. The condition described is Bietti crystalline corneoretinal dystrophy, which is characterized by limbal corneal opacities and tapetoretinal dystrophy. Plaquenil retinopathy typically causes a “bull’s-eye” maculopathy. Synchysis scintillans is a condition of cholesterol crystals within the vitreous. Cystinosis typically causes corneal crystals. Fleck retina of Kandori is a disorder similar to fundus albipunctatus with characteristic night blindness.

248. e. The patient has Irvine-Gass syndrome (e.g., cystoid macular edema [CME] developing after cataract surgery). The incidence of Irvine-Gass is higher after intraoperative complications, but most cases spontaneously resolve. Peak incidence is at 6–10 weeks postoperatively.

### Suggested Readings


Questions

1. What is the definition of severe visual loss?
   a. vision 20/40 or worse.
   b. vision 20/200 or worse.
   c. vision worse than 20/400.
   d. hand motion vision or worse.
   e. no light perception.

2. What is the definition of profound visual loss on the basis of visual acuity?
   a. vision worse than 20/100.
   b. vision worse than 20/200.
   c. vision worse than 20/400.
   d. hand motion vision or worse.
   e. no light perception.

3. What is the definition of blindness as per World Health Organization (WHO) standards?
   a. vision 20/200 or worse.
   b. vision 20/400 or worse.
   c. vision worse than 20/400.
   d. vision worse than counting fingers (CF) at 3 feet.
   e. no light perception.

4. What is the World Health Organization (WHO) definition of low vision?
   a. vision worse than 20/60.
   b. vision worse than 20/100.
   c. vision worse than 20/200.
   d. vision worse than 20/400.
   e. no light perception.

5. What is the definition of legal blindness in the United States?
   a. vision worse than 20/100.
   b. vision 20/200 or worse.
   c. vision worse than 20/400.
   d. vision worse than counting fingers (CF) at 3 feet.
   e. no light perception.

6. Which of the following guidelines is not clearly delineated within the Declaration of Helsinki?
   a. Informed consent should be given voluntarily by a patient.
   b. A study participant has the right to withdraw from the study (unless the withdrawal will significantly affect the results of the study).
   c. Informed consent should be obtained before participation in the study.
   d. An institutional review board can evaluate a study and stop the study if the research is not being conducted according to the highest ethical standards.
   e. All of the above.

7. How many levels of evidence are used in evidence-based eye care?
   a. one.
   b. three.
   c. five.
   d. seven.
   e. nine.
Questions

8. Which of the following is false with regard to levels of evidence?
   a. Level 5 characterizes a case report.
   b. Level 4 characterizes a case series.
   c. Level 3 characterizes a nonrandomized clinical trial.
   d. Level 2 characterizes a randomized clinical trial (RCT) with a greater power than level 1.
   e. Level 1 characterizes an RCT with a power of >80%.

9. In which of the following periods was a randomized controlled clinical trial (RCT) first published?
   a. in the 1700s.
   b. in the 1850s.
   c. in the 1940s.
   d. in the 1980s.
   e. in the year 2002.

10. What is the definition of statistical power?
    a. the probability of obtaining a false-positive result from a randomized clinical trial (RCT).
    b. the probability that an RCT will find a statistically significant difference when a difference exists.
    c. the probability that an RCT will find a statistically significant difference when a difference does not exist.
    d. the probability that an RCT will not find a statistically significant difference when a difference does not exist.
    e. none of the above.

11. T or F Serious adverse events of investigational pharmaceuticals are often not reported in large randomized clinical trials (RCTs).

12. Which of the following is not a legitimate reason to stop a randomized clinical trial (RCT) that is in progress?
    a. when interim data indicates a treatment benefit beyond a reasonable doubt (p <0.001).
    b. when the treatment being studied is clearly found to be ineffective.
    c. when the treatment being studied is found to be potentially fatal.
    d. when the treatment being studied is found to be teratogenic.
    e. when the treatment being studied will not change clinical practice.

13. T or F The Consolidated Standards of Reporting Trials (CONSORT) statement provides a set of guidelines for literature reviews in journals.

14. What is one of the main reasons for inadequate metaanalysis?
    a. small sample size.
    b. small power.
    c. selection bias.
    d. large type I error.
    e. none of the above.

