

PRACTICING OPHTHALMOLOGISTS  
CURRICULUM 2014-2016

# Core Ophthalmic Knowledge

\*\*\*

## **Practicing Ophthalmologists Curriculum Disclaimer and Limitation of Liability**

As a service to its members and American Board of Ophthalmology (ABO) diplomates, the American Academy of Ophthalmology has developed the Practicing Ophthalmologists Curriculum (POC) as a tool for members to prepare for the Maintenance of Certification (MOC) -related examinations. The Academy provides this material for educational purposes only.

The POC should not be deemed inclusive of all proper methods of care or exclusive of other methods of care reasonably directed at obtaining the best results. The physician must make the ultimate judgment about the propriety of the care of a particular patient in light of all the circumstances presented by that patient. **The Academy specifically disclaims any and all liability for injury or other damages of any kind, from negligence or otherwise, for any and all claims that may arise out of the use of any information contained herein.**

References to certain drugs, instruments, and other products in the POC are made for illustrative purposes only and are not intended to constitute an endorsement of such. Such material may include information on applications that are not considered community standard, that reflect indications not included in approved FDA labeling, or that are approved for use only in restricted research settings. The FDA has stated that it is the responsibility of the physician to determine the FDA status of each drug or device he or she wishes to use, and to use them with appropriate patient consent in compliance with applicable law.

The Practicing Ophthalmologists Curriculum is intended to be the basis for MOC examinations in 2014, 2015 and 2016. However, the Academy specifically disclaims any and all liability for any damages of any kind, for any and all claims that may arise out of the use of any information contained herein for the purposes of preparing for the examinations for MOC.

THE AMERICAN ACADEMY OF OPHTHALMOLOGY DOES NOT WARRANT OR GUARANTEE THAT USE OF THESE MATERIALS WILL LEAD TO ANY PARTICULAR RESULT FOR INDIVIDUALS TAKING THE MOC EXAMINATIONS. THE AMERICAN ACADEMY OF OPHTHALMOLOGY DISCLAIMS ALL DAMAGES, DIRECT, INDIRECT OR CONSEQUENTIAL RELATED TO THE POC.

**Any questions or concerns related to the relevance and validity of questions on the MOC examinations should be directed to the American Board of Ophthalmology.**

COPYRIGHT © 2014  
AMERICAN ACADEMY OF OPHTHALMOLOGY  
ALL RIGHTS RESERVED

## **Practicing Ophthalmologists Curriculum**

### **Authors and Financial Disclosures**

The Practicing Ophthalmologists Curriculum was developed by a group of dedicated ophthalmologists reflecting a diversity of background, training, practice type and geographic distribution.

Jeffrey A. Nerad, M.D., American Academy of Ophthalmology Secretary for Knowledge Base Development, serves as the overall project director for the acquisition and review of the topic outlines.

#### **Practicing Ophthalmologists Curriculum Panel**

Jeffrey A. Nerad, M.D., Chair  
Anthony J. Aldave, M.D.  
Jason Bacharach, M.D.  
James T. Banta, M.D.  
Christopher L. Blanton, M.D.  
Michael H. Goldstein, M.D.  
Richard J. Grostern, M.D.  
Jeffrey D. Henderer, M.D.  
Leon W. Herndon, M.D.  
Erich P. Horn, M.D.  
Matthew Dean Kay, M.D.  
Louise A. Mawn, M.D.

Ramana S. Moorthy, M.D.  
Christie L. Morse, M.D.  
Timothy W. Olsen, M.D.  
Rajesh K. Rajpal, M.D.  
Russell W. Read, M.D.  
Stuart R. Seiff, M.D.  
Brian D. Sippy, M.D.  
Mitchell B. Strominger, M.D.  
Roger E. Turbin

#### **Financial Disclosures**

The Academy's Board of Trustees has determined that a financial relationship should not restrict expert scientific clinical or non-clinical presentation or publication, provided appropriate disclosure of such relationship is made. All contributors to Academy educational activities must disclose significant financial relationships (defined below) to the Academy annually.

#### **Contributors who have disclosed financial relationships:**

Anthony J. Aldave, M.D.  
Consultant/Advisor: Allergan, Bausch Lomb  
Grant Support: National Eye Institute  
Lecture Fees: Alcon Laboratories, Inc., Allergan,  
Jason Bacharach, M.D.  
Consultant/Advisor: Allergan, Sucampo  
Lecture Fees: Allergan, Lumenis, Inc., Merck & Co., Inc.  
Christopher L. Blanton, M.D.  
Consultant/Advisor: Abbott Medical Optics, Allergan  
Lecture Fees: Abbott Medical Optics, Allergan  
Grant Support: Abbott Medical Optics  
Leon W. Herndon, M.D.  
Consultant/Advisor: Alcon Laboratories, Inc., Sight Sciences  
Lecture Fees: Alcon Laboratories, Inc.  
Louise A. Mawn, M.D.  
Grant Support: Research to Prevent Blindness

Christie L. Morse, M.D.

Grant Support: AAPOS EVP

Timothy W. Olsen, M.D.

Grant Support: Abraham J. and Phyllis Katz Foundation, National Eye Institute, Research to Prevent Blindness, The Fraser Parker Foundation, The R. Howard Dobbs Jr. Foundation

Patent/Royalty: A Tissue Support Structure, Scleral Depressor

Rajesh K. Rajpal, M.D.

Consultant/Advisor: Abbott Medical Optics, Alcon Laboratories, Inc., Allergan, Inc., Avedro, Bausch Lomb  
Equity Owner: Avedro

Lecture Fees: Alcon Laboratories, Inc., Allergan, Inc., Avedro, Bausch Lomb

Grant Support: Allergan, Inc., Avedro

Russell W. Read, M.D.

Consultant/Advisor: Allergan, Eleven Biotherapeutics, Santen, Inc.

Grant Support: Eyegate Pharmaceuticals, Inc., EyeSight Foundation of Alabama, International Retinal Research Foundation, Matthews Family Foundation, Research to Prevent Blindness

Brian D. Sippy, M.D.

Grant Support: Regeneron, Inc.

**Contributors who state they have no significant financial relationships to disclose:**

James T. Banta, M.D.

Richard J. Grostern, M.D.

Jeffrey D. Henderer, M.D.

Erich P. Horn, M.D.

Matthew Dean Kay, M.D.

Ramana S. Moorthy, M.D.

Jeffrey A. Nerad, M.D.

Stuart R. Seiff, M.D.

Mitchell B. Strominger, M.D.

Roger E. Turbin, M.D.

**Background on Maintenance of Certification (MOC)**

Developed according to standards established by the American Board of Medical Specialties (ABMS), the umbrella organization of 24 medical specialty boards, Maintenance of Certification (MOC) is designed as a series of requirements for practicing ophthalmologists to complete over a 10-year period. MOC is currently open to all Board Certified ophthalmologists on a voluntary basis; time-limited certificate holders (ophthalmologists who were Board Certified after July 1, 1992) are required to participate in this process. All medical specialties participate in a similar process.

The roles of the American Board of Ophthalmology (ABO) and the American Academy of Ophthalmology relative to MOC follow their respective missions.

The mission of the American Board of Ophthalmology is to serve the public by improving the quality of ophthalmic practice through a process of certification and maintenance of certification that fosters excellence and encourages continual learning.

The mission of the American Academy of Ophthalmology is to advance the lifelong learning and professional interests of ophthalmologists to ensure that the public can obtain the best possible eye care.

The role of the ABO in the MOC process is to evaluate and to certify. The role of the Academy in this process is to provide resources and to educate.

### **Background on the Practicing Ophthalmologists Curriculum (POC)**

At the request of the ABO, the Academy developed the Practicing Ophthalmologists Curriculum (POC), a knowledge base that identifies and defines areas of knowledge important to the delivery of quality eye care as a basis for the content of examinations for the MOC process. The content in the POC is comprised of the information deemed as the most relevant clinical information for a practicing ophthalmologist.

The ABO has agreed that their Periodic Ophthalmic Review Test (PORT) and closed-book Demonstration of Ophthalmic Cognitive Knowledge (DOCK) examinations will be based on the POC. The ABO is solely responsible for creating the PORT and DOCK exams and for certifying MOC candidates. The Academy has developed study tools based on the POC to assist doctors preparing to meet these MOC requirements.

### **Organization of the POC**

The Practicing Ophthalmologists Curriculum comprises 10 practice emphasis areas (PEA), plus Core Ophthalmic Knowledge. The ABO has designated the following as practice emphasis areas:

- Comprehensive Ophthalmology
- Cataract/Anterior Segment
- Cornea/External Disease
- Glaucoma
- Neuro-Ophthalmology and Orbit
- Oculoplastics and Orbit
- Pediatric Ophthalmology/Strabismus
- Refractive Management/Intervention
- Retina/Vitreous
- Uveitis

In addition to two practice emphasis areas of choice, every diplomate sitting for the DOCK examination will be tested on Core Ophthalmic Knowledge. Core Ophthalmic Knowledge is defined as the fundamental knowledge every practicing ophthalmologist must have whatever their area of practice.

Each PEA is categorized into topics presented in an outline format for easier reading and understanding of the relevant information points by the reader. These outlines are based on a standard clinical diagnosis and treatment approach found in the Academy's Preferred Practice Patterns.

For each topic, there are Additional Resources that may contain journal citations and reference to textbooks. These resources are supplemental to the topic outline, and should not be necessary for MOC exam preparation purposes.

### **Creation of the POC**

The POC was developed by panels of practicing ophthalmologists in each of the ten practice emphasis areas. The panels reflect a diversity of background, training, practice type and geographic distribution, with more than 90 percent of the panel members being time-limited certificate holders.

The panels ranked clinical topics (diseases and procedures) in terms of clinical relevance to the subspecialist or comprehensive ophthalmologist. The panelists created outlines for the topics deemed Most Relevant, based on what an ophthalmologist in a specific practice emphasis area needs to know to provide competent, quality eye care (i.e., directly related to patient care). These outlines were reviewed by subspecialty societies and the American Board of Ophthalmology.

### **Revision Process**

The POC is intended to be revised every three years. The POC panels will consider new evidence in the peer-reviewed literature, as well as input from the subspecialty societies, the American Board of Ophthalmology and the Academy's Self-Assessment Committee, in revising and updating the POC.

Prior to a scheduled review the POC may be changed only under the following circumstances:

- A Level I (highest level of scientific evidence) randomized controlled trial indicates a major new therapeutic strategy
- The FDA issues a drug/device warning
- Industry issues a warning

\*\*\*

# Core Ophthalmic Knowledge

## Cataract/Anterior Segment

### Types of Cataracts

- 1. Adult cataract..... 17
- 2. Traumatic dislocation and subluxation..... 20

### Anesthesia/Infection Prophylaxis/Viscosurgical Devices

- 3. Anesthesia for cataract eye surgery ..... 22

### Surgery

- 4. Cataract surgery in the adult ..... 25

## Cornea/External Disease

### Anatomy and Basic Concepts

- 5. Anatomy of the cornea ..... 29
- 6. Universal precautions for minimizing transmission of bloodborne pathogens and surface infectious agents..... 30

### Diagnostic Tests

- 7. Anterior segment examination ..... 32
- 8. Tear film evaluation ..... 34
- 9. Corneal topography ..... 35

### Ocular Surface Disorders

- 10. Aqueous tear deficiency, Sjögren syndrome and Mucin deficiency..... 37
- 11. Hordeolum and chalazion ..... 41
- 12. Exposure keratopathy ..... 43
- 13. Recurrent corneal erosion ..... 46

### Infectious Diseases

- 14. Acute conjunctivitis..... 48



15. Chronic conjunctivitis.....	50
16. Herpes simplex virus keratitis .....	52
17. Varicella-zoster virus keratitis.....	54
18. Adenoviral conjunctivitis and keratoconjunctivitis.....	57
19. Blepharitis .....	61
20. Bacterial conjunctivitis.....	63
21. Bacterial keratitis .....	67
<b>Immune-Mediated Disorders</b>	
22. Allergic conjunctivitis .....	69
23. Episcleritis .....	71
<b>Corneal Dystrophies</b>	
24. Fuchs corneal dystrophy .....	73
25. Keratoconus .....	75
<b>Degenerative Disorders</b>	
26. Pterygium.....	77
<b>Toxic and Traumatic Injuries</b>	
27. Chemical (alkali or acid) injury of the conjunctiva and cornea.....	79
28. Toxic medication injury of the cornea (medicamentosa) .....	81
29. Subconjunctival hemorrhage .....	82
30. Conjunctival foreign body .....	84
31. Corneal foreign body .....	87
32. Traumatic corneal abrasion .....	88
33. Corneal laceration.....	90
34. Corneal perforation.....	92

## **Glaucoma**

## **Diagnostic Tests**

35. Intraocular pressure .....	94
36. Gonioscopy.....	97
37. Examination of the optic nerve.....	99
38. Standard automated static perimetry .....	102
39. Corneal pachymetry .....	104

## **Open-Angle Glaucomas**

40. Primary open-angle glaucoma .....	105
41. Normal tension glaucoma .....	109
42. Primary open-angle glaucoma suspect .....	111
43. Secondary open-angle glaucomas .....	113
44. Hyphema .....	119
45. Corticosteroid-induced glaucoma.....	121

## **Angle-Closure Glaucomas**

46. Acute angle-closure glaucoma .....	123
47. Neovascular glaucoma .....	125

## **Medical Management of Glaucoma**

48. Glaucoma medications .....	127
--------------------------------	-----

## **Neuro-ophthalmology and Orbit**

### **Diagnostic Tests and Pupils**

49. Visual field testing.....	137
50. Examination of the pupil.....	140
51. Horner syndrome .....	142
52. Neuroimaging.....	144

## **Optic Neuropathy**

53. Demyelinating optic neuritis .....	147
54. Anterior ischemic optic neuropathy .....	149
55. Giant cell arteritis.....	152
56. Papilledema.....	154
57. Optic atrophy .....	156

## **Anatomy**

58. Anatomy of the visual sensory system .....	158
--	-----

## **High Intracranial Pressure/Headache**

59. Headache .....	160
--------------------	-----

## **Motility**

60. Restrictive strabismus .....	163
61. Third (oculomotor) cranial nerve palsy/paresis.....	165
62. Fourth (trochlear) cranial nerve palsy/paresis.....	167
63. Abducens nerve palsy .....	168
64. Myasthenia gravis .....	170

## **Orbit**

65. Orbital tumor causing neuro-ophthalmic manifestations .....	172
66. Orbital hemorrhage .....	173

## **Oculoplastics and Orbit**

### **Anatomy**

67. Periorbital anatomy .....	175
-------------------------------	-----

### **Diagnostic Tests**

68. External examination of the adnexa and orbit .....	180
--	-----

## **Infectious/Inflammatory Disorders**

69. Preseptal/orbital cellulitis .....	183
--	-----

70. Thyroid eye disease (Graves ophthalmopathy, thyroid orbitopathy) .....	186
--	-----

## **Trauma**

71. Orbital fractures.....	189
----------------------------	-----

72. Periorbital soft tissue trauma .....	192
--	-----

## **Eyelid Disorders**

73. Neoplasms of the eyelid and ocular surface.....	195
---	-----

74. Ectropion .....	198
---------------------	-----

75. Entropion .....	200
---------------------	-----

76. Ptosis .....	202
------------------	-----

## **Periocular Malpositions/Involution**

77. Facial nerve palsy .....	205
------------------------------	-----

## **Disorders of the Nasolacrimal System**

78. Lacrimal system: evaluation of epiphora.....	208
--	-----

79. Acquired nasolacrimal duct obstructions and dacryocystitis in adults.....	211
---	-----

## **Pediatric Ophthalmology/Strabismus**

### **Diagnostic Tests**

80. Tests of ocular alignment.....	214
------------------------------------	-----

### **Amblyopia**

81. Amblyopia .....	216
---------------------	-----

### **Esodeviations**

82. Pseudoesotropia .....	218
---------------------------	-----

### **Infectious Diseases**

83. Acute neonatal conjunctivitis .....	219
---	-----

### **Eyelid disorders/lacrimal system**

84. Congenital nasolacrimal duct obstruction .....	221
--	-----

### **Glaucomas and Cataracts**

85. Congenital glaucoma.....	223
------------------------------	-----

86. Congenital cataracts.....	225
-------------------------------	-----

### **Ocular Tumors/Phakomatoses**

87. Rhabdomyosarcoma .....	227
----------------------------	-----

88. Retinoblastoma .....	229
--------------------------	-----

### **Ocular Trauma**

89. Shaken baby syndrome/non-accidental trauma.....	230
---	-----

## **Refractive Management/Intervention**

### **Diagnostic Tests**

90. Visual acuity measurement.....	232
------------------------------------	-----

91. Refraction: manifest and cycloplegic .....	235
--	-----

92. Lens notation and conversion.....	237
---------------------------------------	-----

### **Refractive Errors**

93. Myopia .....	238
------------------	-----

94. Hyperopia.....	240
--------------------	-----

95. Anisometropia.....	242
------------------------	-----

96. Aniseikonia .....	244
-----------------------	-----

97. Regular astigmatism.....	246
------------------------------	-----

98. Irregular astigmatism .....	247
---------------------------------	-----

99. Presbyopia.....	249
---------------------	-----

### **Contact Lenses/Spectacles**

100. Ocular surface problems related to contact lens wear .....	251
---	-----

101. Eyeglasses .....	253
-----------------------	-----

## **Vision Rehabilitation**

102. Visual disability/visual impairment.....	256
---	-----

## **Retina/Vitreous**

### **Anatomy**

103. Anatomy of the neurosensory retina.....	260
--	-----

### **Diagnostic Tests**

104. Fluorescein angiography .....	262
105. Indications for optical coherence tomography.....	264

### **Macular Diseases**

106. Age-related macular degeneration.....	266
107. Ocular histoplasmosis .....	269
108. Idiopathic central serous chorioretinopathy .....	271
109. Epiretinal membrane .....	273

### **Retinal Vascular Diseases**

110. Diabetic retinopathy .....	275
111. Branch retinal vein occlusion .....	278
112. Central retinal vein occlusion.....	280
113. Branch retinal artery occlusion.....	282
114. Central retinal artery occlusion.....	284
115. Cystoid macular edema .....	286

### **Diseases of the Vitreous**

116. Spontaneous vitreous hemorrhage .....	289
117. Posterior vitreous detachment.....	292

### **Peripheral Retinal Abnormalities**

118. Retinal tears .....	295
--------------------------	-----

119. Retinal detachment.....	298
------------------------------	-----

### **Posterior Segment Trauma**

120. Closed-globe blunt trauma .....	301
--------------------------------------	-----

121. Open-globe injury .....	304
------------------------------	-----

122. Intraocular foreign body .....	307
-------------------------------------	-----

123. Endophthalmitis .....	310
----------------------------	-----

### **Tumors**

124. Melanoma of the ciliary body or choroid .....	313
--	-----

### **Uveitis**

#### **Diagnostic Tests**

125. Clinical approach to uveitis .....	316
---	-----

#### **Non Infectious Uveitis**

126. Acute anterior uveitis.....	319
----------------------------------	-----

127. Persistent (chronic) iridocyclitis .....	322
---	-----

128. Juvenile idiopathic arthritis.....	324
---	-----

129. Sympathetic ophthalmia .....	326
-----------------------------------	-----

130. Scleritis .....	328
----------------------	-----

#### **Infectious Uveitis**

131. Herpes zoster (varicella-zoster virus) iritis.....	330
---	-----

132. Necrotizing herpetic retinitis: acute retinal necrosis.....	332
--	-----

133. Toxoplasmic retinochoroiditis.....	334
---	-----

#### **Masquerade Syndromes**

134. Masquerade syndromes .....	336
---------------------------------	-----

#### **Acquired Immune Deficiency Syndrome related disease**

135. Ocular manifestations of acquired immune deficiency syndrome.....	340
--	-----

## **Medical Therapy**

136. Cycloplegics in uveitis .....	342
137. Non-steroidal anti-inflammatory drugs .....	344
138. Corticosteroids.....	346

## **Additional Core Topics**

### **Ethics**

139. Unique competence of the ophthalmologist.....	349
140. Assimilating new technology .....	351
141. The impaired ophthalmologist.....	353
142. Providing appropriate care .....	355
143. Informed consent.....	357
144. Confidentiality.....	359
145. Clinical investigation .....	360
146. Conflicts of interest.....	363
147. Expert witness testimony.....	365

### **Patient Safety and Risk Management**

148. Patient safety .....	367
149. Risk management.....	370



# Adult cataract

## Cortical cataract

### I. Describe the approach to establishing the diagnosis

#### A. Describe the etiology of this disease

1. Opacification of the cortical lens fibers
2. Swelling of the lens cortex creates intumescent cataract

#### B. List the pertinent elements of the history

1. Progressive loss of vision, often rapid
2. Glare
3. Monocular diplopia

#### C. Describe pertinent clinical features

1. Initially vacuoles and water cleft in the lens cortex
2. Wedge shaped opacities or cortical spokes
3. Progresses to form white intumescent cortical cataract

### II. Define the risk factors

- A. Smoking
- B. UV light exposure
- C. Diabetes mellitus
- D. Poor nutrition
- E. Trauma

## Nuclear cataract

### I. Describe the approach to establishing the diagnosis

#### A. Describe the etiology of this disease

1. Hardening and yellowing of the central lens
2. Advanced nuclear sclerosis leads to dense brunescent nucleus

#### B. List the pertinent elements of the history

1. Gradual progressive loss of vision

2. Glare
3. Monocular diplopia
4. Decreased color discrimination

**C. Describe pertinent clinical features**

1. Central yellow to brown discoloration of the lens
2. Myopic shift
3. Bilateral (typically)

## **II. Define the risk factors**

- A. High myopia**
- B. Malnutrition**
- C. Smoking**
- D. Intraocular surgery**
- E. Diabetes mellitus**
- F. Ultraviolet (UV) light exposure**
- G. Trauma - asymmetric cases**

Posterior subcapsular cataract

## **I. Describe the approach to establishing the diagnosis**

**A. Describe the etiology of this disease**

1. Axial opacity of the posterior cortical material

**B. List the pertinent elements of the history**

1. Progressive loss of vision, sometimes rapid
2. Glare
3. Monocular diplopia
4. Near vision worse than distance vision
5. Vision in bright light worse than dim light

**C. Describe pertinent clinical features**

1. Central opacity of the posterior cortical fibers
2. Glare testing
  - a. Posterior subcapsular cataract (PSC) often induces glare disability while good visual acuity is preserved until cataract is very advanced

## **II. Define the risk factors**

- A. Diabetes mellitus**
- B. Radiation**
- C. Corticosteroids**
- D. Uveitis,**
- E. Smoking**
- F. Trauma**
- G. Intraocular surgery (particularly vitrectomy)**
- H. Family history**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 11: Lens and Cataract, 2013-2014.
2. AAO, Cataract Surgery and Intraocular Lenses, 2nd edition, p.16-18.

# Traumatic dislocation and subluxation

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Displacement of the lens
  - a. Subluxated - partially dislocated
  - b. Luxated with total zonular dehiscence
  - c. Rapid expansion of equatorial globe loosens zonular fibers

### B. List the pertinent elements of the history

1. Decreased near and distance vision
2. Monocular diplopia
3. Glare
4. Trauma
5. Family history

### C. Describe pertinent clinical features

1. Subluxation or total luxation of the lens
2. Phacodonesis
3. Lenticular astigmatism
4. Iridodonesis
5. Impaired accommodation
6. Intraocular pressure (IOP) measurement
7. Vitreous prolapse into the anterior chamber

## II. List the most common or critical entities in the differential diagnosis

### A. Primarily ocular

1. Pseudoexfoliation
2. Simple ectopia lentis
3. Ectopia lentis et pupillae

### B. Systemic

1. Marfan syndrome
2. Weil-Marchesani syndrome

3. Homocystinuria
4. Spherophakia

### **III. Describe patient management in terms of treatment and follow-up**

#### **A. Describe medical therapy**

1. Eyeglasses or contact lenses for subluxated lenses
2. Aphakic contact lens with marked subluxation or luxation

#### **B. Describe surgical therapy**

1. Lensectomy, with or without vitrectomy for selected cases (vitrectomy is not always needed when a dislocated lens is surgically removed)

### **IV. Describe disease-related complications**

#### **A. Pupillary block glaucoma**

#### **B. Traumatic angle-recession glaucoma**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 11: Lens and Cataract, 2013-2014.

# Anesthesia for cataract eye surgery

## I. Introduction

- A. The basis for any form of ocular anesthesia is to provide comfort and safety for the patient; monitoring of vital signs and administration of varying degrees of sedation, as needed, are performed by appropriately trained personnel. Presurgical anesthetic evaluation should include tests appropriate for the age and health of the patient and for the intended anesthesia. Components of local ocular anesthesia are**

1. Ocular anesthesia
2. Akinesia of the extraocular muscles
3. Akinesia of the orbicularis oculi muscle

## II. Methods/Materials

- A. Topical tetracaine, lidocaine, bupivacaine, proparacaine with or without intracameral anesthesia (non-preserved lidocaine HCL 1%)**

1. Topical anesthesia with or without adjunctive intracameral augmentation is reserved for procedures, typically cataract surgery, that are of short duration, provide a self-sealing incision, and are generally free of complications
2. Surgeons generally communicate with patients during surgery, requiring that the surgeon and patient speak in the same language and that the patient is capable of hearing and responding to the instructions of the surgeon
3. The surgeon is responsible for explaining the nature of the procedure to the patient and for giving the patient an advance understanding of the surgical experience, including the visual experiences induced by the microscope light
4. Topical anesthesia with or without intracameral augmentation is contraindicated in patients with coarse nystagmus, total deafness, severe claustrophobia, and those that require lengthy and complex procedures
5. Advantages of topical intracameral anesthesia
  - a. Avoid orbital and systemic risks of injection anesthesia
  - b. Avoid damage to globe and optic nerve that may occur with injection anesthesia
  - c. Avoid cosmetic blemish of lid ecchymoses and subconjunctival hemorrhage
  - d. Early return of visual function after surgery
6. Disadvantages of topical intracameral anesthesia
  - a. Globe movement by patient may be unexpected and undesirable
  - b. Patient more aware of surgical events and may sense more discomfort than with other forms of anesthesia

- c. Possible dilutional/labeling errors with corneal endothelial toxicity

**B. Injection methods for ocular local anesthesia may be employed as a matter of surgeon preference, but are also useful in cases that are likely to be prolonged, complex or complicated**

1. All sharp needle injection methods have the following risks:
  - a. Lid ecchymoses
  - b. Orbital hemorrhage
  - c. Globe/orbital injury
  - d. Optic nerve injury
  - e. Transient amaurosis
  - f. Extraocular muscle damage and diplopia
  - g. Systemic and central nervous system (CNS) complications including apnea and death
2. Agents include lidocaine, mepivacaine and bupivacaine alone or in combination and with or without additional epinephrine
  - a. Hyaluronidase is a useful spreading agent
3. Retrobulbar
  - a. A relatively long and sharp needle is intended to penetrate the "muscle cone" and provide rapid onset ocular anesthesia, ocular akinesia and variable amaurosis
  - b. Typically it is administered in the lower lid at the junction of the outer third and inner two thirds of the eyelid
  - c. Advantages
    - i. Profound and rapid onset which will persist for several hours, varying with the anesthetic agent
  - d. Disadvantages
    - i. Risks associated with sharp needle injections
    - ii. Inability to use the eye for several hours after surgery, as patching is required until the effects have ameliorated
    - iii. Elongated eyes are at particular risk for injury with this form of anesthesia
    - iv. Generally requires sedation and monitoring for best case administration
4. Peribulbar
  - a. Similar to the retrobulbar method; however the intent is to inject the anesthetic agent outside the muscle cone in an attempt to reduce the risks of optic nerve injury, orbital hemorrhage, and systemic or CNS complications
  - b. Administered by injection with a shorter needle than that used for retrobulbar injection
  - c. Generally, the onset of action is considerably slower than for retrobulbar injections
  - d. Advantages

- i. Purported increased safety, although orbital, ocular, and optic nerve injuries have all been reported
- e. Disadvantages
  - i. Slower onset of action
  - ii. Need for supplementation
- 5. Sub-Tenons infiltration
  - a. In an attempt to eliminate the risks of blindly passed sharp needles in the orbit, anesthetic agents are administered directly into the posterior subconjunctival and Sub-Tenons space with a specifically designed blunt cannula after incising the conjunctiva
  - b. Advantages
    - i. Less anesthetic agent is required
    - ii. Onset is rapid
    - iii. The blunt nature of the cannula provides a safety margin when compared with sharp needle orbital injections
    - iv. IV sedation is generally not necessary
  - c. Disadvantages
    - i. Subconjunctival hemorrhage is common with this method
    - ii. Prior conjunctival scarring may contraindicate its use
  - d. It is particularly valuable for those patients who are at risk for injection anesthesia, high myopes as an example
  - e. This method may be used as primary strategy or as a supplement to topical anesthesia during surgery should conditions dictate, and as long as the incisions are sealed

### **C. Orbicularis akinesia**

- 1. Can be achieved by direct injection of the eyelid muscles or by achieving cranial nerve (CN) VII nerve blockade near the stylomastoid foramen or the tragus

### **D. General anesthesia**

- 1. In view of greater morbidity, mortality and expenses, general anesthesia is reserved for those patients who cannot cooperate for any form of local anesthesia
- 2. Care must be taken to assure that the patient's anesthesia and akinesia will remain adequate during the entire procedure

### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course, Section 11: Lens and Cataract, 2013-2014.



# Cataract surgery in the adult

## I. Define the condition

- A. A cataract is a progressive opacification or discoloration of the normally clear and colorless crystalline lens**
- B. Ocular examination should determine**
  - 1. The effect of cataract formation on visual function
  - 2. Appropriate optical correction (refraction)
  - 3. The type and extent of cataract formation
  - 4. The presence of contributory ocular co-morbidities such as macular degeneration, diabetic retinopathy, or optic neuropathy
  - 5. The prognosis for improved visual function should cataract surgery be contemplated
- C. Although the refractive state of the eye may change periodically with the evolution of a cataract, and eyeglass or contact lenses should be updated as needed, eventually the value of such changes will be limited by concomitant reduction in visual function and cataract surgery will be indicated**

## II. Indications for cataract surgical management

- A. Functional impairment in the patient attributable to a reduction in overall visual function secondary to cataract formation or significant lens subluxation in one or both eyes**
  - 1. The visual deficit is considered by the patient to be inconsistent with his/her visual needs, and the examination of the patient suggests that cataract surgery has a strong likelihood of restoring visual function and reducing his/her functional impairment
  - 2. Evaluation consists of a complete eye examination and use of ancillary tests as necessary
    - a. Snellen visual acuity
    - b. Glare disability
- B. Indication for second eye surgery is the same as for the first eye**
- C. Significant anisometropia**
- D. Opacification of crystalline lens sufficient to interfere with evaluation and/or treatment of posterior segment disorders in which there is likelihood for improved visual function**
- E. Lens induced glaucoma/inflammation**
  - 1. Phacomorphic
  - 2. Narrow angle glaucoma
  - 3. Phacolytic
- F. Subluxation of cataractous or clear crystalline lens sufficient to reduce visual function which cannot be adequately aided by eyeglasses or contact lenses**

### **III. Contraindications**

- A. Patient not desirous of improvement in visual function**
- B. Patient is medically or mentally unstable for elective surgery**
- C. Patient/provider not available for postoperative examinations and follow up care**

### **IV. Methodology**

#### **A. Infection prophylaxis**

1. Patient's periocular flora is most common source of microbial contamination in infectious endophthalmitis
  - a. Careful draping of lid margins and eyelashes is appropriate
2. Povidone iodine is antiseptic agent of choice for the lid surfaces and the conjunctival cul-de-sac
3. Topical, intracameral, or subconjunctival antibiotics are commonly used for infection prophylaxis. Efficacy of topical or subconjunctival antibiotics has not been proven by any randomized study

#### **B. Anesthesia (See Anesthesia for cataract eye surgery)**

#### **C. Incision**

1. Size must allow for nucleus extraction and/or IOL insertion
2. Usually placed superior or temporal
3. Location may be clear corneal, limbal, or scleral (pocket)
4. Smaller incision permits faster visual and physical rehabilitation, and is associated with less surgically-induced astigmatism

#### **D. Anterior capsulotomy**

1. Capsulorrhexis or can-opener

#### **E. Removal of the lens nucleus - options**

1. Ultrasonic fragmentation (phacoemulsification)
2. Manual large incision extracapsular cataract extraction (ECCE)
3. Intracapsular cataract extraction (ICCE)
4. Pars plana lensectomy

#### **F. Intraocular lens (IOL) implantation**

1. Posterior chamber IOL
  - a. Placed behind the iris
    - i. Preferred placement inside capsular bag
    - ii. May be placed in ciliary sulcus
    - iii. May be suture fixated to sclera or iris in absence of capsular support

- b. Rigid - polymethylmethacrylate (PMMA)
  - c. Foldable - allows implantation through smaller incision
    - i. Silicone or acrylic material
    - ii. Implanted with injector or folding forceps
  - d. May be monofocal, multifocal, or accommodating
- 2. Anterior chamber IOL
  - a. Angle-supported, placed anterior to iris
  - b. Used in the absence of capsular support
  - c. Made of polymethylmethacrylate (PMMA)

**G. Incision closure**

- 1. Watertight closure
- 2. May be sutured or sutureless, with number of sutures depending upon incision architecture and size

## **V. Intraoperative complications of cataract surgery**

- A. Posterior capsular rupture**
- B. Zonular disinsertion**
- C. Loss of vitreous**
- D. Retained lens material**
  - 1. In anterior segment
  - 2. In posterior segment
- E. Iris/pupil deformity**
- F. Surgical trauma - hyphema**
- G. Retrobulbar hemorrhage**
- H. Choroidal effusion**
- I. Suprachoroidal hemorrhage**
- J. Introduction of microorganisms, toxic substances or unintended inert foreign material into the eye**

## **VI. Postoperative complications of cataract surgery**

- A. Postoperative elevated intraocular pressure**
- B. Endophthalmitis**
  - 1. Increased risk with vitreous loss (10x) and prolonged surgery
- C. Retinal detachment**

1. Increased risk with vitreous loss and in young highly myopic patients

**D. Cystoid macular edema**

1. Increased risk with vitreous loss
2. Often treated prophylactically with non-steroidal anti-inflammatory drops

**E. IOL decentration and dislocation**

**F. Wound leak**

**G. Posterior capsular opacification**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 11: Lens and Cataract, 2013-2014.
2. AAO, Preferred Practice Pattern Committee, Cataract and Anterior Segment Panel. Cataract in the Adult Eye Preferred Practice Pattern, 2006.

# Anatomy of the cornea

## I. Describe relevant aspects of corneal anatomy

**A. Diameter: 11-12mm horizontally, 10-11mm vertically**

**B. Epithelium**

1. Thickness: 50 microns
2. Stratified squamous epithelial cells
3. Limbal stem cells (found in palisades of Vogt) are source of continuous proliferating basal epithelial cells
4. Basement membrane is secreted by basal epithelial cells

**C. Bowman layer: acellular compact layer of anterior stroma (8-12 microns thick)**

**D. Stroma**

1. Made up of regularly arranged flattened collagen lamellae and proteoglycans synthesized by keratocytes
2. Water content 78%

**E. Descemet membrane**

1. Basement membrane secreted by the corneal endothelium
2. Increases in thickness from 3 microns at birth to 10-12 microns in adults

**F. Endothelium**

1. Closely interdigitated cells arranged in a mosaic pattern of mostly hexagonal shapes
2. Cell density is typically 2400-3200 cells/mm<sup>2</sup> in adults
3. Human endothelial cells do not proliferate in vivo
4. Cell loss results in enlargement and spread of neighboring cells to cover the defective area
5. Pump function is critical to keep cornea compact and transparent. Both Na<sup>+</sup> K<sup>+</sup> ATPase and carbonic anhydrase are important in this process

## Additional Resources

1. AAO, Basic and Clinical Science Course. External Disease and Cornea: Section 8, 2013-2014.

# Universal precautions for minimizing transmission of bloodborne pathogens and surface infectious agents

## I. Routine ophthalmic office visits

### A. Hand hygiene

1. Reduces risk of transmitting infection during examinations
2. Perform between patient exams and after procedure involving contact with tears, even after using gloves because gloves can become perforated
3. Methods
  - a. Alcohol-based hand rubs
  - b. Soap and water, with complete drying

### B. Examination safeguards

1. Use disposable gloves if blood or blood-contaminated fluid is present
2. Use disposable gloves if there are open sores on hands
3. Change gloves and wash hands after contact with each patient

### C. Eyedropper bottles

1. Avoid direct contact of dropper tip with skin, lashes, tears or conjunctiva
2. Discard bottle if tip does contact the ocular surface
3. Discard after expiration date

### D. Disinfection of instruments

1. Tonometer tip, goniolens, and lens used for laser treatment
  - a. Wipe clean and disinfect with diluted bleach, 3% hydrogen peroxide, 70% ethanol, or 70% isopropanol for at least 5 minutes
2. Trial-fitting contact lens
  - a. Use hydrogen peroxide-containing or chlorhexidine-containing disinfection system for hard or rigid gas-permeable lens
  - b. Use hydrogen peroxide-containing or heat disinfection system for soft contact lens

## II. Procedures

### A. Handling of sharp instruments

1. Measures to prevent accidental needle-stick injury

- a. Do not recap needles
- b. Do not bend or break needles by hand
- c. Do not remove needles from disposable syringes
- d. Dispose contaminated sharp objects in puncture-resistant container
- e. Use needles with automatic retraction or safety cover if available

**B. IV injections**

- 1. Individuals who may come in contact with blood should wear gloves and if splashing is anticipated, they should wear protective eyewear

### **III. Ophthalmic surgery**

**A. Handling of instruments**

- 1. Manipulate suturing needles with forceps or needle holders, rather than by gloved fingers
- 2. If an instrument punctures a glove or skin, remove from the operative field and sterilize it
- 3. If the physician or medical personnel has a skin puncture:
  - a. The individual should notify appropriate personnel in operating room
  - b. Notify employee health service or infectious disease service to help coordinate patient testing, evaluation of disease transmission, and possible prophylactic medications.
  - c. Cleanse the wound with antiseptic solution and bandage
  - d. Rescrub and reglove

### **IV. Management of exposures of health care personnel**

- A. Employers subject to Occupational Safety and Health Administration (OSHA) regulations are required to make available the hepatitis B vaccine to all employees who have occupational exposure**
- B. Employers subject to OSHA regulations are required to establish exposure control plans, which include post-exposure follow-up and incident reporting**

**Additional Resources**

- 1. Infection Prevention in Eye Care Services and Operating Areas and Operating Rooms. American Academy of Ophthalmology, San Francisco, August 2012 <http://one.aao.org/clinical-statement/infection-prevention-in-eye-care-services-operatin> (<http://one.aao.org/clinical-statement/infection-prevention-in-eye-care-services-operatin>)
- 2. Centers for Disease Control and Prevention: Guidelines for Hand Hygiene in Health Care Settings. October 25, 2002. Accessed on 1/31/05: <http://www.cdc.gov/mmwr/preview/mmwrhtml/rr5116a1.htm> (<http://www.cdc.gov/mmwr/preview/mmwrhtml/rr5116a1.htm>)

# Anterior segment examination

## I. External examination

- A. Observation in ambient light
- B. Focal illumination, such as with illuminator
- C. Palpation of the eyelid and ocular adnexa, to evaluate lesion
- D. Palpation to detect enlarged lymph node

## II. Slit-lamp biomicroscopy

### A. Instrumentation

- 1. Viewing arm
  - a. Magnification depends on eyepieces (oculars) and objective lens setting
  - b. Eyepieces should be adjusted to ensure clear focus of an object at the point of coaxial alignment
- 2. Illuminating arm with slit beam
  - a. Light beam can be varied in length, width, and orientation
  - b. Light filters include neutral gray filter, cobalt blue filter, and red-free filter
  - c. Illumination arm is linked with the viewing arm to give a parfocal and isocentric light beam
- 3. Instrument base and patient positioning frame

### B. Illumination methods

- 1. Diffuse illumination
  - a. Broad or diffuse light beam helps to give overview of the eye
- 2. Direct focal illumination and slit illumination
  - a. Narrow slit beam illuminates an optical section of the cornea and anterior chamber
- 3. Indirect illumination
  - a. Light beam directed toward a different but adjacent site as the viewing arm's direction
  - b. Helps to highlight abnormalities by light scattering and by internal reflection
- 4. Retroillumination
  - a. Helps to highlight darkened abnormalities against an illuminated background, or to highlight an illuminated abnormality against a darkened background

## III. Staining of the ocular surface



**A. Fluorescein**

1. Water-soluble dye that pools within tear film and can stain ocular tissues and compartments
2. Visibility enhanced using cobalt blue filter
3. Stained epithelial defect

**B. Rose Bengal (or Lissamine Green)**

1. Helps to highlight epithelial abnormalities of the corneal and conjunctival surface
2. May be observed using white light
3. Red-free filter may enhance visibility of staining with Rose Bengal
4. Abnormal epithelial cells devoid of mucin layer e.g., desiccated, keratinized or dysplastic epithelial cells

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Tear film evaluation

## I. Describe the instrumentation and technique

- A. Observe amount and quality of the precorneal tear film and tear meniscus**
- B. Apply stain such as fluorescein or Rose Bengal and observe pattern and extent of ocular surface staining**
- C. Tear break-up time**
  - 1. Time from the last blink until the tear film thins and "breaks up"
  - 2. Normally greater than 10 seconds
  - 3. Low value suggests abnormal tear film
- D. Schirmer test**
  - 1. Schirmer strip or thread is placed across the lower lid
  - 2. After 5 minutes, the strip is removed and the amount of wetting measured
  - 3. Low value suggests aqueous tear deficiency
  - 4. Test may be performed without a topical anesthetic; testing with anesthetic aims to estimate basal tear secretion

## Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
- 2. AAO, Focal Points: Corneal Complications of Contact Lens, Module #2, 1993.

# Corneal topography

## I. List the indications/contraindications

### A. Indications

1. Preoperative management
  - a. Refractive surgery
  - b. Corneal surgery
2. Postoperative management
  - a. Refractive surgery
  - b. Penetrating keratoplasty
  - c. Post-cataract surgery astigmatism
3. Evaluation of corneal ectasia, including keratoconus
4. Irregular astigmatism
5. Evaluation of cornea scars (trauma or previous infection)
6. Contact lens fitting
7. Evaluation of unexplained decreased vision

### B. Contraindications

1. Inability to cooperate with testing

## II. Describe the instrumentation and technique

### A. Computerized corneal topography: Placido disc

1. Digitally captures keratoscopic images and analyzes with computer
2. Placido disc-based computerized topography
  - a. Collects reflected data points from the concentric rings and creates a map of the cornea
  - b. Uses color-coded map to present the data with warmer (red and orange) colors representing steeper curvature of the cornea and cooler (blue and green) colors representing flatter curvature
3. Maps that can be obtained
  - a. Power maps
  - b. Simulated keratometry
  - c. Probability of having keratoconus, etc.
  - d. Elevation maps

- e. Corneal wavefront maps

### **III. Describe the considerations in interpretation for this diagnostic procedure**

#### **A. Computerized corneal topography: Placido disc**

1. Useful in detecting irregular astigmatism or multifocal corneas- irregular corneal reflex, scissoring reflex
2. Useful for contact lens fitting and intraocular lens power calculation
3. Critical screening tool for refractive surgery
4. Identifies changes in astigmatism over time or after surgery
5. Identifies corneal ectasias
6. Identifies contact lens induced warpage
7. Useful in postoperative management of corneal transplant patients
8. Helpful in determining etiology for unexplained decreased vision or unexpected post-surgical results including: under corrected aberrations, induced astigmatism, decentered ablations, irregular astigmatism, etc.
9. Requires skill to interpret test results
10. Quality and reproducibility of images is operator dependent and dependent on quality of tear film
11. Non-standardized data maps; user can manipulate appearance of data by changing scales; colors may be absolute or varied (normalized)
12. Other limitations: misalignment may lead to error

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Computerized Corneal Analysis, Module #5, 1996.
3. Arce CG, Martiz J, Alzamora JB, et al. Sectorial and annular quantitative area pachymetry with the Orbscan II. J Refract Surg 2007;23:89-92. Review.
4. Cairns G, McGhee CN. Orbscan computerized topography: attributes, applications, and limitations. J Cataract Refract Surg 2005;31:205-20. Review.

# Aqueous tear deficiency, Sjögren syndrome and Mucin deficiency

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Decreased tear production due to
  - a. Localized lacrimal gland disease
    - i. Idiopathic inflammation
    - ii. Trauma
    - iii. Infiltrative disorders
    - iv. Scarring with obliteration of lacrimal ducts and atrophy of lacrimal gland
  - b. Autoimmune disorders affecting the lacrimal glands
    - i. Sjögren syndrome with or without systemic illness
  - c. Medications
  - d. Decreased corneal sensation
    - i. Trigeminal nerve dysfunction
    - ii. Contact lens wear
    - iii. Post-surgical (e.g., laser in situ keratomileusis)
2. Excessive tear evaporation
  - a. Meibomian gland dysfunction
  - b. Lid/globe congruity disorders (ectropion)
  - c. Lid closure and blinking disorders
    - i. Bells palsy
    - ii. Parkinson disease
3. Decreased mucin production from chemical or inflammatory destruction of conjunctival goblet cells

### B. Define the relevant aspects of epidemiology of this disease

1. Prevalence increases with age
2. More common in women
3. More common among people with arthritis
4. Common after some refractive surgery

### C. List the pertinent elements of the history

1. Dryness, irritation, foreign body sensation, burning, light sensitivity, blurred vision, excessive tearing
2. Symptoms may increase as the day progresses or wax and wane conditions
3. Dry mouth
4. Medication use

**D. Describe pertinent clinical features**

1. Decreased tear meniscus
2. Rapid tear film breakup time
3. Reduced Schirmer test
4. Ocular surface
  - a. Interpalpebral conjunctival staining with vital dyes
  - b. Interpalpebral and/or inferior corneal staining, using fluorescein, rose bengal, or lissamine green
  - c. Relative mucus excess, filaments and plaques

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

1. Tear film break up time less than 10 seconds
2. Schirmer Test
3. Lab testing if suspect Sjögren syndrome

**II. Define the risk factors**

- A. Hormonal effects (e.g. menopause in women)
- B. Age
- C. Connective tissue disease
- D. Conjunctival scarring
- E. Prior corneal surgery/refractive surgery

**III. List the differential diagnosis**

- A. Neurotrophic keratopathy
- B. Exposure keratopathy
- C. Toxicity of topical medications/preservatives

**IV. Describe patient management in terms of treatment and follow-up**

- A. Define medical therapy options

1. Tear replacement therapy
  - a. Preserved artificial tears in milder cases
  - b. Preservative-free artificial tears when frequent application is necessary
  - c. Gel or ointment in severe cases
  - d. Artificial tear pellet
2. Reduce medications contributing to dry eye or ocular surface irritation
3. Reduce evaporation
  - a. Room humidification
  - b. Side shields to eyeglasses
  - c. Avoid drafts
4. Suppress ocular surface inflammation
  - a. Topical cyclosporine
  - b. Topical corticosteroid

**B. Define surgical therapy options**

1. Increase tear retention
  - a. Punctal occlusion: plugs or cauterization
2. Decrease tear evaporation
  - a. Correction of eyelid position abnormalities or lagophthalmos
  - b. Tarsorrhaphy

**V. Describe disease-related complications**

- A. Loss of epithelial integrity: punctate epithelial erosions or large epithelial defect**
- B. Microbial keratitis**
- C. Sterile corneal ulceration**
- D. Corneal thinning, neovascularization, scarring, or perforation**
- E. Corneal calcific deposits and band keratopathy**

**VI. Describe appropriate patient instructions**

- A. Proper administration of topical medications**
- B. Appropriate frequency and timing for use of topical lubricants**
- C. Advantages/disadvantages of different lubricants**
  1. Preserved vs. non-preserved
  2. Viscosity, retention on the ocular surface, and blurring

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 11: Lens and Cataract, 2013-2014.
2. AAO, Preferred Practice Pattern Committee, Cornea and External Disease Panel: Dry Eye Syndrome Preferred Practice Pattern, 2008.



