

PRACTICING OPHTHALMOLOGISTS
CURRICULUM 2014-2016

Oculoplastics/ Orbit

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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.

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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

Oculoplastics/Orbit

Orbital Anatomy

1. Orbital roof and orbital apex (superior orbital fissure and optic canal)	13
2. Medial wall of orbit	16
3. Floor of orbit	16
4. Nasolacrimal canal	20
5. Optic canal	22
6. Intraorbital optic nerve	24
7. Extraocular muscles	27
8. Intranasal anatomy	33

Evaluation of Orbital Disorders

9. Patient history and examination for orbital pathology	35
10. Computed tomography	47
11. Magnetic resonance imaging	52

Infectious/Inflammatory Disorders

12. Preseptal cellulitis	56
13. Orbital cellulitis	60
14. Thyroid eye disease	67
15. Idiopathic orbital inflammatory syndrome (nonspecific orbital inflammation, orbital pseudotumor)	73
16. Sarcoidosis	78
17. Giant cell arteritis	82
18. Wegener granulomatosis	85
19. Oculoplastic manifestations of AIDS	90

Periorbital Cysts and Orbital Neoplasms

20. Dermoid cysts.....	96
21. Infantile hemangioma (capillary hemangioma).....	98
22. Cavernous hemangioma	101
23. Rhabdomyosarcoma	104
24. Lymphoid hyperplasia and lymphoma.....	107
25. Pleomorphic adenoma (benign mixed tumor) of the lacrimal gland.....	111
26. Adenoid cystic carcinoma of the lacrimal gland	113
27. Optic nerve sheath meningioma.....	116
28. Sphenoid wing meningioma.....	119
29. Systemic corticosteroid therapy for orbital disorders	122
30. Surgical approaches to orbital tumors.....	125

Orbital Trauma

31. Zygomatic fractures	132
32. Medial orbital wall fractures.....	135
33. Orbital floor fractures	139
34. Orbital roof fracture.....	143
35. Orbital hemorrhage	147
36. Dural cavernous fistula and traumatic carotid cavernous fistula	150

Management of Socket and Fornices

37. Enucleation	154
38. Evisceration.....	160
39. Exenteration.....	165
40. Orbital implant options.....	169
41. Socket contracture.....	173

42. Congenital socket malformations	175
-------------------------------------------	-----

Periocular Soft Tissues Anatomy

43. Facial anatomy	178
44. Eyelid skin and subcutaneous tissue.....	193
45. Protractors	194
46. Orbital septum	196
47. Orbital fat.....	197
48. Eyelid retractors.....	199
49. Tarsus	201
50. Conjunctiva	202
51. Suborbicularis fat pads.....	203
52. Eyelid margin.....	204
53. Eyelashes.....	205
54. Meibomian glands.....	206
55. Nerve supply of eyelids.....	207

Eyelid Disorders

56. Chalazion.....	209
57. Blepharitis	212
58. Epidermal inclusion cysts.....	216
59. Actinic keratosis	218
60. Nevus	221
61. Lentigo maligna (Hutchinson melanotic freckle)	224
62. Basal cell carcinoma of the eyelid.....	226
63. Squamous cell carcinoma of the eyelid	230
64. Sebaceous adenocarcinoma of the eyelid	233

65. Malignant melanoma of the eyelid.....	236
66. Masquerading syndrome for eyelid tumors	239
67. Mohs micrographic surgery for malignant eyelid tumors	241
68. Blunt trauma to the eyelids	244
69. Penetrating trauma to the eyelids.....	247
70. Eyelid defects not involving the eyelid margin	250
71. Concepts of upper eyelid reconstruction	253
72. Concepts of lower eyelid reconstruction.....	257
73. Lateral canthal defects.....	261
74. Medial canthal defects.....	264
Periocular Malpositions/Involution	
75. Involutional ectropion.....	267
76. Paralytic ectropion	271
77. Cicatricial ectropion	274
78. Facial nerve palsy	278
79. Full-thickness block resection and repair	283
80. Tarsorrhaphy.....	285
81. Involutional entropion	288
82. Cicatricial upper and lower eyelid entropion	291
83. Trichiasis.....	296
84. Congenital myogenic ptosis	299
85. Acquired myogenic ptosis.....	305
86. Aponeurotic ptosis.....	308
87. Oculomotor (cranial nerve III) palsy.....	311
88. Horner syndrome	315

89. Myasthenia gravis with ptosis.....	318
90. Pseudoptosis	321
91. Floppy eyelid syndrome	324
92. Anterior lamellar deficiency.....	329
93. Middle lamellar deficiency.....	333
94. Posterior lamellar deficiency	336
95. Benign essential blepharospasm	339
96. Hemifacial spasm	343
97. Dermatochalasis.....	346
98. Upper eyelid blepharoplasty.....	349
99. Lower eyelid blepharoplasty.....	353
100. Brow ptosis	358
101. Involutional and gravitational changes of the face.....	361
102. Botulinum toxin use in oculoplastics	371

Lacrimal System-Anatomy

103. Main lacrimal gland and lacrimal ductules	378
104. Superior and inferior puncta and canaliculi.....	381
105. Medial canthal tendon.....	382
106. Nasolacrimal duct (NLD) and Valve of Hasner	383

Disorders of the Nasolacrimal System

107. Congenital nasolacrimal obstruction	384
108. Acquired nasolacrimal duct obstruction in adults	388
109. Diagnostic probing (palpation) of the upper system	391
110. Nasal endoscopy	393
111. Evaluation of reflex tearing.....	395

112. Punctal ectropion.....	398
113. Punctal stenosis and atresia	401
114. Canalicular obstruction	404
115. Canaliculitis	406
116. Lacrimal outflow system trauma	409
117. Dacryocystitis	413
118. Endonasal dacryocystorhinostomy for acquired nasolacrimal duct obstruction	415
119. External dacryocystorhinostomy for acquired nasolacrimal duct obstruction	419
120. Silicone intubation of the nasolacrimal drainage system	423

Orbital roof and orbital apex (superior orbital fissure and optic canal)

I. Describe relevant aspects of the anatomy

A. Orbital roof

1. Osteology
 - a. Frontal bone
 - b. Variable contributions of sphenoid bone
2. Clinical correlation
 - a. Separates anterior cranial fossa from orbit
 - b. Defects in the orbital roof can cause pulsatile proptosis

B. Superior orbital fissure

1. Osteology
 - a. Bounded medially by lesser wing of sphenoid
 - b. Bounded laterally by greater wing of sphenoid bone
2. Clinical correlation
 - a. Transmits cranial nerves
 - i. Lacrimal nerve (CN V 1)
 - ii. Frontal nerve (CN V 1)
 - iii. Trochlear nerve (CN IV)
 - iv. Superior division CN III
 - v. Nasociliary nerve (CN V 1)
 - vi. Abducens nerve (CN VI)
 - vii. Inferior division (CN III)
 - b. Venous drainage from orbit passes through this fissure through superior ophthalmic vein to cavernous sinus