15. Which of the following is a type of observational study?
    a. prospective cohort study.
    b. case-control study.
    c. outcome study.
    d. case series.
    e. all of the above.

16. What is the most common cause of disability-adjusted life-years worldwide?
    a. communicable, perinatal/maternal, and nutritional disorders.
    b. injury and trauma.
    c. noncommunicable diseases.
    d. human immunodeficiency virus (HIV)/acquired immunodeficiency syndrome (AIDS).
    e. myocardial infarction (MI).

17. T or F Average uncorrected distance vision for a 50-year-old person is better than 20/20.

18. T or F The letters on a Snellen chart are equally spaced apart both horizontally and vertically.

19. Which of the following statements about prevalence is false?
    a. Prevalence identifies the presence of a disease at a specific point in time.
    b. Prevalence can give a researcher an estimate of the probability of developing the disease.
c. Prevalence can be assessed by examining cross-sectional studies.
d. Prevalence studies can provide valuable information about the impact that a particular disease may have on a specific population.
e. Prevalence can give a researcher an estimate of the magnitude of the disease.

20. Which of the following types of bias would occur if participants with a disease are more accurate historians than participants without a disease?
   a. nonresponse bias.
   b. selection bias.
   c. recall bias.
   d. observation bias.
   e. none of the above.

21. T or F Worldwide, more adults than children are blind.

22. Which of the following types of ophthalmic disease is/are the most common cause of pediatric blindness in the world?
   a. congenital glaucoma.
   b. cataracts.
   c. corneal scarring and retinal diseases.
   d. disorders of the entire globe.
   e. none of the above.

23. Which of the following geographic regions in the world has the highest prevalence of pediatric blindness?
   a. Africa.
   b. China.
   c. India.
   d. Russia.
   e. The Middle East.

24. T or F Lower socioeconomic status increases the incidence of ophthalmia neonatorum.

25. T or F Ethnic background can affect the incidence of ophthalmia neonatorum.

26. What is the most common etiology for ophthalmia neonatorum?
   a. Chlamydia trachomatis.
   b. Neisseria gonorrhea.
   c. Staphylococcus aureus.
   d. Streptococcus pneumoniae.
   e. herpes simplex virus (HSV).

27. Which of the following organisms typically causes the most severe complications of ophthalmia neonatorum?
   a. Chlamydia trachomatis.
   b. Neisseria gonorrhea.
   c. Staphylococcus aureus.
   d. Streptococcus pneumoniae.
   e. herpes simplex virus (HSV).

28. Retinopathy of prematurity (ROP) was previously known as:
   a. persistent fetal vasculature.
   b. familial exudative vitreoretinopathy.
   c. retrolental hyperplasia.
   d. septooptic dysplasia.
   e. none of the above.

29. In the United States, in which zone is retinopathy of prematurity (ROP) most observed?
   a. zone I.
   b. zone II.
   c. zone III.
   d. zone IV.
   e. zone V.

30. T or F The medication(s) needed for treatment of pediatric xerophthalmia is/are usually exorbitantly expensive.

31. What is the earliest symptom of xerophthalmia?
   a. dry eyes.
   b. night blindness.
   c. Bitot’s spots on the conjunctiva.
   d. “salt-and-pepper” retinopathy.
   e. corneal scarring.

32. Which viral infection can cause an increased risk of corneal xerophthalmia in a malnourished child?
   a. herpes virus.
   b. measles virus.
   c. smallpox.
   d. human immunodeficiency virus (HIV).
   e. mumps virus.
33. Which of the following statements about ocular trauma is true?
   a. A penetrating injury occurs when the globe is penetrated by a foreign body, which then exits 180 degrees from the site of entry.
   b. A contusion only occurs when the globe has ruptured.
   c. Lamellar lacerations are usually closed-globe injuries.
   d. A perforating injury usually occurs after an eyeball contusion.
   e. None of the above.

34. T or F Men have a much higher incidence of ocular trauma than women.

35. Approximately how many people in the world currently have profound visual loss?
   a. 1 million.
   b. 10 million.
   c. 45 million.
   d. 85 million.
   e. 125 million.