# Hordeolum and chalazion

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Hordeolum
  - a. Inspissation with inflammation or infection of sebaceous glands
  - b. Anterior eyelid margin
    - i. External hordeolum or styne
  - c. Posterior eyelid margin
    - i. Internal hordeolum
  - d. *Staphylococcus aureus* is most common pathogen
  - e. May lead to chalazion
2. Chalazion
  - a. Extrusion of sebum from Meibomian glands into adjacent tissue
  - b. Sterile granulomatous inflammation

### B. List the pertinent elements of the history

1. Hordeolum
  - a. Rapid onset, painful, tender
  - b. Typically resolves in 1-2 weeks
  - c. May produce conjunctival discharge
2. Chalazion
  - a. Generally has slow onset
  - b. May drain externally
  - c. Typically nontender nodule of eyelid margin or tarsus,

### C. Describe appropriate testing and evaluation for establishing the diagnosis

1. Histopathologic examination of material from recurrent chalazia to rule out neoplasia

## II. Define the risk factors

### A. Rosacea

### B. Chronic blepharitis

### **III. List the most common or critical entities in the differential diagnosis**

- A. Benign eyelid tumor**
- B. Malignancy of the eyelid margin or ocular surface, including sebaceous gland carcinoma**

### **IV. Describe patient management in terms of treatment and follow-up**

#### **A. Describe medical therapy options**

1. Observation with warm compresses
2. Topical antibiotics of questionable efficacy, unless acute infection is present. If patient has cellulitis, then oral antibiotics are indicated.
3. Systemic tetracycline derivative may be considered as age appropriate to treat accompanying rosacea and reduce the risk of recurrent chalazion

#### **B. Describe surgical therapy options**

1. Intralesional corticosteroid injection and/or incision/excision or curettage

### **V. List the complications of treatment, their prevention and management**

#### **A. Depigmentation of overlying skin after corticosteroid injection**

#### **B. Eyelid margin notching, misdirected eyelashes and damage to lacrimal punctum after surgical drainage**

1. Use caution when excising chalazion adjacent to these structures

### **VI. Describe disease-related conditions**

#### **A. Distortion of eyelid margin**

#### **B. Recurrent chalazion**

#### **C. Induced astigmatism**

### **VII. Describe appropriate patient instructions**

#### **A. Eyelid hygiene and cleansing**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Exposure keratopathy

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Inadequate eyelid closure (lagophthalmos)
  - a. Cranial Nerve (CN) VII palsy, including Bells palsy
  - b. Decreased blinking (e.g., Parkinson disease)
  - c. Ectropion
  - d. Eyelid deformity
  - e. Trachoma
  - f. Blepharoplasty
  - g. Altered mental status
2. Proptosis
  - a. Thyroid eye disease
  - b. Orbital pseudotumor
  - c. Retrobulbar tumor

### B. List the pertinent elements of the history

1. Dryness, irritation, foreign body sensation, burning
2. Tearing
3. Blurred vision
4. Photophobia
5. Redness
6. Symptoms worse on awakening (nocturnal lagophthalmos)
7. History of eyelid surgery

### C. Describe pertinent clinical features

1. Incomplete eyelid closure and/or proptosis
2. Dilated conjunctival vasculature
3. Punctate epithelial erosions
4. Epithelial defects of varying size
5. Lesions preferentially involving inferior third of cornea and conjunctiva, in exposure area and usually conjunctiva below the limbus

## **II. Define the risk factors**

- A. Decreased blinking associated with visually attentive tasks**
- B. Low humidity**

## **III. List the differential diagnosis**

- A. Neurotrophic keratopathy**
- B. Keratoconjunctivitis sicca**
- C. Toxicity of topical medications/preservatives**

## **IV. Describe patient management in terms of treatment and follow-up**

### **A. Describe medical therapy options**

1. Treatment of underlying disease
2. Tear supplementation with frequent preservative-free lubricants
3. Ointment at bedtime
4. Reduce evaporative tear loss
  - a. Goggles, moisture shields
  - b. Taping lid shut at bedtime
  - c. Humidifier
5. Treatment of any concomitant dry eye (See Aqueous tear deficiency, Sjögren syndrome and Mucin deficiency)

### **B. Describe surgical therapy options**

1. Punctal occlusion
2. Surgical correction of eyelid position
3. Orbital decompression for proptosis

## **V. List the complications of treatment, their prevention and management**

### **A. Topical lubricants**

1. Complications: epithelial toxicity of preservatives
2. Prevention and management: preservative-free lubricants

### **B. Surgical**

## **VI. Describe disease-related complications**

- A. Corneal scarring
- B. Microbial keratitis
- C. Sterile corneal ulceration
- D. Corneal perforation

## **VII. Describe appropriate patient instructions**

- A. Proper administration of topical medications
- B. Use of moisture shields, taping of lids
- C. Advantages/disadvantages of different lubricants
- D. When to seek further care

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Recurrent corneal erosion

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Poor adhesion of the corneal epithelium because of underlying abnormalities with the attachment to the corneal epithelial basement membrane

### B. List the pertinent elements of the history

1. Prior history of a traumatic corneal abrasion
2. Prior history of corneal epithelial basement membrane dystrophy
3. Prior history of ocular surface or corneal disorder (e.g., corneal dystrophy)
4. Symptoms
  - a. Sudden onset of eye pain, often at night or upon first awakening lasting from several minutes to several days

### C. Describe the pertinent clinical features

1. Corneal epithelial defect at time of erosion
2. Corneal epithelial changes such as "microcysts" and "fingerprint lines"
3. Loosely attached corneal epithelium to the underlying basement membrane

## II. Describe patient management in terms of treatment and follow up

### A. Management of epithelial defect

1. Pain control
  - a. Topical nonsteroidal anti-inflammatory drug (NSAID) may have some role
2. Encourage epithelial healing
  - a. Patching or bandage contact lens may be effective

### B. Prevention of subsequent erosion

1. Lubrication, especially while asleep
  - a. Petrolatum ointment or hypertonic saline ointment may be useful
2. Protection by bandage soft contact lens has been used
3. Control of concomitant ocular surface condition, if present, such as dry eye syndrome, rosacea blepharitis, neurotrophic keratopathy, or corneal dystrophy

### C. Refer for surgical intervention and for preventive strategies of recurrent cases

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Acute conjunctivitis

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Conjunctival discharge
2. Eye redness
3. Eyelid swelling
4. Discomfort

### B. Describe pertinent clinical features

1. Conjunctival hyperemia with chemosis; conjunctival papillae or follicles may develop depending upon cause
2. Watery, serous, mucoid, or purulent discharge depending upon cause
3. Preauricular lymph node enlargement, depending upon cause
4. Conjunctival petechiae or hemorrhage
5. Conjunctival pseudomembrane or membrane in severe inflammation
  - a. Membranes permeate the superficial layers of the conjunctival epithelium, are firmly adherent and bleed when removed
  - b. Pseudomembranes are more superficial, not firmly adherent, and have little or no bleeding when removed
6. Punctate epithelial erosions of cornea

## II. List the most common or critical entities in the differential diagnosis

- A. Viral conjunctivitis
- B. Bacterial conjunctivitis
- C. Allergic conjunctivitis
- D. Toxic conjunctivitis

## III. Describe appropriate patient instructions

- A. Precautions to avoid spreading an infection to the fellow eye or to other people

### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.



2. AAO, Focal Points: Antibiotic Use in Corneal and External Eye Infections, Module #10, 1997, p.10.
3. AAO, Preferred Practice Patterns Committee, Cornea and External Disease Panel. Conjunctivitis Preferred Practice Pattern, 2008.

# Chronic conjunctivitis

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Conjunctival discharge, ocular discomfort, and intermittent blurred vision
2. Eye redness lasting for several weeks

### B. Describe pertinent clinical features

1. Conjunctival injection and chemosis
2. Conjunctival papillary or follicular reaction may be present depending on underlying etiology
3. Mild to moderate mucoid or mucopurulent discharge depending on underlying etiology
4. Corneal changes may be present

## II. List the most common or critical entities in the differential diagnosis

- A. Chronic allergic conjunctivitis (including atopic keratoconjunctivitis, vernal conjunctivitis, contact lens-associated conjunctivitis)
- B. Keratoconjunctivitis sicca
- C. Superior limbic keratoconjunctivitis
- D. Toxic conjunctivitis, including medication toxicity
- E. Cicatrizing conjunctivitis, such as mucous membrane pemphigoid
- F. Chlamydial conjunctivitis

## III. Describe the management in terms of treatment and follow-up

- A. Treatment depends upon underlying cause
- B. Oral anti-chlamydial agent for chlamydial conjunctivitis
- C. Topical antibacterial agent for bacterial blepharoconjunctivitis
- D. Eyelid lesion excision for molluscum or other inflammatory nodule
- E. Topical antihistamine, mast cell stabilizer, corticosteroid, and/or cyclosporine for ocular allergy

## IV. List complications of the disease

- A. Conjunctival scarring and symblepharon
- B. Trichiasis
- C. Tear deficiency
- D. Corneal vascularization or scarring

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Herpes simplex virus keratitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Primary or recurrent herpes simplex virus (HSV)

### B. Describe the pertinent clinical features of HSV anterior segment disease

1. Conjunctiva may be involved
2. Epithelial Disease
  - a. Punctate epithelial keratitis
  - b. Dendritic epithelial keratitis
  - c. Geographic epithelial keratitis
  - d. Neurotrophic keratitis
3. Stromal keratitis
  - a. May be superficial or deep
  - b. May be necrotizing or non-necrotizing
  - c. May be associated with trabeculitis or endotheliitis
4. Endotheliitis
  - a. With associated stromal edema and keratic precipitates
5. Uveitis
  - a. Anterior chamber reaction may be mild or severe with hypopyon
  - b. IOP may be elevated (trabeculitis)

## II. List the most common or critical entities in the differential diagnosis

### A. Epithelial keratitis

1. Pseudo-dendrites due to toxicity
2. Epithelial regeneration line
3. Herpes Zoster

### B. Stromal keratitis

1. Microbial keratitis, including fungal keratitis or *Acanthamoeba* keratitis
2. Interstitial Keratitis (including Herpes Zoster, Syphilitic keratitis, and Cogan Syndrome)

### **III. Describe appropriate patient instructions**

- A. Stress importance of compliance and need for follow up**
- B. Awareness of symptoms that may represent toxicity**
- C. Awareness of symptoms that may represent worsening of disease (stromal keratitis) or recurrence**

#### **Additional Resources**

1. AAO. Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Applications of New Laboratory Diagnostic Techniques in Cornea and External Disease, Module #9, 2002, p.5-7.
3. AAO, Focal Points: Herpes Simplex Virus Eye Disease, Module #2, 1997.

# Varicella-zoster virus keratitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Viral infection of conjunctiva and corneal epithelium after reactivation of latent varicella-zoster virus (VZV) in trigeminal ganglion

### B. Define the relevant aspects of epidemiology of the disease

1. Occurs in about half of patients with ophthalmic zoster

### C. List the pertinent elements of the history

1. Prior history of chicken pox
2. Viral prodrome (fever, malaise)
3. Tingling (pain and burning) in affected dermatome may precede rash
4. Vesicular rash on face
5. Lid edema
6. Acute neuralgia (93% of patients)

### D. Describe pertinent clinical features

1. Varicella (chicken pox)
  - a. Usually mild, self-limited disease
  - b. Punctate or dendritic epithelial keratitis may occur concurrently with the skin lesions
  - c. Subepithelial infiltrates, stromal keratitis, disciform keratitis rare
2. Zoster
  - a. Vesicles on the tip of the nose (nasociliary involvement) increase chance of ocular involvement)
  - b. Corneal changes are common in patients with ocular involvement
  - c. Punctate epithelial keratitis ( early)- peripheral, multiple, raised, focal
  - d. Pseudodendrites - broader than HSV, plaque-like
  - e. Corneal mucous plaques (late)
  - f. Neurotrophic keratopathy possible after corneal involvement
  - g. Stromal keratitis
  - h. Endotheliitis
  - i. Lipid keratopathy (very late)
  - j. Sclerokeratitis

k. Exposure keratopathy

l. Uveitis

**E. Describe appropriate diagnostic testing for establishing the diagnosis**

1. Anterior segment examination
2. Dilated fundus examination

**II. Define the risk factors**

**A. Varicella - non-immunized status**

**B. Zoster**

1. Increasing age (#1 risk factor), most patients are 60-90 years old
2. Immunosuppressed patients (human immunodeficiency virus, etc.)
3. Patients with malignancy
4. Patients undergoing chemotherapy or radiation therapy

**III. Describe patient management in terms of treatment and follow-up**

**A. Consider topical antibacterial therapy to prevent superinfection**

**B. Consider topical corticosteroids to decrease inflammation and immune reaction**

1. Concomitant antiviral therapy may be unnecessary

**C. Consider oral corticosteroids**

**D. Consider referral for pain management**

**E. Debridement of corneal epithelial lesions**

**F. Preservative free ophthalmic lubricants**

**G. Follow-up in 3-7 days depending on severity of presentation**

**IV. List the complications of treatment, their prevention and management**

**A. Systemic antiviral therapy is very well tolerated**

1. Rarely, hepatotoxicity

**B. Topical corticosteroids may lead to prolonged need for treatment and more frequent recurrences**

**C. Cataracts**

**D. Glaucoma**

**E. Prevention and management**

1. Judicious use of corticosteroids in acute disease
2. Taper medications as appropriate

## **V. Describe disease-related complications**

- A. Loss of vision**
- B. Corneal scarring**
- C. Rarely, corneal perforation**
- D. Neurotrophic keratopathy**
- E. Exposure keratopathy**
- F. Glaucoma**

## **VI. Describe appropriate patient instructions**

- A. Emphasize importance of compliance with therapy and follow-up**
- B. Patients should be counseled to call if increasing pain develops or the vision changes**
- C. Avoid contact with non-vaccinated individuals, immunocompromised individuals and pregnant women**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Applications of New Laboratory Diagnostic Techniques in Cornea and External Disease, Module #9, 2002.



# Adenoviral conjunctivitis and keratoconjunctivitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Adenovirus
  - a. Simple follicular conjunctivitis without or with punctate epithelial keratitis: several serotypes
  - b. Epidemic keratoconjunctivitis
  - c. Pharyngoconjunctival fever

### B. Define the relevant aspects of epidemiology of the disease

1. Epidemic outbreaks
  - a. Transmission via close contact with infected persons (ocular or respiratory secretions)
    - i. Populations living in close quarters
    - ii. Contaminated instrument/supplies in physicians' offices
2. Sporadic cases

### C. List the pertinent elements of the history

1. Follicular conjunctivitis - self-limited, transient, mild if any epithelial keratitis symptoms
2. Pharyngoconjunctival fever - fever, headache, pharyngitis, follicular conjunctivitis, preauricular adenopathy, mild if any epithelial keratitis symptoms
3. Epidemic keratoconjunctivitis - majority bilateral; possible preceding upper respiratory infection, ocular symptoms 7-10 days after exposure to infected person/contaminated fomites
4. Photophobia
5. Epiphora
6. Foreign body sensation
7. Possibly reduced visual acuity (associated with subepithelial infiltrates)

### D. Describe pertinent clinical features

1. Acute conjunctivitis
  - a. Papillary conjunctivitis
  - b. Follicular conjunctivitis: tarsal conjunctival follicles
  - c. Bulbar conjunctival hyperemia and chemosis
  - d. Petechial hemorrhages
  - e. Pseudomembranes/membranes

2. Epithelial keratitis
  - a. First week of infection
    - i. Minute punctate epithelial opacities
    - ii. Fine punctate epithelial keratitis
  - b. Second week of infection
    - i. Coarse deep epithelial granular infiltrates
    - ii. Punctate epithelial keratitis resolves
    - iii. Possible central geographic erosions
    - iv. Subepithelial infiltrates
3. Extraocular
  - a. Preauricular adenopathy

**E. Describe appropriate laboratory testing for establishing the diagnosis**

1. Diagnosis usually made clinically

**II. Define the risk factors**

- A. Exposure to infected individual or contaminated fomite
- B. Physician office, operating room

**III. List the differential diagnosis**

- A. Non-adenoviral viral conjunctivitis
  1. Herpes simplex virus
  2. Herpes zoster virus
  3. Molluscum contagiosum-associated virus
- B. Bacterial conjunctivitis
- C. Allergic conjunctivitis
- D. Toxic keratoconjunctivitis

**IV. Describe patient management in terms of treatment and follow-up**

- A. Describe medical therapy options
  1. Supportive
    - a. Cool compresses
    - b. Artificial tears

- c. Topical vasoconstrictor
- 2. Pseudomembranes/membranes
  - a. Mechanical removal
  - b. Topical corticosteroid
- 3. Subepithelial infiltrates
  - a. Topical corticosteroid (if concerned about central scarring or effecting vision)

## **V. List the complications of treatment, their prevention and management**

### **A. Topical corticosteroid**

- 1. May exacerbate herpetic keratoconjunctivitis or bacterial conjunctivitis in case of misdiagnosis or coinfection
- 2. May prolong shedding of adenovirus
- 3. May be challenging to taper off topical steroids

### **B. May use topical non-steroidal anti-inflammatory drugs for discomfort**

## **VI. Describe disease-related complications**

### **A. Following pseudomembrane/membrane formation**

- 1. Conjunctival scarring
- 2. Forniceal foreshortening
- 3. Symblepharon formation

### **B. Following chronic recurrent subepithelial infiltrates**

- 1. Photophobia or glare
- 2. Reduced visual acuity

## **VII. Describe appropriate patient instructions**

### **A. Avoidance of transmission during period of viral shedding (10-14 days after onset of clinical signs and symptoms)**

- 1. Avoidance of direct transmission
  - a. Frequent handwashing
  - b. Not touching eyes
  - c. Leave from work or school while shedding
- 2. Avoidance of indirect transmission
  - a. Cleaning linens

- b. Not sharing objects with others

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Update on Ocular Allergy, Module #5, 1994.
3. AAO, Preferred Practice Patterns Committee, Cornea and External Disease Panel. Conjunctivitis Preferred Practice Pattern, 2008.

# Blepharitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Chronic inflammatory condition of the eyelid margins
2. Most common causes include
  - a. Staphylococcal blepharitis
  - b. Meibomian gland dysfunction or seborrheic blepharitis
  - c. Some cases may be associated with Demodex infestation

### B. List the pertinent elements of the history

1. Burning, irritation, foreign body sensation
2. Crusting of the eyelids, particularly on awakening
3. Eyelid margin redness

### C. Describe pertinent clinical features

1. Scales and crusts on eyelids and lashes
2. Abnormal tear film
3. Papillary conjunctivitis
4. Corneal punctate epithelial erosions
5. Corneal marginal infiltrate and neovascularization (pannus)

## II. List the most common or critical entities in the differential diagnosis

- A. Allergic or atopic dermatoblepharitis
- B. Toxic blepharoconjunctivitis
- C. Eyelid neoplasm
- D. Lice infestation of eyelashes

## III. Describe patient management in terms of treatment and follow-up

- A. Lid hygiene cleansing
- B. Artificial tears for abnormal tear film
- C. Control of associated skin disorder such as rosacea

1. AAO. Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Preferred Practice Patterns Committee, Cornea and External Disease Panel. Blepharitis Preferred Practice Pattern, 2008.

# Bacterial conjunctivitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Transmission by direct contact
  - a. Contaminated source
  - b. Infected individual
2. Self-infection from colonizing microorganisms
  - a. Nasal mucosa
  - b. Genital mucosa
  - c. Other sites

### B. List the pertinent elements of the history

1. Purulent or mucopurulent discharge
2. Matting of the eyelashes and crusting of eyelids
3. Eye redness
4. Eyelid swelling
5. Blurry vision
6. Foreign body sensation
7. Eye discomfort

### C. Describe pertinent clinical features

1. Acute purulent conjunctivitis
  - a. Pneumococcal and *Haemophilus*
    - i. Moderate purulent discharge
    - ii. Eyelid edema
    - iii. Conjunctival chemosis and injection
    - iv. Conjunctival papillary reaction
    - v. Conjunctival hemorrhage (sometimes)
    - vi. Corneal stromal infiltrates or ulceration (uncommon)
  - b. Staphylococcal
    - i. Usually less severe discharge
    - ii. May be associated with blepharoconjunctivitis
2. Gonococcal conjunctivitis

- a. Transmitted sexually (direct genital-to-hand-to-eye transmission) or from mother to baby during vaginal delivery
  - b. Severe, hyperacute conjunctivitis
  - c. Massive exudation and purulent discharge
  - d. Conjunctival injection and chemosis
  - e. Conjunctival inflammatory membranes
  - f. Corneal marginal infiltrates, ulceration, melting, and perforation
  - g. Enlarged preauricular lymph nodes
- 3. Chlamydial conjunctivitis
  - a. Preauricular lymphadenopathy
  - b. Follicular conjunctivitis
  - c. May develop mild keratitis with fine epithelial and subepithelial infiltrates and micropannus

**D. Describe appropriate laboratory testing to establish the diagnosis**

- 1. Culture of the conjunctiva
  - a. Compromised host
  - b. Severe purulent discharge
- 2. Consider nasal and throat swab if pharyngitis is present
- 3. Obtain Giemsa stain and/or immunofluorescent antibody tests of the conjunctiva to rule out chlamydia conjunctivitis in chronic cases

## **II. List the differential diagnosis**

- A. Viral conjunctivitis**
- B. Toxic conjunctivitis**
- C. Allergic conjunctivitis**
- D. Exposure keratopathy**
- E. Cat-scratch disease and other causes of Parinaud oculoglandular syndrome**

## **III. Describe patient management in terms of treatment and follow-up**

- A. Describe medical therapy options for acute purulent conjunctivitis**
  - 1. Mild conjunctivitis may be self-limiting, but a topical antibiotic speeds clinical improvement and microbiologic remission
  - 2. Empiric broad-spectrum topical antibiotics four times a day for 5-7 days
  - 3. If a compromised host, severe purulence, or in refractory cases, then obtain culture



4. Systemic antibiotics reserved for acute purulent conjunctivitis with pharyngitis, for conjunctivitis-otitis syndrome, and for *Haemophilus* conjunctivitis in children. Referral to a primary care physician may be necessary, if other tissues or organs are involved
5. Follow-up 5 to 7 days later

**B. Describe medical therapy options for gonococcal conjunctivitis**

1. Systemic antibiotics - oral, intramuscular or intravenous depending on extent of corneal involvement or if neonate
2. Refer patients and their sexual partners for full medical evaluation
3. Consider topical erythromycin, bacitracin, gentamicin, tobramycin or a fluoroquinolone for conjunctivitis
4. Irrigation of the eye with normal saline can remove inflammatory material that may contribute to corneal melting
5. Follow-up daily until conjunctivitis and corneal ulceration resolve
6. If gonococcal conjunctivitis is confirmed, refer for treatment for chlamydia infection (up to a third of patients may have both). Gonococcal infection and other sexually transmitted diseases are reported to the health department

**C. Describe medical therapy options for chlamydial conjunctivitis**

1. Azithromycin oral and/or topical
2. Tetracycline or doxycycline orally

**IV. List the complications of treatment, their prevention, and management**

- A. Ocular surface toxicity from topical antibiotics
- B. Allergic reaction from topical antibiotics
- C. Bacterial resistance to topical antibiotics
- D. Side effects related to systemic antibiotics

**V. Describe appropriate patient instructions**

- A. Precautions to avoid spreading the infection to the fellow eye or other people
  1. Use separate towels and washcloths, and wash hands frequently
- B. Discontinue contact lens wear until conjunctivitis resolves
- C. School or work release (usually after at least 24 hours of treatment with topical antibiotics)

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Antibiotic Use in Corneal and External Eye Infections, Module #10, 1997,

p.10-11.

# Bacterial keratitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Disruption of epithelial integrity, often from trauma, contact lens wear, or preexisting corneal surface disorder, with bacterial invasion of the corneal stroma
2. Common etiological agents: *Pseudomonas aeruginosa*, *Staphylococcus aureus*, *Staphylococcus* species (including *Staphylococcus epidermidis*), *Streptococcus* species

### B. Define the relevant aspects of epidemiology of the disease

1. Increased risk with contact lens wear, especially overnight wear

### C. List the pertinent elements of the history

1. Duration of symptoms
2. Identification of risk factors
3. Current ocular medications (particularly steroids)

### D. Describe pertinent clinical features

1. Epithelial defect
2. Stromal infiltrate
3. Stromal ulceration
4. Hypopyon

## II. List the most common or critical entities in the differential diagnosis

### A. Herpes simplex virus (HSV) stromal keratitis

### B. Fungal keratitis

### C. Immune (non-infectious) keratitis

## III. Describe disease-related complications

### A. Corneal thinning and perforation

### B. Corneal opacification and neovascularization and irregular topography, with loss of vision

#### Additional Resources

1. AAO, Focal Points: Antibiotic Use in Corneal and External Eye Infections, Module #10, 1997, p.11-13.
2. AAO, Preferred Practice Patterns Committee, Cornea and External Disease Panel. Bacterial

Keratitis Preferred Practice Pattern, 2008.

# Allergic conjunctivitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Hypersensitivity reaction

### B. List the pertinent elements of the history

1. Ocular itching is key feature
2. Often associated with ocular redness, and tearing
3. History of concomitant allergic rhinitis
4. Seasonal variation
5. Personal or family history of atopy, including asthma, eczema and seasonal allergies

### C. Describe pertinent clinical features

1. Conjunctival injection
2. Chemosis
3. Eyelid edema
4. Subepithelial infiltrates may be present

### D. Describe appropriate laboratory testing for establishing the diagnosis

1. History and clinical findings typically establish diagnosis
2. Skin testing by an allergist may provide definitive diagnosis and identify the offending allergen(s)

## II. Define the risk factors

### A. History of atopy including allergic rhinitis

### B. Seasonal variation - exposure to the offending antigen

### C. Awareness of common triggers

## III. List the differential diagnosis

### A. Toxic conjunctivitis

### B. Infectious conjunctivitis

1. Bacterial
2. Viral

## IV. Describe patient management in terms of treatment and follow-up

**A. Describe medical therapy options**

1. Cool compresses
2. Refrigerated artificial tears (preferably non preserved)
3. Topical vasoconstrictors
4. Topical histamine receptor antagonist
5. Topical nonsteroidal anti-inflammatory drugs
6. Topical mast cell stabilizers: inhibit mast cell degranulation
7. Combination agents (antihistamine, mast cell stabilizer, inhibition of inflammatory mediators)
8. Mild topical corticosteroids for a limited time
9. Oral antihistamines

**V. List the complications of treatment, their prevention and management**

- A. Possible reaction to medication or preservative (benzalkonium chloride)**
- B. Awareness of drug toxicity/contact allergy - discontinuation of treatment as indicated**
- C. Rebound from vasoconstrictor use may occur**
- D. Corticosteroids - elevation of intraocular pressure, glaucoma, increased risk of infection**

**VI. Describe disease-related complications**

- A. Rarely associated with severe vision-threatening complications**
- B. Itching, tearing, and injection can be very frustrating resulting in marked discomfort and dissatisfaction**

**VII. Describe appropriate patient instructions**

- A. Stress awareness of common offending antigens and avoidance of potential triggers**
- B. Educate patient regarding chronic nature of disease and reassure patient regarding long term visual prognosis**
- C. Discuss therapeutic options and outline appropriate management**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Update on Ocular Allergy, Module #5, 1994.
3. AAO, Focal Points: Nonsteroidal Anti-Inflammatory Drugs, Module #6, 1997, p. 4.
4. AAO, Preferred Practice Patterns Committee, Cornea and External Disease Panel. Conjunctivitis Preferred Practice Pattern, 2008.

# Episcleritis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Idiopathic (most common)
2. Post traumatic, post surgical

### B. Rarely associated with underlying systemic condition

### C. List the pertinent elements of history

1. Acute onset or recurrence of mild ocular discomfort
2. Localized or diffuse conjunctival injection

### D. Describe pertinent clinical features

1. Localized injection of the bulbar conjunctival and episcleral vessels is more common than diffuse involvement
  - a. Blanching with topical phenylephrine 2.5% or 10%
2. Excessive tearing
3. Conjunctival nodule (in nodular form)
4. Small peripheral corneal opacities
5. Mild anterior cell and flare

### E. Describe the appropriate laboratory testing for establishing the diagnosis

1. None, unless unusual case

## II. List the differential diagnosis

- A. Viral conjunctivitis
- B. Superior limbic keratoconjunctivitis
- C. Anterior scleritis
- D. Conjunctival abrasion
- E. Allergic conjunctivitis
- F. Pingueculitis
- G. Iritis
- H. Staphylococcal blepharoconjunctivitis

## III. Describe patient management in terms of treatment and follow-up

**A. Describe medical therapy options**

1. Treat underlying disorder, if present
2. Judicious use of vasoconstrictors
3. Mild - artificial tears
4. Moderate - topical nonsteroidal or mild corticosteroid
5. Severe, recalcitrant, or recurrent - stronger topical corticosteroid +/- oral non-steroidal anti-inflammatory drug (NSAID)

**IV. Describe disease-related complications**

- A. Usually no visually significant sequelae**
- B. Persistent dilation of conjunctival or episcleral vessels may be of cosmetic concern**

**V. Describe appropriate patient instructions**

- A. Self-limiting disease**
- B. Relapses may occur, in the same or contralateral eye**
- C. Use of topical corticosteroids, NSAIDs and vasoconstrictors should be limited to decrease incidence of associated side effects**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Scleritis and Episcleritis: Diagnosis and Management, Module #9, 1995.



# Fuchs corneal dystrophy

## I. Describe the approach for establishing the diagnosis

### A. Describe the etiology of this disease

1. Familial
2. Accelerated loss of endothelial cells with abnormal production and thickening of Descemet membrane

### B. List the pertinent elements of the history

1. Decreased vision, often worse in the morning after awakening
2. Light sensitivity and discomfort in later stage, associated with epithelial edema (microcysts and bullae)

### C. Describe the pertinent clinical features

1. Bilateral central corneal guttae
  - a. Nodular excrescences of Descemet membrane
  - b. Mottled appearance
    - i. Posterior corneal surface, with variable pigmentation
  - c. Disrupted endothelial cell mosaic pattern
2. Corneal edema
  - a. Stromal edema, with progressive increase in corneal thickness that can be measured by corneal pachymetry
  - b. Epithelial edema in the setting of advanced endothelial dysfunction

## II. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Topical hyperosmotic agent, such as 5% sodium chloride, used primarily for epithelial edema

### B. Describe surgical therapy options

1. Endothelial or penetrating keratoplasty

## III. Describe disease-related complications

### A. Increased risk of corneal endothelial decompensation following intraocular surgery

### B. Epithelial edema may lead to subepithelial scarring

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Keratoconus

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Likely multifactorial, with genetic and environmental contributions
  - a. Associated with genetics disorders, such as Down syndrome, as well as atopy and eye rubbing

### B. List the pertinent elements of the history

1. Vision not fully correctable with eyeglasses
2. Vision better with rigid contact lenses than with glasses
3. Frequent changing of glasses or contact lens prescriptions

### C. Describe pertinent clinical features

1. Onset typically in second decade of life
2. Progression noted over 1-2 decades, followed by stabilization
3. Bilateral, but may be asymmetric
4. Irregular retinoscopic reflex (scissoring)
5. Central or inferior paracentral corneal thinning and ectasia
6. Apical corneal scarring
7. Vogt striae
8. Iron ring outlining the area of conical corneal deformation
9. Bulging of the lower lid when the patient looks down

### D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Corneal topography - inferior or central corneal steepening

## II. Describe patient management in terms of treatment and follow-up

### A. Non-surgical treatment options

1. Eyeglasses
2. Contact lenses
  - a. Rigid gas-permeable contact lenses
  - b. Toric soft contact lenses
  - c. Custom contact lens options

### B. Surgical treatment options

1. Intracorneal ring segments
2. Deep anterior lamellar or penetrating keratoplasty

### **III. Describe disease-related complications**

- A. Hydrops**
- B. Corneal scarring**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Pterygium

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Related to ultraviolet B exposure
2. Microtrauma from a windy or sandy environment may play a role, in addition

### B. List the pertinent elements of the history

1. Ocular redness, irritation, and size of visible lesion
2. Visual distortion or decreased acuity
3. Interference with contact lens wear
4. Significant sun exposure

### C. Describe pertinent clinical features

1. Fibrovascular growth extending onto the cornea in the horizontal meridian
2. Usually nasal; often bilateral
3. May be quiescent with few dilated vessels and little growth or "active" with dilated vessels and progressive growth onto the cornea

### D. Risk factors

1. Sun exposure

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Refraction - may be useful in determining degree of induced astigmatism
2. Corneal topography - evaluate degree of associated corneal distortion

## II. Describe patient management in terms of treatment and follow-up

### A. Medical therapy

1. Artificial tears/ocular lubricants for drying over the pterygium or for dellen central to the leading edge
2. Topical nonsteroidal or steroid drop for associated inflammation

### B. Surgical intervention

1. Indications
  - a. Associated decreased vision or irregular astigmatism
  - b. Recurrent associated irritation/inflammation not responsive to medical therapy
  - c. Interference with contact lens wear

- d. To differentiate from a dysplastic conjunctival lesion
- 2. Procedure
  - a. Excision with conjunctival flap, graft, amniotic membrane transplant or primary closure
  - b. Intraoperative mitomycin C use for aggressive or recurrent pterygia

### **III. List the complications of treatment, their prevention and management**

- A. Recurrence following excision**
- B. Conjunctival scarring with secondary gaze restriction**
- C. Mitomycin C toxicity to the ocular surface**

#### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
- 2. AAO, Focal Points: Surgical Treatment of Pterygium, Module #12, 1996.

# Chemical (alkali or acid) injury of the conjunctiva and cornea

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Chemical exposure to the ocular surface by alkali or acid

### B. List the pertinent elements of the history

1. Chemical substance and its pH
2. Duration of exposure and time to treatment
3. Immediate measures taken including irrigation and removal of particulate material

### C. List the pertinent clinical features

1. Extent of ocular surface injury, including conjunctival blanching or ischemia
2. Presence and degree of skin and eyelid burns
3. Residual chemical matter on the ocular surface
4. Assess intraocular pressure

## II. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Minimize ongoing exposure to offending agent
  - a. Irrigation of the ocular surface to normalize pH of the conjunctival sac
  - b. Removal of particulate matter from the ocular surface
2. Encourage reepithelialization, with lubrication and other means

## III. Describe disease-related complications

- A. Ocular surface disease/dry eye state
- B. Corneal stromal scarring and vascularization
- C. Corneal perforation
- D. Glaucoma
- E. Limbal stem cell deficiency
- F. Secondary infection
- G. Symblepharon formation and lid position abnormalities

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.



# Toxic medication injury of the cornea (medicamentosa)

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Dose-dependent cytotoxicity involving the corneal epithelium and limbal stem cells in some instances

### B. Describe the relevant aspects of epidemiology of the disease

1. Prolonged use of topical medications or topical anesthetics
2. Predisposing ocular surface disorders, such as aqueous tear deficiency or delayed tear clearance

### C. List the pertinent elements of the history

1. Number, type and duration of topical medications
  - a. BAK-containing medications
  - b. Aminoglycosides
  - c. Topical antivirals
2. Topical anesthetic abuse

### D. List the pertinent clinical features

1. Conjunctival injection, especially of the inferior conjunctiva
2. Punctate epithelial erosions involving either a portion of or the entire cornea
3. Inferior corneal epithelial defect

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Subconjunctival hemorrhage

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Direct injury to ocular surface
2. Sudden increase in venous pressure, such as with coughing, vomiting, or straining
3. Hemovascular abnormalities, such as impaired clotting or uncontrolled hypertension

### B. List the pertinent elements of the history

1. Ocular or head trauma
2. Recent Valsalva maneuver
3. Systemic medications with antiplatelet/anticoagulant effect

### C. Describe pertinent clinical features

1. Extravasated blood within and/or beneath the conjunctiva

### D. Describe appropriate testing and evaluation for establishing the diagnosis

1. If recurrent subconjunctival hemorrhages and features of a bleeding diathesis (easy bruising, bleeding from the gums, nose or bowels), may consider
  - a. Hematocrit
  - b. Prothrombin time or partial thromboplastin time
  - c. Blood pressure measurement

## II. List the differential diagnosis

### A. Conjunctival laceration

### B. Acute conjunctivitis

### C. Episcleritis

## III. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. No treatment needed in most cases
2. Address underlying systemic condition, if present

## IV. Describe disease-related complications

### A. No sequelae from the subconjunctival hemorrhage itself

- B. Localized drying, dellen, or exposure keratopathy if large and protuberant**

## **V. Describe appropriate patient instructions**

- A. Reassurance that no treatment is needed**
- B. Spontaneous resolution expected**
- C. Hemorrhage may change color during resolution**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Conjunctival foreign body

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. History of trauma
  - a. Timing
  - b. Nature of injury, possible alkali exposure (e.g., cement)
2. Foreign body sensation
3. Discomfort with blinking
4. Tearing
5. Photophobia
6. Blurred vision

### B. Describe pertinent clinical features

1. Conjunctival erythema and/or hemorrhage
2. Lid edema may be present
3. Visible foreign body
4. Secondary epithelial erosions
  - a. Linear pattern of fluorescein staining is highly suggestive of foreign body on corresponding tarsal conjunctiva

## II. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Topical antibiotic prophylaxis, drops or ointments
2. Follow-up to evaluate healing if epithelial keratopathy or abrasion present

### B. Describe surgical therapy options

1. Removal usually possible with slit lamp biomicroscope in office
  - a. Apply topical anesthesia
  - b. Always evert upper lids. May use Desmarres retractor or bent paper clip to visualize foreign body
  - c. Double-evert lid if multiple foreign bodies or particulate matter suspected
  - d. Wipe away superficial foreign body(ies) with cotton-tipped applicator
  - e. Remove embedded foreign body(ies) with jewelers forceps or needle tip

- f. If no foreign body visualized, or if multiple foreign bodies present or suggested by history, irrigate fornix and sweep with cotton-tipped applicator
2. Patients with multiple, extensive foreign bodies or who are uncooperative may need exploration in operating room
3. Take meticulous care in removal of all foreign bodies, particularly in cases of wet cement or other alkali-containing materials
4. Topical antibiotic prophylaxis, drops or ointments following removal

### **III. List the complications of treatment, their prevention and management**

#### **A. Allergy to topical medications**

1. Prevention
  - a. History
2. Management
  - a. Cessation of medication, consider use of topical corticosteroids if clean and healing

#### **B. Infection**

1. Prevention
  - a. Antibiotic prophylaxis
2. Management
  - a. Culture and sensitivity
  - b. More intensive topical antibiotic therapy, directed at specific organisms once known
  - c. Systemic antibiotics if infectious scleritis develops

### **IV. Describe disease-related complications**

- A. Secondary epithelial erosion**
- B. Microbial keratitis**
- C. Allergic conjunctivitis**
- D. Chemical burn if significant acid or alkali content**

### **V. Describe appropriate patient instructions**

- A. Proper use of topical medications**
- B. When to seek further care**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Corneal foreign body

## **I. Describe the approach to establishing the diagnosis**

### **A. List the pertinent elements of the history**

1. History of trauma
  - a. Timing
  - b. Activities prior to development of foreign body sensation
  - c. Nature of material (if known)
2. Pain and foreign body sensation

### **B. Describe pertinent clinical features**

1. Nature, location, and size of corneal foreign body
2. Corneal epithelial defect and/or possible rust ring
3. Rule out intraocular penetration

## **II. Describe patient management in terms of treatment and follow-up**

- A. Remove superficial corneal foreign body**
- B. Remove rust ring**
- C. May observe deep, inert foreign body**
- D. Topical antibiotic and pain medication as indicated**

### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Traumatic corneal abrasion

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. History of trauma
  - a. Circumstances
    - i. Nature of injury
    - ii. Contact lens wear
2. Pain
3. Blurred vision if visual axis involved

### B. Describe pertinent clinical features

1. Epithelial defect on slit-lamp biomicroscopic examination
  - a. Positive fluorescein staining
  - b. Size of defect may be measured to follow healing
2. Presence or absence of stromal edema or infiltrate
3. Presence or absence of corneal or tarsal conjunctival foreign body

## II. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Pressure patching
  - a. Does not accelerate healing
  - b. May help comfort for patient with large abrasion
  - c. Potential risk of exacerbating microbial keratitis in abrasions associated with contact lens wear
2. Therapeutic contact lens for comfort
3. Topical cycloplegic agent for comfort
4. Topical antimicrobial drop or ointment often used for prophylaxis
5. Follow-up to evaluate for microbial keratitis especially if:
  - a. Associated with contact lens
  - b. Secondary to trauma with dirty or organic material
6. May require pain control



### **III. List the complications of treatment, their prevention and management**

#### **A. Allergy to topical medication**

1. Prevention
  - a. History of previous allergy
2. Management
  - a. Cessation of medication

#### **B. Delayed healing**

1. Prevention
  - a. Minimize use of topical medication that may have epithelial toxicity
2. Management
  - a. Cessation or dosage reduction
  - b. Treat concomitant ocular surface disease

### **IV. Describe disease-related complications**

#### **A. Microbial keratitis**

#### **B. Recurrent corneal erosion**

### **V. Describe appropriate patient instructions**

#### **A. Proper use of topical medications**

#### **B. When to seek further care**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Corneal Complications of Contact Lens, Module #2, 1993.

# Corneal laceration

## **I. Describe the approach to establishing the diagnosis**

### **A. List the pertinent elements of the history**

1. Mechanism and nature of injury
2. Time of last oral intake
3. Status of tetanus prophylaxis

### **B. Describe the pertinent elements of the examination**

1. External exam to assess orbital trauma with inspection of involved eye
2. Slit-lamp biomicroscopic examination with Seidel testing if indicated.
3. Assessment for afferent pupillary defect

### **C. Describe the appropriate testing and evaluation for establishing the diagnosis**

1. Diagnostic imaging - localization of foreign body if suspected
  - a. No magnetic resonance imaging if metallic foreign body suspected

## **II. List the initial management**

- A. Consider protective shield, bandage soft contact lens or glue for small lacerations with minimal wound gape**
- B. Surgical closure with removal or reposition of incarcerated iris or vitreous**
- C. Antimicrobial prophylaxis**

## **III. List the complications**

- A. Postoperative infection including keratitis and endophthalmitis**
- B. Persistent wound leak**
- C. Abnormal intraocular pressure (IOP)**
- D. Hyphema**
- E. Postoperative irregular astigmatism**
- F. Cataract**
- G. Iridocorneal adhesion**

## **IV. Describe appropriate patient instructions**

- A. Discuss physical restrictions, importance of eye protection, and plans for emergency care**

**B. Frequency of post-operative visits depends on control of inflammation and IOP**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Repair of the Traumatized Anterior Segment, Module #1, 1992.

# Corneal perforation

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Loss of the corneal epithelium and underlying stroma
2. Corneal inflammation and necrosis

### B. List the pertinent elements of the history

1. "Gush of fluid" or sudden increased tearing
2. Presence and type of preexisting corneal or ocular surface disorder or recent ocular trauma
3. Current ophthalmic and systemic medications

### C. Describe the pertinent clinical features

1. Flat or shallow anterior chamber
2. Low intraocular pressure
3. Positive Seidel test
4. Descemetocoele
5. Folds in Descemet membrane emanating from the thinned area
6. Iris plugging or prolapse

### D. Describe the appropriate testing and evaluation for establishing the diagnosis

1. Culture cornea if infection is suspected

## II. Define the risk factors

### A. Infection (bacterial, fungal, viral, or protozoal)

### B. Connective tissue disorder or systemic ischemic vasculitis

### C. Xerosis

### D. Exposure keratopathy

### E. Neurotrophic keratopathy

### F. Trauma

## III. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Shield orbit
2. Consider bandage (therapeutic) soft contact lens

3. Consider cycloplegia and mydriasis

**B. Describe surgical therapy options**

1. Application of tissue glue or amniotic membrane to cornea
2. Keratoplasty: penetrating or lamellar

**C. Patient follow-up**

1. Follow patient closely until the integrity of the globe is reestablished

**IV. List the complications of treatment, their prevention and management**

- A. Failure to seal wound
- B. Progression of primary process

**V. Describe disease-related complications**

- A. Loss of vision
- B. Increasing size of the perforation
- C. Progression or development of corneal infiltrate or corneal infection
- D. Choroidal detachment
- E. Endophthalmitis
- F. Corneal scarring and neovascularization
- G. Angle-closure glaucoma

**VI. Describe appropriate patient instructions**

- A. Close follow-up until the tectonic integrity of the globe is reestablished
- B. Advise patients to call as soon as possible should they develop increasing pain, loss of vision, increasing tearing, or another gush of fluid
- C. Shield/eye protection
- D. No eye rubbing

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.