C. Optic Canal

1. Transmits optic nerve, ophthalmic artery
2. Osteology: orbital bones
 - a. Lesser wing of sphenoid
3. Landmarks

- a. Canal lies inside annulus of Zinn
- b. Roof of canal is floor of anterior cranial fossa
- c. Medial wall of canal is the lateral wall of the sphenoid sinus
- d. Dimensions
 - i. 6 mm diameter and 8mm length
- e. Optic nerve surrounded by periorbita
- f. Pathway
 - i. Entrance in the superior and medial orbital apex
 - ii. Optic nerve leaves orbit via optic canal to middle cranial fossa
 - iii. Canal course is medial and superior as it travels posteriorly towards chiasm
- g. Optic strut of lesser wing of sphenoid
 - i. Forms lateral wall of the canal
 - ii. Guards carotid artery behind it
- h. Anterior and posterior ethmoid foramina lie at the level of the cribriform plate and form a line leading to the optic canal

II. Describe clinical correlations-orbital roof and apex

A. Site of optic nerve compression

- 1. Trauma
 - a. Hemorrhage
 - b. Optic canal/apex fractures
 - c. Traumatic optic neuropathy
- 2. Tumors
- 3. Inflammatory processes
- 4. Infective processes

B. Often site of ophthalmoplegia

- 1. Trauma
- 2. Tumors
- 3. Inflammatory processes
- 4. Infective processes

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 9. Surgical Anatomy of the Ocular Adnexa, 1996.
3. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit, and Lacrimal System, Volume 1, 1993.

Medial wall of orbit

I. Describe relevant aspects of the anatomy

A. Osteology - orbital bones

1. Lamina papyracea of ethmoid
2. Lacrimal bone
3. Frontal process of superior maxilla
4. Sphenoid bone

B. Landmarks

1. Frontoethmoid suture
 - a. Border of medial wall and roof
 - b. Site of anterior and posterior ethmoidal arteries and veins
 - c. Cribriform plate superior and medial to frontoethmoid suture line
 - d. Clinical landmark to avoid cerebrospinal fluid (CSF) leak
2. Lamina papyracea
 - a. Thinnest of orbital bones
 - b. Weak point, medial wall blowout fracture occurs
3. Nasolacrimal fossa
 - a. Located in the anterior inferomedial orbital wall
 - b. Comprised of lacrimal bone and frontal process of superior maxilla. Superior orbital rim extends inferiorly to become posterior lacrimal crest. Inferior rim extends superiorly to become anterior lacrimal crest
 - c. Bounded by anterior and posterior lacrimal crest. Attachment sites of anterior and posterior limbs of medial canthal tendon
 - d. Nasolacrimal sac lies within nasolacrimal fossa
 - e. Nasolacrimal canal runs from lacrimal sac fossa to the inferior meatus below the inferior turbinate (See Nasolacrimal canal)
4. Ethmoid Sinus Variants that influence the medial wall and lacrimal fossa
 - a. Haller cells: ethmoid air cells that extend to the medial roof of the maxillary sinus
 - b. Agar Nasi: anterior ethmoid air cells
5. Trochlea: cartilaginous structure redirecting superior oblique muscle tendon located at the anterior junction of the orbital medial wall and roof

II. Describe the clinical correlation

- A. Adjacent to ethmoid, sphenoid sinus and nasal cavity medially**
- B. Sinusitis can spread contiguously into medial orbit as a subperiosteal abscess and cause orbital cellulitis.**
- C. Sinus tumors, and frontal sinus mucocoeles can spread contiguously to invade medial wall**
- D. Medial wall fracture can result in air in the orbit (emphysema)**
 - 1. There is continuity with ethmoid sinus
 - 2. Traumatic telecanthus can occur from lateral displacement of medial wall
- E. Telecanthus can also be a soft tissue widening of the medial canthal angles where there is no underlying bony abnormality. In contrast, hypertelorism is a wider than normal separation of the medial orbital walls**
- F. Medial wall orbital decompression is a common approach for thyroid orbitopathy. Deep dissection to adequately relieve pressure on the optic nerve.**
- G. Orbital tumors rarely arise directly from the orbital bones**
 - 1. Fibrous dysplasia may cause orbital dystopia
 - 2. Secondary orbital tumors from the brain or sinuses may extend into the orbital bones and cause secondary orbital signs and symptoms
- H. Approach to medial wall**
 - 1. Transnasal
 - 2. Transconjunctival
 - 3. Transcaruncular
 - 4. Transcutaneous (Lynch incision)
- I. Medial orbital wall can be breached during endoscopic sinus surgery**
- J. Bony dehiscence can occur naturally or pathologically, examples include clefts or encephaloceles**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
- 2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.33, 38-40.

Floor of orbit

I. Describe relevant aspects of the anatomy

A. Osteology - orbital bones

1. Palatine
2. Zygomatic
3. Superior maxilla

B. Landmarks

1. Forms roof of maxillary sinus
2. Infraorbital nerve
 - a. Maxillary division of infraorbital nerve
 - b. Travels within/just below orbital floor along with infraorbital vessels
 - c. Runs within infraorbital groove and exits anteriorly at the infraorbital foramen
 - d. Injury in anterior orbital/rim fractures result in numbness of cheek
 - e. Anterior superior alveolar branch can be injured in posterior floor fracture causing numbness of teeth
 - f. Emissary vessel off the floor to the inferior muscle complex is often cauterized during orbitotomy
3. Inferior orbital fissure
 - a. Separates floor from lateral orbital wall
 - b. Divides maxillary sinus and greater wing of sphenoid
 - c. Transmits cranial nerve (CN) V2, zygomatic nerve, inferior ophthalmic vein and pterygoid plexus
4. Inferior oblique muscle (See Extraocular muscles)
 - a. Attaches on periosteum of orbital floor lateral to nasolacrimal duct or adjacent to the posterior lacrimal crest
 - b. Injury can cause diplopia
5. Nasolacrimal duct
 - a. Anterior medial corner of orbital floor
 - b. Travels inferiorly from the nasolacrimal fossa. (See Nasolacrimal canal)

II. Describe clinical correlations

A. Contiguous spread of maxillary sinusitis or tumors can occur through the thin orbital floor

B. Orbital floor fracture

1. Posterior-medial maxillary bone most common sight of orbital "blowout" fractures
 - a. Thin bone
2. Signs and symptoms of orbital floor fracture
 - a. Enophthalmos
 - b. Diplopia
 - c. Restriction of extraocular muscle (EOM)
 - d. Fluid in maxillary sinus
 - e. Paresthesia of cheek, lip, teeth
 - f. Dystopia

C. Orbital decompression of floor common approach for treatment of thyroid orbitopathy

1. "Strut" separating medial wall and floor is often preserved during decompression to decrease the incidence of postoperative diplopia

D. Surgical approaches to orbital floor

1. Transcutaneous
2. Subciliary
3. Transconjunctival with lateral canthoplasty
4. Transantral

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Nasolacrimal canal

I. Describe relevant aspects of the anatomy

A. Bony canal which contains the nasolacrimal duct

1. Located at anterior medial orbital floor
2. Nasolacrimal fossa formed by maxillary bone forming the anterior lacrimal crest, and the lacrimal bone, forming the posterior lacrimal crest

B. Canal (2 cm) travels inferiorly and posteriorly in lateral wall of nose

1. Nasolacrimal duct (NLD) travels within the canal
2. NLD is 12mm in length in adults
3. Known to be narrower in women

C. Empties into inferior meatus under the inferior turbinate

1. Valve of Hasner
 - a. Distal valve as duct opens into inferior meatus
 - b. Site of congenital NLD obstruction in infants