36. T or F The prevalence of severe visual loss is higher in the developing world.

37. T or F The most common cause of vision impairment in the world is cataract.

38. What is the most common irreversible cause of blindness in the United States in individuals aged 65 years and older?
   a. cataracts.
   b. age-related macular degeneration.
   c. diabetic retinopathy.
   d. refractive error.
   e. glaucoma.

39. What is the most common cause of blindness in the Middle East?
   a. glaucoma.
   b. cataracts.
   c. diabetic retinopathy.
   d. age-related macular degeneration.
   e. none of the above.

40. What percentage of global blindness is either avoidable or treatable?
   a. 20%.
   b. 30%.

41. Which of the following complications is typically not associated with high myopia?
   a. retinal detachment.
   b. choroidal neovascularization.
   c. posterior staphyloma.
   d. angle-closure glaucoma.
   e. degenerative changes in the vitreous.

42. T or F In the near future, refractive surgery will likely become the modality of choice for treatment of refractive error in developing countries.

43. What is the most common cause of blindness in the world?
   a. refractive error.
   b. cataracts.
   c. onchocerciasis.
   d. trachoma.
   e. none of the above.

44. T or F Cataract surgery is not cost effective, especially in the developing world; the long-term economic gain from cataract extraction does not outweigh the cost of doing surgery.

45. What is the most prevalent type of cataract in white individuals?
   a. cortical.
   b. posterior subcapsular.
   c. nuclear.
   d. anterior polar.
   e. lamellar.

46. How much more common is primary open angle glaucoma (POAG) in African Americans when compared to white Americans?
   a. twice as common.
   b. three to six times as common.
   c. 10 times as common.
   d. 20 times as common.
   e. 50 times as common.

47. Which of the following is not a risk factor for primary open angle glaucoma (POAG) in the African American population?
   a. a family history of POAG.
   b. myopia.
48. Which of the following nations has the highest incidence of primary angle-closure glaucoma?
   a. Singapore.
   b. United States.
   c. Finland.
   d. Japan.
   e. Israel.

49. T or F The fellow eye of an eye with angle closure should usually undergo prophylactic iridotomy.

50. The first large, multicenter, prospective randomized clinical trial (RCT) in ophthalmology was the:
   a. Diabetes Control and Complications Trial (DCCT).
   b. Diabetic Retinopathy Study (DRS).
   c. Early Treatment Diabetic Retinopathy Study (ETDRS).
   d. Diabetic Retinopathy Vitrectomy Study (DRVS).
   e. Central Vein Occlusion Study (CVOS).

51. T or F One of the methods to control trachoma is to ensure face cleanliness.

52. What age group has the highest prevalence of trachoma?
   a. infants.
   b. young children.
   c. teenagers.
   d. middle-aged adults.
   e. older people.

53. How many people in the world have ocular chlamydial infection?
   a. 10 million.
   b. 20 million.
   c. 30 million.
   d. 50 million.
   e. 150 million.

54. Which is the most common method by which trachoma is diagnosed?
   a. polymerase chain reaction (PCR) testing.
   b. microbial culturing.
   c. microscopic examination.
   d. clinical examination.
   e. none of the above.

55. Which of the following microbes most commonly cause microbial keratitis worldwide?
   a. Streptococcus pneumoniae.
   b. Streptococcus viridans.
   c. Pseudomonas aeruginosa.
   d. Fusarium.
   e. Bacillus cereus.

56. What percentage of individuals with human immunodeficiency virus (HIV) live in sub-Saharan Africa and South and Southeast Asia?
   a. 10%.
   b. 20%.
   c. 50%.
   d. 75%.
   e. >90%.

57. T or F Onchocerciasis vaccination has greatly reduced the incidence of ocular onchocerciasis.

58. What is the treatment of choice for onchocerciasis?
   a. suramin.
   b. diethylcarbamazine.
   c. ivermectin.
   d. all of the above.
   e. none of the above.

59. Cuban epidemic optic neuropathy (CEON) is similar to which of the following disorders in presentation?
   a. Leber’s hereditary optic neuropathy (LHON).
   b. morning glory disc anomaly.
   c. optic disc drusen.
   d. optic nerve gliomas.
   e. multiple sclerosis (MS).