# Intraocular pressure

## I. Tonometry: clinical measurement of intraocular pressure (IOP)

### A. Indications and contraindications

1. Indications
  - a. Measurement of IOP for screening and monitoring of glaucoma
2. Relative contraindications
  - a. Corneal laceration
  - b. Flat anterior chamber

### B. Pre-procedure evaluation

1. Slit lamp biomicroscopy to evaluate cornea for abnormalities that may affect IOP measurement accuracy
2. Pachymetry to measure corneal thickness
3. Assess patient for things that might affect tonometry accuracy
  - a. Abdominal or thoracic obesity
  - b. Tight collar or necktie
  - c. High astigmatism
  - d. Breath holding or Valsalva
  - e. Contact lens
  - f. Extraocular muscles acting on restricted globe
  - g. Lid squeezing
  - h. Narrow interpalpebral fissure

### C. Different techniques to measure IOP

1. Goldmann tonometry
  - a. Measures IOP by applanating (flattening) the cornea
  - b. Calculates IOP by flattening the cornea without accounting for its biomechanical properties, which may affect measurement accuracy
    - i. A thicker normal cornea is harder to applanate and may cause falsely high IOP readings. A thinner cornea is easier to applanate and may cause falsely low IOP readings
    - ii. A thicker cornea due to edema is soft and easier to applanate and may cause falsely low readings
    - iii. A stiff cornea is harder to applanate than a soft cornea, regardless of its thickness (e.g. corneal scar, band keratopathy)

2. Tono-Pen: handheld tonometer that contains a strain gauge and produces an electrical signal as the tip applanates a very small area of the cornea
  - a. Helpful in patients with nystagmus and corneal irregularity
  - b. Underestimates in eyes with higher IOP and overestimates in eyes with lower IOP compared with Goldmann
3. Digital palpation
  - a. May be useful in in uncooperative patients
  - b. With keratoprosthesis, useful to compare with other eye
  - c. Can be used intraoperatively

#### **D. Tonometry instrumentation and technique**

1. Goldmann tonometry
  - a. The patient is instructed to relax, keep the eye still and lids open and avoid breath-holding
  - b. Applanation is defined as when the inside edges of the prism-split circular meniscus just touch at the midpoint of their pulsations
  - c. Grams of force to applanate are read from the tonometer dial. IOP in mmHg = 10 (grams of force to applanate)
  - d. Tonometer tip is cleaned with antiseptic solution
2. Tono-Pen tonometry
  - a. Can be used with the patient supine or upright
  - b. Tonometer tip is covered with rubber cover
  - c. Tonometer tip is touched to the central cornea repeatedly until sufficient measurements are taken by the instrument and the average read from the digital display
  - d. Tonometer tip cover is discarded

#### **E. Complications of the procedure, their prevention and management**

1. Complication of contact tonometry
  - a. Corneal abrasion
    - i. Prevent by slow careful applanation and encouraging patient to maintain steady head and eye position
    - ii. Most applanation induced abrasions heal overnight without treatment
  - b. Antiseptic toxicity to the epithelium
    - i. Tonometer tip should be allow to fully dry between patients
  - c. Potential for transmission of infection

#### **F. Considerations in interpretation of this procedure**

1. IOP distribution

- a. Mean IOP 15.5 mmHg
  - b. Non-gaussian distribution skewed toward higher IOPs
- 2. Central corneal thickness may affect IOP measurements by Goldmann and Tono-Pen
  - a. Pachymetry measurements should be used to help interpret IOP measurements, but not to "convert" measurements. In general, Goldmann IOP measurements in eyes with CCT>600 should be considered falsely high, and CCT<500 falsely low
  - b. CCT should be re-measured after corneal surgery
- 3. A single IOP measurement is only a random sample of a dynamic picture. IOP fluctuates throughout the day and night
- 4. Technician or clinician influences on IOP measurement
  - a. Pressure from fingers holding lids may be transmitted to globe and elevate IOP
  - b. Excess fluorescein (thick mires) may cause overestimation of IOP
  - c. Inadequate fluorescein (thin mires) may cause underestimation of IOP
  - d. Improper vertical alignment of mires may cause overestimation of IOP
  - e. Repeated applanation tonometry reduces IOP readings

#### Additional Resources

- 1. AAO Basic and Clinic Science Course, Section 10 Glaucoma, 2013-2014.
- 2. Allingham, RR et al. Shields Textbook of Glaucoma 6th Edition 2011.
- 3. Tonometry in Adults and Children. Eisenberg, et al. Ophthalmology 1998; 105:1173-1181
- 4. Positive Correlation between Tono-Pen Intraocular Pressure and Central Corneal Thickness. Dohadwala et al. Ophthalmology 1998; 105 1849-1854
- 5. Intraocular Pressure difference in Goldmann Applanation tonometry vs Perkins Hand-held Applanation tonometry in Overweight Patients. Gonzaga dos Santos et al. Ophthalmology 105; 2260-2263
- 6. Sources of Error with Use of Goldmann-type Tonometers. Whitacre et al. Survey of Ophthalmology 1993; 38(1):1-30



# Gonioscopy

## I. List the indications/contraindications

### A. Overcomes problem of total internal reflectivity to see angle structures

### B. Essential diagnostic tool in glaucoma (viewing the iridocorneal angle)

1. Most common cause of incorrect diagnosis is omission of gonioscopy
  - a. Overlooking of secondary glaucomas and other glaucomas
2. Identification of
  - a. Angle recession
  - b. Foreign bodies
  - c. Abnormal pigmentation
  - d. Tumors
  - e. Angle neovascularization
  - f. Angle synechiae

### C. Contraindications

1. Inability of patient to cooperate
2. Corneal abrasion or disease precluding application of diagnostic contact lens

## II. Describe the pre-procedure evaluation

### A. Slit-lamp biomicroscopic examination

1. Estimate anterior chamber depth (can use Van Herrick grading)
2. Look for rubeosis (neovascularization) of the iris
3. Iris transillumination defect

## III. Describe the instrumentation and technique

### A. Indirect gonioscopy

1. Produces inverted image 180° away from origination
2. Two types of gonio lenses are in common use
  - a. Goldmann type
    - i. Topical anesthetic
    - ii. Goldmann lens requires clear fluid to fill space between cornea and goniolens

- iii. Lens is brought toward patient's eye and tipped forward quickly enough to trap the clear fluid
  - b. 4 mirror type
    - i. Rests solely on cornea/ tear film
    - ii. Requires only drop of anesthetic
    - iii. Indentation gonioscopy can be performed

**B. Direct gonioscopy**

- 1. Koeppel lens

**IV. List the complications of the procedure, their prevention and management**

**A. Corneal abrasion**

- 1. Prevention
  - a. Moisten cornea, and minimize movement of lens on cornea

**V. Describe the considerations in interpretation of this diagnostic procedure**

**A. Normal angle landmarks**

- 1. Anterior to posterior: cornea, Schwalbe line, non-pigmented trabecular meshwork (TM), pigmented TM, scleral spur, ciliary band, iris root

**Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
- 2. AAO, Atlas: Color Atlas of Gonioscopy, 2000, p. 16, 20-5, 27-38.
- 3. AAO, Glaucoma Medical Therapy: Principles and Management, 1999, p.186, 193-195.

# Examination of the optic nerve

## I. List the indications/contraindications

### A. Indications

1. To examine the optic nerve head for clinical signs of glaucoma or other optic neuropathy
2. Contraindications
3. No absolute contraindications
4. Difficult to perform in cases of very small pupils, dense media opacities, and poor patient cooperation

## II. Describe the pre-procedure evaluation

### A. Evaluate pupil function

1. Specifically look for relative afferent papillary defect (RAPD)

### B. Measure IOP

### C. Evaluate anterior segment

### D. Evaluate angle by gonioscopy if glaucoma suspected

### E. Evaluate color vision if optic neuropathy suspected

### F. Evaluate visual field

### G. Dilate pupil

## III. List the techniques

### A. Handheld ophthalmoscope (limited by monocular view)

### B. Contact lens (center of a gonio lens)

### C. Indirect ophthalmoscopy

### D. Direct ophthalmoscopy

### E. Hruby lens

## IV. Describe the instrumentation and technique

### A. The slit-lamp biomicroscope and indirect and direct ophthalmoscopy are used to provide illumination and magnification, enabling a sense of contour of the optic nerve

### B. Binocular viewing is easier through a dilated pupil, but with experience one can see the optic nerve through an undilated pupil, though usually manoscopically

### C. A fixation target helps to stabilize and to manipulate the position of the eye

## **V. List the complications of this procedure, their prevention and management**

### **A. Corneal abrasion (only a complication of contact lens use)**

### **B. Complications of dilation**

1. Pupillary block attack may develop as pupil dilatation wears off and iris goes into mid-dilated position
  - a. The risk of this occurring can be diminished by accurately assessing angles by gonioscopy and avoiding dilation in critically narrow angles
  - b. Perform laser iridotomy before dilation in at-risk eyes
  - c. Reversing drops (such as Dapiprazole) can get iris to move through mid-dilated position faster
2. Accommodation will be temporarily lost and vision may be blurred for several hours

## **VI. Describe the considerations in interpretation for this diagnostic procedure**

### **A. Look for signs of glaucoma**

1. Generalized
  - a. Large optic cup
  - b. Asymmetry of the cups
  - c. Progressive enlargement of the cup
2. Focal
  - a. Narrowing (notching) of the rim
  - b. Vertical elongation of the cup
  - c. Cupping to the rim margin
  - d. Regional pallor
  - e. Splinter hemorrhage
  - f. Nerve fiber layer loss

### **B. Look for evidence of disc edema**

1. Hyperemia of the disc
2. Flame hemorrhages at or near the disc
3. Obscuration of the nerve fiber bundle
4. Obscuration of vessels as they leave the disc
5. Cotton wool spots
6. Loss of spontaneous venous pulsations (SVP)

**C. Optic atrophy**

1. Pallor of the disc
  - a. Generalized or
  - b. Focal
2. Arterial narrowing
3. Optociliary shunt vessels in cases of optic nerve compression
4. Reduced color vision, RAPD and visual field defects will also be seen on examination

**VII. Describe appropriate patient instructions**

**A. Standard dilation instructions**

**B. If eye pain occurs post-dilation let the office know immediately**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 10, Glaucoma 2013-2014.
2. AAO, Optic Nerve Disorders, 1996, p. 26-27.

# Standard automated static perimetry

## **I. List indications/contraindications**

### **A. Diagnosis of disease**

1. Suspected diagnosis of glaucoma (suspicious disc, ocular hypertension)
2. Neurologic visual field loss
3. Subjective visual field loss
4. Macular/retinal disease
5. Eyelid-related field loss

### **B. Monitoring of disease process**

1. Interval follow-up of suspected or established visual field loss

### **C. Disability determination**

### **D. Testing for motor vehicle license**

## **II. Describe the pre-procedure evaluation**

### **A. Ensure patient can understand and follow instructions**

### **B. Correct refractive error**

### **C. Check head and eyelid position**

## **III. List the alternatives to this procedure**

### **A. Goldmann kinetic perimetry**

### **B. Confrontation visual fields**

### **C. Tangent visual field**

### **D. Amsler grid (to test central visual field)**

### **E. Frequency doubling technology**

## **IV. Describe the considerations in interpretation for this diagnostic procedure**

### **A. Assess patient reliability**

### **B. Review threshold values, global indices, total and pattern deviation plots**

### **C. Correlate test results with ophthalmoscopic and other findings**

### **D. Rule-out artifactual field loss**

1. Corrective lens rim artifact

2. Incorrect refractive correction used for test
3. Upper lid effect

**E. Compare to prior tests**

1. Repeat testing takes into account learning effect
2. Retesting establishes presence of scotomas and fluctuation level of patient's responses

**Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 10: Glaucoma, 2013-2014.
2. AAO, Focal Points: A Primer on Automated Perimetry, Module #8, 1993.
3. AAO, Focal Points: Glaucoma and Automated Perimetry, Module #9, 1993.
4. AAO, Focal Points: Advances to Automated Perimetry, Module #10, 2002.
5. AAO, Ophthalmic Technology Assessment Committee, Glaucoma Panel: Automated Perimetry, Ophthalmic Technology Assessment, *Ophthalmology* 2002; 109:2362-74.

# Corneal pachymetry

## **I. List the indications/contraindications**

### **A. Indications**

1. Glaucoma assessment
2. Monitoring corneal thickness in evaluation of corneal endothelial disease
3. Monitor progression of corneal ectatic diseases

## **II. Describe the considerations in interpretation for this diagnostic procedure**

- A. Thinner corneas may be a result of disease, dystrophy, degeneration or ectatic condition**
- B. Thicker corneas may indicate corneal edema due to corneal endothelial dysfunction**
- C. Thinner corneas underestimate intraocular pressure (IOP) while thicker corneas overestimate IOP**
  1. This can be naturally occurring or post refractive surgery

## **III. Describe appropriate patient instructions**

- A. Explanation of how central corneal thickness could alter course of ocular hypertension for glaucoma treatment**
- B. Corneal pachymetry could aid in planning anterior segment surgery**

### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.



# Primary open-angle glaucoma

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Elevated intraocular pressure (IOP) is the principal risk factor

### B. Define the relevant aspects of epidemiology of the disease

1. Prevalence in African Americans is higher
2. Prevalence increases with age
3. Most frequent cause of irreversible blindness in Hispanic and African-Americans

### C. List the pertinent elements of the history

1. Age
2. Race
3. Symptoms
  - a. Usually asymptomatic until late in disease
4. Family history of glaucoma
5. Cardiovascular disease, diabetes
6. Refractive error
7. Medications
8. Rule out secondary causes (i.e., corticosteroids)
9. Previous eye injury and surgery

### D. Describe pertinent clinical features

1. Usually insidious onset
2. Slowly progressive visual loss without symptoms
3. Painless
4. Usually bilateral, can be asymmetric
5. Central acuity unaffected until late in the disease
6. Elevated intraocular pressure (IOP)
  - a. Can be intermittent (diurnal fluctuation)
  - b. Subset who never have high IOP (normal-tension glaucoma)
7. Consider corneal pachymetry measurement when assessing the accuracy of applanation tonometry
8. Open and normal angle by gonioscopy

9. Optic disc appearance
  - a. Large cup/disc ratio
  - b. Asymmetry of the neuroretinal rim area or cupping
  - c. Focal thinning or notching of the neuroretinal rim
  - d. Optic disc hemorrhage (this could be a "non-specific" finding)
10. Visual fields
  - a. Scotoma can precede visible optic nerve damage
  - b. Visual field defects may not be detectable by standard perimetry
  - c. Typical glaucoma defects
    - i. Generalized depression (early, non-specific)
    - ii. Paracentral scotoma
    - iii. Arcuate scotoma
    - iv. Nasal step
    - v. Central island in far advanced cases

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

1. Visual fields
  - a. Automated static perimetry
2. Optic nerve description
3. Central corneal thickness measurement

**II. Define the risk factors**

- A. Older age
- B. Race (higher prevalence in African-Americans)
- C. Elevated IOP
- D. Positive family history

**III. List the most common or critical entities in the differential diagnosis**

- A. Non-glaucomatous disc abnormalities
- B. Other glaucomas
  1. Secondary open-angle glaucoma
  2. Angle-closure glaucoma
  3. Normal tension glaucoma

4. Elevated episcleral venous pressure
  - a. Etiology
    - i. Obstruction of venous outflow
    - ii. Thyroid eye disease (thyrotropic ophthalmopathy)
    - iii. Elevated EVP may occur in severe cases with marked proptosis and orbital congestion associated with an intraorbital infiltrative process
    - iv. Superior vena cava syndrome
    - v. Lesions of upper thorax may obstruct venous return from head
    - vi. Retrobulbar tumors
    - vii. Cavernous sinus thrombosis

#### **IV. Describe patient management in terms of treatment and follow-up**

##### **A. Describe medical therapy options**

1. Beta-adrenergic antagonists
2. Carbonic anhydrase inhibitors
3. Adrenergic agonists (sympathomimetics)
4. Prostaglandin analogues
5. Cholinergics

##### **B. Describe surgical therapy options**

1. Laser trabeculoplasty
2. Trabeculectomy (with or without antimetabolites)
3. Aqueous shunt surgery (glaucoma drainage tube implants)
4. Ciliary body ablation

#### **V. Describe disease-related complications**

##### **A. Limitations due to visual field loss**

##### **B. End stage glaucoma and blindness**

#### **VI. Describe appropriate patient instructions**

##### **A. Discussion of medications and surgical treatments**

1. Side effects
2. Instructions relating to compliance

- a. Appropriate drop timing
- b. Nasolacrimal occlusion, passive lid closure
- c. Prevention of washout effect by spacing drop therapy

**B. Discussion regarding quality of life issues**

- 1. Support groups
- 2. Career issues
- 3. Financial issues regarding treatment

**C. Importance of periodic follow-up**

**Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
- 2. AAO, Focal Points: Dysfunctional Glaucoma Filtration Blebs, Module #4, 2002.
- 3. AAO, Focal Points: Automated Perimetry, Module #10, 2002.
- 4. AAO, Focal Points: Normal Tension Glaucoma, Module #12, 2000.
- 5. AAO, Focal Points: Techniques of Glaucoma Filtration Surgery, Module #6, 2000.
- 6. AAO, Focal Points: Aqueous Shunting Procedures, Module #3, 2002
- 7. AAO, Focal Points: Update on Glaucoma Clinical Trials, Module #9, 2003.
- 8. AAO, Focal Points: Medical Treatment of Open-Angle Glaucoma, Module #10, 1993.
- 9. AAO, Preferred Practice Patterns Committee, Glaucoma Panel. Primary Open Angle Glaucoma Preferred Practice Pattern, 2010.

# Normal tension glaucoma

## I. Describe the approach to establishing the diagnosis

### A. Definition of this disease

1. Optic nerve changes characteristic of primary open-angle glaucoma (POAG) but intraocular pressure (IOP) in the normal range without treatment

### B. Describe pertinent clinical features

1. No clear difference from optic nerve cupping seen in POAG
2. Normal IOP

### C. Describe appropriate testing and evaluation for establishing the diagnosis

1. Repetitive diurnal measurement of IOP
2. Visual field
3. Gonioscopy
4. Optic disc evaluation
5. Measurement of corneal thickness (pachymetry)

### D. Risk factors

1. Family history
2. Migraine

## II. List the most common or critical entities in the differential diagnosis

### A. Undetected glaucoma

1. POAG with large diurnal pressure variation
2. Intermittent elevation of IOP caused by another type of glaucoma
3. Thin central cornea

### B. Nonglaucomatous optic nerve disease resembling glaucoma

1. Compressive lesion of the optic nerve
2. Anterior ischemic optic neuropathy
3. Compromised ocular blood flow

## III. Describe appropriate patient instructions

### A. Need for regular follow-up and monitoring of IOP

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
2. AAO, Focal Points: Normal-Tension Glaucoma, Module #12, 2000.
3. AAO, Focal Points: Normal (Low) Tension Glaucoma, Module #12, 1991.
4. AAO, Preferred Practice Pattern Committee, Glaucoma Panel: Primary Open-Angle Glaucoma Preferred Practice Pattern, 2010.

# Primary open-angle glaucoma suspect

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Increased intraocular pressure (IOP)
2. An optic nerve that is suspicious for glaucoma even with normal IOP

### B. Define the relevant aspects of epidemiology of this disease

1. Number of primary open angle glaucoma (POAG) suspects exceeds the number of people diagnosed with glaucoma

### C. List the pertinent elements of the history

1. Previous history of elevated IOP
2. History of vision loss
3. Family history of glaucoma
4. Age
5. Ethnicity (e.g., African descent)
6. Recent medications
7. History of vasospastic disease

### D. Describe pertinent clinical features

1. Optic disc or nerve fiber layer damage suggesting glaucoma
  - a. Enlarged cup/disc ratio
  - b. Asymmetric cup/disc ratio
  - c. Notching or narrowing of the neural rim
  - d. Disc hemorrhage
  - e. Diffuse or local abnormality in the nerve fiber layer
2. Visual fields suspicious for early glaucomatous damage
3. IOP consistently above 21 mm Hg (i.e., ocular hypertension) in the context of central corneal thickness measurements
4. Normal open angle on gonioscopy with absence of secondary causes

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Determination of central corneal thickness
2. Evaluation of optic nerve head and retinal nerve fiber layer
3. Visual field testing and analysis

4. Documentation of optic nerve head appearance

## **II. Define the risk factors**

- A. Elevated IOP
- B. Family history of glaucoma
- C. Advancing age
- D. Race and ethnicity (e.g., African descent, Hispanics)
- E. Associated disease states (systemic hypertension, cardiovascular disease, diabetes mellitus)

## **III. List the most common or critical entities in the differential diagnosis**

- A. POAG
- B. Normal tension glaucoma
- C. Corticosteroid responder
- D. Previous history of trauma (i.e., angle recession glaucoma)
- E. Previous or current uveitis
- F. Nonglaucomatous causes (e.g., compressive lesions, ischemic episodes) of abnormal optic disc appearance
- G. Congenitally large optic cup

## **IV. Describe appropriate patient instructions**

- A. Need for periodic follow-up

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
2. AAO, Preferred Practice Patterns Committee, Glaucoma Panel. Primary Open-Angle Glaucoma Suspect Preferred Practice Pattern, 2010.



# Secondary open-angle glaucomas

## I. Pseudoexfoliation (Exfoliation syndrome)

### A. Describe the etiology of this disease

1. Deposition of exfoliative material in several organs including the anterior segment of the eye

### B. Describe pertinent clinical features

1. Deposits of exfoliative material on anterior lens surface
2. Transillumination defects at pupillary margin
3. Poor pupillary dilation
4. May develop intraocular pressure (IOP) elevation
5. Zonular weakness, phacodonesis

## II. Pigmentary glaucoma

### A. Describe the etiology of this disease

1. Concave peripheral iris configuration, usually in myopic eye with deep anterior chamber
  - a. Posterior iris surface comes into contact with lens zonule and with physiologic dilation/constriction of pupil, pigment rubbed free from iris disperses in aqueous humor
  - b. Collection of pigment within angle/trabecular meshwork (TM) occurs during normal aqueous circulation and causes obstruction to outflow and may lead to IOP elevation

### B. Describe pertinent clinical features

1. Classic triad of pigmentary glaucoma
  - a. Krukenberg spindle
  - b. Pigmentation of the TM
  - c. Mid-peripheral iris transillumination defects
2. Gonioscopy
  - a. Dark pigmentation of TM
  - b. May have pigmentation on or anterior to Schwalbe line

## III. Inflammatory glaucoma (secondary open-angle and angle-closure glaucoma)

### A. Describe the etiology of this disease

1. Open-angle glaucoma
  - a. Intraocular inflammation in the anterior segment (anterior uveitis) leads to trabecular

damage and obstruction of trabecular meshwork outflow by:

- i. Edema of trabecular meshwork
- ii. Trabecular endothelial cell dysfunction
- iii. Fibrin and elevated aqueous protein from breakdown of the blood-aqueous barrier
- iv. Inflammatory cells and/or inflammatory nodules

**B. Describe the pertinent clinical features**

1. General features
  - a. Ciliary flush
  - b. AC reaction (cell and flare)
  - c. KP (granulomatous vs. non-granulomatous)
  - d. Iris inflammatory nodules
  - e. Heavy angle pigmentation, especially inferiorly
  - f. Fibrin
  - g. Sterile hypopyon
  - h. Posterior synechiae
    - i. If extensive can lead to iris bombé
  - i. PAS

## **IV. Lens-induced glaucoma**

**A. List the pertinent elements of the history**

1. Phacolytic
  - a. Phakic status
  - b. Elderly, long-standing poor vision, sudden onset of pain, conjunctival hyperemia
2. Lens particle
  - a. Following disruption of lens integrity
    - i. Surgical: following cataract extraction (Nd:YAG) capsulotomy
    - ii. Non-surgical: following lens injury
    - iii. Usually within weeks but may be months or years later
3. Phacoanaphylaxis
  - a. Following CE with or without vitreous loss
  - b. Following CE in one eye, with subsequent CE or leaking hypermature cataract in other involved eye

- c. After traumatic or spontaneous rupture of lens capsule
- 4. Ectopia lentis
  - a. Visual disturbances due to lens subluxation
  - b. Family or personal history
    - i. Simple ectopia lentis
    - ii. Ectopia lentis et pupillae
    - iii. Marfan Syndrome
    - iv. Homocystinuria
      - i) Thrombotic vascular occlusions
      - ii) Mental retardation
    - v. Weill-Marchesani syndrome
  - c. Trauma
  - d. Prior ocular surgery
  - e. Pseudoexfoliation syndrome (See Pseudoexfoliation (exfoliation syndrome))
- 5. Phacomorphic
  - a. Elderly, long-standing decreased vision
  - b. Acute or subacute onset

**B. Define the relevant aspects of epidemiology of the disease**

- 1. Phacolytic
  - a. Secondary to leakage of high molecular weight lens protein, usually in eyes with mature or hypermature cataract
  - b. Sudden onset of open-angle glaucoma
  - c. Usually monocular, not reported in children, rare in young adulthood
  - d. These patients may have long-standing poor vision in the affected eye
- 2. Lens particle
  - a. Occurs when lens cortex material obstructs the trabecular meshwork
  - b. Often delay of days to weeks between trauma and onset of glaucoma
  - c. Similar form may occur years after CE with lens material freed into anterior chamber
  - d. Can occur in children and adults
  - e. Can occur immediately after Nd:YAG capsulotomy
- 3. Phacoanaphylaxis
  - a. Uncommon granulomatous inflammation in response to liberated lens material
  - b. Rarely causes glaucoma, more likely to be associated with hypotony

- c. Almost always unocular, although some reports of sympathizing reaction
- 4. Ectopia lentis
  - a. Autosomal dominant inheritance
    - i. Simple ectopia lentis
    - ii. Weill-Marchesani syndrome
    - iii. Marfan syndrome
  - b. Autosomal recessive inheritance
    - i. Ectopia lentis et pupillae
    - ii. Weill-Marchesani syndrome
    - iii. Homocystinuria
  - c. Early age of onset in inherited conditions
- 5. Phacomorphic
  - a. Older age
  - b. Multifactorial mechanism
  - c. Pupillary block often contributory
  - d. Intumescent lens narrows angle, causing closure

**C. Describe pertinent clinical features**

- 1. Phacolytic
  - a. Open angle
  - b. Intact capsule
  - c. Elevated intraocular pressure (IOP) (30-50 mmHg not uncommon)
  - d. Microcystic corneal edema
  - e. Cell and flare without keratic precipitates (KP)
  - f. Cellular debris or hypopyon in anterior chamber (AC)
  - g. White particles (clumps of lens protein) may be in AC or on lens capsule
  - h. Mature, hypermature or morgagnian cataract (often with wrinkled anterior lens capsule)
  - i. Microscopic findings of an AC tap: macrophages containing lens material (phase microscopy)
- 2. Lens particle
  - a. Open-angle
  - b. Open capsule
  - c. Free cortical material in AC
  - d. Elevated IOP

- e. Moderate AC reaction
  - f. Microcystic corneal edema
  - g. Late: development of posterior and peripheral anterior synechiae
3. Phacoanaphylaxis
- a. Variable clinical presentation
  - b. Most common clinical picture:
    - i. Open capsule
    - ii. Moderate AC reaction with KP on corneal endothelium and anterior lens surface
    - iii. Low grade vitritis
    - iv. Peripheral anterior and/or posterior synechiae
4. Ectopia lentis
- a. Lens subluxated or completely dislocated (into vitreous cavity or AC)
  - b. Body habitus
    - i. Tall, slender with joint laxity and arachnodactyly (Marfan syndrome and homocystinuria)
    - ii. Short, stocky with brachydactyly (Weill-Marchesani syndrome)
  - c. Variable refractive error
  - d. Unilateral lens displacement in trauma
  - e. Bilateral lens displacement in inherited conditions
    - i. Superotemporal in Marfan syndrome and simple ectopia lentis
    - ii. Inferotemporal in homocystinuria
  - f. Small, round lens (microspherophakia) in Weill-Marchesani syndrome
    - i. Consider microspherophakia in high myopes with pupillary block
  - g. Trauma-related ocular injuries
  - h. Phacodonesis
    - i. Shallow AC
    - j. Iris bombé configuration
  - k. Enlarged globe with long axial length (Marfan syndrome)
  - l. Vitreous prolapse
  - m. Thin sclera (Marfan, Ehlers-Danlos syndrome)
  - n. Retinal detachment
5. Phacomorphic
- a. Advanced cataract

- b. Shallow AC
- c. Angle remains narrow despite patent iridotomy

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
2. AAO, Focal Points: Exfoliation Syndrome, Module #9, 1994.
3. AAO, Focal Points: Pigmentary Glaucoma: New Insights, Module #2, 1998.
4. AAO, Atlas: Color Atlas of Gonioscopy, 2000, p. 40, 47, 49, 103.

# Hyphema

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Trauma
2. Intraocular surgery
3. Rubeosis iridis (iris neovascularization)
4. Anterior uveitis
5. Retinoblastoma and other tumors
6. Child abuse

### B. List the pertinent elements of the history

1. History of ocular trauma or surgery
2. History of ocular or systemic disorders associated with spontaneous hyphemas
3. Recent medications

### C. Describe pertinent clinical features

1. Decreased visual acuity
2. Elevated intraocular pressure (IOP)
3. Blood in anterior chamber
  - a. Circulating red blood cells (RBCs)
  - b. Layered hyphema
  - c. Total hyphema

### D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Sickle cell hemoglobin screening in African-American patients
2. Coagulation studies where indicated, e.g., CBC in patients with severe thrombocytopenia since they may need platelet therapy and a prothrombin time (PT)/international normalized ratio (INR) in patients on warfarin.

## II. Define the risk factors

- A. Risk of increased IOP is greater with a larger hyphema and following rebleeding after a traumatic hyphema
- B. Rebleeding usually occurs during the first week after initial hyphema

## III. Describe patient management in terms of treatment and follow-up

- A. Cycloplegic agent
- B. Topical and/or systemic corticosteroid useful in treating inflammation
- C. Topical or systemic IOP-lowering medications as needed
- D. Persistently elevated IOP for persistent hyphema may necessitate consideration of surgical therapy
- E. Analgesics and antiemetic medications as needed
- F. Elevate head of bed
- G. Limit activity

#### **IV. List the complications of treatment, their prevention and management**

- A. Carbonic anhydrase inhibitors may increase sickling tendency in patients with sickle cell hemoglobinopathy

#### **V. Describe appropriate patient instructions**

- A. Compliance with prescribed medical regimen
- B. Use of protective eye shield
- C. Quiet activity or bedrest
- D. Provide safe home environment
- E. Reliable follow-up
- F. No aspirin or oral nonsteroidal anti-inflammatory drugs (NSAIDs)

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.



# Corticosteroid-induced glaucoma

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Caused by a reduction in facility of outflow

### B. List the pertinent elements of the history

1. May develop at any time during long-term corticosteroid administration, but intraocular pressure (IOP) elevation typically occurs within a few weeks with potent corticosteroids, or in months with the weaker corticosteroids
2. Routes of administration
  - a. Topical or local corticosteroid therapy is more often associated with IOP rise than is the case with systemic administration
  - b. IOP elevation may occur in response to subconjunctival, sub-Tenons, intravitreal or retrobulbar injections of corticosteroid

### C. Describe pertinent clinical features

1. Clinical presentation typically resembles primary open-angle glaucoma with an open, normal-appearing anterior chamber angle

## II. Define the risk factors

- A. Individuals with primary open-angle glaucoma or a family history of the disease are more likely to respond to chronic corticosteroid therapy with a significant rise in IOP
- B. High myopes, diabetic patients, and patients with connective tissue diseases have been reported to have a higher predisposition to corticosteroid-induced glaucoma compared to normal patients

## III. List the most common or critical entities in the differential diagnosis

- A. Primary open-angle glaucoma
- B. Secondary open-angle glaucoma, such as exfoliation syndrome, pigmentary glaucoma or ocular inflammation

## IV. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Discontinuation of the corticosteroid is the first treatment option and is often all that is required
2. Some patients experience a more chronic form of this disease where the IOP normalizes in 1-4 weeks, while some experience a more acute form where the IOP typically resolves within days of stopping the corticosteroid

3. In rare cases, the glaucoma may persist despite stopping all corticosteroids
4. Glaucoma medical therapy, as per treatment of primary open-angle glaucoma

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
2. AAO, Glaucoma Medical Therapy: Principles and Management, 1999, p.202-203.
3. Allingham RR, Shields MB, ed. Shields' Textbook of Glaucoma. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins, 2005.

# Acute angle-closure glaucoma

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Physiologic pupillary block
  - a. Excessive iris-lens apposition impedes flow of aqueous humor from posterior chamber to anterior chamber
  - b. Secondary forward bowing of peripheral iris results in occlusion of the trabecular meshwork

### B. Describe the pertinent clinical features

1. Symptoms
  - a. Acute onset of brow ache and eye pain
  - b. Blurred vision
  - c. Haloes around light
  - d. Nausea and vomiting
2. Signs
  - a. High intraocular pressure (IOP)
  - b. Mid-dilated, sluggish pupil
  - c. Corneal epithelial edema
  - d. Congested episcleral and conjunctival vessels
  - e. Shallow anterior chamber
  - f. Appositional angle-closure
  - g. Iris bombé

## II. Define the risk factors

- A. Hyperopia
- B. Family history of angle closure
- C. Older age
- D. Female gender
- E. Cataract (lens swelling)
- F. Asian ethnicity

## III. Describe patient management in terms of treatment and follow-up

**A. Describe medical therapy options**

1. Medical therapy used to lower the IOP and allow clearing of corneal edema in preparation for laser iridotomy
  - a. Beta-adrenergic antagonists
  - b. Alpha-adrenergic agonists
  - c. Carbonic anhydrase inhibitors
  - d. Miotics - 1-2% pilocarpine after IOP starts to normalize
  - e. Prostaglandin analogues
  - f. Oral hyperosmotic agents
2. Pushing on cornea with cotton tip applicator or with gonios lens occasionally opens the angle

**B. Describe surgical therapy options**

1. Laser peripheral iridotomy
2. Prophylactic iridotomy in fellow eye
3. Incisional iridectomy when laser iridotomy is not possible
4. Primary filtering surgery may be required if extensive synechial closure is present
5. Cataract surgery once IOP normalized

**IV. Describe disease-related complications**

- A. Corneal edema**
- B. Iris atrophy**
- C. Posterior synechiae**
- D. Cataract formation**
- E. Optic nerve damage**
- F. Retinal vascular occlusion**

**Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 10: Glaucoma, 2013-2014.
2. AAO, Preferred Practice Patterns Committee, Glaucoma Panel. Primary Angle Closure Preferred Practice Pattern, 2010.
3. AAO, Glaucoma Medical Therapy: Principles and Management, 1999, p.195-198.

# Neovascular glaucoma

## I. Describe approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Pain
2. Photophobia
3. Markedly reduced vision
4. Diabetes mellitus
5. Hypertension
6. Arteriosclerosis

### B. Describe pertinent clinical features

1. Early
  - a. Tufts of new vessels at pupillary margin
  - b. Fine vessels crossing scleral spur
2. Late
  - a. High intraocular pressure (IOP)
  - b. Florid iris neovascularization with ectropion uveae
  - c. Fibrovascular membrane over iris and angle structures
  - d. Variable synechial angle-closure
  - e. Anterior chamber flare

## II. Define the risk factors

### A. Retinal hypoxia

### B. Central retinal vein occlusion

### C. Proliferative diabetic retinopathy

### D. Post cataract extraction, vitrectomy

1. Particularly with breached posterior capsule

### E. Carotid occlusive disease

1. May have normal or low IOP

### F. Central retinal artery occlusion

## III. Describe the patient management in terms of treatment and follow-up

**A. Early**

1. Medical therapy of elevated IOP
2. Topical corticosteroids and atropine
3. Intraocular anti-VEGF agents
4. Panretinal photocoagulation (PRP)
5. Panretinal cryotherapy (when poor visualization does not permit PRP)

**B. Late**

1. Medical therapy of elevated IOP
2. Topical corticosteroids and atropine
3. Intraocular anti-VEGF agents
4. PRP
5. Surgery
  - a. Filtration surgery with adjunctive antimetabolite and usually after pre-treatment with anti-VEGF
  - b. Aqueous shunt
6. Cyclodestruction
  - a. Laser surgery
  - b. Cryotherapy

**IV. Describe disease-related complications**

**A. Absolute glaucoma with blindness**

**V. Describe appropriate patient instructions**

**A. Medication and surgical discussion**

**B. Referral for panretinal photocoagulation, surgical intervention, and/or cyclodestructive procedure**

**C. Referral to primary care physician for care related to etiology**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
2. AAO, Focal Points: Neovascular Glaucoma, Module #7, 1997.

# Glaucoma medications

Beta-adrenergic antagonists

## **I. List the indications and contraindications**

### **A. Indications**

1. First line or adjunctive therapy to lower intraocular pressure (IOP) in the following circumstances
  - a. All types of glaucoma
  - b. Before or after laser surgery
  - c. After cataract surgery

### **B. Contraindications**

1. Chronic obstructive pulmonary disease (non-selective)
2. Asthma (non-selective)
3. Congestive heart failure (check with cardiologist)
4. Bradycardia
5. Hypotension
6. Greater than first degree heart block

## **II. List the alternatives to this therapy (note: all listed drugs can be used as adjunctive therapy also)**

- A. Prostaglandin analogues**
- B. Carbonic anhydrase inhibitors (topical and oral)**
- C. Alpha-adrenergic agonists**
- D. Parasympathomimetic agents**

## **III. Describe the follow-up care**

- A. Consider therapeutic one-eyed trial**
- B. Evaluate response to therapy in 2-6 weeks**
- C. Inquire about drug-related side effects (ocular and systemic)**
- D. Look for evidence of ocular toxicity**

## **IV. Describe appropriate patient instructions**

**A. Discuss potential side effects**

1. Corneal toxicity
2. Allergic reactions
3. Congestive heart failure (CHF) (classic teaching, although cardiology uses beta-blockers as first line treatment in CHF)
4. Bronchospasm (seen with nonselective)
5. Bradycardia
6. Depression
7. Impotence

**B. Discuss/demonstrate nasolacrimal occlusion and passive eyelid closure**

**C. If on more than one topical medication, instruct patient to wait at least 5 to 10 minutes between eye drop administration**

Alpha-adrenergic agonists

**I. List the indications and contraindications**

**A. Indications: non-selective adrenergic agonists**

1. Mydriasis in cataract surgery
2. Reduce bleeding in oculoplastic surgery
3. Reduce bleeding in trabeculectomy
4. Slow absorption of local anesthetics

**B. Indications: selective adrenergic agonists**

1. IOP lowering
  - a. Open-angle glaucoma/ocular hypertension
  - b. Prevention of postoperative pressure spikes
    - i. Prior to or immediately after laser treatment (laser trabeculoplasty, laser peripheral iridotomy, neodymium yttrium-aluminum-garnet (Nd:YAG) capsulotomy)
    - ii. Cataract surgery
  - c. Acute angle-closure glaucoma
2. Miosis after refractive surgery (off-label use)

**C. Contraindications**

1. Monoamine oxidase inhibitor therapy
2. Infants and children younger than 2 years old



## **II. List the alternatives to this therapy**

- A. Prostaglandin analogues**
- B. Parasympathomimetics**
- C. Carbonic anhydrase inhibitors**
- D. Osmotic agents**
- E. Laser trabeculoplasty**
- F. Incisional glaucoma surgery**
- G. Beta-adrenergic antagonists**
- H. Cyclodestruction**

## **III. Describe the follow-up care**

- A. Consider therapeutic one-eyed trial**
- B. Question patient about possibility of side effects**
- C. Examination emphasis**
  - 1. External (lids)
  - 2. Slit-lamp biomicroscopy (conjunctival and corneal exam)
  - 3. Tonometry and optic nerve assessment
- D. Consider checking IOP after starting medication**

## **IV. Describe appropriate patient instructions**

- A. Discuss possible side effects with patient to increase level of awareness**
  - 1. Conjunctival injection
  - 2. Allergic reactions
- B. Demonstrate nasolacrimal occlusion or lid closure**
- C. Schedule regular follow-up visits**

## **V. Describe the dosing technique**

- A. Nasolacrimal occlusion**
- B. Passive lid closure**

Parasympathomimetic agents

## **I. List the indications and contraindications**

### **A. Indications**

1. Treatment of increased intraocular pressure (IOP) in patients with at least some open filtering angle
2. Prophylaxis
  - a. For angle-closure prior to iridotomy
  - b. To decrease iridozonular contact in pigment dispersion glaucoma (may not be tolerated in this younger patient population because of induced myopia)

### **B. Contraindications**

1. Neovascular, uveitic, or malignant glaucoma
2. Need to regularly assess fundus

## **II. List the alternatives to this therapy**

### **A. Prostaglandin analogues**

### **B. Beta-adrenergic antagonists**

### **C. Adrenergic agonists**

### **D. Carbonic anhydrase inhibitors**

### **E. Osmotic agents**

### **F. Laser iridotomy**

### **G. Laser iridoplasty**

### **H. Laser trabeculoplasty**

### **I. Filtering surgery**

## **III. Describe the follow up care**

### **A. Follow IOP**

### **B. Follow gonioscopy**

1. Particularly watch for forward shift of the lens-iris diaphragm and paradoxical angle closure with initiation of medication or with increases in medication strength, in eyes judged to have a relatively narrow but non-occludable angle (such as in pseudoexfoliation glaucoma with an early to moderate cataract and potentially loose zonules).

## **IV. Describe appropriate patient instructions**

### **A. Schedule patient return for follow up IOP check and to review any side effects experienced from the drug**

### **B. Describe symptoms of retinal detachment**

**C. Explain possible side effects**

1. Increased myopia
2. Eye or brow ache/pain
3. Decreased vision
4. Cataract
5. Periocular contact dermatitis
6. Corneal toxicity
7. Paradoxical angle closure

**D. Explain the need to contact the ophthalmologist if they have symptoms of angle-closure glaucoma such as halos around lights**

Carbonic anhydrase inhibitors

**I. List the indications and contraindications**

**A. Indications**

1. Reduction of chronically elevated intraocular pressure (IOP) in adults and children
  - a. Monotherapy
  - b. Additive therapy
    - i. Can help lower diurnal and nocturnal IOP when added to a prostaglandin analog
2. Prophylaxis of elevated IOP after a surgical intervention
3. Reduction of acutely elevated IOP

**B. Contraindications**

1. Sulfonamide allergy
2. Kidney stones
3. Aplastic anemia
4. Thrombocytopenia
5. Sickle cell disease

**II. Describe patient management in terms of treatment and follow-up**

**A. Describe medical therapy options**

1. Oral or topical carbonic anhydrase inhibitors
2. Topical beta-adrenergic antagonists
3. Alpha-adrenergic agonists

4. Parasympathomimetic agents
5. Prostaglandin analogues

**B. Describe surgical therapy options**

1. Laser trabeculoplasty
2. Trabeculectomy
3. Glaucoma drainage devices
4. Cyclodestructive procedures

**III. Describe the follow-up care**

- A. Consider therapeutic one-eyed trial**
- B. Check IOP 2-4 weeks after starting therapy to allow for IOP lowering and side effects to manifest themselves**
- C. Monitor side-effects**
- D. Follow IOP, optic nerve appearance, and visual fields**

**IV. Describe appropriate patient instructions**

- A. Discuss possible side effects and their signs and symptoms**
  1. With topical route
    - a. Metallic taste
    - b. Allergic
    - c. Dermatitis/conjunctivitis
  2. With oral route
    - a. Stevens-Johnson syndrome
    - b. Malaise, anorexia, depression
    - c. Serum electrolyte abnormalities
    - d. Renal calculi
    - e. Blood dyscrasias (aplastic anemia, thrombocytopenia)
    - f. Metallic taste
- B. Discontinue treatment for any serious side effect**
- C. Check serum potassium and use a potassium supplement, especially if already taking a potassium-wasting diuretic for hypertension (this applies to patients taking systemic CAIs)**

Prostaglandin Analogues

## **I. List the indications and contraindications**

### **A. Indications**

1. Initial and adjunctive therapy to lower intraocular pressure (IOP)
  - a. Open-angle glaucoma and ocular hypertension
  - b. Primary angle closure
  - c. Secondary glaucoma

### **B. Contraindications**

1. Macular edema
2. History of herpetic keratitis

## **II. List the alternatives to this therapy**

- A. Beta-adrenergic antagonists**
- B. Alpha-adrenergic agonists**
- C. Carbonic anhydrase inhibitors (topical and oral)**
- D. Parasympathomimetic agents**
- E. Laser surgery**
- F. Surgical therapy**

## **III. Describe the follow-up care**

- A. Evaluate response to therapy in 4-6 weeks**
- B. Inquire about side effects**
- C. Examine for evidence of ocular toxicity**

## **IV. Describe appropriate patient instructions**

- A. Educate patient regarding possible side effects, especially hyperemia and iris, lash, eyelid and skin pigmentary changes**
  1. Cystoid macular edema
  2. Conjunctival injection
  3. Increased eyelash growth
  4. Periocular pigmentary change
  5. Iris color change
  6. Uveitis

7. Possible herpes virus activation

**B. Instruct patient to wipe excess medication off peri-ocular skin**

**C. Discuss shelf-life of agent:**

1. Once opened, latanoprost may be stored at room temperature for up to 6 weeks. Unopened bottles should be refrigerated. Other agents are stored at room temperature.
2. Tafluprost preservative-free should be stored in the refrigerator in the original pouch. Once the pouch is opened, the single-use containers may be stored in the opened foil pouch for up to 28 days at room temperature. Unused containers must be discarded after 28 days.

## Hyperosmotic agents

### I. List the indications/contraindications

**A. Indications**

1. Short-term or emergency treatment of elevated intraocular pressure (IOP)
2. Useful in acute conditions of elevated IOP (e.g., angle-closure glaucoma)
3. Effective when elevated IOP renders iris non-reactive to agents which combat pupillary block such as the parasympathomimetic agents (e.g., pilocarpine)
4. Used to lower IOP and/or reduce vitreous volume prior to initiation of surgical procedures

**B. Contraindications**

1. Should not be used for long-term therapy (becomes ineffective with repeated dosing)
  - a. Glycerin may increase blood sugar levels (may be contraindicated in patients with diabetes)
  - b. Long-term use may perturb electrolytes
2. Of limited value when blood-ocular barrier is disrupted
3. May cause rebound elevation in IOP if agent penetrates eye and reverses osmotic gradient
4. Congestive heart failure
5. Renal failure

### II. List the alternatives to this therapy

- A. Aqueous suppressants (i.e., beta-adrenergic antagonists, topical and/or oral carbonic anhydrase inhibitors, alpha-adrenergic agonists)**
- B. Outflow enhancers (i.e., prostaglandin analogues, sympathomimetic agents, epinephrine-like agents)**
- C. Laser surgery procedures to correct acute glaucoma (e.g., iridotomy and/or iridoplasty for acute angle-closure glaucoma)**
- D. Paracentesis**

- E. Glaucoma surgical procedure (e.g., trabeculectomy, tube shunts, etc.)**

### **III. List complications of this therapy, their prevention and management**

#### **A. Complications: worse with mannitol**

1. Headache
2. Backache
3. Nausea and vomiting (oral agents)
4. Urination frequency and retention
5. Cardiac (chest pain, pulmonary edema, congestive heart failure)
6. Renal impairment
7. Neurologic status (lethargy, seizures, obtundation)
8. Subdural and subarachnoid hemorrhages
9. Hypersensitivity reactions
10. Hyperkalemia or ketoacidosis (when glycerin is given to patients with diabetes)

#### **B. Prevention**

1. Consideration of alternative therapies
2. Cautious use in patients with known compromised cardiac or renal status
3. Avoid use of glycerin in patients with diabetes
4. Close observation for complications

#### **C. Management**

1. Discontinue medication
2. Symptomatic relief of side effect until resolution if applicable
3. Consider urinary catheter (if IV mannitol given preoperatively)

### **IV. Describe the follow-up care**

- A. Close monitoring of IOP (to determine efficacy of hyperosmotic agents)**
- B. Discontinuation of therapy as soon as possible**
- C. Close monitoring of ocular and systemic symptoms and exam**

### **V. Describe appropriate patient instructions (post-op care, vision rehabilitation)**

- A. Alerting physician of any complications**
- B. Substitution of IOP-lowering agents when hyperosmotic agents no longer needed**

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma 2013-2014.



# Visual field testing

## **I. List indications/contraindications**

- A. Functions as a quantification of central and peripheral visual fields**
- B. Visual field indicated as part of afferent visual pathway assessment for screening, cases of unexplained visual loss, or suspected abnormality of afferent visual function**

## **II. List the techniques/alternatives to this procedure**

- A. Confrontation visual field testing**
- B. Kinetic perimetry**
- C. Automated static perimetry**
- D. Tangent screen perimetry**
- E. Amsler grid testing (for central ten degrees)**

## **III. Describe the instrumentation and technique**

### **A. Confrontation visual field testing**

1. Examiner compares patient's visual field to his own
  - a. Patient is seated opposite the examiner and directed to cover one eye and fixate on examiner's opposite eye or nose
  - b. Patient asked to quantify fingers presented in each of the four quadrants in a plane between the patient and examiner
  - c. Patient may be asked to add or to compare targets simultaneously presented in opposing quadrants (double simultaneous stimulation)
  - d. Aphasic, uncooperative, sedated, intubated or very young patients can use finger mimicry, pointing, visual tracking (preferential look) or reflex blink to respond and allow gross appraisal of visual field integrity
2. List advantages/disadvantages
  - a. Advantages
    - i. Rapid screening
    - ii. Additional instrumentation not necessary
  - b. Disadvantages
    - i. Insensitive to subtle visual field loss
    - ii. Poor patient comprehension or cooperation may limit usefulness
    - iii. Poor interexaminer reliability

## **B. Kinetic perimetry**

1. Place appropriate corrective lens corrected for refractive error for near vision
2. Patient head is placed in testing apparatus
  - a. Automated or manual strategies introduce varying size or intensity targets
  - b. Isopter plots of different sensitivity generated based on varying size or intensity of kinetic stimulus
3. List advantages/disadvantages
  - a. Advantages
    - i. Technician may test/retest problem areas
    - ii. Technician can monitor patient for reliability/responsiveness and adjust testing speed accordingly
    - iii. Patient-observer interaction may benefit patients with more difficulty performing longer testing
  - b. Disadvantages
    - i. Technician dependent
      - i) Decreased interexaminer reliability compared with automated perimetry
    - ii. Instrumentation availability increasingly less common

## **C. Automated static perimetry**

1. Patient's head is placed in testing apparatus
  - a. Place appropriate corrective lens corrected for refractive error for near vision
  - b. Automated strategies introduce static stimuli of varying intensity
  - c. The computer records and estimates threshold at each test location in decibels
  - d. These threshold values are compared with age-matched normal values at each point, along with a statistical evaluation of the probability that each point value is abnormal and plotted on topographic grids
  - e. If a patient responds to a stimulus presented in the assumed physiologic blind spot location, a fixation loss is recorded
  - f. A false-positive response indicates the patient hit the buzzer when no light stimulus was presented
  - g. If a patient does not hit the buzzer when a stimulus of identical location and greater intensity to one previously detected is presented, this is considered a false-negative response
  - h. The mean deviation is a location-weighted summation of the differences from age-corrected normals at all tested points
  - i. Mild ptosis or dermatochalasis can be associated with depression of the superior visual field.
  - j. Increasing age, media opacities and pupillary miosis may cause diffusely decreased

sensitivity

2. List advantages/disadvantages

a. Advantages

- i. Reproducible
- ii. Standard strategies minimize inter-technician variability
- iii. Sensitive to subtle field loss and general depression
- iv. Can increase stimulus size in patients with severe visual loss

b. Disadvantages

- i. May require several tests to establish reliable baseline
- ii. Duration of test, physical, and cognitive requirements may be difficult for certain patient populations
- iii. Instrumentation expensive

**D. Tangent screen perimetry**

1. Patient placed opposite tangent screen with a single eye covered and appropriate near corrective lens

- a. Different size or color test objects introduced by examiner in either a static or kinetic manner

2. List advantages/disadvantages

a. Advantages

- i. Rapid testing
- ii. Patient-observer interaction may benefit patients with more difficulty performing longer testing

b. Disadvantages

- i. Reproducibility less than in automated tests
- ii. Intermediate sensitivity between confrontation field testing and other forms of perimetry

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-ophthalmology, 2013-2014.
2. Oxford University Press, Optic Nerve Disorders, 2nd Edition, 2007.
3. AAO, Focal Points: A Primer on Automated Perimetry, Module #8, 1993.
4. AAO, Focal Points: Automated Perimetry in Neuro-Ophthalmology, Module 6, 1995.