II. Describe clinical correlates

A. Anatomic or functional obstruction of nasolacrimal system

1. Infectious
 - a. Dacryocystitis
 - b. Orbital cellulitis
 - c. Abscess
2. Inflammatory
 - a. Allergic rhinitis
 - b. Chronic sinusitis
 - c. Wegener granulomatosis
3. Tumors
 - a. Intranasal tumor
 - b. Lymphoma
 - c. Inverting papilloma
 - d. Tumors of the lacrimal sac
4. Chronic inhalation cocaine can destroy the NLD, leading to obstruction and symptomatic epiphora

and dacryocystitis

5. Other: poor lacrimal pump function (cranial nerve (CN) VII paralysis, age), trauma
6. Obstruction can be characterized by
 - a. Epiphora mucous discharge
 - b. Tender erythematous mass inferior to the medial canthal tendon (acute dacryocystitis)
 - c. Mucous collection in non inflamed distended sac (chronic dacryocystitis)
 - d. Non inflamed mass (mucocele)
 - e. Nasal speculum exam may show absence of nasal septum and inferior turbinate
 - f. Nasolacrimal irrigation with reflux of saline or blood (tumor)
 - g. Deviated nasal septum
 - h. History of trauma (i.e. broken nose) , previous sinus surgery
 - i. High doses of I131

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.39, 45.

Optic canal

I. Describe relevant aspects of the anatomy

A. Transmits optic nerve, ophthalmic artery

B. Osteology: orbital bones

1. Lesser wing of sphenoid

C. Landmarks

1. Entrance to the optic canal lies inside annulus of Zinn
2. Roof of canal is floor of anterior cranial fossa
3. Medial wall of canal is the lateral wall of the sphenoid sinus
4. Dimensions
 - a. 6 mm diameter and 8mm length
5. Optic nerve surrounded by periorbita
6. Pathway
 - a. Entrance in the superior and medial orbital apex
 - b. Optic nerve leaves orbit via optic canal to middle cranial fossa
 - c. Canal course is medial and superior as it travels posterior towards chiasm

D. Optic strut of lesser wing of sphenoid

1. Forms lateral wall of the canal
2. Guards carotid artery behind it

II. Describe clinical correlates

A. Site of optic nerve compression from trauma

1. Hemorrhage
2. Optic canal/apex fractures
3. Traumatic optic neuropathy
4. Tumors
5. Optic nerve glioma
6. Optic nerve sheath meningioma
7. Schwannoma
8. Lymphoma

9. Intracranial tumor spread (sphenoid wing meningioma, intracranial meningioma, glioma)
10. Fibrous dysplasia: narrowing of optic canal

B. Adjacent Inflammatory processes

1. Cavernous sinus thrombosis
2. Tolosa Hunt syndrome
3. Wegener granulomatosis

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.40, 42.

Intraorbital optic nerve

I. Describe the relevant aspects of the anatomy

A. Dimensions

1. Approximately 30 mm length from orbital apex to the globe
2. Approximately 4 mm diameter
3. Traverses 2 cm distance from optic canal to posterior globe

B. Central nervous system white matter tract

1. Axons originate from retinal ganglion cell layer with a myelin coat
2. Surrounded by pia, arachnoid and dura mater

C. S-Shaped configuration of the normal optic nerve is present to allow mobility of the eye

1. Extreme proptosis causes globe to move forward and tether the globe by the optic nerve
2. On imaging, characterized as a straightened optic nerve, causing tenting of posterior globe

D. Blood supply

1. Pial branches of ophthalmic artery along entire intraorbital course
2. Central retinal artery
 - a. Enters nerve posterior to globe on ventral surface
 - b. Supplies central fibers

II. Describe clinical correlations

A. Site of optic nerve compression from trauma

1. Orbital hemorrhage
2. Optic canal/apex fracture

B. Tumors

1. Optic nerve glioma
 - a. Most common type: juvenile pilocystic astrocytoma (spindle shaped)
 - b. Associated with neurofibromatosis
2. Optic nerve sheath meningioma
 - a. Slow growing tumor arises from arachnoid cells
 - b. Meningoendothelial cell proliferation with sparse psammoma bodies
 - c. Optociliary shunt vessels sometimes seen
3. Schwannoma

- a. Neurilemoma from Schwann cells
- 4. Lymphoma
 - a. B-cell type most common
- 5. Vascular lesions
 - a. Varix
 - b. Cavernous hemangioma
 - c. Hemangiopericytoma
 - d. Lymphangioma- can occur in intraconal space adjacent to optic nerve.
- 6. Retinoblastoma
 - a. Most common primary intraocular malignancy in children
 - b. Extension into the choroids or optic nerve gives a worse prognosis
 - c. Inheritance
 - i. 94% sporadic
 - ii. 6% autosomal dominant

C. Inflammatory processes

- 1. Optic neuritis (ON)
 - a. Enhancement on magnetic resonance imaging (MRI), T-1 weighted with fat suppression
- 2. Thyroid orbitopathy
 - a. Enlarged extraocular muscles at apex can cause optic nerve compression
 - b. An increase in orbital volume may push the eye forward stretching the optic nerve
- 3. Wegener granulomatosis
 - a. Necrotizing granulomatous vasculitis
- 4. Orbital cellulitis
- 5. Idiopathic intracranial hypertension (Pseudotumor cerebri)
 - a. Signs and symptoms
 - b. Signs and symptoms of increased intracranial pressure (headaches, nausea, vomiting, transient visual obscuration, papilledema)
 - c. Awake and alert patients
 - d. No localizing neurologic signs other than unilateral or bilateral abducens nerve paresis
 - e. Documented increased cerebrospinal fluid (CSF) pressure (>200 mm of H₂O in non-obese and >250 mm of H₂O in obese) with normal CSF cytology and chemistry
 - f. Normal neuroimaging studies except for small ventricles or empty sella
 - g. No other cause of intracranial hypertension present

- h. Treatment:
 - i. Weight loss
 - ii. Acetazolamide
 - iii. Optic nerve sheath decompression
 - i. Shunt
- 6. Orbital pseudotumor
 - 7. Sarcoidosis
 - a. Non caseating granuloma
 - b. Autoimmune disorder

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
- 2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.66.
- 3. AAO, Focal Points: Management of Orbital Trauma, Module #7, 1994, p.11.