60. What is the most common method of cataract extraction in the world?
   a. phacoemulsification.
   b. extracapsular cataract extraction.
   c. intracapsular cataract extraction.
   d. laser cataract removal.
   e. couching.
61. Which of the following statements about cataract surgery performed in developing countries is incorrect?
   a. Cataract surgery can cost as little as $11 per eye to the patient when performed.
   b. The postoperative infection rate at institutions such as the Aravind Eye Institute in India is comparable to rates in the United States.
   c. The surgical skills of most eye surgeons in developing countries are typically inferior to the skills of US surgeons.
   d. The equitable development model of eye care in developing countries aims to maximize quality and increase patient flow while still being profitable.
   e. Some institutions in developing countries manufacture their own intraocular lenses that are of consistent quality.

62. What is the best way to prevent retinopathy of prematurity (ROP)?
   a. regular screening of infants to identify ROP early.
   b. discontinuing supplemental oxygen use in infants as soon as medically possible.
   c. prophylactic laser treatment in infants with a birth weight <1500 g and who are <36 weeks’ gestation.
   d. prophylactic laser treatment in infants with a birth weight <2000 g and who are <36 weeks’ gestation.
   e. none of the above.

63. Which of the following surgeons was the earliest to perform cataract surgery?
   a. Sushruta.
   b. Kenneth M. Goins.
   c. Albrecht von Graefe.
   d. Ammar.
   e. Sir William Osler.

64. T or F Despite the efforts of the world ophthalmic community, the prevalence of blindness continues to rise.
12. Teratogenicity by itself is not necessarily a reason for a randomized clinical trial (RCT) to be stopped. All of the other answers are legitimate reasons to stop a randomized clinical trial (RCT) (many menopausal women do not care about teratogenicity).

13. The Consolidated Standards of Reporting Trials (CONSORT) statement, which is ascribed to by leading peer-reviewed journals, sets a high standard of quality in the reporting of randomized clinical trials (RCTs) in order to allow the reader to adequately judge the quality of the study.

14. Selection bias is one of the main problems with metaanalysis. Selection bias can be based on several factors, including language bias (e.g., only choosing articles published in English for the metaanalysis) and publication bias (e.g., only choosing articles from a particular set of journals).

15. All of the answers are types of observational studies—they usually lack most of the controls in a randomized clinical trial (RCT) and consequently can be confounded by several sources of bias.

16. Communicable, perinatal/maternal, and nutritional disorders account for >45% of all worldwide disability-adjusted life-years.

17. True. Average visual acuity is significantly better than 20/20 in individuals younger than 60 years.

18. False. This was one of the drawbacks of the original Snellen chart. Bailey and Lovie created a chart that had standard spacing. The Early Treatment Diabetic Retinopathy Study (ETDRS) is based on the Bailey and Lovie chart.

19. True. Incidence, not prevalence, will provide an estimate of the probability of developing the disease.

20. False. This is a classic example of recall bias. Individuals without a particular disease may not remember exposure to particular offending agents. This can lead to significant bias in a study.

21. True. Even large randomized clinical trials (RCTs) do not always report rare, serious adverse events. This is one of the drawbacks of RCTs.

22. True. Pediatric blindness most commonly occurs from corneal scarring and retinal diseases.

23. Africa has the most cases of pediatric blindness, affecting approximately half a million children.

24. True. If the mother has a history of lower socioeconomic status or a prior history of sexually transmitted diseases, then the risk of developing ophthalmia neonatorum is higher for the fetus.

25. False. Studies have shown that ethnicity does not affect the incidence of ophthalmia neonatorum, but socioeconomic status certainly does.

26. Chlamydia trachomatis is the most common cause of ophthalmia neonatorum.

27. Neisseria gonorrhoea can cause severe inflammation, hyperpurulence, endophthalmitis, and corneal perforation. It should be presumed in an infant with hyperpurulence until proven otherwise.

28. Retinopathy of prematurity (ROP) was formerly called retrolental fibroplasia in the 1940s.

29. Most retinopathy of prematurity (ROP) is observed in zone II. ROP is classified into three zones (I, II, and III). Note that this is not necessarily true in developing countries where ROP is often diagnosed at more advanced stages.

30. Children with xerophthalmia can be treated with high-dose oral vitamin A capsules, which are inexpensive (<$0.10 per dose).