# Examination of the pupil

## I. List the indications

### A. Indications

1. A key component of the examination of the efferent and afferent visual system

## II. Describe the examination

### A. Assessment of size

1. Size in ambient light conditions
2. If anisocoria or abnormally reactive pupil, also assess:
  - a. Size at near and in the dark
    - i. Horner syndrome
  - b. Eyelid function
    - i. Ptosis e.g. third nerve palsy or Horner syndrome
  - c. Ocular motility
  - d. Is there light-near dissociation of response?

### B. Assessment of shape

### C. Assess for presence of

1. Pharmacologic dilation
  - a. Known or unknown exposure
  - b. Widely dilated pupil without eyelid ptosis, without accommodative response, and without ocular motility disturbance
  - c. Pharmacologic testing available
2. Surgical or traumatic change or damage
3. Nodules
4. Synechiae
5. Iris atrophy
6. Iris transillumination
7. Heterochromia

### D. Assessment for presence of relative afferent pupillary defect (RAPD)

1. Relative difference in afferent light input because of unilateral or asymmetric optic nerve dysfunction results in difference between direct and consensual light response

2. Hallmark of unilateral or asymmetric optic nerve disease
3. Relative afferent pupillary defect may occasionally be seen in amblyopic eye (low amplitude RAPD) or large asymmetric retinal conditions
4. Describe pertinent clinical features
  - a. Swinging flashlight test in which pupillary escape or early redilation is demonstrated when a light is brought to the eye with the damaged optic nerve and constriction occurs when the light is returned to the "good" eye
    - i. Relax accommodation with a distant target
    - ii. May be performed despite unilateral iris abnormality or unilateral dilation

### **III. Describe the instrumentation needed**

- A. Intense diffuse variable light source such as transilluminator**
- B. Ruler or pupil gauge to measure pupils helpful**

### **IV. List other tests**

- A. Be aware of availability of existence of eye drop testing for:**
  1. Adie tonic pupil
  2. Pharmacologic blockade of the pupil
  3. Horner syndrome
- B. Slit-lamp biomicroscopy of the pupil**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5, Neuro-Ophthalmology, 2013-2014.

# Horner syndrome

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Disturbance of the sympathetic pathway

### B. Define the relevant elements of the epidemiology of this disease

1. Idiopathic
  - a. Common with third order (postganglionic) Horner syndrome
2. Secondary
  - a. Cause determined with appropriate use of imaging

### C. List the pertinent elements of the history

1. Anisocoria - miosis of affected pupil
2. Complaints that the eye "looks smaller"
3. Anhydrosis
4. Presence of neck and/or facial pain or headache (suggestive of carotid dissection)
5. Recent neck trauma or vigorous cervical manipulation (potential cause of carotid dissection)
6. Birth trauma

### D. Describe pertinent clinical features

1. Anisocoria -miosis of affected pupil with dilation lag
  - a. Worse 1-2 seconds after darkness
2. Ptosis of upper eyelid or slight elevation of lower eyelid causing eye to appear smaller
3. Anhydrosis
4. Possible iris heterochromia (congenital) with less pigment of the affected side)

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Cocaine or Apraclonidine to confirm or rule out Horner syndrome confirm
2. Paredrine (hydroxyamphetamine test) to differentiate pre- and post-ganglionic Horner syndrome
3. If face of neck pain is present imaging is required to exclude carotid dissection

## II. Define the risk factors

### A. Recent history of neck or chest trauma/surgery, including central line placement

### B. History of cancer

### C. Vasculopathic risk factors

- D. Cluster headache**

### **III. List the most common or critical entities in the differential diagnosis**

- A. Essential (physiologic) anisocoria with unrelated ptosis**
- B. Adie tonic pupil**
- C. Pharmacological mydriasis/miosis**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.

# Neuroimaging

## Computed tomography

### I. List indications/contraindications

#### A. Indications

1. Globe and orbital trauma
2. Assessment of bony abnormalities including fractures
3. Detection of calcification in lesions
4. Assessment of intracranial hemorrhage
5. Orbital lesions
6. Assessment of extraocular muscles in patients with suspected thyroid eye disease (thyroid orbitopathy)
7. When magnetic resonance imaging (MRI) is contraindicated
  - a. Ferromagnetic foreign body
  - b. Pacemaker
  - c. Metallic cardiac valves
  - d. Incompatible intracranial aneurysm clips
  - e. Cochlear implants
  - f. Claustrophobia
  - g. Morbid obesity

#### B. Contraindications

1. Contrast media relatively contraindicated in setting of renal insufficiency or allergy to contrast dye
2. It is controversial whether pregnancy is a contraindication
  - a. If test is needed, discuss with neuroradiologist

#### C. Disadvantages

1. Soft tissue details can be lost when in close proximity to bony structures such as the orbital apex and optic canal and in the posterior fossa
2. Extensive dental fillings/appliances may cause artifacts
3. Potential deleterious effects of radiation exposures, especially in children with younger individuals at increasingly higher risk



## **II. Describe the considerations in interpretation of this diagnostic procedure**

- A. Provision of details of patient clinical information, differential diagnosis and expected location of pathology to radiologist will usually assure that the correct imaging sequences will be performed**

Magnetic resonance imaging

### **I. List indications/contraindications**

**A. Definition**

1. Magnetic Resonance Imaging (MRI) is a method to generate cross-sectional images of the body based on nuclear magnetic resonance without using ionizing radiation

**B. Indications**

1. Lesions requiring detailed tissue contrast
  - a. Demyelinating disease
  - b. Infarction
  - c. Neoplasia
  - d. Inflammation
  - e. Infection
  - f. Structural/developmental abnormalities
  - g. Extraocular muscle inflammation or infiltration pathology
  - h. Parasellar lesions

**C. Contraindications**

1. Trauma causing injury that could be exacerbated by manipulation of patient in scanner
2. Unstable medical condition
3. Metallic foreign body
4. Pacemaker
5. Claustrophobia
6. Large body size

**D. Disadvantages**

1. High expense
2. Dental filling and appliances may cause artifacts
3. Weight limit to most scanners

## **II. Describe the considerations in interpretation of this diagnostic procedure.**

- A. Provision of details of patient clinical information, differential diagnosis and expected location of pathology to radiologist helps to assure that the correct imaging sequences will be performed**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Optic Nerve Disorders, 1996, p.48, 60-63, 82-83, 95-98, 102, 104, 107, 112-113, 118, 121, 203-206.
3. AAO, Focal Points: Multiple Sclerosis and Optic Neuritis, Module #12, 2003, p.3-5.
4. AAO, Focal Points: Neuroimaging for the General Ophthalmologist, Module #8, 1998.

# Demyelinating optic neuritis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Demyelination of the optic nerve

### B. Define the relevant aspects of epidemiology of the disease

1. Typically patients are predominantly female in their 3<sup>rd</sup> to 5<sup>th</sup> decade

### C. List the pertinent elements of the history

1. Acute visual loss
2. Usually pain on eye movement
3. May have history of demyelinating symptoms or known diagnosis of multiple sclerosis (MS)

### D. Describe pertinent clinical features

1. Decreased visual acuity
2. Decreased color vision
3. Visual field defect
4. Afferent pupillary defect (if unilateral or asymmetric bilateral)
5. Majority of cases do not have disc edema

### E. Describe appropriate diagnostic/laboratory testing

1. Neuro-imaging

## II. Define the risk factors

### A. Female predominance

### B. History of MS

## III. List the differential diagnosis

### A. Anterior ischemic optic neuropathy

### B. Maculopathy in the presence of a normal appearing fundus

### C. Infiltrative optic neuropathy

### D. Compressive optic neuropathy

### E. Leber hereditary optic neuropathy

### F. Infectious optic neuropathy

#### **IV. Describe patient management in terms of treatment and follow-up**

##### **A. Magnetic resonance imaging (MRI) scan**

1. Identify white matter abnormalities consistent with demyelinating lesions
2. No treatment versus IV corticosteroids and immunomodulating agents based upon MRI findings supporting a diagnosis of demyelinating disease
  - a. Corticosteroids may enhance rate of recovery but not visual outcome

##### **B. Referral to a multiple sclerosis specialist for the management of neurological disorders if abnormal MRI scan or consistent neurological examination and history**

##### **C. Oral prednisone in standard doses contraindicated**

#### **V. List the complications of treatment, their prevention and management**

##### **A. Complications of corticosteroids**

#### **VI. Describe disease-related complications**

##### **A. Failure to recover vision is uncommon**

##### **B. High rate of progression to MS**

#### **VII. Describe appropriate patient instructions**

##### **A. Medication instructions**

##### **B. Discussion of relation to MS, risk factors, and possible immunomodulation**

##### **C. Referral to neurologist**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-ophthalmology, 2013-2014.
2. AAO, Optic Nerve Disorders, 1996, p.33-35, 55-73, 85.
3. AAO, Focal Points: Multiple Sclerosis and Optic Neuritis, Module #12, 2003, p.6-7.
4. Beck RW, Gal Mt, Bhatti MT. Visual function more than 10 years after optic neuritis: experience of the opt neuritis treatment trial. Am J Ophthalmol. 2004;137:77-83.

# Anterior ischemic optic neuropathy

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Vasculopathic damage to the anterior optic nerve
  - a. Non-arteritic ischemic optic neuropathy (NAION)
  - b. Arteritic ischemic optic neuropathy (a-AION) (most often associated with giant cell arteritis (GCA))

### B. Define the relevant aspects of the epidemiology of the disease

1. Non-arteritic
  - a. Age typically greater than 45 years
2. Arteritic
  - a. Disease of the elderly, rarely less than 60 years

### C. List the pertinent elements of the history

1. Acute, typically painless, visual loss
2. Symptoms of GCA (e.g. headache, jaw claudication, scalp tenderness, fever, weight loss, fatigue, myalgias, diplopia, antecedent amaurosis fugax, tenderness over the temporal arteries)

### D. Describe the pertinent clinical features

1. Decreased visual acuity
2. Relative afferent pupillary defect if unilateral or markedly asymmetric
3. Visual field defects
  - a. Typically altitudinal or arcuate
4. Optic nerve edema with or without associated peripapillary hemorrhages or cotton wool spots
5. Posterior pole ischemia (e.g., retinal edema, hemorrhages, cotton wool spots) in giant cell arteritis
6. Pallid optic nerve edema may be seen in a-AION
7. Small to absent optic nerve cup in fellow eye in NAION ("disc at risk")
8. Cup : disc > 0.2 should raise concern for a-AION/GCA
9. Visual recovery
  - a. Improvement of visual function limited

### E. Describe the appropriate testing and evaluation for establishing the diagnosis

1. Serologic
  - a. Erythrocyte sedimentation rate (ESR)

- b. C-reactive protein (CRP)
- 2. Assessment for vasculopathic risk factors
- 3. Systemic
  - a. Assess blood pressure
- 4. Temporal artery biopsy if elevated ESR or CRP or if symptoms/ signs suggestive of GCA

## **II. Define the risk factors**

### **A. Non-arteritic**

- 1. Small to absent optic cup
- 2. Vasculopathic risk factors
  - a. Diabetes mellitus
  - b. Hypertension
  - c. Abnormal lipid profile

### **B. Arteritic**

- 1. Polymyalgia rheumatica

## **III. List the most common or critical entities in the differential diagnosis**

- A. Optic neuritis**
- B. Papillitis**
- C. Disc drusen / pseudopapilledema**
- D. Central retinal vein occlusion (CRVO)**

## **IV. Describe patient management in terms of treatment and follow-up**

### **A. Describe medical therapy options**

- 1. Non-arteritic
  - a. No proven therapy
- 2. Arteritic
  - a. Immediate institution of high dose oral or intravenous corticosteroids generally tapered over about one year
  - b. Temporal artery biopsy as soon as is possible
  - c. Referral to physicians experienced in the care of the systemic vasculitic disease manifestations and complications of chronic steroid therapy

## **V. Describe disease related complications**

### **A. Non-arteritic**

1. Vision loss generally ranges from 20/20 to counting fingers
2. Generally milder compared to a-AION
3. Many patients will develop second eye involvement many months to years after initial eye involvement

### **B. Arteritic**

1. Vision loss ranges from 20/20 to no light perception
2. Generally more severe compared to NAION
3. Many patients develop permanent severe visual loss
4. Second eye involvement occurs in majority of untreated patients and usually occurs within days to weeks of first eye involvement
5. Systemic effects of arteritis
6. Systemic effects of chronic steroid therapy

### **Additional Resources**

1. AAO, Basic and Clinic Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.

# Giant cell arteritis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Inflammatory vasculitis
  - a. Affects any of the branches of the arterial tree supplying the eye
    - i. Ophthalmic artery branches
    - ii. Retinal
    - iii. Choroidal

### B. Define the relevant aspects of epidemiology of the disease

1. Generally age greater than 60 years
2. Caucasian predominance

### C. List the pertinent elements of the history

1. Acute visual loss typically without eye pain
2. Symptoms of giant cell arteritis (GCA) (e.g., headache, jaw claudication, scalp tenderness, fever, weight loss, fatigue, myalgias diplopia, antecedent amaurosis fugax)
  - a. History of polymyalgia rheumatica

### D. Describe pertinent clinical features

1. Decreased visual acuity
  - a. Often severe visual loss
  - b. May be bilateral at presentation, however onset usually sequential
2. Diplopia
3. Antecedent amaurosis fugax
4. Relative afferent pupillary defect if unilateral or significantly asymmetric
5. Visual field defect
6. Pallid optic nerve edema with or without associated peripapillary hemorrhages or cotton wool spots
7. Retinal ischemia (e.g., edema, hemorrhages, cotton wool spots, arterial occlusive disease)

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Serologic
  - a. Erythrocyte sedimentation rate (ESR) and/or C-reactive protein (CRP)
    - i. Typically elevated



2. Temporal artery biopsy if elevated acute phase reactants (ESR, CRP) or symptoms/signs suggestive of GCA
  - a. Obtain biopsy as soon as possible
  - b. Do not delay corticosteroids because of biopsy
  - c. Obtain adequate sample of tissue

## **II. Define the risk factors**

- A. History of polymyalgia rheumatica

## **III. List the most common or critical entities in the differential diagnosis**

- A. Non-arteritic anterior ischemic optic neuropathy
- B. Papillitis
- C. Disc drusen (pseudopapilledema)
- D. Visual loss from other vasculitides

## **IV. Describe patient management in terms of treatment and follow-up**

- A. Describe medical therapy options
  1. Oral intravenous corticosteroids started immediately (i.e. never postpone corticosteroids pending TABx), then generally tapered over many months
  2. Referral to specialists involved in management and care of the disease and long term complications of therapy

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Optic Nerve Disorders, 1996, p. 79-81.
3. AAO, Focal Points: Temporal Artery Biopsy, Module #7, 2003.
4. AAO, Focal Points: Neuro-Ophthalmic Diagnosis, Module #9, 1999, p. 4-5.

# Papilledema

## I. Describe the approach to establishing the diagnosis

### A. Definition

1. Bilateral disc edema (secondary to elevated intracranial pressure)
2. Other forms of disc edema not related raised intra cranial pressure typically have visual loss and may be related to inflammation i.e. papillitis

### B. List the pertinent elements of the history

1. Risk factors for increased intracranial pressure (ICP) depend on cause
  - a. Hydrocephalus or brain tumor
  - b. Idiopathic intracranial hypertension (pseudotumor cerebri)
    - i. Typically overweight
    - ii. Female predominance
    - iii. Vitamin A intake
    - iv. Medications (antibiotics, corticosteroids, isotretinoin)
  - c. Infectious meningitis
    - i. Recent exposure to virus, tuberculosis or bacteria
2. Symptoms of increased ICP
  - a. Headache
  - b. Transient visual obscurations
  - c. Intracranial noises (humming or ringing)
  - d. Diplopia
  - e. Decreased vision

### C. Define the relevant aspects of epidemiology of the disease

1. Hydrocephalus or brain tumor
  - a. Any age or gender possible
2. Pseudotumor cerebri
  - a. Women of childbearing age
3. Infection
  - a. Any age or gender possible

### D. Describe pertinent clinical features

1. Bilateral nerve fiber layer opacification

2. Lack of spontaneous venous pulsations
3. Hemorrhages, exudates, macular edema present in fully developed papilledema
4. Central visual function usually normal (central visual acuity, color, pupils and visual fields) early in course
5. Peripheral vision may be abnormal early in course
6. Cranial nerve VI palsy may be present
7. Optic nerve head pallor may be present if chronic
8. Small telangiectatic vessels on disc surface may be present

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

1. Neuroimaging (magnetic resonance imaging (MRI) preferred)
2. Lumbar puncture to demonstrate elevated intracranial pressure; check opening pressure, cells, protein and glucose if no mass on scan

**II. List the most common or critical entities in the differential diagnosis**

- A. Pseudopapilledema (optic nerve head drusen, hyperopic or anomalous discs)**
- B. Malignant hypertension**
- C. Bilateral disc edema not due to increased ICP, e.g., papillitis typically associated with early visual loss**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Optic Nerve Disorders, 1996, p.40-41.

# Optic atrophy

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Pace of visual loss
  - a. Gradually progressive
    - i. May indicate a compressive or infiltrative lesion
  - b. Sudden
    - i. May indicate
      - i) Ischemic
      - ii) Inflammatory
      - iii) Leber hereditary optic atrophy
      - iv) Pituitary apoplexy
2. Associated pain
  - a. Optic neuritis
3. Associated disorders
  - a. Vasculopathic diseases
  - b. Multiple sclerosis
  - c. Rheumatologic disease
  - d. Parasellar benign or malignant tumors with compression
4. Family history of visual loss
5. Drug history
6. Diet history
7. Smoking history
8. Previous trauma (blood loss, hypotension, cardiac arrest, prolonged surgery)

### B. Describe pertinent clinical features

1. Decreased visual acuity
2. Dyschromatopsia
3. Visual field defect
4. Relative afferent pupillary defect in unilateral or asymmetric cases
5. Nerve fiber layer loss

### C. Describe appropriate testing and evaluation for establishing the diagnosis

1. Color vision testing
2. Visual field testing
3. Neuroimaging of the optic nerve and occasionally brain
  - a. MRI is typically more sensitive but CT also has a role
4. Serologic
  - a. Tailor based upon age, history, clinical features, associated diseases
5. Lumbar puncture
  - a. Infrequently required

## **II. List the most common optic neuropathies in the differential diagnosis**

- A. Post-ischemic**
- B. Compressive**
- C. Post-inflammatory/demyelinating**
- D. Infiltrative**
- E. Toxic/metabolic**
- F. Posttraumatic**
- G. Hereditary (e.g. Leber hereditary optic, neuropathy, dominant optic atrophy)**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Electrophysiologic Testing, 2nd ed., 2001, p.101, 219-223, 252.
3. AAO, Optic Nerve Disorders, 1996, p. 31-32, 42-43, 46, 79, 93-95, 152-153.

# Anatomy of the visual sensory system

## I. Anatomy of pupillary light reflex

### A. Clinically relevant anatomic correlations

1. Afferent limb of pupillary light reflex
  - a. Optic nerve
  - b. Hemidecussation of pupillary fibers at optic chiasm to enter optic tract
    - i. greater than 50% of pupillary fibers cross
      - i) **Significance is** an optic tract lesion produces a **contralateral APD**
  - c. Optic tract: pupillary fibers leave optic tract just **prior to** the **lateral geniculate** nucleus and enter brainstem ultimately synapsing with bilateral Edinger-Westphal nuclei in midbrain
    - i. **Significance is** light input to one eye produces bilateral pupillary constriction (i.e. direct and consensual response)
2. Efferent limb of pupillary light reflex
  - a. Edinger-Westphal nucleus (part of CN III nuclear complex in midbrain)
  - b. Ciliary ganglion (**synapse**)
  - c. Pupillary sphincter muscle

## II. Chiasmal/perichiasmal pathology

### A. Visual field loss with or without loss of visual acuity

1. Bitemporal hemianopia
  - a. **Significance is** suggests **compressive lesion** of chiasm (e.g. pituitary tumor, meningioma, craniopharyngioma) though can also occur with inflammatory, traumatic, and infiltrative processes
2. Junctional scotoma- Central scotoma with contralateral superior temporal defect
  - a. **Significance is** localizes lesion to **prechiasmal site** involving optic nerve ipsilateral to the central scotoma
3. Homonymous hemianopia when optic tract involved

## III. Disorders of the lateral geniculate and optic tract

### A. Anatomic structure relates to clinical findings

1. Lateral geniculate and optic tracts contain visual sensory fibers from contralateral hemifield of each eye which originate from retinal ganglion cell

2. The organization of these fibers becomes increasingly more congruous as fibers course from retina to the geniculate and posteriorly to the occipital cortex.
  - a. **Significance is** topographical arrangement leads to a variety of homonymous field cuts which helps to localize disease processes based upon congruity
  - b. **Significance is** homonymous defects indicate retrochiasmal pathology and require an imaging study of brain (preferably MRI with contrast)

#### IV. Disorders of the retrogeniculate visual pathway

##### A. In the retrogeniculate structures, fibers contain visual information from contralateral hemifield of each eye

1. **Significance is** lesions of these pathways produce homonymous field cuts that respect the vertical midline.

##### B. Anatomy of occipital lobe

1. Calcarine cortex
  - a. Located in the posterior occipital lobe
  - b. Contains homonymous fibers from contralateral visual field

##### C. Pathophysiology

1. Vascular - most common
  - a. Infarction of arterial vessels
  - b. Thromboembolic
  - c. Hemorrhagic events
2. Neoplastic - rare
  - a. Primary
  - b. Metastatic
3. Trauma - rare
  - a. Surgically iatrogenic
  - b. Closed head injury
  - c. Penetrating trauma
4. Inflammatory - rare (e.g. demyelinating disease)

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-ophthalmology, 2013-2014.

# Headache

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Daily pattern (AM or PM)
2. Location (unilateral, bilateral, localized or diffuse)
3. Precipitating factors
4. Patient over 60 years of age: consider giant cell arteritis (GCA) (See Giant cell arteritis)
5. Migraine
  - a. Recurrent typical patterns
  - b. Nature (unilateral throbbing)
  - c. Nausea
  - d. Light sensitivity, sound sensitivity
  - e. Fortification scotoma/visual aura
6. Headache associated with elevated intracranial pressure
  - a. Diffuse, constant headache
  - b. May awaken patient from sleep
  - c. Worsened with Valsalva maneuvers
  - d. Transient visual obscurations
  - e. Intracranial noises (humming or ringing)
  - f. Double vision
  - g. Decreased vision
  - h. Emesis may occur without nausea
  - i. Changes in mentation
  - j. Focal neurologic signs
7. Cluster headache
  - a. Excruciating bouts of pain localized behind one eye
  - b. Pain may awaken patient from sleep
  - c. Occurs in clusters over days or weeks, then remits for months or years

### B. Describe pertinent clinical features

1. Features of giant cell arteritis



- a. See outline
- 2. Migraine
  - a. May be induced by hormonal changes, certain foods or wines or other stresses
  - b. Migraine with aura (classic migraine)
    - i. Preceded by aura
    - ii. Imagery scintillating positive phenomena
    - iii. Followed by severe contralateral throbbing headache
    - iv. Last several hours
- 3. Migraine without aura (common migraine)
  - a. No preceding neurologic symptoms
- 4. Raised intracranial pressure headache
  - a. Papilledema
  - b. Other neurologic signs/symptoms
  - c. Non-localizing sixth nerve palsy
- 5. Cluster headache
  - a. Ipsilateral tearing
  - b. Conjunctival injection
  - c. Rhinorrhea
  - d. Ipsilateral Horner syndrome

## **II. Describe patient management in terms of treatment and follow-up**

### **A. Specific to type of headache**

- 1. If suspect GCA, then
  - a. Obtain immediate ESR, CRP
  - b. Initiate immediate corticosteroids
  - c. Orchestrate further evaluation
- 2. Otherwise, may consider referral to neurologist for evaluation
  - a. Neuro-imaging may be indicated
- 3. If optic nerve edema associated with headache, proceed with urgent further evaluation and management
- 4. If bilateral optic disc edema, proceed with further urgent evaluation and management

### **III. Describe appropriate patient instructions**

- A. Report change in character, frequency or duration of symptoms**
- B. Report new visual or neurologic symptoms**
- C. Eliminate precipitating or contributing factors**

#### **Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 5: Neuro-Ophthalmology 2013-2014.

# Restrictive strabismus

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Thyroid eye disease
2. Orbital disease
3. Space-occupying lesion
4. Orbital fractures
5. Previous ocular, conjunctival or orbital surgery, trauma and subsequent surgery
6. Long standing non-restrictive strabismus
7. Congenital fibrosis

### B. List the pertinent elements of the history

1. Double vision
2. Blurry vision
3. Pain with eye movement

### C. Describe pertinent clinical features

1. Incomitant ocular deviations
2. Evidence of orbital involvement
  - a. Enophthalmos
  - b. Proptosis
  - c. Eyelid position abnormality

### D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Forced duction testing
2. Duction and version testing
3. Alternate cover testing
4. Orbital neuroimaging in selected cases
5. Gaze evoked changes in intraocular pressure

## II. Define the risk factors

### A. Previous surgery

### B. Previous trauma

### C. Longstanding strabismus

- D. Orbital inflammation
- E. Thyroid disease

**III. List the most common or critical entities in the differential diagnosis**

- A. Paralytic strabismus
- B. Decompensated phoria

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.

# Third (oculomotor) cranial nerve palsy/paresis

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Binocular diplopia common
2. Ptosis of the ipsilateral upper eyelid typical
3. May be painful
4. Onset usually sudden, but may be gradually progressive
5. Pertinent medical history:
  - a. Vasculopathic disease risk factors
    - i. Diabetes mellitus
    - ii. Systemic hypertension
    - iii. Abnormal serum lipids
  - b. Cerebrovascular disease
  - c. Recent head trauma
6. Other neurological symptoms and history
  - a. Brain tumors
  - b. Aneurysms
  - c. Systemic malignancy

### B. Describe pertinent clinical features

1. Ipsilateral limitation of elevation, depression and adduction in the affected eye
2. Ptosis of the ipsilateral upper eyelid
3. Pupil findings
  - a. Dilated or sluggish pupil
    - i. Rule-out compressive lesions (e.g. aneurysm, tumor)
    - ii. Isolated pupillary dilation without accompanying ptosis and/or diplopia is essentially never a manifestation of CN III palsy in an awake adult
  - b. Pupil sparing
    - i. Normal pupil in the setting of a complete third nerve palsy (i.e. complete ptosis and complete ophthalmoplegia of extraocular muscles innervated by the third cranial nerve) virtually excludes aneurysmal compression

- c. Third nerve palsies may arise from damage or lesions within the brainstem subnuclei and are termed "nuclear." Other forms also exist
- d. Nuclear nerve palsies may produce ptosis and motility deficit in both eyes

**C. Describe appropriate testing and evaluation for establishing the diagnosis**

1. Observation of an isolated unilateral (i.e. no other neurologic or ophthalmologic findings) third nerve palsy for 3-4 months is appropriate in adults  $\geq 55$  years old only if:
  - a. Complete ptosis
  - b. Complete external ophthalmoplegia of extraocular muscles innervated by the third cranial nerve
  - c. The affected eye has a normally reactive pupil without pathologic anisocoria
  - d. No history of systemic malignancy
    - i. Prompt referral if criteria above not met, for the development of additional neurologic signs/symptoms, or if fails to resolve over 3-4 months
    - ii. Imaging typically indicated
2. Assess for vasculopathic disease risk factors (See II.A.1-3)

**II. Define the risk factors**

**A. Vasculopathic disease risk factors**

1. Diabetes mellitus
2. Hypertension
3. Abnormal serum lipids

**B. Cerebrovascular disease**

**C. Systemic disease known to be associated with the formation of aneurysms**

**D. Systemic malignancy**

**III. List the most common or critical entities in the differential diagnosis**

**A. Myasthenia gravis**

**B. Adie pupil**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Focal Points: Third, Fourth and Sixth Cranial Nerve Palsies, Module #8, 1996.

# Fourth (trochlear) cranial nerve palsy/paresis

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Binocular vertical or oblique diplopia
2. May describe torsion
3. Head trauma

### B. Describe pertinent clinical features

1. Hyperdeviation increasing on contralateral gaze
2. Head tilt may be present
3. Increased vertical fusional amplitudes indicates congenital

### C. Describe appropriate laboratory testing for establishing the diagnosis

1. Neuroimaging of the head and orbits in selected patients where not clearly isolated, post-traumatic, microvascular, or congenital in origin

## II. Define the risk factors

### A. Cerebrovascular disease

### B. Cardiovascular disease risk factors (e.g. diabetes, hypertension)

### C. Trauma

## III. List the most common or critical entities in the differential diagnosis

### A. Myasthenia gravis

### B. Skew deviation

### C. Thyroid eye disease

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology 2013-2014.
2. AAO, Focal Points: Third, Fourth, and Sixth Nerve Palsies, Module #8, 1996, p. 5-8.

# Abducens nerve palsy

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Binocular horizontal diplopia worse in ipsilateral gaze
2. Onset may be sudden or gradual
  - a. Congenital form does exist
3. Pertinent medical history
  - a. Vasculopathic disease risk factors
    - i. Diabetes mellitus
    - ii. Systemic hypertension
    - iii. Abnormal lipid profile
  - b. Cerebrovascular disease
  - c. Multiple sclerosis
  - d. Symptoms of raised intracranial pressure (ICP)
4. Elicit neurological symptoms and history that might suggest
  - a. Other cranial nerve involvement (II - VIII)
  - b. Presence of symptoms suggesting giant cell arteritis (GCA)

### B. Describe pertinent clinical features

1. Limitation of abduction ipsilateral to the side of the lesion
2. Esotropia that is greatest with ipsilateral gaze (i.e. when looking toward the side of the lesion)
3. Patient may have a head turn toward the side of the lesion
4. Impairment of cranial nerves (CN) II and III, IV and/or V suggests an orbital apex lesion
5. Impairment of adjacent CN (V, VII, VIII) suggests a lesion of the cerebellopontine angle
6. Involvement of CN III, IV, and/or V suggests cavernous sinus involvement

### C. Describe appropriate testing and evaluation for establishing the diagnosis

1. If palsy is truly isolated, initial observation is reasonable in adults greater than 50 who do not have a history of malignancy
2. Referral for additional testing and management for patients with other neurologic signs/symptoms, progressive motility disturbance, or persistent palsy
  - a. Neuro-imaging may be indicated
3. For patients with systemic symptoms suggesting GCA, an erythrocyte sedimentation rate and C reactive protein may be indicated



## **II. Define the risk factors**

### **A. Vasculopathic risk factors**

1. Diabetes mellitus
2. Hypertension
3. Abnormal lipid profile

### **B. Cerebrovascular disease**

### **C. Multiple sclerosis**

## **III. List the most common or critical entities in the differential diagnosis**

### **A. Neural causes**

### **B. Duane syndrome**

### **C. Myopathic causes**

1. GCA
2. Thyroid eye disease (Graves ophthalmopathy, thyroid orbitopathy)

### **D. Myasthenia gravis**

## **IV. Describe patient management in terms of medical treatment and follow up**

### **A. Management depends upon the underlying condition**

1. Lesions due to small vessel ischemia will generally resolve within several months without specific treatment
  - a. These patients should be referred to their primary care physician for cardiovascular risk factor assessment
2. Occluder to alleviate double vision
3. Prism if relatively comitant
4. Muscle surgery for chronic palsy if stable

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Focal Points: Third, Fourth and Sixth Cranial Nerve Palsies, Module #8, 1996, p.8-10.

# Myasthenia gravis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Immunological disorder characterized by weakness with repetitive movements of a muscle group
2. Acetylcholine receptor sites are blocked

### B. List the pertinent elements of the history

1. Diplopia
2. Asymmetric eyelid droop
3. Difficulty with swallowing
4. Systemic weakness
5. Shortness of breath
6. Fatigability
7. Variability

### C. Describe pertinent clinical features

1. Ophthalmic signs & symptoms
  - a. Ptosis
  - b. Diplopia
  - c. Extraocular muscle (EOM) motility disturbance
  - d. Orbicularis weakness
  - e. Fatigability
  - f. Normal pupil
  - g. Absence of pain
2. Neurologic signs & symptoms
  - a. Muscle weakness
3. Associated conditions
  - a. Thyroid eye disease (thyroid orbitopathy)
  - b. Other autoimmune diseases

### D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Edrophonium chloride ("Tensilon") test
2. Ice test or sleep test
3. Acetylcholine receptor antibody test

4. Imaging scan of chest to rule out thymoma or other thymus abnormality

## **II. Describe disease-related complications**

- A. Progression of ocular to systemic myasthenia**

## **III. Describe appropriate patient instructions**

- A. Advise of need for assessment by neurologist and/or neuro-ophthalmologist**
- B. Call immediately for**
  1. Shortness of breath
  2. Difficulty swallowing
- C. Inform physicians that you have myasthenia gravis**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Focal Points: Myasthenia Gravis, Module #4, 2003.
3. AAO, Focal Points: Third, Fourth and Sixth Cranial Nerve Palsies, Module #8, 1996.

# Orbital tumor causing neuro-ophthalmic manifestations

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Diplopia
2. Decreased vision
3. Pain
4. Proptosis
5. Sinus symptoms
6. Numbness
7. History of malignancy

### B. Describe pertinent clinical features

1. Proptosis
2. Restrictive strabismus
3. Decreased vision

### C. Describe appropriate testing and evaluation for establishing the diagnosis

1. Neuroimaging of the orbit and disease directed surrounding tissue
  - a. Magnetic resonance imaging (MRI) typically requires fat suppressed images with contrast to provide adequate differentiation and resolution of lesion against orbital fat
  - b. Computed tomography (CT)
2. Orbital ultrasound

## II. List the most common or critical entities in the differential diagnosis

### A. Non-specific orbital inflammation (also known as orbital pseudotumor)

### B. Vascular malformations

1. Carotid cavernous fistula (CCF)
2. Dural arteriovenous malformation (AVM)

### C. Orbital manifestation of systemic disease (e.g. thyroid eye disease)

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.

# Orbital hemorrhage

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Traumatic
  - a. Accidental
  - b. Iatrogenic - following retrobulbar injection or intraoperative or postoperative
2. Anticoagulation—exogenous or disease state
3. Spontaneous bleeding of a vascular anomaly

### B. Describe pertinent clinical features

1. Pain and proptosis
2. Conjunctival hemorrhage
3. Signs of optic neuropathy
  - a. Reduction in
    - i. Visual acuity
    - ii. Color vision
    - iii. Visual field
  - b. Presence of relative afferent pupillary defect (RAPD)
4. Restrictive strabismus
5. Possible IOP elevation

### C. Describe appropriate testing and evaluation for establishing the diagnosis

1. Diagnostic tools
  - a. Imaging of orbit
    - i. Avoid magnetic resonance imaging (MRI) for ferromagnetic foreign bodies, unknown composition of penetrating object, or other contraindications
  - b. Assessment of bleeding diathesis in appropriate clinical situation

## II. Define the risk factors

- A. Injury
- B. Bleeding diathesis
- C. Underlying orbital disease including orbital vascular anomaly
- D. Anticoagulation

### **III. List the most common or critical entities in the differential diagnosis**

- A. Orbital mass**
- B. Orbital cellulitis**
- C. Orbital inflammatory disease**
- D. Carotid cavernous sinus fistula**

### **IV. Describe patient management in terms of treatment and follow-up**

- A. Emergent canthotomy/cantholysis depending on examination findings**
- B. Patient assessment will determine need for appropriate neuro/orbital imaging study if necessary**
- C. Consider surgical or medical therapy if significant visual loss or other indication such as vision threatening proptosis**
  - 1. Corticosteroids may be a useful adjunct in certain cases
  - 2. Manage intraocular pressure
- D. Return to or contact ophthalmologist immediately if visual problems progress**

#### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 7: Neuro-Ophthalmology, 2013-2014.
- 2. AAO, Focal Points: Management of Orbital Trauma: Fractures, Hemorrhage, and Traumatic Optic Neuropathy, Module #7, 1994, p. 6-8.

# Periorbital anatomy

## I. Introduction

- A. Knowledge of periorbital anatomy is essential to understand the etiology and treatment of oculoplastic and orbital diseases
- B. A brief description of the important anatomy structures followed with related clinical application is included in this section
- C. More detail can be found regarding individual disorders described later in the outline

## II. Eyelid - structures identified from superficial to deep

### A. Skin

1. Anatomically thin, its structure allows rapid eyelid blinking
2. The eyelid skin contains structures known as the ocular adnexa. These include sweat glands, sebaceous glands, mucin producing glands and hair follicles
3. Susceptible to sun damage and an important risk factor for malignancy. Basal cell carcinoma is the most common skin cancer of the eyelid
4. Redundant skin in the upper lid is called dermatochalasis. Blepharoplasty is the surgical removal of the skin (and often underlying muscle and fat). Blepharoplasty may be performed for functional reasons such as interference of upper visual field as well as for cosmetic purposes

### B. Orbicularis muscle

1. The muscle is responsible for passive and forced eyelid closure
2. The orbicularis muscle extends from the eyelid margin to the eyebrow above and the cheek below. The muscle fibers interdigitate with the muscles of the forehead (the frontalis muscle) and the muscles of the glabella
3. Sagging of the eyebrow is known as brow ptosis
4. The orbicularis muscle is innervated by branches of the facial nerve (cranial VII), as are all muscles of facial expression
5. Weakness of the orbicularis can occur with generalized muscle problems or with facial nerve palsy. Lagophthalmos or incomplete blinking can result in corneal exposure
6. Over activity of the facial muscles
  - a. Isolated muscle fibers causing a quick twitch - myokymia
  - b. One side of the entire face in spasm - hemifacial spasm
  - c. Both eyes exhibiting uncontrolled closure - essential blepharospasm
  - d. Botulinum toxin can be used to decrease spasm of the orbicularis muscle
7. Botulinum toxin can be used to treat disorders of facial musculature over-activity as well as a

variety of cosmetic disorders

**C. Orbital septum**

1. The orbital septum is a fibrous layer separating the orbit from the eyelid
2. The septum extends from the eyelid to attach along the bony orbital rims
3. Infection of the eyelid, often due to external introduction of a pathogen (scrape or bite), is known as preseptal cellulitis. Orbital cellulitis, in contrast, is an infection affecting the deeper orbital tissues. Proptosis and motility disturbances are often present. This usually arises from an adjacent sinusitis

**D. Orbital fat**

1. The deep fat pads in the upper and lower eyelid lie posterior to the orbital septum and superficial to the levator aponeurosis in the upper eyelid and the eyelid retractors in the lower lid
2. Laxity of the septum with age allows the orbital fat to prolapse forward creating full or puffy eyelids

**E. Levator muscle**

1. The levator muscle retracts the upper eyelid. It is under voluntary control and is innervated by cranial nerve III
2. The muscle originates in the orbital apex and extends anteriorly into the eyelid. The eyelid portion of the levator muscle is broad tendon known as the levator aponeurosis that inserts on to the tarsal plate
3. Drooping of the upper eyelid is known as blepharoptosis or ptosis
4. Thinning of the levator aponeurosis is the assumed mechanism for most involutional ptosis. An abnormal development of the levator muscle is the etiology of most congenital ptosis
5. Correction of ptosis can be performed by directly shortening the levator muscle through the skin. Removal of a portion of conjunctiva and Müllers' muscle from the posterior aspect of the eyelid can be performed to correct ptosis

**F. Müllers muscle**

1. This thin muscle is under sympathetic control. It lies posterior to the levator aponeurosis and anterior to the conjunctiva
2. Loss of sympathetic innervation to the eyelid is known as a Horner's syndrome resulting in a mild upper eyelid ptosis

**G. Lower eyelid retractors**

1. Like the upper eyelid, the lower eyelid moves up and down with eye movements. The lower eyelid retractors are analogous to the levator and Müllers muscle in the upper eyelid
2. Laxity of the lower eyelid retractors plays an etiologic role in involutional entropion of the lower eyelid

**H. Tarsal plates**

1. The tarsal plates are fibrous structures. The orbicularis muscle is tightly attached to the anterior surface of the upper tarsal plate inferior to the which forms the skin crease, and gives that skin crease giving that portion of the eyelid a flat platform



2. Within the tarsal plates are the meibomian glands, specialized robust sebaceous glands, which contribute to the oil layer of tears
3. Inflammation and obstruction of the meibomian glands contributes to poor ocular lubrication and can cause eye irritation
4. A hordeolum results from acute blockage of a meibomian gland presenting with erythema and swelling of the affected portion of the eyelid. Eventually the inflammation is either resolved or sequestered into a localized mass known as a chalazion

#### **I. Canthal tendons**

1. The lateral canthal tendon attaches the eyelids to the lateral orbital rim
2. The medial canthal tendon attaches the eyelids to the medial orbital rim. This tendon has anterior and posterior heads that surround the lacrimal sac. Normal blinking opens and closes the sac to promote drainage of tears. This mechanism is known as "the lacrimal pump". Loss of normal blinking, as in facial nerve palsy, may delay tear drainage
3. Laxity of the canthal tendons results in horizontal eyelid laxity (eyelids are loose)
  - a. This laxity may result in lower eyelid ectropion
  - b. Tightening the lateral canthal tendon via a variety of surgical techniques is the treatment for lower eyelid ectropion

#### **J. Blood supply to the eyelids**

1. The eyelids are well vascularized. Rich anastomoses form from both the internal carotid artery and external carotid arteries (via the facial tissues) and contribute blood supply

#### **K. Anterior and posterior lamellae of the eyelid**

1. At the eyelid margin, the eyelids can be separated into two layers
  - a. The anterior lamella - skin and orbicularis muscle
  - b. The posterior lamella - tarsus and conjunctiva
2. The concept of an anterior and posterior lamella is important in understanding some eyelid disorders and their surgical correction
  - a. Scarring or shortening of the anterior lamella results in a cicatricial ectropion
    - i. Lengthening the anterior lamella using a full thickness skin graft corrects this
  - b. Scarring or shortening of the posterior lamella results in a cicatricial entropion
    - i. Lengthening the posterior lamella using a mucous membrane graft corrects this

### **III. Lacrimal**

#### **A. Lacrimal gland**

1. Aqueous tears are produced by the lacrimal gland, which sits in the lacrimal gland fossa of the superior and temporal corner of the orbit
2. Aqueous fluid tears drain into the orbit through ductules entering the superior conjunctival fornix

3. The gland may become inflamed, infected, obstructed or involved in benign or malignant neoplastic processes

**B. Accessory lacrimal glands**

1. Accessory lacrimal glands are found in the conjunctiva contributing to the aqueous basal lubrication of the eye

**C. The tear film**

1. Three layers of fluid coat the eye for protective lubrication
  - a. A mucous layer from conjunctival goblet cells
  - b. An aqueous layer arises from the main accessory lacrimal glands
  - c. An oil layer covers the aqueous layer preventing evaporation. The oil layer arises from the meibomian glands and other sebaceous glands along the eyelid margin
2. A deficiency in quality or quantity of any layer is disruptive to normal lubrication and can result in eye irritation

**D. Tear drainage**

1. Canaliculus
  - a. Tears enter the drainage system through an opening in the medial upper and lower eyelids known as the lacrimal puncta
  - b. Each punctum extends into a horizontal canaliculus. The upper and lower canaliculi typically unite in a common canaliculus adjacent to the nasolacrimal sac
  - c. Obstruction of one or both puncta or any portion of the canaliculus can cause tearing. Infection of the canaliculus, canaliculitis, results in a discharge and a swollen and erythematous medial eyelid. Treatment typically includes curettage
2. Lacrimal sac
  - a. The sac lies in the bony lacrimal sac fossa
  - b. The sac extends inferiorly to become the membranous portion of the nasolacrimal duct
3. Nasolacrimal duct
  - a. The duct refers to both the bony canal extending into the nose and the membranous tissue forming the duct
  - b. The opening of the duct is in under the inferior turbinate of the nose
  - c. Congenital nasolacrimal duct obstruction results when the nasal end of the duct (the valve of Hasner) fails to open after birth
  - d. Acquired nasolacrimal duct obstruction may result in tearing or signs of acute or chronic obstruction. Acute dacryocystitis is a painful swelling of the sac due to infection
  - e. Adult acquired nasolacrimal duct obstruction is usually treated with a dacryocystorhinostomy (DCR) operation

## IV. Orbit

### A. Bony orbit

1. The bony orbit is a pyramidal shaped space formed from four walls. The bones of the medial wall and floor of the socket are very thin. Trauma to the orbit may result in increased intraorbital pressure causing the medial wall or floor to "blowout"
2. Small fractures of the orbit can trap tissue and cause a restriction of motility and diplopia. Large fractures allow displacement of orbital tissues and can cause shifts in the position of the eye, such as enophthalmos
3. Several openings into and out of the bony orbit, foramina, exist
4. The optic canal extends out the posterior aspect of the orbit. The optic nerve (cranial nerve II) courses through the optic canal. Direct or indirect trauma to the canal can result in an optic neuropathy
5. The maxillary branch of the infraorbital nerve (cranial nerve V<sub>2</sub>) travels from the space posterior to the orbit into the orbit through the inferior orbital fissure and out to the cheek through the infraorbital foramen. Fracture of the orbital floor results in numbness of this nerve
6. Structures extending into or out of the orbit to the brain travel through the superior orbital fissure. These structures include cranial nerves III, IV, V and VI

### B. Orbital tissues

1. Aside from the globe, the orbit contains many structures including the extraocular muscles, the optic nerve, and the lacrimal gland. These tissues are enveloped in orbital fat. The orbital fat is organized by a fibrous architecture. Many blood vessels and nerves course through the orbit
2. Proptosis is the hallmark of orbital disease. An enlarged orbital structure or a space-occupying lesion will displace the eye
3. The most common cause of proptosis is thyroid eye disease. The proptosis is due to enlarged extraocular muscles, fat and connective tissue
4. A wide variety of orbital benign and malignant neoplasms are seen in the orbit. The most common benign neoplasm of the adult orbit is a cavernous hemangioma

### Additional Resources

1. AAO, Basic Clinical and Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

# External examination of the adnexa and orbit

## I. Eyelid

### A. Eyelid examination

1. Eyelid position
  - a. The normal upper eyelid rests 1-2 mm below the limbus; the normal lower eyelid rests at the limbus
  - b. Blepharoptosis is present when the upper eyelid resting position is low
  - c. Lid retraction is present when the upper eyelid rests above or the lower lid rests below the limbus
    - i. MRD - margin reflex distance
  - d. Eyelid excursion (levator function) is the movement of the upper eyelid from downgaze to upgaze.
  - e. The lid margin should be in contact with the eyeball without inversion (entropion) or eversion (ectropion)
  - f. Horizontal eyelid laxity is an etiologic factor in involutional ectropion and entropion
2. Eyelashes
  - a. Normal directed eyelashes are present from the punctum to the lateral canthus
  - b. Mucocutaneous junction should be identified to detect any subtle inversion of the eyelid that may misdirect the eyelashes (marginal entropion)
  - c. Misdirected eyelashes (trichiasis) that curve toward the ocular surface and produce erosion or inflammation of the cornea or conjunctiva should be epilated, destroyed, or redirected, depending on the etiology and severity of the problem

## II. Lacrimal examination

### A. Eyelid tension, medial or lateral canthal tendon laxity

### B. Cranial nerve (CN) VII function

1. Blink rate
2. Completeness blink
3. Presence of lagophthalmos

### C. Punctal position and patency

### D. Palpation of lacrimal sac

### E. Lacrimal gland position and size

**F. Lacrimal testing**

1. Dye disappearance testing
2. Canalicular palpation
3. Lacrimal irrigation
  - a. Probing of nasolacrimal duct is not a diagnostic procedure

**G. Ocular surface**

1. Tear film
2. Tear meniscus
3. Tear break up time
4. Corneal sensation

**III. Orbit**

**A. Position of the globe in orbit**

1. Proptosis is the hallmark of orbital disease
2. Laterally
3. Primary orbital tumors are usually unilateral
4. Bilateral orbital masses are usually lymphoid
5. Thyroid eye disease (Graves disease) is the most common cause of unilateral or bilateral proptosis
6. Direction of globe displacement
  - a. A mass pushes the eye opposite the direction of the mass
  - b. Downward (globe ptosis) - lacrimal gland mass, frontal mucocele
  - c. Lateral - ethmoid mucocele or subperiosteal abscess
  - d. Upward - rare, maxillary sinus tumor
  - e. Axial - intraconal mass or extraocular muscle enlargement (thyroid eye disease)
7. Enophthalmos
  - a. Increased orbital volume - fracture
  - b. Sclerosing orbital tumor - breast cancer
  - c. Small eye
  - d. Contralateral proptosis
8. Measurement of globe position by exophthalmometer

**B. Palpation**

1. Heat

- a. Infection
  - b. Inflammation
- 2. Tenderness
  - a. Infection
  - b. Inflammation
- 3. Shape
  - a. Cystic or irregular
  - b. Localized or diffuse
  - c. Fixed or mobile
  - d. "Bag of worms" plexiform neuroma

**C. Pulsation**

- 1. Vascular orbital tumor with arterial component
- 2. Bony orbital defect transmitting brain pulsation
- 3. Valsalva maneuver may increase proptosis if a varix is present

**D. Periocular skin**

- 1. Erythema
- 2. Edema
- 3. Ecchymosis
- 4. Café au lait spots
- 5. Periorbital skin malignancy

**Additional Resources**

- 1. AAO, Basic Clinical and Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
- 2. Nerad, J: Oculoplastic Surgery in the Requisites. Mosby 2001.