Extraocular muscles

I. Describe relevant aspects of the anatomy

A. Rectus muscles

1. Striated muscles
2. Motor nerves travel through intraconal orbit and innervate rectus muscles at posterior 1/3 of muscle belly
3. Superior rectus
 - a. Origin
 - i. Annulus of Zinn
 - b. Insertion
 - i. 7.7 mm from limbus
 - c. Innervation
 - i. Superior branch of Cranial Nerve (CN) III
 - d. Actions
 - i. Primary: elevation
 - ii. Secondary: intorsion
4. Inferior rectus
 - a. Origin
 - i. Annulus of Zinn
 - b. Insertion
 - i. 6.5 mm from limbus
 - c. Innervation
 - i. Inferior branch of CN III
 - d. Actions
 - i. Primary: depression
 - ii. Secondary: extorsion
5. Lateral rectus
 - a. Origin
 - i. Annulus of Zinn
 - b. Insertion
 - i. 6.9 mm from limbus

- c. Innervation
 - i. CN VI
- d. Action
 - i. Abduction
- 6. Medial rectus
 - a. Origin
 - i. Annulus of Zinn Muscle travels anteriorly, tendon travels through trochlea posterior to rim
 - b. Insertion
 - i. 5.5 mm from limbus
 - c. Innervation
 - i. Inferior branch of CN III
 - d. Action
 - i. Adduction
 - e. Can be injured during functional endoscopic ethmoid or intranasal sinus surgery

B. Oblique muscles

- 1. Superior oblique
 - a. Origin
 - i. Orbital apex above annulus of Zinn
 - ii. Muscle travels anteriorly
 - iii. Tendon travels through trochlea posterior to rim
 - b. Insertion
 - i. Posterior to equator in superotemporal quadrant, ventral to the superior rectus muscle
 - c. Innervation
 - i. CN IV
 - ii. Extraconal
 - iii. Longest cranial nerve
 - d. Action
 - i. Primary: intorsion
 - ii. Secondary: depression
 - e. Can be traumatized during osteoplastic ablation of the frontal sinus
- 2. Inferior oblique

- a. Origin
 - i. Orbital floor lateral to lacrimal sac
- b. Insertion
 - i. Overlying the macula, situated ventral to the inferior rectus
- c. Innervation
 - i. Inferior branch of CN III
- d. Primary action
 - i. Extortion

C. Upper eyelid muscles

1. Levator palpebrae superioris
 - a. Origin
 - i. Lesser wing of sphenoid above annulus of Zinn
 - b. Insertion
 - i. Anterior surface of tarsus and septa of pretarsal orbicularis
 - c. Innervation
 - i. Superior branch of CN III
 - d. Action
 - i. Upper eyelid elevation
 - e. Muscle length approximately 40mm, aponeurosis length approximately 15mm
2. Müller muscle
 - a. Origin
 - i. Under surface of levator muscle
 - b. Insertion
 - i. Anterior-superior border of tarsus
 - c. Innervation
 - i. Sympathetic nerve
 - d. Action
 - i. Approximately 2 mm of eyelid elevation
 - e. Smooth retractor muscle

D. Lower eyelid retractors

1. Capsulopalpebral fascia-analogous to levator muscle in upper eyelid
 - a. Origin

- i. Capsulopalpebral head and at inferior rectus muscle
 - b. Encircles inferior oblique
 - c. Insertion
 - i. Inferior tarsal border and some fibers in fornix
 - d. Action
 - i. Retraction
2. Inferior tarsal muscle
- a. Smooth muscle
 - b. Equivalent of Müller muscle
 - c. Origin
 - i. Posterior to capsulopalpebral fascia
 - d. Insertion
 - i. Inferior fornix
 - e. Innervation
 - i. Sympathetic
 - f. Action
 - i. Lower eyelid retractor

E. Vascular supply

- 1. Extraocular muscles (EOM)
 - a. Arterial supply
 - i. Muscular branches of ophthalmic artery, lacrimal artery and infraorbital artery
 - b. Venous system
 - i. EOMs drain into superior and inferior orbital veins
 - c. Lymphatic drainage
 - i. Superior and inferior ophthalmic veins into cavernous sinus
- 2. Eyelids
 - a. Arterial supply
 - i. Internal carotid artery to ophthalmic artery to supraorbital and lacrimal branches
 - ii. External carotid artery to angular and temporal branches
 - iii. Extensive collateral circulation
 - b. Venous drainage
 - i. Pretarsal: angular vein medially and superficial temporal vein laterally

- ii. Post tarsal: orbital veins and deeper anterior facial veins into pterygoid plexus
- c. Lymphatic drainage
 - i. Medially: submandibular lymph nodes
 - ii. Laterally: superficial preauricular nodes and deeper cervical chain

F. Eyelid sensory innervation

- 1. CN V1- supraorbital nerve
 - a. Forehead and lateral periocular region
- 2. CN V2- maxillary branch
 - a. Lower eyelid and cheek, leaving orbit via inferior orbital fissure, into brain via foramen rotundum

II. Describe the clinical correlation

A. Tumors

- 1. Lymphoma
- 2. Metastasis
 - a. For example, breast, lung, prostate, leukemia
 - i. Consider checking PSA
 - b. Associated bony destruction often

B. Inflammatory

- 1. Thyroid orbitopathy
 - a. Cellular infiltrate of mononuclear cells, lymphocytes, plasma cells, mast cells, and fibroblasts and mucopolysaccharides
 - b. Tendons not involved
- 2. Orbital myositis
 - a. Idiopathic orbital inflammation
 - b. Polymorphous inflammatory response in muscle and tendon (neutrophils, plasma cells, lymphocytes, macrophages,)

C. Neuromuscular disorders

- 1. Chronic progressive external ophthalmoplegia (CPEO)
 - a. Mitochondrial disorder
 - b. Ragged red fibers on muscle biopsy
 - c. Associated with Kearns-Sayre syndrome (retinal pigment changes, cardiac conduction defects, CPEO)
- 2. Myasthenia gravis

- a. Acetylcholine receptor antibodies

D. CN palsy

1. Hypertension, diabetes mellitus (DM), cerebrovascular accident (CVA) causing a CN III or CN VI palsy resulting in ptosis, or diplopia
2. CN IV susceptible in intracranial injury
 - a. Results in superior oblique (SO) palsy, diplopia (3 step test)

E. Direct trauma to muscle and orbital septae can result in fibrosis, restriction

1. Inferior rectus muscle entrapment seen in orbital floor fracture
2. Lost muscle during strabismus surgery or sinus surgery
3. Positive forced duction testing
 - a. Restriction and diplopia on upgaze
4. Understand the relationship between EOM and the intricate network of fibrous septae

F. EOM enlargement secondary to carotid-cavernous sinus fistula

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 5: Neuro-Ophthalmology; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Intranasal anatomy

I. Describe relevant aspects of the anatomy

A. Nasal cavity

1. Medial nasal wall
 - a. Bisects nasal cavity
2. Nasal septum
 - a. Anterior cartilaginous midline structure
3. Vomer
 - a. Posterior bony vertical plate of ethmoid plate

B. Lateral nasal wall

1. Turbinates/meatus
 - a. Horizontal ridges with corresponding meatus below each
 - b. Superior turbinate/meatus
 - c. Middle turbinate/meatus
 - i. Site of osteotomy for dacryocystorhinostomy (DCR) at lacrimal fossae
 - ii. Arises from roof of nose at the cribriform plate posteriorly
 - iii. Nasolacrimal sac at anterior tip
 - iv. Hiatus semilunaris- superior to middle turbinate-ostium of maxillary sinus
 - v. Meatus receives drainage from ethmoids, frontal and maxillary sinuses
 - vi. Uncinate process-curved ridge within middle meatus
 - d. Inferior turbinate/meatus
 - i. Site of ostium of nasolacrimal duct
 - ii. Nasolacrimal duct (NLD) drains into inferior meatus
2. Uncinate Process
 - a. Entry into ethmoid sinus
 - i. Anterior located ethmoid air cell can be found medial to the nasolacrimal fossa
 - b. Reside just posterior to the nasolacrimal fossa