31. True. Night blindness is the earliest manifestation of xerophthalmia.

32. The cataractic state associated with measles infection can lead to severe corneal xerophthalmia.

33. Lamellar lacerations are usually closed-globe injuries. A perforating injury involves two full-thickness lacerations (as with answer a.). A contusion occurs with blunt trauma when the globe has not ruptured.
Answers

34. True.
35. c. Worldwide, 135 million people have vision <20/60, and approximately 45 million have vision <20/400.
36. True. The prevalence of severe visual loss is significantly higher in the developing world because of lack of preventive measures and lack of adequate care.
37. False. Refractive error is the most common cause of vision impairment in the world.
38. b. Macular degeneration is the most common cause of blindness in US seniors.
39. b. Cataracts accounted for >50% of the etiology for blindness in one survey.
40. e. Eighty percent of global blindness either can be treated or avoided.
41. d. Angle-closure glaucoma is usually associated with hyperopia >2D.
42. False. Because of its current cost, refractive surgery will probably not replace the use of spectacles to treat refractive error in the developing world.
43. b. Approximately 15 million people in the world are blind from cataracts.
44. False. Cataract surgery is a very cost-effective intervention, and the economic benefits in productivity easily outweigh the cost of the surgery.
45. c. Nuclear cataracts are the most common form in white individuals.
46. b. Primary open angle glaucoma (POAG) is three to six times as common in African Americans than in white Americans.
47. d. The effect of body size as a risk factor for primary open angle glaucoma (POAG) is unclear. In fact, the Barbados Eye Study found that lean body mass may be a risk factor for POAG in the African American population.
48. a. The Singaporean Chinese community, in particular, has a very high incidence of primary angle-closure glaucoma.
49. True. There is clear evidence that fellow eyes that do not undergo prophylactic iridotomy have a high risk of developing angle closure.
50. b. The Diabetic Retinopathy Study (DRS) was a landmark trial demonstrating that laser photoagulation of the retina significantly reduces the risk of visual loss in patients with proliferative diabetic retinopathy.
51. True. The SAFE (surgery, antibiotics, face cleanliness, and environmental improvements) program is a program instituted by the World Health Organization (WHO) to reduce the incidence of trachoma.
52. b. Children aged 3 to 6 years have the highest prevalence of trachoma.
53. e. Worldwide, 150 million people are infected with ocular chlamydia.
54. d. Clinical (e.g., slit-lamp) examination is the most common method to diagnose trachoma. Polymerase chain reaction (PCR) and culturing are time consuming and not cost effective.
55. a. Streptococcus pneumoniae is one of the most common causes of microbial keratitis worldwide.
56. e. Human immunodeficiency virus (HIV) prevalence varies greatly within different geographic areas of the world.
57. False. No vaccination against onchocerciasis is currently available as of 2005.
58. c. Ivermectin is the treatment of choice, given its relatively fewer side effects, when compared to either suramin or diethylcarbamazine.
59. a. Cuban epidemic optic neuropathy (CEON) was similar to Leber’s hereditary optic neuropathy (LHON) in clinical presentation and was most likely caused by malnutrition and toxicity leading to profound visual loss. CEON primarily affected men and became an epidemic in Cuba, affecting approximately 50,000 individuals in 1993.
60. b. Extracapsular cataract extraction is the most common method to remove cataracts in the world. Although phacoemulsification surgery has become the method of choice in patients from the United States, patients from developing areas of the world are far more likely to undergo extracapsular cataract extraction.
61. c. The surgical skills of many eye surgeons in developing countries are quite outstanding and in no way are inferior to the skills of surgeons in the United States. The authors (RDJ and JCL) have had incredible experiences observing and performing surgery abroad and would encourage any ophthalmologist-in-training to consider exposure to international ophthalmic care at some point in their future.
62. e. There is currently no effective way to prevent retinopathy of prematurity (ROP). Regular screening is useful to provide treatment, which reduces the incidence of ROP progression. Unfortunately, a significant number of infants in developing countries are only diagnosed when their ROP is very advanced.
63. a. Sushruta, the first known surgeon, performed cataract surgery in India by couching. Couching was performed in India as early as 800 BC.
64. True. Unfortunately over the last 25 years, the prevalence of blindness has risen from an estimated 30 million individuals to 50 million today. A more concerted effort among ophthalmologists worldwide will be needed to eradicate blindness in the world.

Suggested Readings