# Preseptal/orbital cellulitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Spread from structures
  - a. Sinus infection (especially ethmoiditis)
  - b. Orbital fracture
  - c. Dental abscess
  - d. Periorcular skin infection
2. Post-trauma or postsurgical
3. Systemic vascular spread
  - a. Bacteremia

### B. List the pertinent elements of the history

1. Onset
2. Duration
3. Malaise
4. Fever
5. Pain
6. Visual loss
7. Diplopia
8. Recent upper respiratory infection
9. History of sinus infection or sinus surgery
10. Facial trauma
11. Recent orbital surgery
12. Diabetes mellitus
13. Immunocompromised status

### C. Describe pertinent clinical features

1. Assessment of ocular function is essential because infection extending into the orbit may cause ischemia and permanent visual loss
2. Distinguish preseptal from orbital cellulitis
  - a. Preseptal cellulitis
    - i. Lid erythema

- ii. Edema
  - iii. Tenderness
  - iv. Full range of motion
  - v. Vision intact
  - vi. Pupil intact
- b. Orbital cellulitis may have the following:
  - i. Proptosis
  - ii. Chemosis
  - iii. Restricted extraocular movement
  - iv. Afferent system dysfunction (decreased acuity, visual field defect, afferent pupillary defect, optic disc changes)

**D. Describe appropriate testing and evaluation for establishing the diagnosis**

- 1. Complete blood count (CBC) with differential, blood cultures (especially if febrile or systemic illness known)
- 2. Neuro-imaging
  - a. Used to diagnose the source (typically sinus disease) and extent of infection
  - b. Subperiosteal abscess
    - i. Extension of an ethmoiditis into orbit, but contained by the periorbital
    - ii. May require drainage
  - c. Orbital abscess can be seen when pus collects within the orbital tissues
    - i. A less common but more dangerous situation

**II. Define the risk factors**

- A. Recent infection, tooth abscess trauma or surgery**
- B. Immunocompromised or patient with diabetes**
  - 1. At risk for fungal orbital or sinus infection

**III. List the most common or critical entities in the differential diagnosis**

- A. Orbital inflammations**
- B. Rapidly growing orbital tumors, typically metastatic**
- C. Orbital vascular abnormality (carotid cavernous fistula, superior ophthalmic vein thrombosis)**
- D. Other ocular pathology, including endophthalmitis or necrotic intraocular tumor**



#### **IV. Describe patient management in terms of treatment and follow-up**

##### **A. Describe medical therapy options**

1. Systemic antibiotic
2. Consider ear, nose, and throat (ENT) consultation and co-management

##### **B. Describe surgical therapy options**

1. If vision or orbital compromise, refer for possible surgical drainage and exploration
2. If unresponsive to medical therapy, refer for possible surgical drainage and exploration
3. Orbital or subperiosteal abscess should be referred for possible drainage

#### **V. Describe disease-related complications**

##### **A. Progression of preseptal cellulitis to orbital abscess**

##### **B. Visual loss**

##### **C. Intracranial involvement**

##### **D. Death**

#### **VI. Describe appropriate patient instructions**

##### **A. Oral or IV antibiotic therapy**

##### **B. Adequate follow-up with other specialists: e.g., ear, nose, and throat (ENT) for contiguous infections/etiology and prevention treatment**

##### **C. Monitoring of vision, symptoms and follow-up with ophthalmologist**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Orbital Cellulitis, Module #11, 1991.

# Thyroid eye disease (Graves ophthalmopathy, thyroid orbitopathy)

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Autoimmune disorder
  - a. May have other systemic autoimmune manifestations/signs such as myasthenia gravis
  - b. Most often hyperthyroid, but may be euthyroid or hypothyroid

### B. Describe the epidemiology

1. Female preponderance

### C. List the pertinent elements of the history

1. Symptoms of systemic hyperthyroidism or hypothyroidism
2. Smoking history
3. Family history
4. Discomfort
5. Vision loss
6. Ptosis
7. Diplopia
8. Rate of progression of visual symptoms
9. Presence of diurnal variation in symptoms

### D. Describe pertinent clinical features

1. Signs of orbital inflammation/congestion
2. Lid retraction
3. Lid lag on downgaze
4. Proptosis
5. Conjunctival edema
6. Corneal exposure
7. Extraocular motility restriction
  - a. Inferior and medial recti are two most commonly affected muscles
8. Intraocular pressure elevation with eyeball excursion
9. Compressive optic neuropathy can occur

- a. May occur without overt external signs
- b. May lead to a visual field loss

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

- 1. Endocrine evaluation
- 2. Orbital imaging
  - a. Computerized tomography (CT) scan
  - b. Magnetic resonance imaging (MRI) scan
  - c. Orbital ultrasonography

**II. Define the risk factors**

- A. Smoking**
- B. Female**
- C. Autoimmune diathesis**
- D. Family history**

**III. List the differential diagnosis**

- A. Orbital and preseptal inflammation/infection**
- B. Orbital tumors causing proptosis**
- C. Carotid cavernous fistula, dural arteriovenous malformation (AVM), cavernous sinus or orbital vein thrombosis**
- D. Myasthenia**

**IV. Describe patient management in terms of treatment and follow-up**

- A. Referral for metabolic evaluation and management**
- B. Prioritize management based on ocular symptoms and clinical signs**
  - 1. If optic neuropathy is suspected, referral is urgent
  - 2. Surface lubrication for comfort and corneal exposure
  - 3. Referral for evaluation regarding
    - a. Orbital decompression
    - b. Strabismus surgery
    - c. Eyelid surgery
- C. For patients who smoke, smoking cessation is necessary**

## **V. Describe disease-related complications**

- A. Decreased vision due to compressive optic neuropathy**
- B. Ocular irritation from exposure**
- C. Decreased vision due to corneal ulceration or perforation**
- D. Diplopia due to extraocular muscle restriction**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System; Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.75, 130.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.134-153.
4. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.13-17, 36-47.
5. AAO, Focal Points: Thyroid-Associated Orbitopathy, Module #1, 1997.

# Orbital fractures

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Caused by blunt orbital / periorbital trauma
2. All orbital fractures are not blow out fractures
3. Fractures of the orbit include
  - a. Fractures of the mid face
    - i. Involves portions of the orbital floor, medial wall, or rim
  - b. Zygomatic complex fractures (tripod fracture)
    - i. Involves inferior and lateral orbital rims, orbital floor
  - c. "Blow out" fractures
    - i. Fracture of orbital floor or medial wall
    - ii. Caused by increased orbital pressure or floor buckling
    - iii. The orbital rims remain intact

### B. List the pertinent elements of the history

1. History of trauma
2. Complaints may include decreased vision
3. Possible diplopia

### C. Describe pertinent clinical features

1. Periorbital swelling and ecchymosis
2. Epistaxis
3. Subcutaneous or orbital emphysema
4. Decreased sensation in distribution of infraorbital nerve (cheek and teeth)
5. Facial deformity
  - a. Orbital rim step
  - b. Flat cheek
6. Initially, exophthalmos is possible due to swelling
7. Incomitant strabismus
  - a. Most typically restrictive hypotropia, but can be variable depending on fracture type and location
8. Associated ocular or periocular trauma

- a. Rule out lacerations, ruptured globe, hyphema, vitreous hemorrhage, optic nerve damage, retinal detachment
- 9. Associated intracranial injury
- D. Describe appropriate testing and evaluation for establishing the diagnosis**
  - 1. Computerized tomography (CT) scan (axial and coronal views, 3mm cuts or finer)
  - 2. Magnetic resonance imaging (MRI) is not study of choice in the management of acute orbital trauma, because of desire to see bony anatomy
  - 3. Motility testing
  - 4. Forced duction testing may be useful

## **II. Define the risk factors**

- A. Patients under age 18 are at greater risk of small "trap door" floor fractures that entrap orbital tissues and lead to rapid infarction and permanent extraocular muscle restriction**
  - 1. Urgent referral is necessary

## **III. List the most common or critical entities in the differential diagnosis**

- A. For diplopia, consider cranial nerve damage or decompensated preexisting phoria**
- B. Forced duction testing will determine if movement problem is restrictive or parietic**

## **IV. Describe the patient management in terms of treatment and followup**

- A. Not all orbital fractures require repair**
- B. Indications for repair of blowout fracture**
  - 1. Enophthalmos of greater than 2mm, or a floor fracture involving greater than 50% of the orbital floor on CT scan
  - 2. Diplopia in primary position, demonstrated to be restrictive
- C. Timing of interventions**
  - 1. In adults, observation for 10 days is reasonable to see if diplopia or enophthalmos improves
  - 2. In children with a small fracture causing diplopia, urgent referral and intervention may limit muscle scarring
- D. Indications for surgical repair of other orbital fractures**
  - 1. Enophthalmos and strabismus indications as in blowout repair
  - 2. Facial deformity

## **V. Describe disease related complications**

- A. Permanent incomitant strabismus with diplopia**
- B. Enophthalmos**
- C. Facial asymmetry**
- D. Malocclusion of jaw (Le Forte)**

## **VI. Describe appropriate patient instructions**

- A. Limit nose blowing**
- B. Compliance with referrals for further evaluation and treatment**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System; Section 5: Neuro-Ophthalmology, 2013-2014.

# Periorbital soft tissue trauma

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Injury to the eyelids, midface orbit, or eye
2. Accidental trauma that may be blunt or sharp

### B. Define the relevant aspects of epidemiology of the disease

1. Participation in any activity or occupation with exposure to potential trauma i.e., motor vehicle accident, personal assault, sports or work- related injury
2. Dog bites in children
3. Surgical excision of malignancy

### C. List the pertinent elements of the history

1. History should include nature and circumstances of the injury
2. Sharp, penetrating injury may extend into globe or brain
3. Associated vision loss or neurologic deficit mandates further evaluation
4. Penetrating injury (e.g. wood or pencil injury) may leave foreign bodies in orbit
5. Child's account of injury may be inaccurate

### D. Describe pertinent clinical features

1. Examination should include evaluation of the globe including:
  - a. Visual acuity
  - b. Relative afferent pupillary defect (RAPD)
  - c. External examination
  - d. Intraocular pressure (IOP) measurement
  - e. Dilated fundus examination
2. Examination should include the following to rule out orbital fractures: (See Orbital fractures)
  - a. Globe malposition
  - b. Reduced motility
  - c. Infraorbital nerve hypesthesia
  - d. Step deformity at rim
  - e. Epistaxis
  - f. Subcutaneous emphysema
3. Neurologic examination to rule out associated head injury including:



- a. Alteration in mentation
  - b. Cerebrospinal fluid rhinorrhea
- 4. Soft tissue examination
  - a. Eyelid margin and canthal integrity
  - b. Trivial appearing lacerations in the medial canthal area may be associated with canalicular lacerations
    - i. History of finger or object engaging lower eyelid with lateral traction and subsequent "laceration" in the medial canthal area usually indicates an avulsive injury causing canalicular disruption
    - ii. Direct sharp trauma may lacerate and can also incise the canaliculus and careful inspection of the lacrimal system needs to be performed in all periocular injuries
  - c. Orbital fat in wound implies orbital involvement
    - i. Inspect for violation of the orbital septum
    - ii. Penetration of the orbital septum by a small instrument (e.g., ice pick) may not show fat in the wound
    - iii. A small laceration without fat in the wound does not rule out deep penetration.

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

- 1. Canalicular irrigation and probe - Examination under anesthesia for a child with a laceration medial to the punctum, if insufficient examination in the office
- 2. Computed tomography (CT) scan to evaluate possible facial and orbital fractures, if clinically indicated
  - a. Intracranial air implies intracranial penetration
- 3. Magnetic resonance imaging (MRI) scan to evaluate possible organic foreign body (after CT or plain films to rule out metallic foreign body)
- 4. Visual field test, if traumatic optic neuropathy suspected

**II. Define the risk factors**

- A. Assault, motor vehicle accidents and sports related injuries most common, but trauma can occur at any time, any place**

**III. List the differential diagnosis**

- A. Self-inflicted injury needs psychiatric evaluation**

**IV. Describe patient management in terms of treatment and follow-up**

- A. Describe medical therapy options**

1. Systemic antibiotic prophylaxis especially for suspected intracranial penetration
2. Tetanus toxoid status - toxoid if indicated

**B. Surgical repair**

- C. Surgery may be delayed 24 - 48 hours to allow resolution of edema and assembly of necessary team and instrumentation**
- D. Stent placement in case of lacerated canaliculus**

**V. List the complications of treatment, their prevention and management**

- A. Epiphora secondary to lacrimal injury**
- B. Traumatic ptosis, observe 6 months for improvement prior to any surgical repair**
- C. Eyelid malposition (ectropion, entropion), frequently cicatricial type, associated with soft tissue loss, may require grafting or complex reconstruction to correct**
- D. Diplopia secondary to orbital cicatricial changes, extraocular muscle injury, or neurologic injury**
- E. Intracranial injury requires urgent neurological evaluation**

**VI. Describe appropriate patient instructions**

- A. If injury is acute, complete ocular or orbital evaluation may be difficult because of hemorrhagic edema - must evaluate status of globe and vision acutely**
1. Repeat evaluation indicated after edema decreases or as soon as possible to assess for any globe injury or muscle paresis

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Management of Eyelid Trauma, Module #10, 1996.
3. AAO, Focal Points: Eyelid Trauma, Module #10, 1991.
4. AAO, Focal Points: Periorbital Animal Bites, Module #3, 1992.

# Neoplasms of the eyelid and ocular surface

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Ultraviolet radiation exposure is a risk factor
2. Most eyelid tumors are benign, some are malignant

### B. Define the relevant aspects of the epidemiology of the disease

1. Most malignancies increase in frequency with advancing age
2. Most often seen in lightly pigmented individuals

### C. List the pertinent elements of the history

1. Lesion growth
2. Change in lesion appearance, especially pigmentation
3. Ulcerating or bleeding
4. Chronic unilateral blepharoconjunctivitis (especially relevant with sebaceous carcinoma)
5. History of previous skin cancer

### D. Describe pertinent clinical features

1. Basal cell carcinoma of the eyelid
  - a. Irregular contours
  - b. Often nodular with telangiectasis
  - c. May have ulceration
  - d. Loss of normal skin or eyelid margin architecture (notch or lash loss)
  - e. Associated actinic skin changes
2. Squamous cell carcinoma of the eyelid
  - a. Flat or nodular skin lesion
  - b. Often similar in appearance to basal cell carcinoma
  - c. Hyperkeratosis is a distinguishing feature
  - d. May have spread to regional lymph nodes
3. Keratoacanthoma
  - a. Hyperkeratotic lesion with raised edges
  - b. Keratin plug in center

- c. Rapid growth
  - d. Tends to be locally destructive without distant spread
- 4. Sebaceous gland carcinoma of the eyelid and ocular surface
  - a. May present as isolated nodular lesion on the eyelid margin but more commonly presents with diffuse infiltration, resembling chronic blepharoconjunctivitis or chalazion
  - b. Frequently has skip areas
  - c. May spread to regional lymph nodes
- 5. Ocular surface squamous neoplasia
  - a. Conjunctival intraepithelial neoplasia, usually at the limbus
  - b. Squamous cell carcinoma of the conjunctiva
- 6. Melanoma and its variants
  - a. Irregularly shaped pigmented lesion
  - b. May have variations in color throughout lesion
  - c. Active growth is associated with bleeding and ulceration
  - d. Primary acquired melanosis of the conjunctiva may precede conjunctival melanoma
  - e. May have distant hematogenous spread

## **II. Define the risk factors**

- A. Advancing age**
- B. Sun exposure**

## **III. Describe the patient management in terms of treatment and followup**

- A. Indications for biopsy and histopathological evaluation**
  - 1. Possibly malignant lesion
  - 2. Cosmetically undesirable or uncomfortable lesion
  - 3. Lesion interfering with vision
- B. Referral for medical, radiation, or surgical management as appropriate based on biopsy**

## **IV. List the complications of treatment**

- A. Altered lid function and/or appearance**
- B. Inadequate resection/removal of tumor**

## **V. Describe disease related complications**

- A. Local tissue destruction**
- B. Regional or distant metastasis, which can lead to death**

## **VI. Describe appropriate patient instructions**

- A. Avoid sun exposure**
- B. Regular ocular examinations**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea. 2013-2014.

# Ectropion

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Involutional form
  - a. Lower eyelid horizontal laxity
    - i. Gravitational effects lead to eversion of the eyelid margin and loss of eyelid apposition to the globe
  - b. Eyelid retractor dehiscence may contribute to the lower eyelid rotation outwards
2. Paralytic form
  - a. Most often associated with facial nerve palsy
3. Cicatricial form
  - a. Scar tissue pulls lid away from the eye

### B. Define the relevant aspects of epidemiology of the disease

1. Advanced age with involutional horizontal eyelid laxity is the most common form of lower eyelid ectropion
2. Paralytic ectropion is most commonly seen accompanied by other signs of facial weakness
3. The most common form of upper eyelid ectropion is related to scar tissue pulling the eyelid away from the eye

### C. List the pertinent elements of the history

1. Chronic ocular exposure symptoms
2. Muroid discharge
3. Foreign body sensation
4. Epiphora
5. Facial nerve palsy

### D. Describe pertinent clinical features

1. Medial and/or lateral canthal tendon laxity
2. Epiphora
  - a. Secondary to reflex lacrimal secretion and/or punctal ectropion
3. Ocular exposure, secondary to lower eyelid malposition
4. Inferior corneal epithelial changes secondary to lower eyelid malposition
5. Palpebral conjunctival hypertrophy and keratinization
6. Scar tissue is seen in cicatricial ectropion

7. Facial sagging and brow ptosis accompany paralytic ectropion

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

1. Suspect malignancy if eyelid margin anatomy altered or loss of lashes
  - a. Biopsy as appropriate

**II. Define the risk factors**

- A. Advanced age
- B. Chronic eyelid rubbing
- C. Facial nerve palsy
- D. Previous trauma
- E. Previous eyelid surgery for skin cancer

**III. List the most common or critical entities in the differential diagnosis**

- A. Floppy eyelid syndrome
- B. Eyelid retraction

**IV. Describe patient management in terms of treatment and follow-up**

**A. Describe therapeutic options**

1. Topical lubrication
2. Horizontal eyelid taping (temporary only)
3. Surgical management as necessary
  - a. Lower eyelid tightening
  - b. Severe cicatricial changes require skin graft
  - c. Associated brow ptosis and facial sagging may require treatment

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.57-67.
3. AAO, Focal Points: Ectropion and Entropion, Module #10, 1994, p.1-2.

# Entropion

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. The eyelid margin rotates inward
  - a. Eyelashes are directed against the globe as the margin turns inward
  - b. This is contrast to other forms of trichiasis where abnormally positioned lashes are directed against the globe, but the lid margin is in a normal position
2. Involutional entropion
  - a. Caused by lower eyelid laxity, overriding orbicularis muscle, and retractor disinsertion
  - b. Seen only in the lower eyelid
3. Cicatricial entropion
  - a. Scarring of the posterior lamella rotates lid margin inward
  - b. Upper eyelid entropion is always cicatricial
    - i. There is no involutional form of upper eyelid entropion
  - c. Most lower eyelid entropion is involutional, but may be cicatricial

### B. Define the relevant aspects of epidemiology of the disease

1. Involutional
  - a. Elderly population
2. Cicatricial
  - a. Trauma
  - b. Conjunctival disease (e.g., trachoma)
  - c. Stevens Johnson Disease

### C. List the pertinent elements of the history

1. Foreign body sensation, pain and epiphora

### D. Describe pertinent clinical features

1. Lower eyelid margin rotated inward against cornea and conjunctiva
2. Lower eyelid horizontal laxity (involutional form)
3. Posterior lamellar scarring (cicatricial form)
4. Corneal and conjunctival irritation

## II. Define the risk factors



- A. Older age
- B. Lower eyelid horizontal laxity
- C. History of conjunctival scar

### **III. List the most common or critical entities in the differential diagnosis**

- A. Spastic entropion
- B. Trichiasis with normal lid position

### **IV. Describe patient management in terms of treatment and follow-up**

#### **A. Describe therapeutic options**

1. Ocular lubrication to protect from lashes
2. Bandage contact lens
3. Surgical management
  - a. Involutional entropion
    - i. Entropion rotation suture (Quickert suture)
    - ii. Retractor reinsertion and lid tightening
  - b. Cicatricial entropion - rotational sutures (temporary) or posterior lamellar grafting

### **V. Describe disease-related complications**

- A. Corneal scarring
- B. Corneal ulcer
- C. Vision loss

### **VI. Describe appropriate patient instructions**

- A. Follow for recurrence

#### **Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Ectropion and Entropion, Module #10, 1994, p.3.

# Ptosis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Congenital blepharoptosis
  - a. Myopathic (weak and fibrotic levator muscle)
2. Structural problems of upper eyelid
  - a. Levator dehiscence (involutional ptosis)
  - b. Contact lens associated
  - c. Neurofibromatosis
  - d. Hemangioma
  - e. Trauma
3. Myopathic, neuropathic, and neuromyopathic conditions
  - a. Myotonic dystrophy
  - b. Cranial nerve (CN) III dysfunction
  - c. Horner syndrome
  - d. Myasthenia gravis
  - e. Other myopathies

### B. Describe the pertinent history elements

1. Onset and progression of ptosis
2. Symptoms of obstruction of vision or peripheral visual field
3. Presence of diplopia
4. Presence of non-ocular muscle weakness
5. Diurnal variation in degree of ptosis
6. History of eyelid or ocular trauma and/or surgery

### C. Describe pertinent clinical features

1. Eyelid measurements
  - a. Interpalpebral fissures
  - b. Lid excursion (levator excursion)
2. Presence of other signs
  - a. Anisocoria
  - b. Lid lag

- c. Proptosis
- d. Fatigue
- e. Orbicularis weakness
- f. Extraocular muscle weakness and/or diplopia
- g. Chin-up head posture

## **II. List the differential diagnosis**

- A. Dermatochalasis**
- B. Pseudoptosis**
- C. Lid retraction of contralateral eye**
- D. Enophthalmos/proptosis (may mimic ptosis)**

## **III. Describe patient management in terms of treatment and follow-up**

- A. Treatment of ptosis is based on the problem and the amount of levator excursion (function)**
- B. Types of ptosis that require special consideration**
  - 1. Myasthenia gravis
  - 2. Neuropathic or myopathic ptosis
  - 3. Mechanical ptosis
  - 4. Pseudoptosis
- C. If the ptosis is associated with systemic disease or other neurological abnormalities, specific evaluation and management of the underlying disorder is necessary**

## **IV. List the complications of treatment, their prevention and management**

- A. Complications**
  - 1. Asymmetry
  - 2. Under or over correction
  - 3. Lagophthalmos
  - 4. Corneal exposure
- B. Exposure can usually be managed with ocular lubrication**
- C. Reoperation is necessary for unacceptable lid position or exposure not responding to medical therapy**

## **V. Describe disease-related complications**

- A. Amblyopia**
- B. Impairment of visual field**
- C. Decreased central acuity**

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System; Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Focal Points: Myasthenia Gravis, Module #4, 2003, p.2-3.
3. AAO, Focal Points: Congenital Ptosis, Module #2, 2001.

# Facial nerve palsy

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Idiopathic (Bell palsy) is most common
2. Congenital
3. Traumatic
4. Inflammatory
5. Compressive
6. Vascular (stroke)
7. Iatrogenic (intracranial or facial surgery)
8. Infectious

### B. Define the relevant aspects of epidemiology of the disease

1. Isolated facial nerve palsy with no clear etiology is termed "Bell palsy"
  - a. Usually unilateral
  - b. Thought to be inflammatory or a sequel to herpes zoster infection

### C. List the pertinent elements of the history

1. Patients may have concurrent anesthetic corneas, making them more likely to develop corneal pathology
2. Ipsilateral dry eye, hearing loss, and vestibular dysfunction are associated with cerebellopontine angle masses
3. Other associated neurological findings should be evaluated
4. Removal of acoustic neuroma, cerebellar tumor, parotid tumor

### D. Describe the pertinent clinical features

1. Lack of movement of the involved side of the face with eyebrow ptosis, lagophthalmos, ectropion, poor blink, and mouth droop
2. In the case of Bell palsy, some recovery of function may be anticipated

### E. Describe appropriate laboratory testing for establishing the diagnosis

1. Neuroimaging is not usually recommended for isolated first episode of Bell palsy
2. Imaging indicated if brainstem cause is suspected, especially of slowly progressive, or if eyelid myokymia is present
3. Consider laboratory testing for Lyme disease if clinically appropriate

## **II. Define the risk factors**

- A. Viral syndrome**
- B. Cerebellopontine angle tumor**
- C. Trauma**
- D. History of stroke, facial surgery, or intracranial surgery**

## **III. Describe patient management in terms of treatment and follow-up**

- A. Topical lubrication**
- B. Taping of eyelids**
- C. Moisture chambers**
- D. Lateral (and rarely medial) tarsorrhaphy**
- E. Ectropion repair**
- F. Consider referral for gold weight implantation or facial reanimation**

## **IV. List the complications of treatment, their prevention and management**

- A. All tightening procedures eventually relax**
- B. Opening of the tarsorrhaphy**

## **V. Describe disease-related complications**

- A. Corneal exposure, ulceration**
- B. Drooling**
- C. Aberrant regeneration**

## **VI. Describe appropriate patient instructions**

- A. In the short term, ocular lubrication is most important**
- B. If Bell palsy, conservative observation with aggressive lubrication is indicated, in anticipation of some recovery**
- C. Surgical correction may be indicated when the palsy is expected to be permanent**

### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System, Section 5: Neuro-Ophthalmology, 2013-2014.**
- 2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p. 4.**
- 3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p. 173-201.**

4. AAO, Focal Points: Essential Blepharospasm and Related Diseases, Module #5, 2000.
5. AAO, Focal Points: Ophthalmic Management of Facial Nerve Paralysis, Module #4, 2000.

# Lacrimal system: evaluation of epiphora

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. The lacrimal system is a dual system
  - a. Secretory via the main and accessory lacrimal glands
  - b. Excretory via the lacrimal outflow system including
    - i. Eyelid pumping mechanisms
    - ii. Puncta
    - iii. Canaliculi
    - iv. Common internal punctum
    - v. Lacrimal sac
    - vi. Nasolacrimal duct (NLD)
    - vii. Valve of Hasner exiting into inferior meatus of the nose under the inferior turbinate
2. Tearing may arise from reflex irritation or from a qualitatively poor tear film
3. Total or partial obstruction of the lacrimal outflow system may cause tearing
  - a. In infants and children, the obstruction is most commonly at the level of the valve of Hasner
  - b. Partial or complete obstruction in adults most commonly occurs in the nasolacrimal duct from inflammation and recurrent infection

### B. Define the relevant aspects of the epidemiology of the disease

1. Infants will manifest congenital obstructions shortly after birth
2. Women are at greater risk for acquired NLD obstruction and dry eyes
3. Trauma or facial nerve weakness may affect the lid position or pumping mechanism

### C. List the pertinent elements of the history

1. Patients with poor ocular surface will report tearing worse on cold mornings, windy days, with air conditioning, in dry environments
2. Lacrimal obstruction patients report tearing all day long

### D. Describe pertinent clinical features

1. Lacrimal outflow obstruction patients may have increased tear lake and prolonged dye disappearance test
2. Nasolacrimal duct obstruction may be "functional" or "anatomic"
  - a. Functional obstruction means epiphora and prolonged dye disappearance, but irrigation into nose is possible



- i. Sometimes called a "partial" obstruction.
- b. Anatomic obstruction means that irrigation is not possible
  - i. Sometimes termed "complete" obstruction

## **II. Define the risk factors**

- A. Infants for congenital NLD obstruction**
- B. Women for both dry eyes and acquired NLD obstructions**
- C. Dry eyes may be associated with Sjögren disease or other autoimmune diseases**
- D. Various chemotherapeutic agents can cause tearing via ocular surface damage or canalicular scarring**
- E. History of trauma to eyelids or canaliculi**
- F. History of facial nerve palsy**

## **III. List the most common or critical entities in the differential diagnosis**

- A. Ocular surface disease causing irritation and reflex tearing**
- B. Hypersecretion**
- C. Lacrimal obstruction**
  - 1. Punctal
  - 2. Canalicular
  - 3. NLD
- D. Congenital glaucoma in infants**

## **IV. Describe the patient management in terms of treatment and followup**

- A. Patients with poor tear film require appropriate ocular moisture and lubrication**
- B. Patients with underlying immunologic disorder need appropriate referral**
- C. Surgical management as appropriate**

## **V. Describe disease related complications**

- A. Severe dry eyes can lead to corneal decompensation**
- B. Persistent NLD obstruction can lead to dacryocystitis**

## **VI. Describe appropriate patient instructions**

- A. Compliance with medical recommendations**

**B. Follow up for surgical management as indicated**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

# Acquired nasolacrimal duct obstructions and dacryocystitis in adults

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Nasolacrimal duct obstruction
  - a. Trauma
  - b. Medications
  - c. Inflammatory disease
  - d. Involutional stenosis
    - i. Most common
    - ii. Women are more frequently affected than men
  - e. Granulomatous disease
    - i. Sarcoidosis
    - ii. Wegener granulomatosis
  - f. Tumors
    - i. Lymphoma
    - ii. Squamous cell carcinoma
  - g. Radiation therapy for malignancy
  - h. Nasal pathology
    - i. Cocaine inhalation abuse
    - ii. Endoscopic sinus surgery
2. Dacryocystitis
  - a. Nasolacrimal duct obstruction
  - b. Chronic tear stasis and retention
  - c. Secondary bacterial infection
  - d. May present as an acute or chronic condition
  - e. Gram positive most common organism
    - i. Consider gram negative organisms in patients with diabetes mellitus, immunocompromised patients or nursing home residents if there is no response to initial treatment

### B. Describe pertinent clinical features

1. Nasolacrimal duct obstruction
  - a. Constant tearing
  - b. May have maceration of skin at medial or lateral canthus from tear overflow
2. Acute dacryocystitis
  - a. Erythema, edema inferior to the medial canthal tendon
  - b. Pain
  - c. Mass (the normal lacrimal sac usually is not palpable)
  - d. Mucopurulent discharge
  - e. Epiphora
3. Chronic dacryocystitis
  - a. Chronic conjunctivitis
  - b. Epiphora may be present if there is normal tear production
  - c. Mucopurulent discharge
  - d. Pressure on sac causes reflux
  - e. Mucocele can form if there is obstruction at common internal punctum
  - f. Nasal speculum examination to exclude intranasal pathology

## **II. Describe patient management in terms of treatment and follow-up**

### **A. Nasolacrimal duct obstruction**

1. Probing/tube placement and balloon dacryoplasty sometimes work
2. Dacryocystorhinostomy (DCR) is often necessary
3. Consider malignancy if blood tinged tears are present
4. Biopsy of lacrimal sac and intranasal lesion if malignancy is suspected

### **B. Dacryocystitis**

1. Do not irrigate or probe during acute infection or if reflux occurs on massage of sac
2. Acute infection
  - a. Warm compresses
  - b. Oral antibiotic
  - c. IV antibiotic if orbital or preseptal cellulitis develops
  - d. Incision and draining of localized abscess with packing of the abscess
  - e. Dacryocystorhinostomy (DCR) once inflammation subsides
3. Chronic infection

- a. Massage to clear sac of contents
- b. Optional use of antibiotic drops
- c. DCR should be performed at earliest patient convenience

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p. 102.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p. 102-103.
4. AAO, Focal Points: Evaluation of the Tearing Adult Patient, Module #8, 2002.
5. AAO, Focal Points: Evaluation and Surgery of the Lacrimal Drainage System in Adults, Module #12, 1995.

# Tests of ocular alignment

## I. List the indications/contraindications

### A. Indications

1. The main purpose of this test is to determine whether a manifest tropia exists

## II. Describe the pre-procedure evaluation

### A. If sensory testing of stereopsis or binocularity is planned, this should be performed first before interrupting fusion with cover testing

### B. Assess visual acuity in each eye before proceeding with full cover testing

1. The presence of subnormal acuity in one eye may indicate amblyopia and may help to determine the eye preferred for fixation in cover testing
2. If visual acuity is very poor in one or both eyes, one cannot do cover testing and must do corneal light reflex testing instead

### C. Assess cooperation level of patient

1. Cover tests preferred if possible
2. Corneal light reflex tests used otherwise

## III. Describe the instrumentation and technique

### A. Instrumentation

1. Accommodative fixation targets at distance and at near
2. Opaque occluder
3. Prisms
4. Hand-held muscle light

### B. Techniques

1. Cover-uncover test
  - a. With patient fixating on an accommodative target at distance, cover one eye and observe
    - i. The fellow eye
      - i) A re-fixation movement indicates a manifest tropia.
    - ii. The covered eye
      - i) If no manifest tropia, observe the covered eye as cover is removed. If covered eye makes a refixation movement, phoria is present.
2. Corneal light reflex tests (Krimsky and Hirschberg tests)

- a. Hold muscle light 14-18 inches from face
  - b. Evaluate location of light reflex using iris or pupil landmarks
  - c. Light displacement opposite of eye shift (i.e., light reflex displaced temporally in esotropia, nasally in exotropia)
  - d. May use prisms to quantitate degree of deviation
  - e. If corneal light reflex is displaced under monocular conditions, then eccentric fixation or abnormal angle kappa is present (or patient is blind)
3. Alternate prism and cover test
    - a. Alternately occlude each eye with prism over one eye
    - b. Apex of prism is placed in direction of deviation/base in opposite direction, i.e., base out to measure esotropia, base in to measure exotropia, etc.
    - c. Continue to adjust prism amount until no ocular movement is discerned when occluder is alternatively moved from one eye to the other
    - d. This test measures the total deviation, the combined phoria and tropia

#### **IV. List the complications of this procedure, their prevention and management**

- A. Inaccurate measurements will be obtained if the patient does not maintain sufficient fixation effort**
  1. Especially in children, communicate with the patient during the procedure to ensure that they are engaged in the process and attentive to the target
  2. Use accommodative targets at distance and at near when possible
- B. Variable measurements may be obtained in patients with large amounts of uncorrected refractive error or in certain myopathic conditions, or if large incomitancies are present and head position is not held constant**
  1. Have patient return for subsequent visits and repeat testing
  2. Repeat measurements after correcting refractive errors

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
2. AAO, Preferred Practice Patterns Committee, Pediatric Ophthalmology Panel. Pediatric Eye Evaluations Preferred Practice Pattern, 2007.

# Amblyopia

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Decreased vision in one or both eyes due to a defect in cortical visual development caused by one or more of the following conditions:
  - a. Strabismus
    - i. Constant, non-alternating strabismus
  - b. Ametropia/Uncorrected bilateral refractive error
    - i. High spherical and/or cylindrical relatively equal refractive errors in both eyes
  - c. Anisometropia
    - i. Unequal refractive error between the two eyes
  - d. Deprivation (occlusion of visual axis)
    - i. Ptosis
    - ii. Corneal opacity
    - iii. Cataract

### B. List the pertinent elements of the history

1. Constant misalignment of the eyes
2. Visually inattentive child
3. Droopy eyelid
4. Abnormal corneal light reflex
5. Abnormal head positioning (chin down, head turn to side)

### C. Describe pertinent clinical features

1. Decreased best-corrected visual acuity
2. One or more of the etiologic factors described above

## II. Define the risk factors

### A. Strabismus

### B. Anisometropia

### C. Family history

### D. Congenital or traumatic abnormalities of the anterior or posterior segment



### **III. List the most common or critical entities in the differential diagnosis**

- A. Other causes of organic visual loss (e.g., retinal or optic nerve disease)**
- B. Functional or non-physiologic visual loss**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
2. AAO, Focal Points: Practical Management of Amblyopia, Module #2, 2000.
3. AAO, Preferred Practice Patterns Committee, Pediatric Ophthalmology Panel. Amblyopia Preferred Practice Pattern, 2007.

# Pseudoesotropia

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Inherited facial features that cause tissue to obscure the nasal sclera
2. This is often associated with a reduced interpupillary distance and wide epicanthal folds

### B. List the pertinent elements of the history

1. Photographs that show the eyes apparently crossed in lateral gaze

### C. Describe the pertinent clinical features

1. Child with no visual complaints and does not demonstrate amblyopia
2. Full ductions and conjugate versions with a cover test at distance and at near that does not show a tropia
3. Well-centered corneal light reflex

## II. Describe appropriate patient instructions

### A. Reassure parents that as child grows, bridge of nose displaces epicanthal folds and pseudoesotropia appearance will improve

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.

# Acute neonatal conjunctivitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Direct contact with the infecting agent during passage through birth canal or organism can ascend to the uterus and infect the infant
2. Most common infection is *Chlamydia*
3. *Neisseria* can cause severe infection
4. *Herpes Simplex* is rarer than *Chlamydia* or *Neisseria*

### B. List the pertinent elements of the history

1. *Neisseria* usually occurs in first week of life
2. *Chlamydia* usually occurs at 5-12 days of life
3. *Herpes Simplex* usually occurs in the second week of life
4. Overlap possible, so cannot diagnose by time course alone

### C. Describe the pertinent clinical features

1. *Chlamydia*
  - a. Mild to moderate lid swelling
  - b. Conjunctival hyperemia
  - c. Watery or filmy discharge
2. *Neisseria*
  - a. Hyperacute and purulent discharge
  - b. More marked lid edema and chemosis
3. Clinical signs of above two may overlap
4. *Herpes Simplex*
  - a. Epithelial vesicle and dendritic ulcers

### D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Gram stain
2. Culture of conjunctival discharge
3. Rapid diagnostic available for *Chlamydia*

## II. List the most common or critical entities in the differential diagnosis

### A. Other infectious conjunctivitis

- B. Chemical conjunctivitis if silver nitrate used
- C. Congenital nasolacrimal duct obstruction

### III. Describe patient management in terms of treatment

- A. *Neisseria* requires rapid institution of systemic (IV) antibiotics to prevent corneal ulceration and perforation
- B. *Chlamydia* should be treated orally; and possibly topically
- C. *Herpes Simplex* treated topically, or orally based upon clinical findings.
- D. Notify appropriate health authorities for *Neisseria gonorrhea* and *Chlamydia trachomatis*

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
2. AAO, Preferred Practice Patterns Committee, Cornea and External Disease Panel: Conjunctivitis Preferred Practice Pattern, 2008.

# Congenital nasolacrimal duct obstruction

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Obstruction of the lacrimal drainage system
2. Most commonly caused by a thin mucosal membrane at the lower end of the nasolacrimal duct (NLD)

### B. Define the relevant aspects of epidemiology of the disease

1. High rate of spontaneous resolution
2. Beyond age 1 year, spontaneous resolution becomes less likely

### C. Describe pertinent clinical features

1. Epiphora and/or mucopurulent discharge with or without an associated conjunctivitis
2. Lower eyelid skin irritation/breakdown
3. Reflux of mucoid material from the puncta with digital pressure over the lacrimal sac

## II. List the most common or critical entities in the differential diagnosis

### A. Conjunctivitis

### B. Congenital glaucoma

### C. Congenital dacryocystocele

### D. Punctal or canalicular atresia

## III. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Digital massage
2. Topical antibiotics
3. Observation

### B. Describe surgical therapy options

1. Probing
  - a. NLD probing is more effective if done before 2 years of age

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus,

2013-2014.

# Congenital glaucoma

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. May be a family history of congenital glaucoma
2. Symptoms may occur at birth or within the first several weeks to months of life
  - a. Epiphora
  - b. Photophobia
  - c. Blepharospasm

### B. Describe pertinent clinical features

1. Corneal edema
2. Striae of Descemet membrane (Haab striae)
3. Corneal enlargement
4. Elevated intraocular pressure
5. Epiphora
6. Optic nerve cupping (may be reversible)
7. Buphthalmos
8. With enlargement of globe, children may develop significant myopia +/- astigmatism

## II. Define the risk factors

### A. Family history of congenital glaucoma

## III. List the most common or critical entities in the differential diagnosis

- A. Hereditary corneal opacities (e.g., dystrophies)
- B. Birth trauma
- C. Megalocornea
- D. Secondary glaucomas (e.g. anterior segment dysgenesis)

## IV. Describe patient management in terms of appropriate treatment and follow up

- A. Referral to an appropriate subspecialist
- B. Treatment is surgical; medical treatment is a temporizing measure

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
2. AAO, Focal Points: Pediatric Glaucoma, Module #5, 1997, p. 1-2.
3. AAO, Focal Points: Childhood Glaucoma, Module #10, 1990, p. 4.



# Congenital cataracts

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Familial / hereditary
2. Metabolic errors (e.g., galactosemia)
3. TORCH infections: toxoplasmosis, rubella, cytomegalovirus, herpes
4. Idiopathic

### B. List the pertinent elements of the history

1. Possible reduced visual attentiveness
2. Abnormal red reflex
3. Possible nystagmus or strabismus

### C. Describe pertinent clinical features

1. Leukocoria or abnormal or absent red reflex
2. Opacity of lens may involve anterior or posterior capsule or subcapsular area, cortex, or nucleus
3. Larger, more central, and more posterior opacities increase likelihood of being visually significant and producing amblyopia

### D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Assess visual acuity
2. Slit-lamp biomicroscopic examination
3. Assessment of red reflex
4. If no view of retina, B-scan ultrasound if available
5. For bilateral cataracts, evaluate for possible infectious etiologies or metabolic errors if there is no family history of early-onset cataracts

## II. Define the risk factors

### A. Family history of cataracts

### B. Congenital intraocular infections

### C. Developmental abnormalities such as persistent fetal vasculature

## III. List the most common or critical entities in the differential diagnosis

### A. Other causes of leukocoria

1. Retinal detachment
2. Retinoblastoma

#### **IV. Describe patient management in terms of treatment and follow-up**

##### **A. Describe medical therapy options**

1. Treat amblyopia and refractive error if present

##### **B. Describe surgical therapy options**

1. Prompt surgery required for visually significant cataracts

#### **V. Describe disease-related complications**

##### **A. Reduced vision**

##### **B. Development of amblyopia**

##### **C. Strabismus**

##### **D. Nystagmus**

##### **E. Glaucoma**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
2. AAO, Focal Points: Childhood Cataracts, Module #1, 1996, p. 1.

# Rhabdomyosarcoma

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Malignant neoplasia of extraocular muscle precursor cells found in orbital soft tissue

### B. Define the relevant aspects of epidemiology of the disease

1. Most common primary orbital malignancy in childhood

### C. List the pertinent elements of the history

1. Sudden onset and rapid evolution of unilateral proptosis
2. Possible vision loss, eyelid swelling, redness, and/or strabismus

### D. Describe pertinent clinical features

1. Unilateral proptosis
2. Globe displacement
3. Ptosis, eyelid edema
4. Mass may be palpable, most commonly in superior nasal orbit
5. Pain is uncommon
6. Conjunctival injection and chemosis
7. Possible decreased vision and/or strabismus

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Orbital imaging: Computed tomography (CT) scan and/or Magnetic resonance imaging (MRI) scan
2. Urgent orbital biopsy

## II. List the most common or critical entities in the differential diagnosis

- A. Orbital pseudotumor
- B. Orbital infection
- C. Other orbital tumors
- D. Orbital trauma

## III. Describe appropriate patient instructions

- A. Precise diagnosis is critical
- B. Tumor biopsy is needed urgently
- C. Multidisciplinary approach

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
2. AAO, Focal Points: Orbital Tumors in Children, Module #9, 1990, p.6-7, 9.

# Retinoblastoma

## **I. Describe the approach to establishing the diagnosis**

### **A. List the pertinent elements of the history**

1. Child not appearing to see well
2. Strabismus
3. Leukocoria
4. Family history

### **B. Describe pertinent clinical features**

1. White retinal and/or intravitreal lesion(s) with possible calcification
2. Strabismus
3. Eye(s) usually not inflamed

## **II. Describe the most common or critical entities in differential diagnosis of leukocoria**

- A. Cataract**
- B. Coats disease**
- C. Persistent fetal vasculature**
- D. Toxocariasis**
- E. Retinopathy of prematurity**
- F. Retinal detachment**

## **III. Describe patient management in terms of treatment**

- A. Prompt referral**
- B. Systemic evaluation for metastases or other malignancies**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
2. AAO, Laser Photocoagulation of the Retina and Choroid, 1997, p.240-242.
3. AAO, Focal Points: Retinoblastoma, Module #1, 1990.

# Shaken baby syndrome/non-accidental trauma

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Non-accidental trauma to child after birth
2. Abrupt acceleration/deceleration injury ruptures intracranial vessels and compresses brain against skull

### B. Define the relevant aspects of epidemiology of this disease

1. Usually under age 3 years

### C. List the pertinent elements of the history

1. Often unreliable history or minor trauma related by caregiver or history inconsistent with clinical findings

### D. Describe pertinent clinical features

1. May have external evidence of trauma
2. May have neurologic findings
3. May have occult fractures (e.g. ribs), sometimes of varying ages
4. Hemorrhages in multiple retinal layers and/or vitreous cavity, typically but not always bilateral
5. Hemorrhages may appear to be of different ages

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Pediatric medical evaluation

## II. Define the risk factors

### A. Trauma under suspicious circumstances

### B. Multiple injuries

## III. List the most common or critical entities in the differential diagnosis

### A. Accidental trauma

### B. Birth trauma

### C. Coagulation and hematological disorders

## IV. Describe patient management in terms of treatment and follow-up

- A. Social service involvement**
- B. Any physician who suspects child abuse is required by law to report this to a designated governmental agency**

## **V. Describe disease-related complications**

- A. Death**
- B. Neurologic complications**
- C. Vision loss**
  - 1. Optic nerve damage
  - 2. Cortical damage
  - 3. Amblyopia

## **VI. Describe appropriate patient instructions**

- A. Child must be removed from at-risk environment**
- B. Child's interests come first**
- C. Recovery depends on severity of injury**

### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
- 2. AAO, Focal Points: Diagnosis and Management of Vitreous Hemorrhage, Module #10, 2000, p. 13.
- 3. AAO, Focal Points: The Ocular Findings in Child Abuse, Module #7, 1998.