C. Cribriform plate

1. Roof of nose/ posterior middle turbinate
2. At level of frontoethmoid suture

II. Describe the functions of the nose

- A. Mucosalized surface, hair**
- B. Filters, warms, moistens air, smell**
- C. Collects secretions from the sinuses and NLD**

III. Describe clinical correlations

A. DCR

1. Osteotomy site at middle meatus removing frontal process of maxilla and lacrimal bone
2. Nasolacrimal obstruction may be secondary to intranasal pathology (See Acquired nasolacrimal duct obstruction in adults)
3. Intranasal surgery
 - a. Functional endoscopic sinus surgery
 - i. Iatrogenic traumatic entry into the orbit
 - b. Septo-rhinoplasty
4. Trauma
 - a. Naso-orbito-ethmoid fracture
5. Sinusitis
6. Sinus, nasal neoplasms
 - a. Polyps
 - b. Malignancy
7. Congenital
 - a. Obstruction of Valve of Hasner
8. Severe septal deviation
9. Inflammatory disorders
 - a. Wegener granulomatosis
10. Inhalation cocaine abuse

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.39, 45.
3. Watkins LM, Janfaza P, Rubin PA. The evolution of endonasal dacryocystorhinostomy. *Surv Ophthalmol* 2003;48:73-84. Comment in: *Surv Ophthalmol* 2004;49:543; author reply 544.

Patient history and examination for orbital pathology

I. The diagnostic procedure uses the standard history and physical examination process

A. History

1. Chief complaint
2. History of present illness (HPI)
3. Past medical history (PMH)
4. Current medications/allergies
5. Family history
6. Review of systems (ROS)

B. Physical examination

1. Visual acuity, uncorrected and corrected
2. Pupils
3. Extraocular muscle motility
4. External examination
5. Visual fields
6. Slit-lamp biomicroscope examination
7. Intraocular pressure
8. Fundus

C. Differential diagnosis

D. Diagnostic laboratory tests and imaging (See Magnetic resonance imaging and Computed tomography)

E. Biopsy

F. Medical or surgical treatment

II. The 6 P's of the orbital history and physical examination are useful in the diagnostic process

A. Pain

B. Proptosis

C. Progression

D. Palpation

- E. Pulsation
- F. Periocular Changes

III. History

A. Chief complaint

1. Decreased vision
2. Double vision
3. Pain/anesthesia/paresthesia
 - a. Pain is typically associated with infection or inflammation
 - b. Pain or paresthesia may be associated with specific tumor types
 - c. Inflammatory disorders
 - i. Idiopathic orbital inflammation
 - i) Pain with extraocular muscle movement
 - ii) Inflammatory response in muscle and tendon
 - ii. Tolosa Hunt syndrome
 - i) "Painful ophthalmoplegia"
 - ii) Inflammation surrounding the orbital apex
 - iii. Wegener granulomatosis
 - i) Chronic subacute pain
 - ii) Necrotizing granulomatous vasculitis
 - iv. Optic neuritis
 - i) Pain with EOM movements, marked decrease in visual acuity
 - v. Ruptured dermoid
 - i) Acute or chronic pain associated with inflammation
 - d. Infection
 - i. Orbital abscess, cellulitis, cavernous sinus thrombosis
 - i) Usually systemically ill with malaise, fever and leukocytosis
 - ii. Herpes zoster - varicella zoster virus
 - i) History of chicken pox
 - ii) Pain prodrome along ophthalmic branch of CN V
 - e. Orbital hemorrhage
 - f. Soft tissue injury

- g. Orbital fracture
- h. Associated facial and systemic injury
- i. Vascular lesion
 - i. Lymphangiomas
 - i) Pain associated with hemorrhage
- j. Malignancy
 - i. Adenoid cystic carcinoma of the lacrimal gland causing pain or paresthesia of several months duration
 - ii. Metastatic carcinoma can be associated with rapid onset of inflammatory conditions
 - iii. Rhabdomyosarcoma may mimic an inflammatory or infectious condition, but generally does not cause pain
 - iv. Squamous cell carcinoma
 - i) Pain or numbness at forehead associated with supraorbital nerve involvement

B. History of present illness (HPI)

- 1. Age and sex of patient
- 2. Onset, quality and duration of symptoms
 - a. Immediate: minutes to hours
 - i. Varix (upon Valsalva)
 - i) Worse with head down position
 - ii. Hemorrhage
 - iii. Lymphangioma
 - i) Exacerbated by upper respiratory infection
 - b. Rapid: (days to weeks)
 - i. Children
 - i) Capillary hemangioma
 - ii) Rhabdomyosarcoma
 - iii) Retinoblastoma
 - iv) Neuroblastoma
 - v) Leukemia
 - ii. Inflammatory disease
 - i) Idiopathic orbital inflammatory disease
 - ii) Thyroid eye disease (thyroid orbitopathy)
 - iii) Recurrent inflamed dermoid

- iii. Infection
 - i) Orbital cellulitis
 - ii) Abscess
 - iii) Cavernous sinus thrombosis
- iv. Trauma
 - i) Post surgical hemorrhage
 - ii) Orbital hemorrhage
- v. Malignancy
 - i) Rhabdomyosarcoma
 - ii) Metastatic tumors
 - iii) Adenoid cystic carcinoma
- vi. Vascular
 - i) Carotid-cavernous (C-C) fistula
 - ii) Lymphangioma
- c. Months to years
 - i. Dermoid cysts
 - ii. Benign mixed tumors
 - iii. Neurogenic tumors
 - iv. Cavernous hemangioma
 - v. Lymphoma
 - vi. C-C fistula
- d. Causes for exacerbation or improvement
- e. Associated changes in vision
 - i. Gaze-evoked amaurosis
- f. Pain and progression

C. Past medical history (PMH)

- 1. General medical diseases, diabetes, hypertension, thyroid abnormalities, cancers, trauma
- 2. Eye diseases

D. Current medications/allergies

- 1. Anticoagulant management pre-, peri- and post-operatively

E. Family history

- 1. Systemic diseases

2. Craniofacial anomalies
3. Genetic diseases

F. Review of systems (ROS)

1. Neurological
2. Cardiovascular
3. Pulmonary
4. Renal
5. Connective tissue

IV. Physical examination

A. General medical examination

B. Eye examination

1. Visual acuity, uncorrected and corrected
 - a. Induced hyperopia
2. Pupils
 - a. Afferent pupillary defect
 - b. Optic Neuropathy
 - i. Compression
 - ii. Ischemia
3. Extraocular muscle motility
 - a. Ophthalmoplegia
 - i. Forced duction testing performed to distinguish restrictive vs. paretic type
 - ii. Causes
 - i) Strabismus
 - ii) Noncomitant motility disturbance
 - (i) Orbital apex syndrome
 - (ii) Paralysis of cranial nerves (CN), III, IV, and VI
 - (iii) Injury
 - (iv) Tumor
 - (v) Orbital fracture
 - (vi) Hemorrhage
4. External examination