# Visual acuity measurement

## **I. List the indications/contraindications**

### **A. Indications**

1. To determine level of central vision quantitatively

### **B. Contraindications**

1. There are no contraindications
2. It can be difficult to obtain in some cases of ocular emergencies, but should always be attempted

## **II. Describe the pre-procedure evaluation**

### **A. Medical and ophthalmic history**

## **III. List the alternatives to this procedure**

### **A. Electrophysiology**

### **B. Preferential looking and other techniques for young children**

### **C. Menace threat reflex (check for reflex withdrawal or blink when patient presented with a threatening stimulus)**

## **IV. Describe the instrumentation and technique**

- A. Standard eye lane with Snellen acuity chart typically placed 20 feet or 6 meters from patient, either directly or with use of mirrors**
- B. If patient cannot understand the letters on the Snellen chart, other types of charts could be used, such as tumbling 'E' and Landolt C.**
- C. Patient uses occluder to test each eye individually and is instructed to detect the smallest possible line**
- D. Attention is directed to maximizing patient effort**
- E. Vision recorded as a fraction, for the smallest line for which the patient is able to read at least half the letters is recorded as the denominator**
- F. For higher accuracy, +/- notation is used to identify the number of letters made on the next smallest line or missed on the recorded line respectively**
- G. The fraction is sometimes reported in metric terms or decimal notation, and sometimes converted to a logMAR (logarithm of the minimal angle of resolution) equivalent**
- H. If vision is too poor for conventional eye chart**
  1. Eye chart moved forward until letters are recognized
  2. Counting fingers at various distances



3. Hand motion
4. Light perception

## **V. Describe the considerations in interpretation of this diagnostic procedure**

- A. There is some level of subjectivity to this test
- B. There are alternative methods for assessing vision (see above).
- C. Consider functional loss/malingering if results seem inconsistent with other clinical assessments

### Near Vision Testing

## **I. List the indications**

- A. Presbyopic patients presenting for refractive surgery evaluation
- B. Patients complaining of near vision symptoms
- C. Patients in the presbyopic age range

## **II. Describe the pre-procedure evaluation**

- A. Ophthalmic and social history with particular regard to vocation, hobbies and lifestyle

## **III. Describe the instrumentation and technique**

- A. A hand-held reading card is typically used at approximately 14 to 16 inches of the patient's preferred reading distance
- B. Ideally, the patient is tested for both the uncorrected and best-corrected state at an appropriate distance as determined by the patient's needs
- C. Another important component of near vision is the accommodative amplitude
  1. Near point of accommodation
  2. Prince rule
- D. Range of accommodation
  1. This measures the useful range of near vision when a certain lens is employed and helps determine the functional capabilities of a near lens

## **IV. Describe the considerations in interpretation of this diagnostic procedure**

- A. Subjective nature of measurement
- B. Range of near point needs will vary from patient to patient

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, 2013-2014.
2. AAO, Low Vision Rehabilitation, 1999, p.26.

# Refraction: manifest and cycloplegic

## **I. List the indications**

- A. To measure the refractive error**
- B. To determine best eyeglass corrected visual acuity**
- C. To prescribe eyeglasses**
- D. Cycloplegic refraction is especially important in younger or hyperopic patients**

## **II. Describe the pre-procedure evaluation**

- A. Medical and ophthalmic history**
- B. Visual acuity without correction**
- C. Retinoscopy, autorefraction, or lensometry of current eyeglasses, if available, to obtain starting point**

## **III. List the alternatives to this procedure**

- A. No real substitute**

## **IV. Describe the instrumentation and technique**

- A. A preliminary refractive error can be determined using old glasses, an autorefraction or retinoscopy**
- B. The refraction is termed a cycloplegic refraction if the patient has received cycloplegic eyedrops to paralyze the accommodative mechanism prior to the refraction**
- C. Refracting in a natural state (i.e. without cycloplegic eye drops) is termed a manifest refraction.**
- D. The patient is placed at the phoropter or in trial frames and then offered a series of choices to correct the spherical component of the refractive error until the best vision is achieved with the least amount of minus (or most plus) sphere**
- E. The cylinder component of the refractive error is determined using the cross-cylinder method**
- F. For manifest refraction, endpoint can be verified**
  - 1. Fogging**
    - a. To minimize chance of accommodation**
  - 2. Red/green test**
    - a. To verify accuracy of spherical correction**

## **V. Describe the considerations in interpretation of this diagnostic procedure**

- A. Test is subjective**
- B. The usual cause of a disparity between the manifest and cycloplegic refraction is accommodation where the cycloplegic sphere will have less minus power than the manifest sphere**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, 2013-2014.

# Lens notation and conversion

## I. Lenses used to correct cylinder are with either plus or minus lens notation

- A. In the plus lens notation, the correcting plus cylinder is aligned along the steepest corneal meridian and noted in plus terminology. For example,  $+1.75+1.50 \times 085^\circ$  indicates a cornea with 1.5 diopters of astigmatism in which the steep meridian is at 85 degrees
- B. In the minus lens notation, the correcting minus cylinder is aligned along the flattest corneal meridian. For example, the above refractive error would be recorded as  $+3.25-1.50 \times 175^\circ$  to indicate a cornea with 1.5 diopters of astigmatism in which the flat meridian is at 175 degrees
- C. To transpose refractive errors within the two notations
  1. Add the sphere and cylinder magnitude arithmetically
  2. Switch the cylinder sign, i.e. make minus plus or make plus minus
  3. Change the axis of the cylinder by 90 degrees

### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, 2013-2014.

# Myopia

## **I. Describe the approach to establishing the diagnosis**

### **A. Describe the etiology of the disease**

1. Genetic factors
2. Axial myopia
3. Disease states such as keratoconus
4. Nuclear sclerotic cataract

### **B. List the pertinent elements of the history**

1. Vision worse at distance than at near

### **C. Describe pertinent clinical features**

1. Uncorrected visual acuity worse at distance than at near

## **II. Define the risk factors**

### **A. Family history**

## **III. List the most common or critical entities in the differential diagnosis**

### **A. Other forms of decreased vision associated with ocular disorder**

## **IV. Describe patient management in terms of treatment and follow-up**

### **A. Describe medical therapy options**

1. Eyeglasses
2. Contact lenses
3. Observation

### **B. Describe surgical therapy options**

1. Refractive surgery

## **V. Describe disease-related complications**

### **A. High myopia is a risk factor for other problems such as glaucoma, retinal detachment, and early cataract**

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, Section 14: Refractive Surgery, 2013-2014.
2. AAO, Focal Points: Choosing the Appropriate PRK Patient, Module #9, 1998.
3. AAO, Preferred Practice Patterns Committee, Refractive Errors Panel. Refractive Errors Preferred Practice Pattern, 2007.

# Hyperopia

## **I. Describe the approach to establishing the diagnosis**

### **A. Describe the etiology of this disease**

1. Genetic factors
2. Axial short eye
3. Flat cornea

### **B. List the pertinent elements of the history**

1. Vision worse at near than at distance

### **C. Describe pertinent clinical features**

1. Hyperopia on refraction
2. Uncorrected visual acuity worse at near than at distance
3. Accommodative esotropia may occur

## **II. Define the risk factors**

### **A. Family history**

## **III. List the most common or critical entities in the differential diagnosis**

### **A. Other forms of decreased vision associated with ocular pathology**

## **IV. Describe patient management in terms of treatment and follow-up**

### **A. Define medical therapy options**

1. Eyeglasses
2. Contact lenses
3. Observation

### **B. Define surgical therapy options**

1. Refractive surgery

## **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses; Section 14, Refractive Surgery, 2013-2014.
2. AAO, Focal Points: LASIK, Module #3, 2000, p.12-13.



3. AAO, Preferred Practice Patterns Committee, Refractive Errors Panel. Refractive Errors Preferred Practice Pattern, 2007.

# Anisometropia

## I. Describe the approach to establishing the diagnosis

- A. Definition: anisometropia is a difference of refractive power between fellow eyes**
- B. Describe the etiology of this disease**
  - 1. Genetic factors
  - 2. Disease states such as keratoconus
  - 3. Postsurgical
    - a. Typically cataract surgery
- C. List the pertinent elements of the history**
  - 1. Vision decreased at different distances in fellow eyes
  - 2. One eye with blurry vision
  - 3. Vision fatigue
  - 4. Symptoms of visual asthenopia
- D. Describe pertinent clinical features**
  - 1. Uncorrected visual acuity difference in fellow eyes
  - 2. Aniseikonia

## II. Define the risk factors

- A. Postsurgical is most common**
- B. Congenital is often well tolerated except in that it may cause amblyopia**

## III. List the most common or critical entities in the differential diagnosis

- A. Cataract**
- B. Corneal disease**

## IV. Describe patient management in terms of treatment and follow-up

- A. Define medical therapy options**
  - 1. Eyeglasses
  - 2. Contact lenses
  - 3. Observation
- B. Define surgical therapy options**

1. Refractive surgery
2. Intraocular lens exchange if following cataract surgery

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, 2013-2014.

# Aniseikonia

## I. Describe the approach to establishing the diagnosis

**A. Definition: aniseikonia is a defect of binocular vision in which fellow eye retinal images of an object differ in size**

**B. Describe the etiology of this disease**

1. A difference in refractive power between eyes
2. A difference in axial length
  - a. Congenital
  - b. Postsurgical (scleral buckling)
3. Asymmetric keratoconus
4. Post surgical anisometropia

**C. List the pertinent elements of the history**

1. History of surgery or ocular condition
2. Symptoms
  - a. Vision decreased at different distances in different eyes, associated with different image size perception by patient, typically with eyeglass correction
  - b. Vision fatigue
  - c. Headaches
  - d. Diplopia

**D. Describe pertinent clinical features**

1. Significant difference in any area of refraction, sphere or cylinder (typically over 1 to 3 diopters) in the refraction between fellow eyes
2. Uncorrected visual acuity (UCVA) testing in fellow eyes, symptoms different between eyeglasses and contact lenses

## II. Define the risk factors

**A. Postsurgical**

## III. Describe patient management in terms of treatment and follow-up

**A. Define medical therapy options**

1. Contact lenses
2. Observation

3. Eyeglasses

**B. Define surgical therapy options**

1. Refractive surgery
2. Intraocular lens exchange if after cataract surgery

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, 2013-2014.

# Regular astigmatism

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Genetic factors
2. Disease states
3. Post cataract and corneal surgery

### B. List the pertinent elements of the history

1. Symptoms, vision decreased at all distances
2. May have monocular diplopia or blurring

### C. Describe pertinent clinical features

1. Astigmatism on refraction
2. Uncorrected visual acuity poor at all distances
3. Excellent best-corrected visual acuity with eyeglass correction

## II. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Eyeglasses
2. Contact lenses
3. Observation

### B. Describe surgical therapy options

1. Refractive surgery

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, Section 14: Refractive Surgery, 2013-2014.
2. AAO, Focal Points: Choosing the Appropriate PRK Patient, Module #9, 1998, p. 11-12.
3. AAO, Focal Points: Contact Lens Fitting for Regular Astigmatism, Module #11, 1997.
4. AAO, Preferred Practice Patterns Committee, Refractive Errors Panel. Refractive Errors Preferred Practice Pattern, 2007.

# Irregular astigmatism

## I. Describe the approach to establishing the diagnosis

### A. Definition

1. Astigmatism in an optical system is defined by the condition where variations in the curvature (typically cornea and lens in the human eye) prevent light rays from focusing to a single point
2. Irregular astigmatism is that astigmatism which cannot be corrected with a spherocylindrical lens

### B. Describe the etiology of this disease

1. Disease states
  - a. Corneal ectasia
    - i. Keratoconus
    - ii. Pellucid marginal degeneration
    - iii. Post-refractive surgery
  - b. Any cause of epithelial irregularity
    - i. EBMD
    - ii. Dry eye
  - c. Degenerative, inflammatory diseases or mass lesions
    - i. Pterygium
    - ii. Terrien marginal degeneration
    - iii. Peripheral ulcerative keratitis
    - iv. Dermoid
    - v. Salzmann nodular degeneration
    - vi. Corneal scar due to any cause
  - d. Post ocular surgery, particularly penetrating keratoplasty
  - e. Adnexal mechanical causes
    - i. Lid mass such as chalazion
    - ii. Orbital mass such as lacrimal gland tumor

### C. List the pertinent elements of the history

1. Vision decreased at all distances
2. Monocular diplopia (uncommon)

### D. Describe pertinent clinical features

1. Vision not correctable with eyeglass refraction

2. Vision is correctable with a rigid contact lens (depends on amount of astigmatism)
3. Uncorrected visual acuity poor at all distances

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

1. Corneal topography showing an irregular corneal surface
2. Gas-permeable contact lens overrefraction with improvement in best corrected visual acuity

**II. Define the risk factors**

- A. Trauma
- B. Family history of keratoconus
- C. Corneal surgery
- D. Refractive surgery complication

**III. List the most common or critical entities in the differential diagnosis**

- A. Exclude other cases of decreased vision associated with ocular pathology

**IV. Describe patient management in terms of treatment and follow-up**

**A. Describe medical therapy options**

1. Gas-permeable contact lenses are the mainstay of medical therapy
2. Eyeglasses and soft contact lenses typically work poorly for high amount of astigmatism
3. Observation
4. Treat primary condition, such as dry eye

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, 2013-2014.
2. AAO, Preferred Practice Patterns Committee, Refractive Errors Panel. Refractive Errors Preferred Practice Pattern, 2007.



# Presbyopia

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Aging
  - a. The accommodative amplitude of the crystalline lens gradually decreases as the lens enlarges with age
  - b. While wearing correction for ametropia, this reduction ultimately impairs reading vision
  - c. An emmetrope, or corrected myope, usually becomes symptomatic during the fifth decade of life
  - d. A latent or uncorrected hyperope is usually symptomatic earlier in life

### B. Define the relevant aspects of epidemiology of this disease

1. All phakic patients develop presbyopia with age

### C. List the pertinent elements of the history

1. Vision worse at near than at distance without correction or with correction in patients with concomitant myopia or hyperopia
2. Latent hyperopia may cause symptoms of premature presbyopia

### D. Describe pertinent clinical features

1. Reduced near vision

## II. List the most common or critical entities in the differential diagnosis

### A. Other forms of decreased vision associated with ocular pathology

## III. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Eyeglasses
  - a. Bifocal lenses, progressive lenses, or reading eyeglasses
2. Contact lenses
  - a. Using monovision with one eye corrected for distance and the other for reading
  - b. Bifocal

### B. Describe surgical therapy options

1. Refractive surgery with monovision correction

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction and Contact Lenses, Section 14: Refractive Surgery, 2013-2014.
2. AAO, Focal Points: Choosing the Appropriate PRK Patient, Module #9, 1998, p.8-9.
3. AAO, Preferred Practice Patterns Committee, Refractive Errors Panel. Refractive Errors Preferred Practice Pattern, 2007.

# Ocular surface problems related to contact lens wear

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Direct mechanical trauma from contact lens
2. More common with soft contact lens wear than with rigid gas-permeable contact lens
3. Immune-mediated response to mechanical trauma
4. Allergic reaction
5. Hypoxic response with metabolic epithelial damage
6. Toxicity from contact lens solution

### B. List the pertinent elements of the history

1. Redness
2. Itching, irritation, mucoid discharge
3. Pain
4. Blurred vision
5. Decreased tolerance or inability to wear contact lenses (common)

### C. Describe pertinent clinical features

1. Conjunctival changes
  - a. Papillary reaction of the superior tarsal conjunctiva
  - b. Giant papillary reaction in severe cases, referred to as giant papillary conjunctivitis (GPC)
  - c. Conjunctival injection and mucoid conjunctival discharge
2. Corneal changes
  - a. Punctate epithelial erosions on the cornea
  - b. Peripheral subepithelial corneal infiltrates
  - c. Corneal neovascularization
  - d. Corneal haze
3. Mild iritis

## II. Define the risk factors

### A. Extended contact lens wear

- B. Overnight contact lens wear**
- C. Lower oxygen permeability (more common with soft contact lenses than gas-permeable contact lenses)**

### **III. List the most common or critical entities in the differential diagnosis**

- A. Bacterial conjunctivitis, including chlamydia**
- B. Allergic conjunctivitis**
- C. Toxic conjunctivitis**
- D. Microbial keratitis, especially bacterial**

### **IV. Describe patient management in terms of treatment and follow-up**

#### **A. Describe medical therapy options**

1. Stop contact lens wear
2. Consider topical corticosteroids, usually low dose (if significant corneal inflammation present)

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Corneal Complications of Contact Lens, Module #2, 1993.

# Eyeglasses

## I. List the indications/contraindications

### A. Refractive errors

1. Eyeglasses are a time-proven, simple and safe method to correct refractive errors

### B. Presbyopia

1. Accommodative effort is increased when a patient with myopia changes from eyeglasses to contact lenses or after refractive surgery

### C. Protection against accidental injury

1. Individuals involved in certain sports and hazardous activities in which there is risk of eye trauma (Refer to section IV.E. Eye protection)
2. Individuals with good vision in only one eye

## II. Describe the evaluation

### A. History

1. Symptoms suggestive of a refractive error
2. Level of visual difficulties
3. Patient's visual needs in terms of occupation, lifestyle, and recreation

### B. Physical examination

1. Distance visual acuity for each eye with current correction
2. Near visual acuity
3. Refraction for each eye
  - a. Distance refraction performed with accommodation relaxed
  - b. Cycloplegic refraction is indicated when:
    - i. Accommodation cannot be relaxed (e.g., in children)
    - ii. Patient's symptoms are not consistent with manifest refractive error

## III. List the alternatives

### A. Contact lenses

### B. Refractive surgery

## IV. Describe considerations in prescribing and fitting eyeglasses

## **A. Myopia**

1. Individuals with asymptomatic myopia may not need eyeglass correction except for activities such as driving or school work
2. Overcorrection can cause asthenopic symptoms due to excessive accommodation
3. Some patients become symptomatic at low levels of illumination and may require increased minus correction for clearer vision at night

## **B. Hyperopia**

1. Slight undercorrection may be desired in young and middle-aged individuals, because of some physiologic accommodative tone
2. As the patient ages, full correction may be needed to provide optimal distance vision and to minimize problems with near vision

## **C. Astigmatism**

1. Individuals with regular astigmatism may not require full correction
2. Adults may not accept full cylindrical correction in their eyeglasses if their astigmatism has been only partially corrected or is at an oblique axis
3. Children usually adapt to their full cylindrical correction

## **D. Presbyopia**

1. Bifocals
2. Trifocals
  - a. Consider for patients with specific intermediate-vision needs
  - b. May be helpful for individuals who use computers
  - c. Top of the segment is set higher than that of a bifocal
3. Progressive addition lenses
  - a. Can be used to increase range of vision
  - b. Cosmetically well accepted
  - c. Major disadvantages
    - i. Peripheral distortion inherent in lens design
    - ii. Can have smaller size of reading zone and intermediate zone compared to bifocals or trifocals
    - iii. Difficulty in properly fitting the lenses
    - iv. May have trouble getting used to if patient has previously used segmented design
    - v. Generally more expensive than bifocals

## **E. Eye protection**

1. Polycarbonate lenses more impact and shatter resistant than other lens materials

## **V. List the difficulties and complications of eyeglass wear**

- A. Incorrect prescription**
- B. Prisms or prism effects**
  - 1. Vertical prism-induced diplopia can be found in presbyopic patients who wear bifocals
- C. Vertex distance**
- D. Spherical and chromatic aberrations**
- E. Lens distortions, including magnification and minification**
- F. Image jump and object displacement**

## **VI. Describe the follow-up care**

- A. If visual symptoms develop, evaluate the patient's eyeglasses and refraction**

### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, 2013-2014.
- 2. AAO, Low Vision Rehabilitation, 1999, p.55-56.
- 3. AAO, Focal Points: Athletic Eye Injuries & Appropriate Protection, Module #8, 1997.
- 4. AAO, Preferred Practice Patterns Committee, Refractive Errors Panel. Refractive Errors Preferred Practice Pattern, 2007.

# Visual disability/visual impairment

## Definitions

Visual Disability describes how the person functions in vision-related activities. Assessment includes: Activities of Daily Living (ADL), functional communication (e.g. facial expressions, reading, writing), personal and household tasks (e.g. cooking, grooming, managing medications), community tasks (e.g. shopping, vocational/avocational activities), and mobility (e.g. walking and vehicle transportation).

Visual Impairment describes how the eyes function. It is a decrease in function of the visual system caused by changes in the eye, the adnexa, or the central nervous system. Measures of visual impairment include visual acuity, visual field, contrast sensitivity, color vision, dark adaptation, etc.

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this "disease"

1. The etiology of the disability is the impairment
  - a. The etiology of the impairment is the underlying ophthalmic or CNS disorder

### B. Define the relevant aspects and epidemiology of this disease

1. Infants
  - a. Often multiple deficits, including brain injury-related cognitive visual impairment (CVI)
2. Elderly
  - a. Often multiple deficits, including hearing loss, mobility

### C. Describe appropriate testing and evaluation for establishing the diagnosis

1. Functional tests
  - a. Reading speed
  - b. Vocational
  - c. Driving

## II. Define the risk factors

### A. For visual impairment

1. Infancy
  - a. Prematurity
2. Adults/elderly
  - a. Age-related disorders



**B. For visual disability: (at any level of impairment)**

1. Social environment
  - a. Living situation
  - b. Educational/vocational parameters
  - c. Community services (e.g., transportation)
2. Personal factors
  - a. Comorbidities
  - b. Depression
  - c. Financial constraints

**III. List the most common or critical in differential diagnosis**

**A. Reversible ophthalmic disease**

1. Treat the underlying disorder

**B. Cognitive visual impairment (CVI) affecting higher visual functions**

**C. Malingering (hysterical or psychogenic visual loss)**

**IV. Describe patient management in terms of treatment and follow-up**

**A. Describe the "medical" therapy options**

1. Counseling
  - a. Address emotional, psychosocial impact, depression
2. Referral
  - a. Appropriate medical, vocational, educational professionals
3. Optical aids
  - a. Spectacle prescription based on low vision refraction
  - b. Magnification
    - i. Enlarge: large print books, cards, clocks, phones
    - ii. Bring closer: high-plus reading spectacles
    - iii. Magnify image: magnifiers, telescopes, video magnifiers
  - c. Prisms for image relocation
4. Lighting and glare control
  - a. Absorptive lenses and visors
  - b. Task oriented illumination control

5. Contrast enhancement
  - a. High contrast materials; selective transmission filters
  - b. Video magnifiers (closed circuit television)
6. Computers
  - a. Reading: screen enlargement software, speech output
  - b. Writing: keyboard, speech input
7. Vision substitution: non-visual adaptations
  - a. Auditory aids, e.g., "talking" glucometers, watches; books on tape (free through Library of Congress)
  - b. Tactile aids, e.g., braille; raised dots for appliances and dials, long cane for mobility
8. Skill development, e.g., reading with devices, activities of daily living (ADL) training, mobility training
9. Community Services, e.g., education, transportation

**B. Follow-up of "medical" therapy**

1. Follow-up essential to assure proper use of aids, reinforce skills, address psychosocial impact, confirm referrals

**C. Describe the surgical therapy options**

1. Currently none are Food and Drug Administration approved

## **V. List the complications of treatment, their prevention and management**

**A. Complications**

1. Failure to acquire devices or master adaptive skills
2. Change in needs with increased impairment and disability (secondary to progressive disease, new comorbidities, and changes in living/employment situation).

**B. Prevention and management**

1. Recognition of psychosocial impact of disability and appropriate treatment (e.g., depression)
2. Adequate training in the use of devices or new skills
3. Follow-up on referrals

## **VI. Describe disease-related complications**

- A. Loss of autonomy**
- B. Social isolation**
- C. Decrease in exercise**
- D. Depression**

- E. Falls and fractures (hip, wrist)**
- F. Medication errors**
- G. Nutritional decline**

## **VII. Describe appropriate patient instructions**

- A. Training in the application of spectacles and other devices to activities, and reassurance that much can be accomplished**
- B. Training in adaptive techniques and alternate strategies for task completion, with occupational therapists, orientation and mobility specialists, rehabilitation teachers, low vision therapists**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 3: Optics, Refraction, and Contact Lenses, 2013-2014.
2. AAO, Archives of Ophthalmology, Special issue on the Epidemiology of Vision Loss and Its Causes, April, 2004.
3. Faye, EE, Albert, DL, Freed, B, et al, The Lighthouse Ophthalmology Resident Training Manual: A New Look at Low Vision Care, Lighthouse Int'l, 2000.
4. Fletcher, DC, Ed, Low Vision Rehabilitation, Caring for the Whole Person, Amer. Academy of Ophthalmology Monograph, 1999.
5. Markowitz SN - Low Vision Rehabilitation, Ophthalmology Rounds, Vol. 1, # 2, Snell Medical Comm., Inc. Montreal, Canada, Nov. 2003.
6. Mogk, L., Mogk M, Macular Degeneration: The Complete Guide to Saving and Maximizing Your Sight, N.Y: Ballantine Books, 2003.
7. AAO, Preferred Practice Pattern Committee, Vision Rehabilitation Committee: Vision Rehabilitation for Adults Preferred Practice Pattern, 2007.

# Anatomy of the neurosensory retina

## I. Describe relevant aspects of the basic anatomy

### A. Neurosensory retina

1. Histological layers have been identified
2. The central retinal artery branches primarily into 4 anatomic quadrants and supports the metabolic demands of the inner neurosensory retina
  - a. Retinal artery ischemia could lead to atrophy of the inner layer only, sparing the outer retinal layers
3. The outer retina metabolic demand is supplied primarily from the choroidal circulation
  - a. Choroidal infarction or ischemia could lead atrophy of the outer layers, sparing the inner layers

### B. Retinal pigment epithelium (RPE)

1. Pigments within the RPE absorb excess light
2. Participates in the photo-transduction visual cycle as well as nutrient transport to the neurosensory retina

## II. Describe clinical correlations

### A. Neurosensory retina

1. Initial visual processing and organization
2. The posterior chamber can be safely entered surgically, anterior to the ora serrata and posterior to the ciliary body (injections, sclerotomies)

### B. Retinal pigment epithelium

1. Genetic defects, drugs, and aging can negatively impact the cellular health and viability of the RPE
2. Reactive changes of the RPE may be seen in various pathologic states such as bone-spicules in retinitis pigmentosa (RP)

### C. The potential space between the neurosensory retina and the RPE is important anatomically and can be critical with OCT interpretation

1. Choroidal neovascularization can lead to tissue and fluid in the sub-retinal space such as in exudative ARMD
2. Sub-retinal fluid or a macular serous detachment can be present in diseases that have exudation into this potential space such as in central serous chorioretinopathy

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular

Tumors, 2013-2014.

2. AAO, Basic and Clinical Science Course. Section 12: Retina and Vitreous, 2013-2014.

# Fluorescein angiography

## **I. List the indications/contraindications**

### **A. Indications**

1. Retinal vascular disease
2. Diabetic retinopathy
3. Macular degeneration and other maculopathies
4. Posterior uveitis
5. Tumors

### **B. Contraindications**

1. Pregnancy (relative contraindication)
2. Known allergy

## **II. Describe the pre-procedure evaluation**

- A. Explain procedure and obtain informed consent**
- B. Ask about any allergic reactions in the past**
- C. Dilate pupils**

## **III. List the alternatives to this procedure**

- A. Optical coherence tomography**

## **IV. Describe the technique**

- A. Intravenous injection of sodium fluorescein**
- B. Timed sequence of photographs (digital or film camera)**

## **V. List the complications of the procedure, their prevention and management**

### **A. Side effects**

1. Yellowing of skin
2. Yellow-orange urine color

### **B. Complications**

1. Extravasation and local tissue necrosis

2. Nausea, vomiting
3. Pruritus, urticaria
4. Vasovagal reaction
5. Severe allergic reaction including respiratory arrest and anaphylaxis
6. Thrombophlebitis
7. Death

#### Additional Resources

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Fluorescein and Indocyanine Green Angiography: Technique and Interpretation, 2nd ed., 1997.
3. AAO, Laser Photocoagulation of the Retina and Choroid, 1997, p.29, 102-103, 112-113, 150-151, 228-229, 238-239.
4. AAO, Focal Points: Advances in Posterior Segment Imaging Techniques, Module #7, 1999, p.1-4.

# Indications for optical coherence tomography

## I. List the indications/contraindications

### A. Indications

1. Macular diseases
  - a. Vitreomacular interface disorders
    - i. Vitreomacular traction (VMT)
    - ii. Epiretinal membrane (ERM)
    - iii. Macular hole
  - b. Intra-retinal or sub-retinal diseases
    - i. Cystoid macular edema (CME) associated with:
      - i) Post-op edema
      - ii) Diabetic retinopathy
      - iii) Retinal vascular disease
    - ii. Subretinal fluid (SRF) such as with
      - i) Central serous chorioretinopathy
    - iii. Age-related macular degeneration
      - i) Non-exudative (dry, drusen)
      - ii) Exudative (wet, CNVM)
2. Optic nerve diseases
  - a. Glaucomatous optic neuropathy

### B. Contraindications

1. Poor media clarity (relative contraindication)
2. Poor patient cooperation (relative contraindication)

## II. Describe considerations in interpretation of this procedure

### A. Acquisition of a good quality OCT scan

1. Clear ocular media
2. Steady patient fixation

### B. Accurate OCT scan interpretation - qualitative information



1. Review cross sectional anatomy
  - a. Identification of morphological changes in tissue layers - atrophy, edema, thickening, distortion
  - b. Interpretation of changes in the relative reflectivity of tissue layers - hyporeflectivity, hyperreflectivity
2. Serial, comparative review and analysis

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. Carl Zeiss Ophthalmic Systems, Inc. Optical Coherence Tomographer - Model 3000, User manual. Germany, 2002.
3. Joel S. Schuman, Carmen A. Puliafito, James G. Fujimoto. Optical Coherence Tomography of Ocular Disease, Second Edition. Slack Incorporated, 2004.
4. Massin P, Girach A, Erginay A, et al. Optical coherence tomography: a key to the future management of patients with diabetic macular oedema. *Acta Ophthalmol Scand* 2006;84:466-74.
5. AAO, Focal Points: Optical Coherence Tomography in the Management of Retinal Disorders, Module #11, 2006.
6. AAO, Preferred Practice Patterns. Diabetic Retinopathy: Diagnostic Procedures, Oct 2011
7. AAO, Preferred Practice Patterns. Age-related Macular Degeneration: Diagnostic Procedures, Oct 2011.
8. AAO, Preferred Practice Patterns. Idiopathic Macular Hole, Sept 2008.

# Age-related macular degeneration

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Reduced visual function
  - a. Metamorphopsia
  - b. Central scotoma
  - c. Blurred vision

### B. Describe pertinent clinical features

1. Non-neovascular age-related macular degeneration (AMD)
  - a. Drusen
  - b. Retinal pigment epithelium (RPE) pigmentary changes
    - i. Hyperpigmentation
    - ii. Depigmentation
  - c. Geographic atrophy
2. Neovascular AMD
  - a. Subretinal fluid
  - b. Subretinal blood
  - c. Hard exudate
  - d. Elevation of the RPE
    - i. Serous RPED
    - ii. Hemorrhagic RPED
    - iii. Fibrovascular RPED
    - iv. Disciform scar

### C. Describe appropriate testing and evaluation for establishing the diagnosis

1. Amsler grid
2. Fluorescein angiography
3. Optical coherence tomography

## II. Define the risk factors

### A. Age is typically >50 years with increasing risk for each decade thereafter

- B. Family history**
- C. Light colored irides**
- D. Smoking**

### **III. Describe patient management in terms of treatment and follow-up**

#### **A. Describe medical therapy options for non-neovascular AMD**

- 1. Micronutrient supplementation

#### **B. Describe medical therapy options for neovascular AMD**

- 1. Anti-vascular endothelial growth factor (VEGF) drugs

#### **C. Describe laser surgery therapy options for neovascular AMD**

- 1. Thermal laser surgery
- 2. Photodynamic therapy

#### **D. Other life-style recommendations**

- 1. No smoking and avoidance of second hand smoke
- 2. Control cardiovascular risk factors and include regular exercise
- 3. Diet with green leafy vegetables plus fresh fruits
- 4. Fish intake (2-3 servings per week)
- 5. Follow the Age-Related Eye Disease Studies (AREDS I and II) recommendations for supplements

### **IV. Describe appropriate patient instructions**

- A. Yearly follow-up, even with no subjective progression**
- B. Use of Amsler grid**
- C. Patient to report any changes in central visual function**
- D. Review risk of fellow eye involvement**
- E. Encourage use of low vision devices and vision rehabilitation**

#### **Additional Resources**

- 1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
- 2. AAO, Laser Photocoagulation of the Retina and Choroid, 1997, p.149-161,165-171,190-191,197-200.
- 3. AAO, Focal Points: Advances in the Treatment of Nonexudative Age-Related Macular Degeneration, Module #5, 2003.
- 4. AAO, Focal Points: Advances in the Treatment of Exudative Age-Related Macular Degeneration, Module #6, 2003.

5. AAO, Focal Points: Age-Related Macular Degeneration, Module #2, 1995.
6. AAO, Preferred Practice Patterns Committee, Retina Panel: Preferred Practice Patterns: Age Related Macular Degeneration, 2006.

# Ocular histoplasmosis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. *Histoplasma capsulatum* causes widespread foci of chorioretinitis that evolve into focal scars
2. Usually, the disease is in a cicatricial phase when diagnosed
3. History should include inquiry about photopsia, metamorphopsia, or central visual disturbance

### B. List the pertinent elements in the history

1. Endemic in the Ohio and Mississippi river valleys
2. Inquire about a history of living in one of these endemic regions
3. Document location and severity of the symptoms, but usually asymptomatic

### C. Describe pertinent clinical features

1. Classic triad
  - a. Peripheral "punched-out" chorioretinal scars
  - b. Peripapillary chorioretinal atrophy or peripapillary pigmentary scarring
  - c. Macular scars with secondary choroidal neovascular membranes (CNV) developing and emanating from edge of chorioretinal scar
2. Absence of inflammatory cells in the anterior chamber and vitreous
3. Bilateral usually

## II. List the differential diagnosis

### A. Age related macular degeneration

### B. Myopic degeneration

### C. Multifocal choroiditis

## III. Describe patient management in terms of treatment and follow-up

### A. The presence of CNV is an indication for referral to a retina or uveitis specialist

### B. Monitor with bilateral, monocular Amsler grid (or equivalent) testing

### C. Educate about the risks of choroidal neovascularization and current treatment options, should symptoms occur.

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.

# Idiopathic central serous chorioretinopathy

## I. Describe the approach to establishing the diagnosis

### A. Mechanism

1. Localized neurosensory retinal detachment
  - a. Occurs in the macular region
  - b. Usually due to one or more areas of serous detachment of the retinal pigment epithelium (RPE)

### B. Define the relevant aspects of epidemiology of the disease

1. Typically unilateral; recurrent in approximately 25% of cases
2. Has been described with systemic corticosteroid use
3. May be associated with emotional stress
4. Most prevalent in patients between 20 - 40 years of age
5. More common in men than women

### C. List the pertinent elements of the history

1. Central visual impairment
  - a. Metamorphopsia
  - b. Blurred vision
  - c. Central scotoma

### D. Describe pertinent clinical features

1. Localized serous retinal detachment
2. Serous RPE detachment (may or may not be visible)
3. Yellow spot in the center of the fovea
4. Small yellow subretinal precipitates

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Fluorescein angiogram
  - a. Focal leakage
  - b. Pooling of dye into the area of serous retinal detachment
  - c. Transmission defects are common in both eyes
2. Optical coherence tomography
  - a. Serous retinal pigment epithelial detachments
  - b. Serous retinal detachment(s)

## **II. Define the risk factors**

- A. Male gender**
- B. Corticosteroid use**

## **III. List the most common or critical entities in the differential diagnosis**

- A. Idiopathic choroidal neovascularization**
- B. Cystoid macular edema**
- C. Exudative retinal detachment**

## **IV. Describe initial patient management in terms of treatment and follow-up**

- A. Observation**
- B. Refer to a retina specialist if chronic or recurrent**

## **V. Describe disease-related complications**

- A. Loss of central vision due to chronic detachment of the macula with pigmentary changes**
- B. Secondary choroidal neovascularization (uncommon)**

### **Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.



# Epiretinal membrane

## I. Describe the approach to establishing the diagnosis

### A. Definition of disease

1. Contraction/ proliferation of glial tissue on the surface of the retina that may or may not cause visual symptoms
2. Other names for this condition include
  - a. Macular pucker
  - b. Cellophane maculopathy
  - c. Surface wrinkling retinopathy

### B. Define the relevant aspects of epidemiology of the disease

1. Etiology
  - a. Typically occurs in a patient with a posterior vitreous detachment, but can be associated with other ocular conditions including
    - i. Trauma
    - ii. Retinal venous occlusion
    - iii. Diabetic retinopathy
    - iv. Uveitis
    - v. Hereditary retinopathy
    - vi. Following retinal detachment
2. May occur at any age, but more common in patients over 50 years

### C. List the pertinent elements of the history

1. Central visual impairment
2. Blurred vision
3. Metamorphopsia

### D. Describe pertinent clinical features

1. Surface of the inner retina has shiny, glistening appearance
2. Retinal striae and/or folds
3. Cystoid macular edema (CME)
4. Membrane opacification
5. Vascular tortuosity or straightening

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Optical coherence tomography
2. Fluorescein angiogram

## **II. Define the risk factors**

- A. Posterior vitreous detachment**
- B. History of retinal abnormalities**
- C. Trauma**

## **III. Describe patient management in terms of treatment and follow-up**

- A. Observation if patient is asymptomatic or symptoms are mild**
- B. Pars plana vitrectomy and membrane peeling for patients with significant visual loss and symptoms**

## **IV. Describe the disease-related complications**

- A. Persistent metamorphopsia**
- B. Loss of central vision**
- C. Chronic CME**

### **Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Focal Points: Evaluation and Management of Diabetic Retinopathy, Module #1, 2003, p.7, 15-16.
3. AAO, Focal Points: Macular Surface Disorders, Module #4, 1996, p.5-7.

# Diabetic retinopathy

## I. Classification

- A. Nonproliferative
- B. Proliferative
- C. Diabetic macular edema

## II. Describe the approach to establishing the diagnosis

### A. Define the relevant aspects of epidemiology of the disease

- 1. Prolonged hyperglycemia as evidenced by chronic increase in Hemoglobin A1c (HbA1c)
- 2. Increased duration of diabetes mellitus
- 3. Elevated blood pressure and lipids

### B. List the pertinent elements of the history

- 1. Duration and type of diabetes mellitus
- 2. Glycemic control (HgA1C)
- 3. Systemic hypertension
- 4. Nephropathy
- 5. Neuropathy

### C. Describe pertinent clinical features

- 1. Microaneurysms
- 2. Dot and blot hemorrhages
- 3. Cotton wool spots
- 4. Intraretinal lipid (hard exudates)
- 5. Neovascularization
- 6. Vitreous hemorrhage
- 7. Tractional retinal detachment

### D. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Fundus photography
- 2. Optical coherence tomography
- 3. Fluorescein angiography

## III. List the differential diagnosis for diabetic retinopathy

- A. Hypertensive retinopathy**
- B. Retinal vein occlusions**

#### **IV. Describe patient management in terms of treatment and follow-up**

- A. Describe the natural history, outcome and prognosis**
  - 1. Progression and severity correlates with glycemic control and duration of disease
- B. Describe medical therapy options**
  - 1. Treat associated medical conditions (refer to primary care provider)
  - 2. Intravitreal injection of anti-VEGF and/or steroids medications
- C. Describe surgical therapeutic options**
  - 1. Laser photocoagulation
    - a. Focal laser for macular edema
    - b. Panretinal photocoagulation (PRP) for proliferative disease
- D. Annual eye exam or sooner if symptomatic**
- E. Referral to retinal specialist**

#### **V. List the complications of treatment, their prevention and management**

- A. Laser**
  - 1. Macular scarring with focal laser
  - 2. Peripheral field loss with PRP laser
- B. Intravitreal injections**
  - 1. Endophthalmitis
  - 2. Cataract
  - 3. Detached retina

#### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 12: Retina and Vitreous, 2013-2014.
- 2. AAO, Preferred Practice Pattern Committee, Retina Panel. Diabetic Retinopathy Preferred Practice Pattern, 2008.
- 3. AAO, Laser Photocoagulation of the Retina and Choroid, 1997, p.63-95.
- 4. AAO, Diabetes and Ocular Disease: Past, Present and Future Therapies, 2000, p.4-5, 8, 13, 20-26, 36-40, 45-49, 69-202, 239-298.
- 5. AAO, Focal Points: Practical Management of Diabetic Retinopathy, Module #3, 2003.

6. AAO, Focal Points: Diabetic Retinopathy: Classification, Progression, and Management, Module #7, 1993.
7. Early Treatment Diabetic Retinopathy Study Research Group. Results from the Early Treatment Diabetic Retinopathy Study. *Ophthalmology*. 1991;98:739-840.
8. Wilkinson CP, Ferris FL 3rd, Klein RE et al. Proposed international clinical diabetic retinopathy and diabetic macular edema disease severity scales *Ophthalmology*. 2003; 110:1677-82.

# Branch retinal vein occlusion

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Exact mechanisms unknown
  - a. The branch retinal vein and artery share a common adventitial sheath
  - b. If arteriosclerotic changes are found in the artery which often lies on top of the branch retinal vein, a thrombosis may occur in the vein

### B. Define the relevant aspects of epidemiology of the disease

1. Generally occur in patients 50 years or older
2. Increased association with
  - a. Diabetes mellitus
  - b. Hypertension
  - c. Hyperlipidemia
3. History of ocular disease
  - a. Primary open-angle glaucoma

### C. List the pertinent elements of the history

1. Systemic hypertension
2. Diabetes mellitus
3. Possible visual acuity impairment

### D. Describe pertinent clinical features

1. Measure intraocular pressure (IOP) in both eyes, and assess for possible glaucoma
2. Possible macular edema, retinal hemorrhages, cotton-wool spots, retinal neovascularization and capillary dropout

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Fundus photographs
2. Fluorescein angiography to assess for
  - a. Cystoid macular edema
  - b. Capillary non-perfusion
  - c. Neovascularization
3. Optical coherence tomography for macular edema

## **II. Define the risk factors**

### **A. Systemic diseases**

1. Hypertension
2. Diabetes mellitus
3. Hyperlipidemia

### **B. Ocular disease**

1. Primary open-angle glaucoma

## **III. List the most common or critical entities in the differential diagnosis**

### **A. Diabetic retinopathy**

#### **Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Laser Photocoagulation of the Retina and Choroid, 1997, p.97, 110-119.
3. AAO, Focal Points: Branch and Central Vein Occlusions, Module #9, 1997, p.1-5.

# Central retinal vein occlusion

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Exact mechanisms unknown, though may involve thrombus at level of anterior optic nerve
2. Rarely may have systemic diseases associated with hypercoagulability, particularly if bilateral

### B. Define the relevant aspects of epidemiology of the disease

1. Occur more commonly in patients 50 years or older
2. Increased association with diabetes mellitus, hypertension
3. Increased association with primary open-angle glaucoma (POAG)

### C. List the pertinent elements of the history

1. History of systemic diseases such as diabetes mellitus, hypertension
2. Impaired visual function

### D. Describe pertinent clinical features

1. Retinal hemorrhages in all four quadrants
2. Vascular dilation, and tortuosity
3. Possible macular edema
4. Cotton-wool spots
5. Capillary dropout
6. Collateral vessels at the disc
7. Retinal and iris neovascularization
8. Optic disc edema
9. Relative afferent pupillary defect

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Fundus photographs
2. Fluorescein angiography

## II. List the most common or critical entities in the differential diagnosis

### A. Diabetic retinopathy

### B. Ocular ischemia from carotid disease

## III. Describe patient management in terms of treatment and follow-up



- A. Careful follow-up for detection of iris and angle neovascularization**
- B. Scatter peripheral laser therapy for iris neovascularization**
- C. Glaucoma may require treatment**

#### Additional Resources

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Laser Photocoagulation of the Retina and Choroid, 1997, p.97-108.
3. AAO, Focal Points: Branch and Central Vein Occlusions, Module #9, 1997, p.5-7.

# Branch retinal artery occlusion

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology/multifactorial nature of this disease

1. Embolization, especially from a carotid or cardiac source
2. Thrombosis of the affected vessel

### B. Define the relevant aspects of epidemiology of the disease

1. Associated age-dependent systemic conditions
  - a. Hypertension
  - b. Diabetes mellitus
  - c. Cardiac valvular disease

### C. List the pertinent elements of the history

1. Unilateral, painless, abrupt onset of segmental visual field defect corresponding to the affected retina
2. History of amaurosis fugax may be present

### D. Describe pertinent clinical features

1. Superficial opacification or whitening along the distribution of a branch retinal artery from edema when acute
2. Narrower irregular caliber of branch retinal artery
3. Emboli may be seen

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Fluorescein angiography
2. Carotid artery evaluation and cardiac echography

## II. Define the risk factors

- A. Smoking
- B. Hypertension
- C. Cardiac valvular disease
- D. Carotid arterial disease
- E. IV drug abuse
- F. Sepsis

## III. Describe patient management in terms of treatment and follow-up

**A. Possible role of ocular massage or anterior chamber paracentesis in moving the embolus downstream**

Additional Resources

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Electrophysiologic Testing in Disorders of the Retina, Optic Nerve and Visual Pathway, 2nd ed., 2001, p. 85-88.
3. AAO, Focal Points: Retinal Arterial Obstruction, Module #1, 1994, p. 3-7.

# Central retinal artery occlusion

## I. Describe the approach to establishing the diagnosis

### A. Describe the common etiologies of this disease

1. Atherosclerosis
2. Embolization
3. Inflammation (giant cell arteritis (GCA))

### B. Define the relevant aspects of epidemiology of the disease

1. Seventh decade most common
2. Retinal vascular occlusive disease prevalence
  - a. Majority are central retinal artery occlusions
  - b. Some are branch artery occlusions

### C. List the pertinent elements of the history

1. Painless, sudden, usually severe visual loss in one eye, usually to counting fingers (CF) or hand movements (HM)
2. May have a history of amaurosis fugax

### D. Describe pertinent clinical features

1. Opaque, edematous retina, particularly in posterior pole (when acute)
2. Cherry red spot (orange color from intact choroidal vasculature beneath fovea; when acute, may take hours to develop)
3. Marked relative afferent pupillary defect (RAPD)
4. Narrowed retinal arterioles and box-carring or segmentation of the blood column in the arterioles (when acute)
5. Retinal arteriolar emboli
6. Patent cilioretinal artery may spare the fovea and central vision

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Fluorescein angiography
2. Immediate erythrocyte sedimentation rate (ESR), complete blood count (CBC), and C-reactive protein (CRP) in patients without visible embolus to exclude associated GCA
3. Check the blood pressure
4. Carotid artery evaluation and cardiac echography

## II. Define the risk factors

- A. Atherosclerotic cardiovascular disease
- B. Smoking
- C. Oral contraceptive use
- D. Hypertension
- E. Cardiac valvular disease
- F. Cardiac septal defects
- G. Other disease associated with emboli
- H. Hypercoagulable states
- I. Intravenous drug use

### **III. List the most common or critical entities in the differential diagnosis**

- A. Arteritic anterior ischemic optic neuropathy

### **IV. Describe patient management in terms of treatment and follow-up**

- A. Describe immediate patient management
  - 1. Ocular massage
  - 2. Anterior chamber paracentesis
  - 3. Erythrocyte sedimentation rate (ESR), CBC and CRP
  - 4. Temporal artery biopsy if indicated to rule out GCA
    - a. If GCA is suspected, begin high-dose systemic corticosteroids while awaiting ESR, CRP, and temporal artery biopsy results

#### **Additional Resources**

- 1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
- 2. AAO, Focal Points: Retinal Arterial Obstruction, Module #1, 1994.