- a. Exophthalmometry
 - i. Normal distance between apex of cornea and lateral orbital rim less than 20 mm
 - ii. Identify asymmetry between eyes. A difference of 2 mm between eyes is abnormal
 - iii. Normal Hertel measurements are higher for African Americans and lower for Asian Americans
- b. Type of proptosis
 - i. Axial displacement
 - i) Retrobulbar intraconal mass
 - (i) Tumors
 - (ii) Symmetric enlargement of extraocular muscles in thyroid eye disease
 - (iii) Hemorrhage
 - ii. Nonaxial displacement: extraconal lesions
 - i) Superior displacement
 - (i) Maxillary sinus tumor
 - (ii) Hemorrhage
 - ii) Inferomedial displacement
 - (i) Dermoid cyst
 - (ii) Lacrimal gland tumor
 - (iii) Lymphoma
 - (iv) Cholesterol granuloma
 - iii) Inferolateral displacement
 - (i) Frontoethmoidal mucocoeles
 - (ii) Encephalocele
 - (iii) Frontal sinus abscess
 - (iv) Osteoma
 - (v) Frontal sinus carcinoma
 - iv) Lateral displacement
 - (i) Subperiosteal abscess or tumor associated with ethmoid sinus
 - v) Unilateral
 - (i) Most commonly thyroid eye disease
 - (ii) Common presentation for most orbital neoplasms
 - (iii) Abscess, hemorrhage, inflammation, or an autoimmune lesion
 - vi) Bilateral

- (i) Most commonly associated with thyroid eye disease
 - (ii) May be seen in pseudotumor, metastatic tumors, carotid-cavernous (C-C) fistula, C-C thrombosis, or systemic conditions like leukemia, neuroblastoma, or Wegener granulomatosis
- iii. Enophthalmos
 - i) Secondary to sclerosing tumors, metastatic breast carcinoma, orbital trauma/fractures, silent sinus syndrome, silent brain syndrome (hydrocephalus)
- iv. Displacement with Valsalva
 - i) Suspect varix
- v. Mastication proptosis
 - i) Dermoid cyst protrudes into temporalis fossa
- c. Pseudoproptosis
 - i. Large globe (axial myopia)
 - ii. Facial asymmetry
 - iii. Contralateral enophthalmos
 - iv. Microphthalmos
 - v. Eyelid retraction
- d. Clinical correlation of proptosis
 - i. Thyroid eye disease is the most common cause of unilateral or bilateral proptosis
 - ii. Primary orbital neoplasms are usually unilateral
 - iii. Bilateral proptosis seen in inflammatory conditions, immune processes, or systemic diseases
 - iv. Inflammatory disorders
 - i) Thyroid disease
 - (i) Most common cause of proptosis
 - (ii) Tendons not involved
 - ii) Orbital pseudotumor
 - iii) Wegener granulomatosis
 - v. Infection (orbital abscess, cellulitis)
 - vi. Vascular
 - i) Orbital hemorrhage
 - ii) Lymphangioma (sudden)
 - iii) C-C fistula
 - iv) Orbital varices-proptosis with Valsalva

- vii. Tumor
 - i) Benign
 - (i) Cavernous hemangioma
 - (ii) Lymphangioma
 - ii) Malignant
 - (i) Adenoid cystic carcinoma
 - (ii) Lymphoma
 - (iii) Glioma
 - iii) Contiguous
 - (i) Sinus
 - (ii) Intracranial nasopharynx
 - (iii) Skin
 - iv) Systemic
 - (i) Lymphoma
 - (ii) Leukemia
 - v) Metastatic
 - (i) Breast (may cause enophthalmos)
 - (ii) Lung
 - (iii) Prostate
 - (iv) Neuroblastoma
 - vi) Rhabdomyosarcoma
 - (i) Sudden or rapid onset of proptosis in a child

- e. Physical findings associated with inspection
 - i. Globe displacement
 - ii. Common clinical manifestation of orbit involvement
 - i) Horizontal
 - ii) Vertical
 - iii. Enophthalmos
 - i) Posttraumatic
 - ii) Metastatic lesions
 - iii) Silent sinus syndrome
 - iv. Periorbital changes

- i) Erythema
 - ii) Edema
 - iii) Ecchymosis
- v. Exorbitism
 - i) Associated craniofacial defects
 - ii) Eyelid defects
- vi. Hypertelorism
 - i) Associated craniofacial defects
 - ii) Eyelid defects
- vii. Telecanthus
 - i) Associated craniofacial defects
 - ii) Eyelid defects
 - iii) Trauma (naso-orbital-ethmoidal fracture)
 - iv) Sulcus defect
 - v) Enophthalmos
 - vi) Fat atrophy
 - vii) Trauma (blowout orbital floor fracture)
 - viii) Silent sinus syndrome
 - ix) Ptosis
 - x) Silent brain syndrome
- viii. Lagophthalmos
- ix. Nasal and sinus infection
 - i) Nasal discharge
 - ii) Tenderness
 - iii) Febrile
- x. Valsalva
 - i) Increase in proptosis, varix
- xi. CN palsy
 - i) Acoustic neuroma
 - ii) Mass
 - iii) Injury
- xii. Eyelid malformations

- i) Capillary hemangioma
 - ii) Neurofibroma (S-shape eyelid deformity)
 - xiii. Exposure keratopathy
 - i) Lagophthalmos
 - ii) Proptosis
 - iii) Paralysis
 - xiv. Vascular findings
 - i) Lymphangioma
 - ii) Capillary hemangioma
 - iii) Hemorrhage
 - iv) Ecchymosis (neuroblastoma)
 - v) Varix
 - xv. Nasal speculum exam for signs of inhalation cocaine abuse
 - xvi. Skin changes
 - i) Café au lait spots
 - ii) Juvenile xanthogranuloma (JXG)
 - iii) Xanthelasma
 - iv) Pigment changes (melanoma nevus)
 - v) Hutchinson sign (Herpes Zoster)
 - (i) Involvement of nasociliary nerve seen by lesion on nasal tip
 - (ii) Suggests corneal involvement
- f. Physical findings associated with palpation
 - i. Character
 - i) Firm
 - ii) Soft
 - iii) Mobile
 - iv) Fixed
 - v) Painful to touch
 - vi) Painless
 - vii) Expands with Valsalva
 - viii) Compressible
 - ix) Globe retropulsion

- x) Fluctuant
- ii. Location of palpable lesion
 - i) Superonasal quadrant
 - (i) Mucocele, mucopyocele, encephalocele
 - (ii) Neurofibroma
 - (iii) Dermoid
 - (iv) Lymphoma
 - (v) Orbital abscess
 - (vi) Rhabdomyosarcoma
 - ii) Superotemporal quadrant
 - (i) Dermoid
 - (ii) Prolapsed lacrimal gland
 - (iii) Lacrimal gland tumor
 - (iv) Lymphoma
 - (v) Idiopathic orbital inflammation
 - (vi) Orbital abscess
 - (vii) Sphenoid wing meningioma
 - iii) Inferonasal quadrant
 - (i) Dacryocystitis
 - (ii) Lacrimal sac tumor
 - (iii) Invasive sinus tumor
 - (iv) Orbital abscess
 - (v) Wegener granulomatosis
 - (vi) Lymphoma
 - iv) Inferotemporal quadrant
 - (i) Invasive sinus tumor
 - (ii) Hemorrhage
 - (iii) Displaced zygomatic fracture
 - (iv) Lymphoma
 - v) Behind equator of globe: usually not palpable
 - (i) Globe may be firm to retropulsion
 - (a) Intraconal masses

- vi) Palpation of orbit, thyroid, regional lymph nodes, and abdomen if suspect systemic or metastatic disease
- iii. Clinical correlation of pulsation in orbit
 - i) With bruits/thrills
 - (i) Cavernous carotid fistula
 - (ii) Orbital arteriovenous fistula
 - (iii) Dural arteriovenous (A-V) fistula
 - ii) Without bruits
 - (i) Meningoencephaloceles
 - (ii) Neurofibromatosis
 - (iii) Orbital roof defect
- 5. Visual fields
 - a. Central scotoma
 - b. Constriction
 - c. Altitudinal defects
- 6. Slit-lamp biomicroscope examination
 - a. Chemosis
 - b. Injection
 - c. Discharge
 - d. Exposure keratopathy
- 7. Intraocular pressure
 - a. Increased
 - b. Higher in upgaze in thyroid eye disease
- 8. Fundus
 - a. Optociliary shunt vessels
 - b. Optic nerve swelling/pallor
 - c. Choroidal Folds

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Computed tomography

I. List the indications/contraindications

A. Indications

1. Soft tissue resolution of orbital structures and fat
2. Assessment of bony abnormalities
3. Detection of calcification in lesions
4. Detection of high density/metallic foreign bodies
5. Imaging study of choice for primary assessment of orbital trauma
6. Assessment of active orbital and intracranial hemorrhage
7. Clinical indications
 - a. Proptosis, globe displacement
 - b. Palpable orbital mass
 - c. Progressive monocular visual loss of nonocular origin
 - d. Monocular optic disc edema
 - e. Unexplained ophthalmoplegia
 - f. Paranasal sinus disease affecting the orbit
 - g. Basofrontal tumors affecting orbit
 - h. Optic nerve tumors
 - i. Thyroid eye disease, orbital inflammation
 - j. Orbital fractures, optic canal fracture
 - k. Extraocular muscle lesions
 - l. Suspected orbital abscess, worsening orbital cellulitis
 - m. Guide for surgical approach to orbitotomy
 - n. Intracranial lesions
 - o. Head injury
 - p. Stroke
8. Indications for CT with IV contrast
 - a. Details the internal characteristics of lesions and distinguishes them from adjacent lesions
 - b. Vascular tumors
 - c. Blood vessel anomalies - aneurysm, arteriovenous (a-v) fistula

B. Contraindications

1. Significant cervical spine disease
2. Excessive dental amalgam
3. Contrast contraindication
 - a. Severe allergy
 - b. Renal failure

II. Describe the pre-procedure evaluation

A. Obtain history

1. IV contrast allergic reaction
2. Dental amalgam
3. Recent cervical disease or fractures
4. Unstable neck or spine
5. Renal disease

B. Blood urea nitrogen (BUN)/creatinine (glomerular filtration rate may be a more specific indicator of subtle renal dysfunction) for IV contrast

III. List the alternatives to this procedure

- A. Magnetic resonance imaging (MRI)**
- B. Ultrasound**
- C. X-ray**
- D. Angiography**

IV. Describe the instrumentation and technique

A. Axial and coronal views in thin slices

1. Finer slices will increase radiation dose
2. 3-Dimensional image is electronically constructed to view orbital and periorbital structures

B. IV contrast

1. Iodine in contrast medium provides increased density or enhancement in vascular structures (e.g. extraocular muscle (EOM), lacrimal gland)
2. Defines
 - a. Vascular lesions
 - b. Thrombosis

- c. Cystic or solid masses
 - d. Extraorbital extension or intracranial tumors
 - e. Varices
 - f. Lymphangiomas
 - g. Arteriovenous malformation (AVM)
 - h. Hemangiomas
 - i. Optic nerve lesions (meningioma and glioma)
3. Should not use IV contrast if planning to treat a hyperthyroid patient with radioactive iodine

C. Spiral CT

- 1. Continuous data is sent as the scanner revolves around patient
- 2. Shortens acquisition times
- 3. Good technique with children

D. 3D scanning: reformats CT into 3D projections of bony orbital walls

- 1. Expensive
- 2. Aids in the analysis of orbital bone changes
- 3. Used in preparation for craniofacial surgery and complex orbital fracture repair

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

A. Cataract

- 1. Treatment
 - a. Cataract surgery
 - b. Avoid excess imaging

B. Radiation complications

- 1. Delayed neurological developmental
 - a. Brain more radio-sensitive in children
 - b. Limit use in children
 - c. Prefer MRI in children when possible
- 2. Malignancy

C. IV allergic reaction

- 1. Treatment
 - a. Premedicate with Benadryl or corticosteroids for allergic reaction
 - b. Avoid IV contrast administration

D. Difficult to position head to obtain planes to be imaged in some patients, increased time for axial,

coronal and sagittal planes

1. Treatment
 - a. CT reformatting
 - b. Images are not as sharp but provide faster, simpler scanning
 - c. Quality of reformatted images are better with thin slices on initial axial images

VI. Describe considerations in interpretation for this diagnostic procedure

A. CT is optimal for analysis of the bony orbit, metallic foreign bodies and calcium

B. CT narrows differential diagnosis for orbital lesions/processes

1. Can help distinguish types of lesions: characteristic radiographic findings
 - a. Cystic vs. solid - e.g., dermoid cyst, eosinophilic granuloma
2. Circumscribed vs. diffuse - cavernous hemangioma vs. metastasis
3. Calcification - meningioma, retinoblastoma
 - a. Tram-tracking-meningioma
4. Ground glass - fibrous dysplasia
5. Diffuse spread - metastasis
6. Ring enhancement - infection
7. Thickened sclera, T-sign- Wegener granulomatosis, orbital pseudotumor (scleritis)
8. Molding vs bony erosion - e.g., benign mixed cell tumor vs. adenoid cystic carcinoma
9. Tendon and muscle enlargement vs. muscle only - orbital pseudotumor vs thyroid disease
10. S-shaped nerve-optic nerve glioma
11. Define lymphangioma characteristics
 - a. Diffuse proliferation of lymphatic vessels, slowly progressive or acute in lid, conjunctiva, orbit, mouth, sinuses.
 - b. Heterogeneous grapelike structures seen on CT-Chocolate cysts- vacuolated spaces with old loculated hemorrhage
12. Dilated superior ophthalmic vein - carotid-cavernous (C-C) fistula

C. Allows surgical planning: locates orbital fractures, planning for orbital decompression, craniofacial disorders, tumor excision

D. May need adjunct imaging: e.g., MRI for apex tumors, magnetic resonance angiography (MRA) or angiography for vascular, intracranial lesions

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology; Section 6: Pediatric

2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.9-10, 40, 45, 61-62, 123, 176-179, 210-211, 263.
3. AAO, Focal Points: Management of Orbital Trauma: Fractures, Hemorrhage, and Traumatic Optic Neuropathy, Module #7, 1994, p. 1-2.
4. Linet MS, Kim KP, Rajaraman P. Children's exposure to diagnostic medical radiation and cancer risk: epidemiologic and dosimetric considerations. *Pediatr Radiol.* 2009;39 (1), S4-26.