# Cystoid macular edema

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Multifactorial
  - a. Inflammation
  - b. Vascular injury
  - c. Congenital vascular leakage
  - d. Loss of intercellular adhesion
2. Loss of microvascular integrity with fluid extravasation into cystoid spaces within the macula

### B. Define the relevant aspects of epidemiology of the disease

1. May be associated with numerous conditions
  - a. Cataract surgery (most common) or other intraocular surgery
  - b. Venous occlusive disease
  - c. Diabetic retinopathy
  - d. Uveitis
  - e. Retinitis pigmentosa
  - f. Medication usage
    - i. Prostaglandin analogs (i.e. topical glaucoma therapy)
    - ii. Systemic medications (i.e. Niacin)

### C. List the pertinent elements of the history

1. Central visual impairment
2. Blurred vision
3. Metamorphopsia
4. Past medical history
  - a. Past surgery
  - b. Systemic disease
  - c. Topical glaucoma therapy

### D. Describe pertinent clinical features

1. Honeycomb-like cystoid spaces surrounding and involving the center of the fovea
2. May appear as intraretinal striae radiating from center of fovea
3. Possibly vitreomacular traction

4. Concurrent findings from associated disease states (diabetic retinopathy, venous occlusive disease, etc.)
5. Retinal pigment epithelium (RPE) hypertrophy, clumping, or atrophy if longstanding
6. Anterior segment vitreous adhesions

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

1. Optical coherence tomography (OCT)
2. Fluorescein angiography

**II. Define the risk factors**

- A. Contralateral post-surgical cystoid macular edema (CME)
- B. Epiretinal membrane (ERM)
- C. Vitreomacular interface abnormalities
- D. Uveitis
- E. Diabetic retinopathy
- F. Multiple intraocular surgeries

**III. Describe patient management in terms of treatment and follow -up**

- A. Observation if mild, as many cases of post-operative CME resolve spontaneously within 6 weeks
- B. Topical nonsteroidal anti-inflammatory drugs (NSAIDs) and topical corticosteroids
- C. Sub-Tenon corticosteroid injection
- D. Oral corticosteroids, particularly if CME is associated with uveitis
- E. Intravitreal corticosteroid injection
- F. Surgery (i.e. association with ERM)

**IV. Describe disease-related complications**

- A. Permanent reduction of central vision if edema fails to resolve
- B. Steroid-related side effects
- C. Surgery-related side effects or complications

**Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Laser Photocoagulation of the Retina and Choroid, 1997, p.55.
3. AAO, Diabetes and Ocular Disease: Past, Present and Future Therapies, 2000, p.148, 211-213.

4. AAO, Focal Points: Evaluation of Impaired Visual Acuity Following Cataract Surgery, Module #6, 1996, p.11-12.
5. AAO, Focal Points: Retinal Complications Following Cataract Surgery, Module #12, 1992, p.4-6.
6. AAO, Focal Points: Optical Coherence Tomography in the Management of Retinal Disorders, Module #11, 2006.



# Spontaneous vitreous hemorrhage

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. During a posterior vitreous detachment (PVD) or with traction
  - a. Avulsion of a retinal vessel
  - b. Traction at an area of neovascularization
  - c. Development of a retinal tear
2. Spontaneous hemorrhage from retinal or disc neovascularization
3. Vascular occlusive disorders

### B. Define the relevant aspects of epidemiology of the disease

1. Common causes
  - a. Proliferative diabetic retinopathy
  - b. Retinal tear without detachment
  - c. Posterior vitreous detachment (PVD)
  - d. Rhegmatogenous retinal detachment
  - e. Retinal neovascularization associated with vascular occlusive disease (e.g., sickle cell disease)
  - f. Peripheral retinal neovascularization
  - g. Subretinal hemorrhage with secondary vitreous hemorrhage (e.g., hemorrhagic exudative process from ARMD with breakthrough vitreous hemorrhage)
  - h. Arterial macroaneurysm

### C. List the pertinent elements of the history

1. Decreased visual acuity
2. New onset floaters
3. Photopsia
4. Visual field defect

### D. Describe pertinent clinical features

1. Vitreous hemorrhage

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Ultrasonography if hemorrhage obscures view of retina
2. Peripheral retinal examination using scleral depression

3. Examine fellow eye
4. Testing for diabetes mellitus if suspected

## **II. List the most common or critical entities in the differential diagnosis**

- A. Unlikely to be mistaken for anything else but following might be confused with an old vitreous hemorrhage**
  1. Vitreous opacity due to chronic uveitis
  2. Vitreous opacity due to chronic endophthalmitis
  3. Asteroid hyalosis
  4. Lymphoma

## **III. Describe patient management in terms of treatment and follow-up**

- A. Observe for clearing of hemorrhage if retina attached and no retinal breaks**
- B. Laser surgery or cryopexy of retinal break if present and visible**
- C. Laser surgery for ischemic, proliferative process**
- D. Refer to retina specialist for other surgical options**

## **IV. List the complications of treatment, their prevention and management**

- A. Complications of laser surgery or cryopexy**
- B. Complications of vitrectomy surgery**

## **V. Describe disease-related complications**

- A. Progression to RD possible if untreated or lost to follow-up**

## **VI. Describe appropriate patient instructions**

- A. Follow up is very important**
  1. May need repeat ultrasonography until hemorrhage clears sufficiently for retinal examination
- B. Patient should be instructed to monitor for and report new onset of visual field defect or loss of vision**
- C. May require vitrectomy if no clearance or retinal detachment is diagnosed**

### **Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.

2. AAO, Focal Points: Diagnosis and Management of Vitreous Hemorrhage, Module #10, 2000, p.9.

# Posterior vitreous detachment

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Initial event: Liquefaction of the central vitreous humor
2. Hole develops in the posterior vitreous cortex
3. Liquefied vitreous passes abruptly into the subhyaloid space
4. The posterior hyaloid rapidly separates from the retina
5. Vitreous gel remains attached to vitreous base
6. Resulting vitreous traction on vitreous base or upon other visible or invisible vitreoretinal adhesions can result in a retinal break

### B. Define the relevant aspects of epidemiology of the disease

1. Prevalence increases with increasing axial length and patient age

### C. List the pertinent elements of the history

1. Many patients are asymptomatic
2. Photopsias
3. Floaters, usually sudden onset
4. Cloudy or hazy vision

### D. Describe pertinent clinical features

1. Weiss ring: glial floater overlying the optic disc
2. Shafer sign: pigment in the anterior vitreous may indicate an associated retinal break

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Slit-lamp biomicroscopy with indirect condensing lens or contact lens
2. Indirect ophthalmoscopy with scleral depression to detect associated retinal breaks
3. B-scan echography, if a vitreous hemorrhage precludes ophthalmoscopy

## II. Define the risk factors

### A. Aphakia or pseudophakia

### B. Post-trauma

### C. Myopia

### D. Uveitis

### E. Age

### **III. List the most common or critical entities in the differential diagnosis**

- A. Vitreous hemorrhage**
- B. Vitreous inflammation**
- C. Infection (e.g., endophthalmitis giving visual haze)**

### **IV. Describe patient management in terms of treatment and follow-up**

#### **A. Describe the natural history, outcome, and prognosis**

- 1. Acute PVD without vitreous hemorrhage
  - a. Low incidence of retinal break
- 2. Acute PVD with vitreous hemorrhage
  - a. High incidence of retinal break
- 3. Vitreous separation may be incomplete, and tears can occur weeks to months following initial symptoms

#### **B. Describe medical therapy options**

- 1. If an associated vitreous hemorrhage prevents adequate examination, bed rest with the patient's head elevated 45 degrees for 1-2 days may clear the vitreous sufficiently to allow breaks in a superior location to be found

#### **C. The Preferred Practice Pattern monograph for PVD recommends follow-up retinal exam in 1-6 weeks for an acute PVD, depending on the extent of traction, vitreous hemorrhage, etc**

### **V. Describe disease-related complications**

- A. Vitreous hemorrhage without a retinal break**
- B. Retinal break with or without vitreous hemorrhage**
- C. Rhegmatogenous retinal detachment**

### **VI. Describe appropriate patient instructions**

#### **A. Natural history description**

#### **B. If any of the following symptoms develop, the patient should return for an eye examination right away (RD precautions)**

- 1. Significant photopsias or flashes
- 2. Any new floaters or change in floaters
- 3. A dark veil or curtain coming over the vision from any direction in the affected eye

#### **C. Monocular screening (affected eye should be checked while fellow eye is covered)**

## Additional Resources

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Retinal Detachment: Principles and Practice, 2nd ed., 1995, p.26-29.
3. AAO, Focal Points: Diagnosis and Management of Vitreous Hemorrhage, Module #10, 2000, p.5-6.
4. AAO, Focal Points: Retinal Complications Following Cataract Surgery, Module #12, 1992, p. 1-2.
5. AAO, Preferred Practice Patterns Committee, Retina Panel. Posterior Vitreous Detachment, Retinal Breaks and Lattice Degeneration Preferred Practice Pattern, 2008.

# Retinal tears

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Vitreoretinal traction in the setting of a posterior vitreous detachment (PVD)
2. Traction concentrated at a point of vitreoretinal adhesion - posterior extension of the vitreous base, cystic retinal tuft, zone of lattice degeneration, chorioretinal scar
3. Typical horseshoe configuration resulting from firm attachment of vitreous body to the vitreous base, with residual vitreous traction on flap of the tear often elevating it anteriorly

### B. Define the relevant aspects of epidemiology of this disease

1. A small percent of eyes with a symptomatic PVD are found to have at least one retinal break
2. Symptomatic retinal flap tears
  - a. higher rate of clinical retinal detachment especially with
    - i. Aphakia
    - ii. Pseudophakia
    - iii. Vitreous hemorrhage
    - iv. Presence of significant subretinal fluid
    - v. Larger breaks
    - vi. Superior breaks
3. Asymptomatic operculated tears much less commonly lead to retinal detachment

### C. List the pertinent elements of the history

1. Symptomatic retinal tear
  - a. Sudden increase in
    - i. Floaters
    - ii. Photopsias
    - iii. Peripheral scotoma may be associated with a retinal detachment
  - b. Tear likely occurred at or near the time of the examination
2. Asymptomatic retinal tear
  - a. Suggests that the tear might have been present for an extended period of time prior to examination

### D. Describe pertinent clinical features

1. Red blood cells or pigment granules on slit-lamp biomicroscopic examination of the anterior vitreous

- a. Cells may sometimes be present in the posterior vitreous
- 2. Full-thickness retinal break with horseshoe configuration on indirect ophthalmoscopy with scleral depression
- 3. Subretinal fluid may be present around the retinal tear
- 4. Pigmentary change around retinal tear is evidence of chronicity (>3 months)
- 5. Retinal vessel sometimes bridging a tear can be a source of vitreous hemorrhage

## **II. Define the risk factors**

- A. Increasing age
- B. History of retinal tear or detachment in contralateral eye
- C. Family history of retinal tear or detachment
- D. Ocular trauma
- E. Axial myopia
- F. Aphakia, pseudophakia
- G. Lattice degeneration
- H. Recent (Nd:YAG) posterior capsulotomy
- I. Retinal detachment in the fellow eye

## **III. List the most common or critical entities in the differential diagnosis**

- A. Operculated retinal tear
- B. Atrophic retinal hole
- C. Serous retinal detachment (different management and evaluation)

## **IV. Describe patient management in terms of treatment and follow-up**

- A. Describe the natural history, outcome and follow-up
  - 1. Most symptomatic retinal tears resulting in retinal detachment occur within 6 weeks of the onset of symptoms
  - 2. Retinal detachment rarely occurs more than 3 months after development of the tear
  - 3. Asymptomatic retinal tears rarely progress to retinal detachment
  - 4. Pigmentary demarcation around retinal tear is evidence of chronicity and, while the demarcation itself is not protective, its presence might reassure that the condition has not been progressive
  - 5. Subretinal fluid extension beyond a pigmentary demarcation or multiple pigmented or depigmented lines implies that the condition is progressive



## **V. Describe disease-related complications**

- A. Progression to retinal detachment**
- B. Epiretinal membrane formation (ERM)**
- C. Vitreous hemorrhage**

### **Additional Resources**

1. AAO, Basic Clinical and Science Course, Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Laser Photocoagulation of the Retina and Choroid, 1997, p.211-212.
3. AAO, Retinal Detachment: Principles and Practice, 2nd ed., 1995, p.12-14, 17-18.
4. AAO, Focal Points: Retinal Breaks, Holes and Tears, Module #3, 1996, p.7-8.
5. AAO, Preferred Practice Pattern Committee, Retina Panel. Posterior Vitreous Detachment, Retinal Breaks and Lattice Detachment Preferred Practice Pattern, 2008.

# Retinal detachment

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Rhegmatogenous retinal detachment
  - a. Vitreous liquefaction
  - b. Posterior vitreous detachment (PVD)
  - c. Vitreous traction
  - d. Retinal break (usually horseshoe tear)
2. Traction retinal detachment
  - a. Vitreous traction on the retina (usually fibrocellular membranes)
  - b. Associated conditions (e.g., proliferative diabetic retinopathy)
3. Exudative retinal detachment
  - a. Accumulation of subretinal fluid in the absence of retinal breaks or traction
  - b. Exudation from a malignant, inflammatory, or vascular process

### B. List the pertinent elements of the history (rhegmatogenous)

1. Curtain or shadow moving over the field of vision
2. Photopsias
3. Floaters
4. Peripheral and/or central visual loss
5. Previous ocular trauma
6. Previous ocular surgery
7. High myopia
8. Family history

### C. Describe pertinent clinical features

1. Rhegmatogenous
  - a. Elevation of retina associated with a break in the retina
2. Tractional
  - a. Elevation of retina at site of fibrovascular traction
  - b. Can be relatively stable if no associated retinal breaks
3. Exudative
  - a. RD with smooth convex surface

- b. Location of fluid is gravity dependent (usually inferior with patient upright)
- c. Choroidal lesions may be present

**D. Describe appropriate testing and evaluation for establishing the diagnosis**

- 1. Indirect ophthalmoscopy with depressed exam if indicated
- 2. B-scan echography (especially if view obscured)

**II. Define the risk factors (rhegmatogenous)**

- A. Myopia
- B. Lattice degeneration
- C. Acute PVD
- D. Prior retinal detachment in the fellow eye
- E. Family history
- F. Recent ocular surgery
- G. Ocular trauma

**III. List the most common differential diagnosis**

- A. Degenerative retinoschisis
- B. Choroidal detachment
- C. Peripheral white without pressure

**IV. Describe patient management in terms of treatment and follow-up**

- A. Rhegmatogenous
  - 1. Surgical therapy is usually indicated
- B. Tractional
  - 1. Often observed if the macula is attached
  - 2. Vitrectomy surgery with progression threatening the macula
- C. Exudative
  - 1. Determination of underlying cause and treat

**V. Describe disease-related complications**

- A. Permanent vision loss
- B. Proliferative vitreoretinopathy (PVR)
- C. Hypotony

- D. Phthisis bulbi (if left untreated)**
- E. Surgery or treatment associated complications (e.g., cataract after vitrectomy)**
- F. Recurrent retinal detachment**

## **VI. Describe appropriate patient instructions**

- A. Discuss warning signs of floaters, photopsias, visual field defect**
- B. Importance of prompt consultation with ophthalmologist or retinal specialist if these signs or symptoms develop**
- C. Eye Protection**
- D. Benefits, risks, alternatives of prophylactic and/or surgical management**

### **Additional Resources**

1. AAO, Basic Clinical and Science Course, Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Diabetes and Ocular Disease: Past, Present and Future Therapies, 2000, p.163-164,170.
3. AAO, Retinal Detachment: Principles and Practice, 2nd ed., 1995, p.7, 77-78.
4. AAO, Focal Points: Retinal Detachment, Module #5, 1998, p.2-6.
5. AAO, Preferred Practice Patterns Committee, Retina Panel: Posterior Vitreous Detachment, Retinal Breaks and Lattice Degeneration Preferred Practice Pattern. 2008.

# Closed-globe blunt trauma

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Direct and indirect concussive injury to the posterior pole may create posterior segment damage due to deformation and energy transfer

### B. Define the relevant aspects of epidemiology of the disease

1. Young males most frequently affected
2. Physical violence, including sports
3. Motor vehicle accidents (e.g., airbag injury)
4. Workplace trauma
5. BB gun or paint ball injuries

### C. List the pertinent elements of the history

1. Circumstances of injury, including mechanism of injury and persons involved
2. Timing of accident and visual loss
3. History of vision prior to accident
4. Use of eyeglasses or protective eye wear at the time of accident

### D. Describe pertinent posterior segment clinical features

1. Vitreous hemorrhage
2. Retinal tears, dialysis, giant retinal tears
3. Commotio retinae (retinal whitening)
4. Macular hole
5. Choroidal rupture

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Complete eye examination
2. Special attention to
  - a. Relative afferent pupillary defect
  - b. Gonioscopy to evaluate for angle recession
  - c. Depressed exam to evaluate peripheral retina
  - d. Possible fundus photographs for documentation
  - e. 'Gentle' ultrasound if posterior pole is obscured
  - f. Computed tomography (CT) scan if ruptured globe or IOFB suspected (avoid MRI scan)

## **II. List the most common or critical entities in the differential diagnosis**

- A. Rule out open globe injury**

## **III. Describe patient management in terms of treatment and follow-up**

- A. Describe the natural history, outcome and prognosis**

1. May have variable degrees of visual loss
2. May have long-term consequences such as glaucoma or retinal detachment

- B. Describe medical therapy options**

1. Topical corticosteroids
2. Cycloplegia
3. Pressure-lowering agents

- C. Describe surgical therapy options**

1. Treatment of retinal breaks
2. Repair if retinal detachment
3. Macular hole surgery
4. Vitrectomy for vitreous hemorrhage

## **IV. Describe disease-related complications**

- A. Loss of sight**

- B. Cataract**

- C. Dislocated lens or zonular weakness**

- D. Glaucoma**

- E. Retinal detachment**

- F. Posterior choroidal rupture**

1. With or without secondary choroidal neovascularization

## **V. Describe appropriate patient instructions**

- A. Medication instructions (topical regimen)**

- B. Monitor vision (monocular screening)**

- C. Periodic follow-up to monitor IOP and ocular status**

- D. Eye protection**

## Additional Resources

1. AAO, Basic and Clinical Science Course. Section 12. Retina and Vitreous, 2013-2014.
2. Williams DF, Mieler WF, Williams GA. Posterior segment manifestations of ocular trauma. *Retina* 1990;10 Suppl 1:S35-44.
3. Martin et al. Treatment and pathogenesis of traumatic chorioretinal rupture (sclopetaria). *Am J Ophthalmol* 117:2:190-200, 1994.

# Open-globe injury

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Perforating globe injury
  - a. Entrance wound plus exit wound
  - b. Usually a missile-like injury
2. Penetrating globe injury
  - a. Entrance wound but no exit wound
  - b. Usually a sharp laceration trauma

### B. Define the relevant aspects of epidemiology of the disease

1. Most common in young males
2. May be work-related or recreational accident

### C. List the pertinent elements of the history

1. Mechanism and nature of injury (specifically time and circumstances of injury, rule out possible intraocular foreign body, high or low-velocity injury, use of eye protection)
2. Previous history of ocular trauma and surgery
3. Timing of accident and visual loss
4. Use of glasses or protective eye wear at the time of accident
5. Level of acuity prior to injury
6. Medications
7. Time of last food intake
8. Status of tetanus prophylaxis
9. Work or occupation related (did employer offer protective eyewear?)

### D. Describe pertinent clinical features (variable)

1. Visual impairment
2. Pain
3. Chemosis and/or conjunctival hemorrhage
4. Corneal, corneoscleral, and/or scleral laceration or rupture
  - a. Partial thickness versus full thickness
  - b. Seidel testing for smaller lacerations
  - c. Length and depth of laceration



- d. Iris prolapse
- 5. Anterior chamber
  - a. Altered depth
  - b. Hyphema
- 6. Cataract and/or dislocated lens
- 7. Altered intraocular pressure
- 8. Vitreous hemorrhage
- 9. Retinal and/or choroidal detachment and hemorrhage
- 10. Possible intraocular or intraorbital foreign bodies (IOFB)
- 11. Relative afferent pupillary defect
- 12. Possible evidence of endophthalmitis at presentation
- E. Describe appropriate testing and evaluation for establishing the diagnosis**
  - 1. Computed tomography (CT) scan
    - a. Check for intraocular or intraorbital foreign bodies
  - 2. 'Gentle' ultrasound, in order to minimize risk of extrusion of intraocular contents

## **II. Define the risk factors**

- A. Occupational injury**
- B. Personal injury, e.g., gunshot wound, motor vehicle accident**
- C. Lack of protective eyewear**
- D. Soldier in modern warfare**

## **III. List the most common or critical entities in the differential diagnosis**

- A. Blunt (closed-globe) injury**
- B. Intraocular foreign body**
- C. Partial thickness ocular laceration**

## **IV. Describe patient management in terms of treatment and follow-up**

- A. Describe medical therapy options (adjunctive role)**
  - 1. Protect eye until time of surgical assessment and intervention
  - 2. Consider tetanus toxoid after perforating injury if not up to date
  - 3. Pain medication when indicated

**B. Describe surgical therapy options**

1. When indicated, primary surgery for exploration, and closure of wounds
2. Refer for secondary evaluation and intervention when appropriate

**V. Describe disease-related complications**

- A. Endophthalmitis**
- B. Retinal tear and detachment**
- C. Sympathetic ophthalmia (rare)**
- D. Toxicity from IOFB**

**VI. Describe appropriate patient instructions**

- A. Recommend physical restrictions, importance of eye protection, and plans for follow-up care**
- B. Discuss expectations for postoperative recovery and visual rehabilitation depending on nature and extent of the injury**

**Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Focal Points: Open-Globe Injuries, Module #7, 2001.
3. Benson WH, Snyder IS, Granus V, Odom JV, Macsai MS. Tetanus prophylaxis following ocular injuries. J Emerg Med 1993; 11:677-83.
4. Mieler WF, Mittra RA. The role and timing of pars plana vitrectomy in penetrating ocular trauma. Arch Ophthalmol. 1997;115:1191-1192.

# Intraocular foreign body

## I. Describe the approach to establishing the diagnosis

### A. Describe etiology of disease

1. Object penetrates globe and all or part remains within the eye
2. Intraocular foreign body (IOFB) is an ocular emergency

### B. Define the relevant aspects of epidemiology of the disease

1. High-risk environments that involve exploding particles or "metal on metal" contact
2. Firearms, BB guns, pellet guns
3. Inadequate eye protection with high-risk activities

### C. List the pertinent elements of the history

1. Eye pain associated with high-risk exposure or event
2. Visual blurring

### D. Describe pertinent clinical features (variable)

1. Subconjunctival hemorrhage
2. Corneoscleral or conjunctival/ scleral laceration
3. Lens trauma (check for sectoral cataract or phacodonesis)
4. Iris trauma (iris defects or iridodonesis)
5. Hyphema
6. Vitreous hemorrhage
7. Retinal tear
8. Subretinal hemorrhage
9. Intraocular inflammation
10. Endophthalmitis
11. Visible foreign matter within globe (anterior or posterior segment)
12. Fibrous encapsulation of IOFB (late)

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Plain film x-ray (especially if CT not available)
2. Computed tomography (CT) scan
3. Ultrasonography (caution if globe is ruptured)
4. Magnetic resonance imaging (MRI) contraindicated if metallic foreign body suspected

## **II. Define the risk factors**

- A. Occupational hazards**
- B. metal on metal hammering**
- C. Unsafe use of guns and other weapons**

## **III. List the most common or critical entities in the differential diagnosis**

- A. Ocular perforation (with or without IOFB)**
- B. Occult scleral rupture**
- C. Blunt trauma**
- D. Closed-globe injury with periocular foreign body**

## **IV. Describe patient management in terms of treatment and follow-up**

### **A. Prognosis**

- 1. Related to type and extent of injury
- 2. Related to location of impact
- 3. Presence/development of endophthalmitis or retinal detachment (RD)
- 4. Type/composition of projectile
- 5. Related to presenting vision and presence or absence of an APD

### **B. Surgical therapy**

- 1. Prompt removal of IOFB may reduce incidence of endophthalmitis
- 2. IOFB removed through sclerotomy or limbus depending on size
- 3. Antibiotics
  - a. Topical and systemic (intraocular for suspected or established endophthalmitis)
- 4. Tetanus prophylaxis (if required)

## **V. List the complications of treatment, their prevention and management**

### **A. Risk of RD and proliferative vitreoretinopathy (PVR)**

- 1. Ophthalmoscopy to detect and treat any retinal tear and detachment

### **B. Infection**

- 1. Intraocular antibiotics necessary for treatment of suspected or established endophthalmitis

## **VI. Describe disease-related complications**

- A. Siderosis (iron)
- B. Chalcosis (copper)
- C. Retinal tear and RD
- D. Endophthalmitis
- E. Cataract
- F. Glaucoma (angle trauma)

## **VII. Describe appropriate patient instructions**

- A. Usual post-vitreectomy instructions
- B. Safety glasses for any high risk activity

### **Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Focal Points: Open-Globe Injuries, Module #7, 2001.

# Endophthalmitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Bacteria or fungi are introduced into the eye
  - a. Post-traumatic
  - b. Post-operative
  - c. Bleb-associated
  - d. Endogenous (bloodborne)

### B. Define the relevant aspects of epidemiology of the disease

1. Postoperative
  - a. Acute
    - i. Gram-positive organisms most common
  - b. Chronic or delayed
    - i. Indolent bacteria or fungal infections
2. Post-traumatic
  - a. May be caused by bacteria from the patient's own flora or from material introduced into the eye from the site of injury
  - b. Bacillus is one of the most common and aggressive organisms
3. Bleb-related
  - a. Can occur with traumatic or surgical filtering blebs
4. Endogenous
  - a. Associated with systemic infection
  - b. Bacterial
  - c. Fungal

### C. List the pertinent elements of the history

1. Detailed history is critical for diagnosis
2. Duration of symptoms
3. Underlying medical condition
  - a. Diabetes mellitus
  - b. Immunocompromised
  - c. Systemic infection

- d. Recent minor procedure (colonoscopy, tattoo, etc)
- e. Intravenous drug abuse (IVDA)
- 4. Recent intraocular surgery or trauma (especially complicated surgery)
- 5. Filtering surgery (especially mitomycin C related)
- 6. Eyelid disease (e.g., severe blepharitis)

**D. Describe the pertinent clinical features**

- 1. Decreased vision
- 2. Pain & photophobia
- 3. Conjunctival injection and chemosis
- 4. Corneal and lid edema
- 5. Anterior chamber reaction (cells, flare, fibrin, hypopyon)
- 6. Floaters
- 7. Vitritis
- 8. Retinal hemorrhages
- 9. Periphlebitis
- 10. Blebitis (when bleb present)

**II. Define the risk factors**

- A. Ocular surgery
- B. Lid, lacrimal or conjunctival infection
- C. Systemic infection
- D. Soil or vegetable matter contaminated injuries
- E. Intraocular foreign body
- F. Immunocompromised host
- G. Filtering bleb

**III. List the most common or critical entities in the differential diagnosis**

- A. Uveitis (sterile, inflammatory)
- B. Lymphoma

**IV. Describe patient management: prevention, treatment and follow-up**

- A. Prophylactic antiseptic surgical preparation at time of surgery
- B. Acute endophthalmitis is an ocular emergency

**C. Prognosis determined by**

1. Type of organism
2. Setting of infection
3. Duration of symptoms
4. Timing of treatment
5. Other associated ocular findings

**D. Describe initial therapy**

1. Anterior chamber and vitreous tap for Gram stain & culture
2. Intravitreal antibiotics
3. Refer for possible early vitrectomy

**V. Describe disease-related complications**

- A. Vision loss**
- B. Vitreous opacification**
- C. Cystoid macular edema**
- D. Epiretinal membrane**
- E. Pupillary membrane**
- F. Corneal decompensation**
- G. Phthisis bulbi**
- H. Loss of eye**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Focal Points: Evaluation of Impaired Visual Acuity Following Cataract Surgery, Module #6, 1996



# Melanoma of the ciliary body or choroid

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. Some choroidal melanomas result from malignant transformation of an underlying nevus

### B. Define the relevant aspects of the epidemiology of the disease

1. Most common primary intraocular tumor in adults
2. Rare (incidence in the United States is 6 to 7 cases per million population)

### C. List the pertinent elements of the history

1. Visual symptoms including blurring, photopsias and visual field loss
2. Recent history of systemic symptoms such as weight loss, cachexia, malaise, jaundice or respiratory difficulties (all rare at presentation)

### D. Describe pertinent clinical features

1. Ciliary body melanoma
  - a. Asymptomatic initially
  - b. Sectoral cataract
  - c. Erosion through iris root, presenting as new iris mass
  - d. Lens displacement, induced astigmatism, angle closure glaucoma
  - e. Sentinel episcleral vessel
  - f. Episcleral pigmentation (trans-scleral extension)
2. Choroidal melanoma
  - a. Macula tumor (may be symptomatic)
  - b. Peripapillary tumor (may be symptomatic)
  - c. Peripheral tumors (usually asymptomatic and detected on routine indirect ophthalmoscopic examination, but may be symptomatic due to associated exudative retinal detachment)

### E. Describe appropriate testing and evaluation for establishing the diagnosis

1. A- and B-scan ultrasound
  - a. Evaluate for features characteristic of melanoma (i.e., dome or mushroom-shaped, low internal reflectivity)
  - b. Evaluate for other etiologies of choroidal mass lesion
  - c. Establish basal dimensions and thickness
  - d. Liver and chest evaluations to monitor for metastatic disease

## **II. List the most common or critical entities in the differential diagnosis**

- A. Iris lesion**
- B. Choroidal nevus**
- C. Melanocytoma**
- D. Metastatic carcinoma (most common non-primary intraocular tumor)**
- E. Atypical or eccentric disciform scar associated with age-related macular degeneration (AMD), hypertrophic subretinal fibrosis**
- F. Congenital hypertrophy of the retinal pigment epithelium (CHRPE)**
- G. Choroidal osteoma**
- H. Choroidal hemangioma**
- I. Suprachoroidal hemorrhage**

## **III. Describe the patient management in terms of treatment and follow-up**

### **A. Describe the natural history, outcome, and prognosis**

- 1. Untreated uveal melanoma will often eventually metastasize
- 2. Most common metastatic sites for uveal melanoma are liver and lung

### **B. Collaborative Ocular Melanoma Study (COMS)**

- 1. Defined tumor size classification (thickness and basal diameter)
  - a. Small
  - b. Medium
  - c. Large
- 2. Small tumors had a low 5 year mortality
- 3. Medium tumors treated with brachytherapy vs. enucleation showed no difference in patient survival
- 4. Large tumor outcomes did not benefit from pre-enucleation external beam radiation

### **C. Describe surgical therapy options**

- 1. Radiotherapy
  - a. Brachytherapy
  - b. Charged particle radiation (proton beam)
- 2. Eye wall resection
- 3. Enucleation
- 4. Transpupillary thermotherapy (TTT)

## **IV. List the complications of treatment, their prevention and management**

- A. Radiation retinopathy, papillopathy and neovascular glaucoma**
- B. Cataract**
- C. Panretinal photocoagulation may be effective in producing regression of neovascularization**
- D. Failure to achieve local tumor control**

## **V. Describe appropriate patient instructions**

- A. Patient should return periodically to retina specialist for assessment of post-treatment stability**
- B. Chest and liver assessment should be performed periodically to monitor for the development of metastatic disease**
- C. Despite excellent local tumor control, pre-existing micrometastasis can progress to gross metastatic disease**

### **Additional Resources**

1. AAO, Basic Clinical and Science Course. Section 12: Retina and Vitreous, 2013-2014.
2. AAO, Focal Points: Choroidal Melanoma Update: Collaborative Ocular Melanoma Study (COMS) Results, Module #4, 2005.

# Clinical approach to uveitis

## I. Describe the symptoms and signs of uveitis

### A. List of symptoms of uveitis

1. Blurred vision
2. Floaters
3. Photophobia
4. Pain
5. Epiphora
6. Redness

### B. List of signs of anterior uveitis

1. Perilimbal or diffuse conjunctival injection
2. Keratic precipitates
3. Anterior chamber flare and cells
4. Iris nodules may be seen in granulomatous anterior uveitis
5. Iris posterior synechiae and/or peripheral anterior synechiae
6. Hypotony or elevated intraocular pressure

### C. List of signs of intermediate uveitis

1. Vitreous inflammatory cells
2. Snowball opacities
3. Exudates over pars plana (snow banking)
4. Vitreous strands

### D. List of signs of posterior uveitis

1. Retinal or choroidal inflammatory infiltrates
2. Inflammatory sheathing of retinal vessels
3. Atrophy or swelling of the retina, choroid or optic nerve

### E. List of signs of panuveitis

1. Anterior, intermediate and posterior segments of uveal tract involved with no predominant site of inflammation (cells)
2. Combinations of signs from anterior, intermediate, and posterior uveitis

### F. List of symptoms and signs of scleritis

1. Severe pain radiating to the jaw or temple (can interfere with sleep because of severity)

- a. Pain is characteristic feature of scleritis and not episcleritis
- 2. Red violaceous hue due to deep scleral injection
  - a. Redness does not blanch with topical Neo-Synephrine like episcleritis or conjunctivitis
- 3. Edema to the episcleral and scleral tissues

**G. List of symptoms and signs of endophthalmitis**

- 1. Pain and visual loss (can be profound)
- 2. Diffuse conjunctival injection
- 3. Lid edema sometimes with ptosis of the upper lid
- 4. Hypopyon often with fibrin
- 5. Vitritis - often dense

## **II. Describe the pre-procedure evaluation**

**A. Comprehensive history and review of systems**

- 1. Medication history - particularly for drug induced uveitis
- 2. Personal history of autoimmune disorders
- 3. Prior ocular surgery
- 4. Travel and exposure history
- 5. Social history

**B. Determine status of tobacco use**

**C. Determine the onset of uveitis, whether sudden or insidious, chronic or acute, unilateral or bilateral, or isolated process or associated with systemic disease**

**D. Evaluate vision**

**E. Evaluate pupil function**

**F. Evaluate anterior segment**

**G. Evaluate intraocular pressure**

**H. Evaluate angle by gonioscopy in patients with elevated intraocular pressure**

**I. Dilate pupils and evaluate vitreous and posterior segment**

## **III. List the instruments and techniques**

**A. Slit-lamp biomicroscopic examination**

**B. Applanation tonometry**

**C. Gonioscopy**

**D. Examination of posterior pole with contact lens**

**E. Indirect ophthalmoscopy**

#### **IV. List the complications of this procedure, their prevention and management**

- A. Complications of pupillary dilation**

#### **V. Describe the considerations in interpretation for this diagnostic procedure**

- A. Look for conjunctival nodules/granuloma**
- B. Look for keratic precipitates, small (non-granulomatous uveitis) or large (granulomatous uveitis)**
- C. Look for the iris color, light color iris is seen in Fuchs heterochromic cyclitis**
- D. Look for cataract and iris synechiae**
- E. Look for structural changes in the anterior chamber angle (peripheral anterior synechiae, rubeosis)**
- F. Look for cells in the anterior chamber and vitreous, snow banking to determine anterior uveitis vs. intermediate uveitis/pars planitis**
- G. Look for posterior uveitis signs - chorioretinal lesions, perivasculitis or vasculitis of retinal vessels, swollen disc**
- H. Look for cystoid macular edema**

#### **VI. Describe the laboratory investigations**

- A. Obtain the investigations based on clinical features (a tailored approach) may include serology for syphilis, purified protein derivative (PPD) and chest x-ray**

#### **VII. Describe appropriate patient instructions**

- A. Standard dilation instructions**
- B. For patients who use tobacco, inform them that smoking increases the risk of uveitis and its complications**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
2. Lin P, Loh AR, Margolis TP, Acharya NR. Cigarette smoking as a risk factor for uveitis. *Ophthalmology*. 2010 Mar;117(3):585-90.

# Acute anterior uveitis

## I. Describe the approach to establishing the diagnosis

### A. List the pertinent elements of the history

1. Sudden onset
2. Pain, redness, photophobia
3. Sometimes decreased vision
4. Patterns
  - a. One episode or recurrent
  - b. Unilateral or bilateral
  - c. Same eye or alternating
5. Post-operative iridocyclitis
  - a. Acute post-operative
    - i. Redness, pain, photophobia
    - ii. May be due to too rapid taper of topical corticosteroids
  - b. Delayed post-operative iridocyclitis
    - i. Can have episodic redness, pain, and photophobia due to exacerbations
    - ii. Usually becomes persistent and unresponsive to topical corticosteroids
    - iii. Infectious cause - most commonly *Propionibacterium acnes*

### B. Describe the pertinent clinical features

1. Ciliary flush
2. Non-granulomatous or granulomatous, keratic precipitates
3. Anterior chamber flare and cells
4. With severe disease, hypopyon may be present
5. Occasionally retrolenticular/anterior vitreous cells (iridocyclitis)

## II. List the most common or critical entities in differential diagnosis

### A. Systemic diseases associated with human leukocyte antigen (HLA)-B27

### B. Sarcoidosis

### C. Infection

1. Viral

- a. Herpes simplex virus
  - b. Varicella zoster virus
- 2. Syphilis
- 3. Endophthalmitis
- D. Trauma**
- E. Intraocular foreign body**

### **III. Describe patient management in terms of treatment and follow-up**

#### **A. Describe medical therapy options**

- 1. Topical corticosteroid
- 2. Cycloplegia
- 3. Therapy for severe or recurrent attacks
  - a. Medical evaluation for recurrent episodes of uveitis
  - b. Periocular corticosteroid injection
  - c. Systemic corticosteroids
  - d. Immunomodulatory agent
- 4. Concomitant systemic or intraocular antibiotic therapy if infectious etiology

### **IV. List the complications of treatment, their prevention and management**

#### **A. Complications of topical corticosteroids**

- 1. Elevated intraocular pressure
- 2. Cataract

### **V. Describe disease-related complications**

- A. Posterior synechiae and pupillary seclusion**
- B. Peripheral anterior synechiae**
- C. Glaucoma**
- D. Cataract**
- E. Cystoid macular edema**

### **VI. Describe appropriate patient instructions**

#### **A. Medication instructions**



- B. Follow-up instructions**
- C. Natural history of the disease**
- D. Appropriate rheumatologic or expert consultation**

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
2. AAO, HIV: HIV/AIDS and the Eye: A Global Perspective, 2002, p.49-50.
3. AAO, Focal Points: Topical Therapy for Iritis, Module #7, 1991.
4. AAO, Focal Points: Diagnosis and Management of Anterior Uveitis, Module #1, 2002, p.6.

# Persistent (chronic) iridocyclitis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Often idiopathic
2. Associated with systemic disease (inflammatory diseases or masquerade syndromes such as lymphoma)
3. Uveitis associated with juvenile idiopathic (rheumatoid) arthritis

### B. List the pertinent elements of the history

1. Onset is usually insidious
2. Blurred vision
3. Floaters
4. May be asymptomatic
5. Pain, redness and photophobia are rare, although may occur at onset

### C. Describe pertinent clinical features

1. Keratic precipitates
2. Anterior chamber cell
3. Anterior chamber flare, may be significant, even with few cells
4. Anterior vitreous cells
5. Iris nodules

## II. Define the risk factors

### A. Systemic inflammatory disease - e.g. sarcoidosis

## III. Describe patient management in terms of treatment and follow-up

### A. Describe medical therapy options

1. Topical corticosteroid
2. Mydriatic/cycloplegic
3. Local injections
4. Systemic therapy
  - a. Oral corticosteroids
  - b. Immunomodulatory therapy

- c. Referral to uveitis specialist if immunomodulatory therapy considered
- 5. Internal medicine consultation for systemic disease, if appropriate, e.g., for pulmonary tests, chest X-ray

#### **IV. List the complications of treatment, their prevention and management**

- A. (See Corticosteroids)

#### **V. Describe disease related complications**

- A. Posterior synechiae are frequent
- B. Peripheral anterior synechia
- C. Ocular hypertension/glaucoma
- D. Hypotony
- E. Iris atrophy
- F. Cystoid macular edema - frequent cause of decreased vision
- G. Cataract is frequent, both because of underlying inflammatory disease and corticosteroid use
- H. Band keratopathy
  - 1. Much more common in children

#### **VI. Describe appropriate patient instructions**

- A. Nature of chronic disease, i.e., we cannot cure the disease, and patient may not be able to be tapered off of drops
- B. Necessity of using drops even in absence of symptoms
- C. Requirement for follow-up, even in absence of symptoms
  - 1. For example, may develop asymptomatic glaucoma secondary to corticosteroid drops

#### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.

# Juvenile idiopathic arthritis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease

1. More commonly associated with oligo-articular subtypes of juvenile idiopathic arthritis (JIA) (formerly known as juvenile rheumatoid arthritis)
2. Need to check antinuclear antibody (ANA) status

### B. List the pertinent elements of the history

1. Asymptomatic
2. Painless loss of vision
3. More common in females

### C. Describe the pertinent clinical features of JIA uveitis

1. Bilateral
2. Non-granulomatous iridocyclitis

## II. Describe patient management in terms of treatment and follow-up

### A. Mild disease

1. Topical corticosteroids
2. Mydriatics

### B. Rheumatology consultation for management of primary disease (JIA)

### C. Early referral to uveitis specialist if inflammation is not controlled

## III. List the complications of treatment, their prevention and management

### A. Topical corticosteroid therapy

1. Cataract
2. Glaucoma

### B. Periocular corticosteroid therapy

1. Cataract
2. Glaucoma
3. Inadvertent globe perforation

### C. Systemic corticosteroid and immunosuppressive therapy: (See Corticosteroids)

## **IV. Describe patient instructions**

### **A. The disease is painless and children may not exhibit any symptoms**

1. Regular follow-up for active disease is required
2. Regular screening for those at highest risk (oligoarticular disease , ANA-positive, girls) is required

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.

# Sympathetic ophthalmia

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Autoimmune response to intraocular antigens

### B. Define the relevant aspects of epidemiology of the disease

1. Uncommon
2. Bilateral, granulomatous, diffuse panuveitis
3. Occurs mostly after penetrating injury to one eye (the exciting eye)
4. Time to onset is variable: 2 weeks to many years following penetrating ocular trauma
  - a. Latent period followed by development of uveitis in both the exciting and uninjured globe (sympathizing eye)
  - b. Most cases occur in first three months

### C. List the pertinent elements of the history

1. Photophobia, bilateral redness and blurring of vision usually 10 days or more after injury to the opposite eye

### D. Describe pertinent clinical features

1. Panuveitis in the injured and opposite eye
2. Mutton fat keratic precipitates (KP)
3. Vitritis
4. Multifocal, yellow-white choroidal lesions
5. Peripapillary choroidal lesions
6. Exudative retinal detachment

## II. List the most common or critical issues in differential diagnosis

- A. Bilateral uveitis following any penetrating or non-penetrating ocular injury or surgery should suggest sympathetic ophthalmia (SO)
- B. Vogt-Koyanagi-Harada syndrome
- C. Sarcoidosis

## III. Describe patient management in terms of treatment and follow-up

- A. Early enucleation usually within first two weeks of trauma may prevent SO
- B. Once SO is established, enucleation most likely makes no difference

- C. With advances in treatment, an exciting eye may turn out to be the eye with the better visual acuity**
- D. Treatment**
  - 1. Topical corticosteroids and cycloplegic agents for anterior chamber inflammation
  - 2. Systemic corticosteroids for posterior segment inflammation
  - 3. Immunomodulation as extended therapy is anticipated in most patients
- E. Refer the patient to specialist for treatment**

#### **IV. Describe appropriate patient instructions**

- A. Follow-up care by a specialist**

#### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.

# Scleritis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Several systemic connective tissue, vasculitis, and infectious diseases may be associated with scleritis

### B. Define the relevant clinical symptoms

1. Most commonly present in fourth to sixth decade
2. Insidious onset moderate to severe penetrating ocular pain
  - a. Frequent radiation to jaw, forehead, sinuses
3. Photophobia, epiphora

### C. Describe relevant clinical signs and features

1. Redness
  - a. Sectoral (interpalpebral) or diffuse
2. Exquisite tenderness to palpation
  - a. Pain absent in scleromalacia perforans
3. Tortuous, congested vessels of the deep episcleral plexus
4. Violaceous appearance to sclera (bluish -red color)
  - a. Blanches incompletely with topical 10% phenylephrine (Neo-Synephrine®)
5. Nodules (non-mobile)
6. Avascular zones with sequestra, uveal show in cases of necrotizing scleritis

## II. Define the risk factors

### A. Systemic inflammatory diseases

1. Rheumatoid arthritis - most common

## III. Describe patient management

### A. Topical corticosteroids - for mild cases

### B. Oral non-steroidal anti-inflammatory agents (NSAIDs)- mild cases

### C. Oral corticosteroids

### D. Immunomodulatory therapy - for severe and necrotizing scleritis

1. Refer to uveitis specialist and rheumatologist



#### **IV. Complications of treatment**

- A. Usual complications from corticosteroid treatment (See Corticosteroids )**
- B. NSAIDs**
  - 1. Nephrotoxicity
  - 2. Peptic ulcers

#### **V. Disease related complications**

- A. Spontaneous perforations rare in necrotizing scleritis**
- B. Glaucoma - due to elevated episcleral venous pressure**

#### **Additional Resources**

- 1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.

# Herpes zoster (varicella-zoster virus) iritis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Associated with herpes zoster ophthalmicus (HZO) (varicella-zoster virus (VZV) involving the first branch of trigeminal nerve)

### B. List the pertinent elements of the history

1. Herpes zoster skin lesions (shingles)
2. Pain, redness, photophobia are common

### C. Describe pertinent clinical features

1. Corneal disease
2. Intraocular pressure (IOP) may be elevated
3. Corneal keratic precipitates (KP)

## II. Define the risk factors

### A. Increased risk with immunosuppression

1. Advanced age
2. Secondary to immunosuppressive agent
3. Leukemia, lymphoma and human immunodeficiency virus (HIV) infection

## III. List the most common or critical entities in the differential diagnosis

### A. Herpes simplex virus iritis

## IV. Describe patient management

### A. Topical corticosteroids

### B. Mydriatic agents (mild)

### C. Systemic antivirals

## V. Complications of therapy

### A. Systemic antivirals

1. Neutropenia
2. Renal toxicity

3. Hallucinations, seizures

## **VI. Disease related complications**

- A. Mydriatic pupil**
- B. Iris atrophy**
- C. Glaucoma**

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
2. AAO, HIV: HIV/AIDS and the Eye: A Global Perspective, 2002, p.46.
3. AAO, Focal Points: Diagnosis and Management of Anterior Uveitis, Module #1, 2002, p.2.

# Necrotizing herpetic retinitis: acute retinal necrosis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of this disease

1. Varicella zoster virus
  - a. Most common agent
2. Herpes simplex virus
3. Cytomegalovirus
  - a. Least common

### B. Describe pertinent clinical features

1. Symptoms
  - a. Decreased vision
  - b. Floaters
  - c. Pain may occur in patients with acute retinal necrosis
2. Signs
  - a. Peripheral retinal necrosis with discrete borders
  - b. Rapid progression in the absence of antiviral therapy
  - c. Circumferential spread
  - d. Cells and flare in anterior chamber and cells in vitreous, with or without optic disc swelling or hyperemia
  - e. In immunocompetent hosts - usually acute retinal necrosis
    - i. More inflammation in vitreous since host can mount an immune response
    - ii. Less rapid progression of retinitis
  - f. In Immunocompromised hosts (i.e. AIDS) - Progressive Outer Retinal Necrosis (PORN)
    - i. Less inflammation in vitreous (especially if low CD4 counts)
    - ii. More rapid progression

## II. Describe patient management in terms of treatment and follow-up

### A. Prompt referral to uveitis or retina specialist

### **III. List the complications of treatment, their prevention and management**

#### **A. Prognosis is good if**

1. Peripheral disease that is caught early and treated aggressively

#### **B. Prognosis is poor if**

1. Large areas of retinitis especially with macular involvement
2. Structural complications - retinal detachment

#### **C. Long term complications**

1. Retinal detachment risk is high (>50%)
2. High likelihood of involvement of fellow eye if antivirals not instituted

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.

# Toxoplasmic retinochoroiditis

## I. Describe the approach to establishing the diagnosis

### A. Describe the etiology of the disease - *Toxoplasma gondii*

1. Exists as inactive scars, latent cysts in retinal cells at borders of scars
2. Reactivated infection produces inflammatory reaction

### B. List the pertinent elements in the history

1. Exposure to undercooked meat from infected animals, exposure to uncooked vegetables grown in soil contaminated with excretions of infected animals
2. Maternal prenatal infection
3. Acquired disease may be more common than previously appreciated

### C. Describe pertinent clinical symptoms

1. Unilateral floaters or blurred vision

### D. Describe pertinent clinical features

1. Unilateral focal chorioretinitis adjacent to healed chorioretinal scar
2. Atypical forms of extensive chorioretinitis can occur in immunocompromised individuals, e.g., patients with acquired immunodeficiency syndrome (AIDS) or those over age 70 years
3. In acquired disease, the typical chorioretinal scar may not be present
4. Active chorioretinitis is yellow-white and slightly elevated
5. Intraocular inflammation
  - a. Iritis
  - b. Vitritis

## II. List the differential diagnosis

### A. Infections

1. Toxocariasis
2. Necrotizing herpetic retinitis
3. Syphilis

### B. Masquerade syndromes

1. Intraocular lymphoma

## III. Describe patient management in terms of treatment and follow-up

- A. Treatment dependent upon location of lesion in the eye**
- B. Medical treatment options include a variety of anti-protozoal agents**
- C. Oral corticosteroids may be used in combination with anti-toxoplasmic therapy**

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
2. AAO, Focal Points: Ocular toxoplasmosis: Clinical presentations, Diagnosis and Therapy, Module #10, 1999.

# Masquerade syndromes

Neoplastic masquerade syndromes: Primary central nervous system-intraocular lymphoma

## I. Describe the approach to establishing the diagnosis

### A. Define the relevant aspects of epidemiology of the disease

1. Lymphoma arising in the eye or associated with the primary central nervous system lymphoma (PCNSL)
  - a. Primary intraocular lymphoma is usually a diffuse large B cell lymphoma
2. Older patients - 6<sup>th</sup> and 7<sup>th</sup> decade
3. Incidence increasing

### B. List the pertinent elements of the history

1. Central nervous system (CNS) signs and symptoms
  - a. Confusion, weakness, deterioration in mental functions
2. Ocular history
  - a. Bilateral, asymmetric
  - b. Decreased vision and floaters
  - c. Eye can be first site of presentation of PCNSL

### C. Describe pertinent clinical features

1. Clumps and sheets of white cells in the vitreous
2. Multifocal subretinal and sub-pigment epithelial infiltrates

### D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Referral to uveitis or retinal specialist
  - a. Work-up including vitreous biopsy

## II. Define the risk factors

A. Age

B. Immunosuppression

C. AIDS

## III. List differential diagnosis

A. Sarcoidosis



- B. Syphilis**
- C. Tuberculosis**
- D. Necrotizing herpetic retinitis**
- E. Toxoplasmosis**
- F. Acute posterior multifocal placoid pigment epitheliopathy**

#### **IV. Describe patient management in terms of treatment and follow-up**

- A. Avoid corticosteroids**
  - 1. Cytolytic to lymphoma cells leading to an apparent response to treatment initially
  - 2. Corticosteroids also decrease the yield of vitreous biopsy
- B. Immediate referral to an oncologist for treatment and staging**
- C. Median survival rate of PCNSL is 3-6 months with supportive care alone**
  - 1. With treatment, longest median survival approaches 40 months

#### **V. Describe disease-related complications**

- A. Death**
- B. Visual impairment or blindness**

Non-Neoplastic Masquerade Syndromes: Retained intraocular foreign body

#### **I. Describe the approach to establishing the diagnosis**

- A. Describe the etiology of this disease**
  - 1. Penetrating ocular injury from high velocity projectile
  - 2. Foreign body irritates uveal tissues
    - a. Anterior chamber and vitreous inflammatory cells
      - i. Can be confused for autoimmune uveitis
    - b. Fibrous encapsulation can occur
    - c. Type of foreign body determines severity of reaction
      - i. Glass relatively inert but can cause inflammation
      - ii. Wood or organic matter can cause endophthalmitis rapidly
      - iii. Metal can cause uveal and ocular toxicity
        - i) Copper Chalcosis
        - ii) Iron Siderosis

**B. Define the relevant aspects of the epidemiology of this disease**

1. Men more commonly affected than women
2. Ages 18-25 highest risk

**C. List the pertinent elements of the history**

1. High velocity metal on metal injury
2. Often no safety glasses
3. Distant history may have been forgotten

**D. Describe pertinent clinical features**

1. Anterior chamber cells
2. Evidence of corneoscleral laceration may be occult
3. Foreign body may be in anterior chamber angle
4. Vitreous cells
5. Peripheral retinal exam foreign body may be in vitreous base

**E. Describe appropriate testing and evaluation for establishing the diagnosis**

1. Thorough history and physical examination
2. Gonioscopy may reveal retained foreign body in angle
3. B-mode ultrasonography or Ultrasound biomicroscopy
  - a. High reflectivity retained foreign body in peripheral vitreous
4. CT of orbits highly sensitive for intraocular foreign bodies

**II. Define the risk factors**

- A. No safety glasses**
- B. High velocity metal on metal injury**

**III. List the differential diagnosis**

- A. Traumatic iridocyclitis**
- B. Chronic infectious endophthalmitis**
- C. Chronic uveitic conditions autoimmune**

**IV. Disease related complications**

- A. Vitreous hemorrhage**
- B. Endophthalmitis especially with wood or organic foreign bodies**
- C. Retinal detachment**

## **V. Describe patient management in terms of treatment and follow-up**

### **A. Surgical removal of intraocular foreign body**

1. If foreign body in posterior segment vitreoretinal consultation

## **VI. Describe appropriate patient instructions**

### **A. Immediately report any sudden loss of vision associated with pain**

### **B. Wear safety goggles when performing high risk activities**

#### Additional Resources

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
2. AAO, Focal Points: Intraocular Lymphoma, Vol XXIII, Number 12, 2005.
3. Choi JY, Kafkala C, Foster CS. Primary intraocular lymphoma: A review. *Semin Ophthalmol* 2006;21:125-33.
4. Nussenblatt RB, Chan CC, Wilson WH and the CNS and Ocular Lymphoma Workshop Group. International Central Nervous System and Ocular Lymphoma Workshop; recommendations for the future. *Ocul Immunol Inflamm* 2006;14:139-44.
5. AAO, Focal Points: Intraocular Lymphoma, Module #12, 2005.

# Ocular manifestations of acquired immune deficiency syndrome

## I. Describe the approach to establishing the diagnosis

### A. Human immunodeficiency virus (HIV) retinopathy

1. Common in HIV-infected individuals and usually no symptoms
2. Cotton-wool spots, retinal hemorrhages and microaneurysms

### B. Cytomegalovirus (CMV) retinitis

1. Most common opportunistic ocular infection in acquired immunodeficiency syndrome (AIDS)
2. Slowly progressive perivascular whitening, hemorrhages and necrosis of the retina
3. CMV retinitis associated with a substantial risk of vision loss despite treatment
  - a. Causes of vision loss
    - i. Retinal detachment
    - ii. Retinitis involving optic nerve or macula (zone 1)
    - iii. Cataract
    - iv. Cystoid macular edema
4. Differential diagnosis
  - a. Acute retinal necrosis (ARN)
  - b. Progressive outer retinal necrosis caused by varicella-zoster virus (VZV) or herpes simplex virus (HSV)
  - c. Toxoplasmic retinochoroiditis

### C. Herpes zoster ophthalmicus (HZO)

1. Painful rash on face
2. Redness
3. Decreased vision
4. Cutaneous vesicular eruption along cranial nerve V1 (ophthalmic division)
5. Corneal epithelial changes
6. Anterior chamber cells and flare
7. Vitreous cells
8. Optic neuritis

## **II. Describe appropriate patient instructions**

### **A. Refer patient to a specialist immediately**

1. Infectious disease specialist for highly active anti-retroviral therapy (HAART) and other systemic therapy
2. Retinal or Uveitis specialist if ocular manifestations

### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
2. AAO, HIV/AIDS and the Eye: A Global Perspective, 2002, p.6-7, 26, 35-36, 62-63, 68-69, 97-87, 103-104.

# Cycloplegics in uveitis

## I. List the indications/contraindications

### A. Indications

1. Anterior chamber involvement in uveitis
  - a. To prevent/reduce formation of posterior synechiae
  - b. To decrease photophobia and pain due to ciliary and sphincter muscle spasm
  - c. To break recently formed posterior synechiae

### B. Contraindications

1. Allergy or insensitivity to agent or to others in its class
2. Occludable angles

## II. Describe the pre-procedure/therapy evaluation

- A. Question for history of allergy or adverse reaction
- B. Check angle depth because of possibility of inducing angle-closure glaucoma

## III. Describe the agents (agents listed in decreasing order of duration of effect)

- A. Atropine 1%
- B. Scopolamine 0.25%
- C. Homatropine 2% and 5%
- D. Cyclopentolate 0.5% and 1%

## IV. List the complications of the procedure/therapy, their prevention and management

- A. Tachycardia
- B. Fever
- C. Urinary retention
- D. Cycloplegia/blurred vision
  1. Can be minimized by use of a short-acting cycloplegic (not indicated if patient is at highest risk for synechiae)
  2. Temporary use of reading glasses (or temporary use of stronger reading glasses)
- E. Psychosis, acute psychotic reaction
  1. Limit dosage to no more than recommended frequency

2. Most case reports are in the pediatric age group, but has been reported in adults
3. Treat with supportive care

**F. Allergic reaction**

**V. Describe the follow-up care**

- A. Routine monitoring of the extent of posterior synechiae and anterior inflammation**

**VI. Describe appropriate patient instructions**

- A. Inform the physician of any new symptoms while on the medication**
- B. Use only as prescribed, not more frequently**
- C. Explain to patient rationale for use, including need to keep pupil moving to prevent synechiae**
- D. Recommend protection from bright light**
- E. Counsel use of reading glasses for near work (with bilateral use)**

**Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
2. AAO, Focal Points: Diagnosis and Management of Anterior Uveitis, Module #1, 2002, p.11.

# Non-steroidal anti-inflammatory drugs

## I. List the indications/contraindications

### A. Indications

1. Noninfectious ocular inflammatory disease, especially scleritis/episcleritis
  - a. Topical nonsteroid anti-inflammatory drugs (NSAIDs) may be of benefit in episcleritis only
2. May be useful as adjunct during tapering of topical corticosteroids
3. Analgesia

### B. Contraindications

1. Renal insufficiency or other kidney disease
2. Allergy or insensitivity to agent or others in its class
3. Peptic ulcer disease
4. Bleeding diathesis

## II. Describe the routes of administration

### A. Oral

### B. Topical

## III. List the complications of the procedure/therapy, their prevention and management

### A. Oral

1. Renal insufficiency
2. Gastritis/peptic ulcer
3. Nausea
4. Decreased clotting ability
5. Abnormal liver enzymes

### B. Topical

1. Corneal epithelial breakdown, thinning, erosion, ulceration
2. Ocular wound healing delay
3. Ocular bleeding
4. Conjunctival hyperemia



#### **IV. Describe appropriate patient instructions**

**A. Inform the physician of any new symptoms while on the medication**

1. Bleeding
2. Increased bruising
3. Changes in stool
4. Changes in urination

**B. Take with food**

**C. Do not use aspirin-containing products concurrently**

#### **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.

# Corticosteroids

## I. List the indications/contraindications

### A. Indications

1. Ocular, systemic, or neuro-ophthalmic inflammatory disease
2. In some situations, may be used cautiously in infections as adjunct agent

### B. Contraindications

1. Infectious etiology, unless covered by appropriate antimicrobial agents, e.g., Toxoplasma retinochoroiditis
2. Poorly controlled or difficult to control diabetes; systemic, but not topical corticosteroids
3. Necrotizing scleritis (for topical, periocular)

## II. List the alternatives to this procedure/therapy for systemic use

### A. Immunosuppressive agents

### B. Periocular, intraocular, topical corticosteroids (for isolated anterior disease)

## III. Describe the instrumentation and technique

### A. Describe the routes of administration

1. Oral
2. Intravenous
3. Intramuscular
4. Sub Tenon
5. Intraocular
  - a. Intravitreal
  - b. Injection of suspension
  - c. Polymer matrix
  - d. Sutured implant
6. Topical

## IV. List the complications of the procedure/therapy, their prevention and management

### A. For all forms

1. Posterior subcapsular cataracts (not reversible)

2. Increased intraocular pressure (often reversible)

**B. Systemic use**

1. Aseptic necrosis of femoral or humeral head
2. Sodium retention, fluid retention, potassium loss, congestive heart failure in susceptible patients, hypertension
3. Infection; reduced symptoms from infection
4. Menstrual irregularities, manifestations of latent diabetes mellitus; increased requirements for insulin or oral hypoglycemic agents in patients with diabetes
5. Increased intracranial pressure, convulsions, emotional disturbances
6. Weight gain
7. Sleep disturbance
8. Impaired wound healing, thin fragile skin, petechiae and ecchymoses
9. Peptic ulcer
10. Pathologic fracture of long bones, muscle weakness, vertebral compression fractures
11. Development of Cushingoid state, suppression of growth of children
12. Fat redistribution, adrenal suppression, accelerated atherosclerosis

**C. Regional use**

1. Unintended injection into the choroidal or retinal circulation or emboli
2. Perforation of the globe with retinal detachment, vitreous hemorrhage and/or permanent loss of vision
3. Ptosis (more common with superior injections)
4. Proptosis
5. Orbital fat atrophy, fibrosis
6. Orbital fat prolapse (with inferior retroseptal injections)
7. Subconjunctival hemorrhage
8. Pain from injection, syncope

**D. Intravitreal use**

1. Intraocular inflammation
  - a. Sterile, toxic-type reaction (with preserved formulations)
  - b. Infectious endophthalmitis
2. Vitreous hemorrhage
3. Retinal detachment

**E. Topical use**

1. Worsening of external infectious disease

2. Increased incidence and frequency of spontaneous subconjunctival hemorrhages

## **V. Describe the follow-up care**

### **A. Systemic use**

1. Long-term use should be minimized

### **B. Topical, regional, intravitreal**

1. Monitor intraocular pressure and cataract status

## **VI. Describe appropriate patient instructions**

### **A. Inform the physician of any new symptoms while on the medication**

### **B. Avoid exposure to infection**

### **C. Maintain physical activity and optimal body weight**

### **D. Persons with diabetes should monitor their blood glucose frequently and adjust treatment accordingly, in conjunction with their primary care physicians**

## **Additional Resources**

1. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
2. AAO, Focal Points: Diagnosis and Management of Anterior Uveitis, Module #1, 2002, p.9-11.

# Unique competence of the ophthalmologist

## **I. Describe professional responsibilities pertaining to the topic**

### **A. Provide background**

1. Comprehensive medical training in the art and science of medicine and ophthalmology
2. Continuing medical education
3. Knowledge of applicable scope of practice regulations

### **B. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics**

1. An ophthalmologist is a physician who is educated and trained to provide medical and surgical care of the eyes and related structures
2. An ophthalmologist must not delegate to an auxiliary those aspects of eye care within the unique competence of the ophthalmologist (which do not include those permitted by law to be performed by auxiliaries)

## **II. Discuss legal implications of noncompliance**

### **A. Regulatory enforcement (fraud and abuse)**

### **B. Civil litigation**

### **C. Antitrust litigation**

## **III. Describe the potential consequences of breach of professional responsibility**

### **A. Unnecessary risk to patients**

### **B. Unnecessary expense to patients and third party payers**

### **C. Loss of trust in physician and ophthalmologic profession**

## **IV. Describe potential corrective actions**

### **A. Formal continuing medical education**

### **B. Cessation of inappropriate delegation of services**

### **C. Correction of any misrepresentation of credentials or experience**

### **D. Education regarding scope of practice of relevant personnel**

## **Additional Resources**

1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))

2. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Delegated Services. <http://www.aao.org/about/ethics/delegated.cfm> (<http://www.aao.org/about/ethics/delegated.cfm>)
3. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Postoperative Care. [http://www.aao.org/about/ethics/upload/ao\\_post\\_op.pdf](http://www.aao.org/about/ethics/upload/ao_post_op.pdf) ([http://www.aao.org/about/ethics/upload/ao\\_post\\_op.pdf](http://www.aao.org/about/ethics/upload/ao_post_op.pdf))
4. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Employment and Referral Relationships between Ophthalmologists and Other Health Care Providers. <http://www.aao.org/about/ethics/upload/Employment-Referral-Relationships-2007.pdf> (<http://www.aao.org/about/ethics/upload/Employment-Referral-Relationships-2007.pdf>)
5. American Academy of Ophthalmology. Policy Statements of the Code of Ethics. An Ophthalmologist's Duties Concerning Postoperative Care. <http://www.aao.org/about/policy/upload/Ophthalmologists-Duties-re-Postoperative-Care-2012.pdf> (<http://www.aao.org/about/policy/upload/Ophthalmologists-Duties-re-Postoperative-Care-2012.pdf>)
6. American Academy of Ophthalmology. Policy Statements of the Code of Ethics. Pretreatment Assessment Responsibilities of the Ophthalmologist. [http://www.aao.org/about/policy/upload/PS\\_Pretreatment\\_Assessment.pdf](http://www.aao.org/about/policy/upload/PS_Pretreatment_Assessment.pdf) ([http://www.aao.org/about/policy/upload/PS\\_Pretreatment\\_Assessment.pdf](http://www.aao.org/about/policy/upload/PS_Pretreatment_Assessment.pdf))
7. American Academy of Ophthalmology. Policy Statements of the Code of Ethics. Joint Position Statement of AAO and ASCRS: Ophthalmic Postoperative Care. [http://www.aao.org/about/ethics/upload/joint\\_position\\_paper.pdf](http://www.aao.org/about/ethics/upload/joint_position_paper.pdf) ([http://www.aao.org/about/ethics/upload/joint\\_position\\_paper.pdf](http://www.aao.org/about/ethics/upload/joint_position_paper.pdf))

# Assimilating new technology

## **I. Describe professional responsibilities pertaining to the topic**

### **A. Provide background**

1. Assimilating new technology is essential to competent practice
2. Some techniques are easily assimilated with informal study
3. Other techniques require a comprehensive and systematic approach to master

### **B. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics**

1. Competence
  - a. As new technology is introduced, an ophthalmologist must become adequately competent to make it available to patients "by virtue of specific training or experience, or is assisted by one who is" incorporating the new technology into practice
    - i. This may include referral to or consultation with another physician

## **II. Discuss legal implications of noncompliance**

### **A. Risk of civil litigation (malpractice)**

### **B. Risk of State Board of Medicine action against license**

## **III. Describe the potential consequences of failure to appropriately assimilate new technology**

- A. Offering only outdated technology for lack of competence in newer, better methods is unethical**
- B. Offering new technology without achieving sufficient competence is unethical**
- C. If not competent to provide a technology, withholding a referral to another who is competent to provide it is unethical**
  1. "Consultations should be obtained if required by the condition"

## **IV. Describe potential proactive actions**

- A. Follow and evaluate relevant literature to learn of new technology as it becomes available**
- B. Seek reliable training in new technology, including formal study, courses, and use of mentor(s) as appropriate**
- C. Choose appropriate patient candidates for new treatment modalities**

Additional Resources

1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))
2. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Learning New Techniques After Residency. [http://www.aao.org/about/ethics/index\\_advisory.cfm](http://www.aao.org/about/ethics/index_advisory.cfm) ([http://www.aao.org/about/ethics/index\\_advisory.cfm](http://www.aao.org/about/ethics/index_advisory.cfm))



# The impaired ophthalmologist

## **I. Describe professional responsibilities pertaining to the topic**

### **A. Provide background**

1. Competence
2. Patient safety

### **B. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics**

1. It is the responsibility of an ophthalmologist to act in the best interest of the patient
2. An impaired ophthalmologist should withdraw from those aspects of practice affected by the impairment
3. It is the duty of other ophthalmologists who know of the impairment to take action appropriate to the specific setting

## **II. Discuss legal implications of noncompliance**

- A. Action by State Medical Board**
- B. National Practitioner Data Bank reporting**
- C. Civil litigation**
- D. Criminal litigation**

## **III. Describe the potential consequences of practicing with an impairment**

- A. Permanent patient injury**
- B. Report to local, state and national authorities**
- C. Loss of American Board of Ophthalmology certification**
- D. Violation of the American Academy of Ophthalmology Code of Ethics, with potential sanction**
- E. Loss of personal and professional respect**

## **IV. Describe potential corrective actions**

- A. Self reporting**
- B. Reporting others**
- C. Voluntary limitations on practice**
- D. Where can help be obtained**
  1. Colleagues
  2. Hospital

3. State medical organizations
4. Local societies

#### Additional Resources

1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))

# Providing appropriate care

## **I. Describe professional responsibilities pertaining to the topic**

### **A. Provide background**

1. Evaluation of patients' needs
2. Subjugation of physician's needs/interests/financial incentives
3. Misrepresentation of services

### **B. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics**

1. An ophthalmologist must not misrepresent the service that is performed or the charges made for that service
2. Ordering unnecessary procedures or materials for a patient, or withholding necessary procedures or materials is unethical
3. An ophthalmologist's clinical judgment and practice must not be affected by professionally related commercial enterprises

## **II. Discuss legal implications of noncompliance**

### **A. Regulatory enforcement (fraud and abuse)**

### **B. Civil litigation**

## **III. Describe the potential consequences of unethical patient care**

### **A. Unnecessary risk to patients**

### **B. Unnecessary expense**

### **C. Loss of trust in physician, profession**

## **IV. Describe potential corrective actions**

### **A. Avoid restrictive contractual obligations**

### **B. Divest of potentially conflicting commercial assets**

### **C. Self-educate in applicable statutes and regulations**

## **Additional Resources**

1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))
2. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Unnecessary

Surgery and Related Procedures. <http://www.aao.org/about/ethics/upload/Unnecessary-Surgery-2008.pdf> (<http://www.aao.org/about/ethics/upload/Unnecessary-Surgery-2008.pdf>)

3. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Appropriate Examination and Treatment Procedures. [http://www.aao.org/about/ethics/exam\\_procedures.cfm](http://www.aao.org/about/ethics/exam_procedures.cfm) ([http://www.aao.org/about/ethics/exam\\_procedures.cfm](http://www.aao.org/about/ethics/exam_procedures.cfm))
4. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Disclosures of Professionally Related Commercial Interests. <http://www.aao.org/about/ethics/upload/Disclosures-of-Commercial-Interest-2009.pdf> (<http://www.aao.org/about/ethics/upload/Disclosures-of-Commercial-Interest-2009.pdf>)
5. American Academy of Ophthalmology. Policy Statements of the Code of Ethics. Gifts to Physicians from Industry. <http://www.aao.org/about/policy/upload/Gifts-to-Physicians-from-Industry-2009.pdf> (<http://www.aao.org/about/policy/upload/Gifts-to-Physicians-from-Industry-2009.pdf>)

# Informed consent

## **I. Describe professional responsibilities pertaining to the topic**

### **A. Describe elements of informed consent process**

1. Respect for patients' autonomy
2. Patients' comprehension
3. Physician truthfulness
4. Absence of factors inappropriately influencing the practice of medicine
5. Discuss risks, benefits, complications and alternatives

### **B. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics**

1. Informed consent
  - a. "The performance of medical or surgical procedures shall be preceded by appropriate informed consent"

## **II. Discuss legal implications of noncompliance**

- A. Procedures performed without adequate informed consent may be legally interpreted as "battery"**
- B. Medical liability risk**

## **III. Describe the potential consequences of failure to obtain informed consent**

- A. Breach of patient trust**
- B. Loss of patient satisfaction**
- C. Unrealistic expectations of outcomes**
- D. Report(s) to local, state and National Practitioner Data Bank**

## **IV. Describe potential proactive actions**

- A. Education regarding ethical requirements**
- B. Education regarding legal requirements**
- C. Practical suggestions regarding approaches to informed consent and adequate documentation**

### **Additional Resources**

1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))

2. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Informed Consent. <http://www.aao.org/about/ethics/upload/Informed-Consent-2008.pdf> (<http://www.aao.org/about/ethics/upload/Informed-Consent-2008.pdf>)

# Confidentiality

## **I. Describe professional responsibilities**

- A. Respect for patient privacy**
- B. Professional tradition of confidentiality (Hippocratic Oath)**
- C. Knowledge of federal statute (Health Insurance Portability and Accountability Act (HIPAA) and Health Information Technology for Economic and Clinical Health Act (HITECH))**
- D. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics**
  - 1. Patient care must be provided with compassion, respect for human dignity, honesty and integrity

## **II. Describe the potential consequences of unethical patient care**

- A. Loss of patient trust**
- B. Loss of respect for the profession**
- C. Violation of governmental regulation or statute (with civil litigation possible)**
- D. Violation of professional organizations' ethical guidelines**

## **III. Describe potential proactive actions**

- A. Holding patient information in confidence**
- B. Secure handling of confidential patient documentation**
- C. Compliance with relevant federal regulations**

### **Additional Resources**

- 1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))
- 2. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Release and Confidentiality of Patient Records. [http://www.aao.org/about/ethics/patient\\_records.cfm](http://www.aao.org/about/ethics/patient_records.cfm) ([http://www.aao.org/about/ethics/patient\\_records.cfm](http://www.aao.org/about/ethics/patient_records.cfm))

# Clinical investigation

## I. Describe professional responsibilities pertaining to the topic

### A. Provide background

1. Clinical investigations occur in academic and private practice.
2. Knowledge of scientific method
  - a. Evaluation and use of existing literature
  - b. Commonly used study methods
  - c. Basics of evidence based medicine (EBM)
    - i. Double masked, placebo controlled studies provide most reliable evidence, when possible
    - ii. Other study designs can provide useful information, including pilot data for further study
3. Knowledge of review procedures
  - a. Investigational Review Boards
  - b. Informed consent for research
  - c. Ethics opinion as appropriate
4. Knowledge of editorial review and publication process

### B. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics

1. Informed consent
  - a. The performance of medical or surgical procedures shall be preceded by appropriate informed consent
2. Clinical trials and investigative procedures
  - a. Clinical trials and investigative procedures shall be approved by adequate review mechanisms
  - b. Appropriate informed consent for these procedures must recognize their special nature and ramifications
3. Medical and surgical procedures
  - a. An ophthalmologist must not misrepresent the service that is performed or the charges made for that service
4. Communications to the public (Advertising the merits of innovative therapies; soliciting research subjects, etc)
  - a. Communications to the public must be accurate
  - b. They must not convey false, untrue, deceptive, or misleading information through



statements, testimonials, photographs, graphics or other means

- c. They must not omit material information without which the communications would be deceptive
  - d. Communications must not appeal to an individual's anxiety in an excessive or unfair way; and they must not create unjustified expectations of results
  - e. If communications refer to benefits or other attributes of ophthalmic procedures that involve significant risks, realistic assessments of their safety and efficacy must also be included, as well as the availability of alternatives and, where necessary to avoid deception, descriptions and/or assessments of the benefits or other attributes of those alternatives
  - f. Communications must not misrepresent an ophthalmologist's credentials, training, experience or ability, and must not contain material claims of superiority that cannot be substantiated
5. Conflict of interest
- a. A conflict of interest exists when professional judgment concerning the well-being of the patient has a reasonable chance of being influenced by other interests of the provider
  - b. Disclosure of a conflict of interest is required in communications to patients, the public, and colleagues.

## **II. Discuss legal implications of noncompliance**

- A. Civil litigation**
- B. Violation of federal and/or state statutes or regulations**

## **III. Describe the potential consequences of unethical clinical investigation**

- A. Creation of flawed "junk" science**
- B. Loss of patient trust**
- C. Loss of respect for the profession**
- D. Bodily or financial harm to patients**

## **IV. Describe potential proactive actions**

- A. Rigorous adherence to consensus clinical research methods**
- B. Avoidance of conflicts of interest (academic, financial, personal)**
- C. Avoidance of misleading communications to colleagues and the public**

### **Additional Resources**

- 1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))

2. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Communications to the Public. 2003. [http://www.aao.org/about/ethics/index\\_advisory.cfm](http://www.aao.org/about/ethics/index_advisory.cfm) ([http://www.aao.org/about/ethics/index\\_advisory.cfm](http://www.aao.org/about/ethics/index_advisory.cfm))
3. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Informed Consent. <http://www.aao.org/about/ethics/upload/Informed-Consent-2008.pdf> (<http://www.aao.org/about/ethics/upload/Informed-Consent-2008.pdf>)
4. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Advertising Claims Containing Certain Potentially Misleading Phrases. [http://www.aao.org/about/ethics/misleading\\_phrases.cfm](http://www.aao.org/about/ethics/misleading_phrases.cfm) ([http://www.aao.org/about/ethics/misleading\\_phrases.cfm](http://www.aao.org/about/ethics/misleading_phrases.cfm))
5. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Disclosures of Professionally Related Commercial Interests. <http://www.aao.org/about/ethics/upload/Disclosures-of-Commercial-Interest-2009.pdf> (<http://www.aao.org/about/ethics/upload/Disclosures-of-Commercial-Interest-2009.pdf>)
6. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Clinical Trials and Investigative Procedures. <http://www.aao.org/about/ethics/clinical.cfm> (<http://www.aao.org/about/ethics/clinical.cfm>)

# Conflicts of interest

## **I. Describe professional responsibilities pertaining to the topic**

### **A. Provide background**

1. Truthfulness
2. Absence of factors inappropriately influencing the practice of medicine
3. Professional responsibility and restraint
4. Disclosure

### **B. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics**

1. Disclosure
  - a. "Disclosure of a conflict of interest is required in communications to patients, the public, and colleagues"
2. Commercial relationships
  - a. "An ophthalmologist's clinical judgment and practice must not be affected by economic interest in, commitment to, or benefit from professionally related commercial enterprises"
3. Informed consent
  - a. "The performance of medical or surgical procedures shall be preceded by appropriate informed consent"

## **II. Discuss legal implications of noncompliance**

### **A. Medical liability risk**

## **III. Describe the potential consequences of failure to disclose conflicts of interest**

- A. Breach of patient trust**
- B. Dishonest communication among colleagues**
- C. Biased and misleading scientific presentations**
- D. Loss of professional credibility**
- E. Impaired relationship with industry**
- F. Potential violation(s) of local, state and federal regulations**
- G. Rendering inappropriate care**
- H. Adverse effect on quality of care provided by colleague**

## **IV. Describe potential proactive (ethics) actions**

- A. Disclose conflicts of interest at appropriate times**
- B. Disclose professionally related commercial interests at appropriate times**
- C. Manage situations where the potential for a conflict of interest exists (e.g. avoidance, recusal)**

#### Additional Resources

1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))
2. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Informed Consent. <http://www.aao.org/about/ethics/upload/Informed-Consent-2008.pdf> (<http://www.aao.org/about/ethics/upload/Informed-Consent-2008.pdf>)
3. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Disclosures of Professionally Related Commercial Interests. <http://www.aao.org/about/ethics/upload/Disclosures-of-Commercial-Interest-2009.pdf> (<http://www.aao.org/about/ethics/upload/Disclosures-of-Commercial-Interest-2009.pdf>)
4. American Academy of Ophthalmology. Policy Statements of the Code of Ethics. Gifts to Physicians from Industry. <http://www.aao.org/about/policy/upload/Gifts-to-Physicians-from-Industry-2009.pdf> (<http://www.aao.org/about/policy/upload/Gifts-to-Physicians-from-Industry-2009.pdf>)

# Expert witness testimony

## **I. Describe professional responsibilities pertaining to the topic**

### **A. Provide background**

1. Valid medical license
2. Comprehensive medical training and true expertise in the topic
  - a. Be actively engaged as a clinical practitioner in the medical specialty that is the focus of the malpractice case
3. Knowledge of applicable ethical guidelines
4. Possess sufficient experience with the treatment modality in question to avoid doubt as to qualifications
5. Careful review of all relevant documents and facts
6. Willing to testify on the standard of care prevailing at the time of the occurrence in question, develop an opinion; have allegiance to facts

### **B. Describe applicable rule of the American Academy of Ophthalmology Code of Ethics**

1. Expert testimony should be provided in an objective manner using medical knowledge to form expert medical opinions
2. Nonmedical factors (such as solicitation of business from attorneys, competition with other physicians, and personal bias unrelated to professional expertise) should not bias testimony
3. It is unethical for a physician to accept compensation that is contingent upon the outcome of litigation
4. False, deceptive or misleading expert testimony is unethical

## **II. Describe the potential consequences of breach of professional responsibility**

- A. Loss of respect for the profession**
- B. Loss of credibility as an expert witness**
- C. Loss of professional society membership**
- D. Perjury**
- E. State Board of Medicine discipline (in some states)**

## **III. Describe potential corrective actions**

- A. Rigorous provision of unbiased expert testimony**
- B. Assurance of expertise in the matter**
- C. Should be willing to have expert testimony peer reviewed**

## Additional Resources

1. American Academy of Ophthalmology: Code of Ethics. [http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm) ([http://www.aao.org/about/ethics/code\\_ethics.cfm](http://www.aao.org/about/ethics/code_ethics.cfm))

# Patient safety

## **I. Providing safe anesthesia**

- A. Select patients by criteria including the American Society of Anesthesiologists (ASA) patient selection Physical Status Classification system**
- B. Have admitting privileges or emergency transfer agreement at a nearby hospital or a transfer agreement with another physician who has admitting privileges at a nearby hospital**
- C. At least one physician, who is credentialed or currently recognized as having successfully completed a course in advanced resuscitative techniques (Advanced Trauma Life Support, Advanced Cardiac Life Support, or Pediatric Advanced Life Support), must be present or immediately available with age- and size-appropriate resuscitative equipment until the patient has met the criteria for discharge**

## **II. Ensuring correct surgical site**

- A. Pre-operative verification process with all relevant documents, including informed consent**
- B. Marking the operative site**
- C. "Time out" immediately before starting the procedure for final verification**

## **III. Ensuring correct patient identification**

- A. Use at least two patient identifiers whenever taking blood samples or administering medications or blood products**
- B. Prior to the start of any surgical or invasive procedure, conduct a final verification process, such as a "time out", to confirm the correct patient, procedure and site, using active- not passive-communication techniques**

## **IV. Avoiding medication errors**

- A. Measures to minimize errors in communication of drug orders**
  - 1. Confusion over drugs with similar names**
    - a. Provide both the generic and brand name in communications of drug orders, if known**
    - b. Write the purpose for the medication on the prescription to help the pharmacist interpret the order accurately**
    - c. Provide patients with written information about their medications, including both brand and generic names**
  - 2. Writing of prescriptions**
    - a. Write in block letters, using upper-case (not cursive)**
    - b. Use the metric system (instead of the apothecary and avoirdupois systems)**

- c. Avoid use of abbreviations and Latin directions for use (e.g., q.d.), and instead write it out, e.g., daily
- d. Use a leading zero if a number is less than one (0.1), and don't use a trailing zero after a decimal (5.0)
- e. Prescriptions should include: date, drug name, dosage, route of administration, frequency of administration, and signature and professional designation of authorized prescriber
- 3. Labelling of syringes
  - a. Strongly advised in certain situations when multiple drugs are in use
- 4. Improve effectiveness of communication among caregivers
  - a. For verbal or telephone orders, verify the order by having the person receiving the information "read-back" the complete order

## V. Communicating patient safety issues

### A. Physician, nurses, and clinic staff should act in the best interests of the patient

### B. Judge the severity of any error

- 1. The patient or legal guardian or relative/person with power of attorney should be informed at the soonest possible time
- 2. The facility's risk management office and/or legal counsel should be consulted
- 3. Discussion, actions, and outcomes should be documented and copies sent to the appropriate risk management office and/or legal counsel, if appropriate or based on the facility's policies

### Additional Resources

- 1. American College of Surgeons. Statement on patient safety principles for office-based surgery utilizing moderate sedation/analgesia, deep sedation/analgesia, or general anesthesia. *Bulletin of the American College of Surgeons* 2004;89:32-34. Supported by the American Academy of Ophthalmology, American Society of Cataract and Refractive Surgery, national medical specialty societies, Accreditation Association for Ambulatory, American Association for Accreditation of Ambulatory Surgery Facilities, Committee for Quality Assurance
- 2. American Society of Anesthesiologists: ASA Physical Status Classification System. Available at <http://www.asahq.org/For-Members/Clinical-Information/ASA-Physical-Status-Classification-System.aspx> (<http://www.asahq.org/For-Members/Clinical-Information/ASA-Physical-Status-Classification-System.aspx>)
- 3. Joint Commission for Accreditation of Healthcare Organizations Universal Protocol for Preventing Wrong Site, Wrong Procedure Wrong Person Surgery™ [http://www.jointcommission.org/assets/1/18/UP\\_Poster1.PDF](http://www.jointcommission.org/assets/1/18/UP_Poster1.PDF) ([http://www.jointcommission.org/assets/1/18/UP\\_Poster1.PDF](http://www.jointcommission.org/assets/1/18/UP_Poster1.PDF)) Compliance required of all Joint Commission accredited organizations effective July 1, 2004.
- 4. Joint Commission on Accreditation for Healthcare Organizations 2010 Ambulatory Care/Office-Based Surgery Patient Safety Goals <http://www.jointcommission.org/assets>



/1/6/2013\_OBS\_NPSG\_final\_10-23.pdf ([http://www.jointcommission.org/assets/1/1/6/2013\\_OBS\\_NPSG\\_final\\_10-23.pdf](http://www.jointcommission.org/assets/1/1/6/2013_OBS_NPSG_final_10-23.pdf))

5. National Coordinating Council for Medication Error Reporting and Prevention. What is a Medication Error? (<http://www.nccmerp.org/index.htm>) <http://www.nccmerp.org/aboutmederrors.htm> (<http://www.nccmerp.org/aboutmederrors.htm>)
6. American Academy of Ophthalmology Patient Safety Bulletin. Minimizing Medication Errors: Communication about drug orders. <http://one.aao.org/patient-safety-statement/minimizing-medication-errors-communication-about-d> (<http://one.aao.org/patient-safety-statement/minimizing-medication-errors-communication-about-d>)
7. American Academy of Ophthalmology Patient Safety Bulletin. Eliminating Wrong Site Surgery. <http://one.aao.org/patient-safety-statement/recommendations-of-american-academy-ophthalmology-> (<http://one.aao.org/patient-safety-statement/recommendations-of-american-academy-ophthalmology->)
8. Joint Commission on Accreditation for Healthcare Organizations. The Official Do Not Use List. [http://www.jointcommission.org/assets/1/18/Do\\_Not\\_Use\\_List.pdf](http://www.jointcommission.org/assets/1/18/Do_Not_Use_List.pdf) ([http://www.jointcommission.org/assets/1/18/Do\\_Not\\_Use\\_List.pdf](http://www.jointcommission.org/assets/1/18/Do_Not_Use_List.pdf))

# Risk management

Disclaimer: This information is intended solely to provide risk management recommendations. It is not intended to constitute legal advice and should not be relied upon as a source for legal advice. If legal advice is desired or needed, an attorney should be consulted

## I. Patient complaints

### A. How the ophthalmologist and the office staff respond to patient complaints may determine whether the patient continues under the physician's care or even decides to pursue legal action

1. Recommendations may include
  - a. Active listening can improve patient-doctor communication (avoid distractions, nod head and ask clarifying questions to indicate interest and attempt at understanding, restate what the patient is saying to confirm understanding)
  - b. Explain the situation (for example, why there is an inequality in lid height after ptosis surgery, or why the doctor is 30 minutes late for a patient's appointment)
  - c. Physician calmness, absence of defensiveness and honesty can help defuse patient unhappiness
  - d. Sometimes, patient complaints can generate impetus to make positive changes to office or physician practice; this is an essential part of feedback loop
  - e. Try not to argue over who is right, but what can be done to make it right
  - f. Recognize that not all complaints can be resolved satisfactorily for all parties involved

## II. Retention of medical records

### A. Retention guidelines

1. Have a written retention and destruction policy in place, and train employees in its use, including legal requirements
2. Consider any material related to patient care to be part of the record (such as billing records, appointment schedules)
3. Other laws and regulations may apply (e.g. Medicare, Occupational Safety and Health Administration (OSHA), managed care organizations)
4. Duties with regard to copying or transferring medical records

### B. Destruction of medical records

1. The physician must approve any decision to purge or to destroy records
2. Keep a list of destroyed records that includes: the patient's name and birthday, the date of the last visit, and the date the records were destroyed

3. The method of destruction must protect the confidentiality of this information, such as shredding or burning

### **III. Informed consent**

#### **A. Obtaining informed consent**

1. In general, the ophthalmologist performing the procedure or surgery must obtain informed consent for it
  - a. The duty to obtain informed consent cannot be delegated in its entirety; however, portions may be delegated to staff
2. Informed consent is an agreement between the ophthalmologist and the patient reached after a discussion
  - a. The discussion includes the condition, recommended treatment or procedure, and the risks, complications, benefits, and alternatives, and observation
  - b. The discussion should include the consequences of refusing the recommended treatment or procedure
3. If the patient has any known risk factors that increase the likelihood of complications, side effects, or poor outcome, the ophthalmologist should discuss these with the patient
4. The informed consent discussion should take place when the patient is awake and aware, free from the effects of any medication that could interfere with the patient's ability to participate in the decision-making process
  - a. For elective surgeries, it should take place before the day of the surgery whenever possible

#### **B. Documentation of the informed consent**

1. The ophthalmologist should document the discussion in the patient's medical record and, if used, on the informed consent document itself
2. If the discussion took place in a language other than English, document the language used, and the name and relationship of any translator
3. Specific questions or concerns raised by the patient should be documented, along with the answers
4. In addition to documenting that the discussion took place, the patient may be asked to sign a consent form
5. Document any materials the patient was given or shown explaining the procedure or treatment

#### **C. Verification of informed consent: the role of the consent form**

1. An informed consent form serves to document and to verify that the informed consent discussion between the ophthalmologist and the patient took place
  - a. It is not a substitute for the informed consent discussion
2. To promote patient understanding and to document the content of the discussion, a procedure-specific consent form is recommended

3. Once the informed consent discussion between the ophthalmologist and the patient has taken place, any designated member of the ophthalmologist's staff can give the patient the form and witness the patient's signature
4. As a risk management measure, ask patients to confirm what procedure will be done and why before asking them to sign the form
5. Keep the original document in the patient's record and give patients a copy of the form, and encourage them to review it at home with their family members, and to call if they have any questions

## **IV. Remote screening of ophthalmic problems**

### **A. Guidelines**

1. Exercise the same care when treating a patient over the telephone or by other electronic means as during an office visit
  - a. To promote both the continuity and defensibility of care
    - i. Gather the information necessary to assess the situation and determine the treatment plan
    - ii. Communicate the assessment and plan to the patient, and
    - iii. Document the encounter and your decision-making process in the medical record
  - b. Safely enlist the office staff's assistance in gathering information by developing and implementing written protocols for telephone screening and treatment that are specific to the patient population, subspecialty, and staff
2. Document all patient-care related calls and communiqués in the patient's medical record
3. After-hours and on-call telephone contacts with patients or other caregivers need to be carefully screened, handled, and documented

### **B. Handling telephone requests for prescription refills**

1. Physicians have a duty to monitor the continued need, effectiveness, and safety of the drugs they prescribe
2. Many states have "good faith prior examination" laws that stipulate the maximum time drugs can be prescribed without another examination
3. Respect scope of practice laws
  - a. Only healthcare providers with the legal authority to order drugs can authorize prescription refills

### **C. E-mail correspondence with patients**

1. When using e-mail, physicians hold the same ethical responsibilities to their patients as they do during other types of encounters
2. Physicians should engage in e-mail communication with notification of e-mail's inherent limitations
3. E-mail should not be used to establish a patient-physician relationship, but rather supplement

other, more personal encounters

## **V. Terminating the physician-patient relationship**

### **A. The physician-patient relationship and allegations of abandonment**

1. Patient abandonment occurs when a physician fails to provide for necessary medical care to a current patient without adequate justification
2. In general, once a physician-patient relationship is established, a physician has an ongoing responsibility to the patient until the relationship is terminated
3. In order to terminate the relationship, the physician must notify the patient sufficiently in advance for the patient to secure the services of another physician
  - a. Prior to terminating the relationship
    - i. Check the provisions of any contract you have signed with the patient's health plan
    - ii. Resolve acute medical conditions
    - iii. Verify that other sources of care are available in your community

### **B. Notify the patient that the relationship will be terminated**

1. Give the patient written notice
2. Give the patient sufficient time to locate another physician
3. You are not required to give a reason for terminating the relationship
4. Inform the patient that upon receipt of a written authorization, you will provide a copy of the medical records to the new treating physician

### **C. Providing care during the notice period**

1. Continue to treat the patient during the notice period
  - a. Do not schedule or perform elective procedures or surgeries
  - b. Remind the patient of the termination date each time care is provided during the notice period
2. Once the notice period has ended, do not provide any care

### **D. Patients seen while on-call to the hospital emergency department**

1. Once a physician has agreed to provide on-call coverage, he/she must comply with state and federal (Emergency Medical Treatment and Active Labor Act (EMTALA)) emergency care and transfer laws
2. Physicians who accept on-call responsibilities need to clarify with the Emergency Department and with the Medical Staff Office whether or not they will provide post-discharge care

#### **Additional Resources**

1. Ophthalmic Mutual Insurance Company. Retention and Destruction of Medical Records.

<http://www.omic.com/retention-and-destruction-of-medical-records/> (<http://www.omic.com/retention-and-destruction-of-medical-records/>)

2. Ophthalmic Mutual Insurance Company. Obtaining and Verifying Informed Consent.  
<http://www.omic.com/informed-consent-obtaining-and-verifying/> (<http://www.omic.com/informed-consent-obtaining-and-verifying/>)
3. Ophthalmic Mutual Insurance Company. Telephone Screenings of Ophthalmic Problems.  
<http://www.omic.com/telephone-screening-of-ophthalmic-problems-sample-contact-forms-and-screening-guideline/> (<http://www.omic.com/telephone-screening-of-ophthalmic-problems-sample-contact-forms-and-screening-guideline/>)
4. American Medical Association Guidelines for Physician-Patient Electronic Communications  
<http://www.ama-assn.org/ama/pub/physician-resources/medical-ethics/code-medical-ethics/opinion5026.page> (<http://www.ama-assn.org/ama/pub/physician-resources/medical-ethics/code-medical-ethics/opinion5026.page>)
5. eRisk Working Group for Healthcare's Guidelines for Online Communication. October 2006  
[http://www.aao.org/education/upload/2006\\_eRisk\\_Guidelines\\_Final\\_2-2.doc](http://www.aao.org/education/upload/2006_eRisk_Guidelines_Final_2-2.doc) ([http://www.aao.org/education/upload/2006\\_eRisk\\_Guidelines\\_Final\\_2-2.doc](http://www.aao.org/education/upload/2006_eRisk_Guidelines_Final_2-2.doc))
6. Ophthalmic Mutual Insurance Company. Terminating the Physician-Patient Relationship.  
<http://www.omic.com/terminating-the-physician-patient-relationship/> (<http://www.omic.com/terminating-the-physician-patient-relationship/>)