Magnetic resonance imaging

I. List the indications/contraindications

A. Indications

1. Provides excellent soft tissue detail of the orbit, apex, canal, cavernous sinus and brain
2. Provides view of orbital apex with no bony artifacts
3. Excellent detail of globe, posterior fossa of brain
4. Poor view of bone or calcium
5. Clinical indications
 - a. Soft tissue or vascular orbital lesions
 - b. Orbital apex, canal, cavernous sinus or intracranial lesions
 - c. Intraocular tumors
 - d. Optic nerve compression from thyroid orbitopathy
 - e. Neural lesions of orbit and optic nerve
 - f. Hemorrhage within a lesion
 - g. Brain/cranial nerve lesions

B. Contraindications

1. Metallic foreign bodies in orbit or periorbital tissue
2. Ferromagnetic vascular clips
3. Non-MRI compatible intracranial aneurysm clips
4. Magnetic intravascular filters
5. Cardiac pacemakers / defibrillator
6. Claustrophobia
7. Acute trauma and an unstable patient
8. Patient unable to cooperate with time or position required to complete MRI study

II. Describe the pre-procedure evaluation

A. Obtain history

1. Metallic foreign bodies
2. Aneurysm or cardiac surgery
3. Claustrophobic or psychiatric disorders

4. Gadolinium allergic reaction, age

B. Certain eye make up, typically mascara, should be removed

III. List the alternatives to this procedure

- A. Computed tomography (CT)**
- B. Angiogram**
- C. Ultrasound**
- D. X-ray**

IV. Describe the instrumentation and technique

- A. No ionizing radiation is used**
- B. No adverse biological effects**
- C. Multiple planes imaged at once**
- D. MRI based on 3 components of nuclear magnetic resonance**
 - 1. Atomic nuclei possessing an electrical charge
 - 2. Radiofrequency waves
 - 3. Powerful magnetic field
- E. The strength of magnetic field is measured in Tesla or Gauss units**
- F. Thin slices in the axial, coronal and sagittal planes are performed with images from T1 and T2 parameters**
 - 1. T1 and T2 weighted images are based on tissue proton density and relaxation times (proton density- number of protons per unit volume of tissue)
 - 2. Each tissue has different T1 and T2 characteristics and proton density which determines signal intensity
 - 3. T1, longitudinal relaxation time: recognized by dark vitreous and dark cerebrospinal fluid (CSF)
 - 4. T2, transverse relaxation time: characteristic bright vitreous and CSF
- G. MRI with IV Gadolinium**
 - 1. Contrast improves imaging by brightening vascular lesions:
 - a. Arteriovenous malformations (AVMs)
 - b. Meningiomas
 - c. Arteriovenous (AV) fistulas
 - d. Vascular malformations
 - e. Cavernous hemangiomas
 - 2. Shows high flow vessels as a signal void, which are the same density as fat

3. Useful for most central nervous system (CNS) lesions

H. Optic nerve coil

1. High resolution images of eye and optic nerve

I. Fat suppression

1. Differentiates orbital lesions from fat and vital structures, e.g., optic neuritis, optic nerve glioma, optic neuritis

J. Magnetic resonance angiography (MRA)

1. Non invasive visualization of medium and large size vessels, not as fine details of angiography
2. Clinically useful for detecting A-V malformations, aneurysms of branches of internal and external looped arteries, venous occlusions and arterial stenosis
3. Relies on the physiology of blood flow instead of actual anatomy, aneurysms can be missed by this modality. Conventional angiography remains the "gold standard" for exclusion of aneurysms

K. Magnetic resonance venography

1. Inject contrast into frontal and angular vein
2. Detects venous abnormality in orbit, cavernous sinus and brain, for example, varices

L. Angiography

1. Better visualization of small- to large-size vessels in fine detail
2. Invasive - cannulate femoral or carotid arteries

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

A. Motion artifact

1. Dental amalgam
2. Patient must lie still
 - a. Sedation sometimes necessary

B. Claustrophobia

1. Some patient unable to complete MRI
 - a. May require sedation

C. Longer test time

1. Higher chance for motion artifact
2. Patient discomfort

D. More expensive than CT

VI. Describe the considerations in interpretation of this diagnostic procedure

A. MRI provides more tissue specificity than other diagnostic modalities depending on signal

intensities

- B. MRI identifies relation of tumor to optic nerve (better than CT)**
- C. MRI omits bone, and CT is the preferred study for good study for fractures, traumas, foreign bodies, bony lesions or calcified lesions**
- D. MRI is used in conjunction with CT and ultrasound or angiogram when indicated**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.9-11, 33,62, 176-179.
3. AAO, Focal Points: Management of Orbital Trauma, Module #7, 1994, p.2.

Preseptal cellulitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Spread from adjacent structures
 - a. Sinus infection (especially ethmoiditis)
 - b. Orbital fracture
 - c. Dental abscess
 - d. Periorbital skin infection
2. Spread from orbital structures
 - a. Dacryocystitis
 - b. Dacryoadenitis
3. Spread from eyelid structures
 - a. Anterior and posterior hordeolum
4. Post trauma
 - a. Eyelid lacerations
 - b. Orbital fractures
5. Organisms
 - a. *Staphylococcus* species
 - i. Beware MRSA
 - ii. May be indolent or aggressive
 - b. *Streptococcus* species
 - c. *Haemophilus influenza*
 - d. Gram negative rods (after trauma)

B. List the pertinent elements of the history

1. Age
2. Onset, duration
3. Symptoms
 - a. Fever
 - b. Eyelid tenderness
 - c. Warmth

4. Recent infections
 - a. Upper respiratory infection
 - b. Ear, nose, throat infection
 - c. Dacryocystitis
5. History of sinus infections or surgery
6. Trauma
7. Skin break, insect bites, eyebrow epilation
8. Recent eyelid or facial surgery

C. Describe pertinent clinical features

1. Eyelid edema, erythema, tenderness, warmth, fever
2. Minimal proptosis, normal extraocular movements, normal vision
3. Minimal conjunctival injection and chemosis

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Orbital imaging of orbit and sinuses with MRI or CT if no direct site of inoculation is identified

II. Define the risk factors

- A. Recent infection (sinus, upper respiratory, dacryocystitis, bacteremia), tooth abscess, trauma, or surgery
- B. Ruptured skin vesicles

III. List the differential diagnosis

- A. Orbital inflammatory disorders
- B. Rapidly growing orbital tumors, typically metastatic
- C. Orbital vascular abnormality (carotid cavernous fistula, superior ophthalmic vein thrombosis, venous malformations)
- 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. Oral antibiotics
2. Warm compresses
3. IV antibiotics, if unresponsive
4. Treatment of sinusitis if present

B. Describe surgical therapy options

1. Drain localized abscesses
2. Necrotizing cellulitis(fasciitis) may require debridement
3. Sinus surgery if indicated

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

A. Complications

1. Medical
 - a. Complications of antibiotics
 - b. Need for surgery due to progression of disease
2. Surgical
 - a. Need for further surgery
 - b. Spread of infection

B. Prevention

1. Serial clinical exams to monitor for deterioration
2. Adequate imaging studies to identify etiology
3. Aggressive medical treatment
4. Appropriate surgical approach and location of infection

C. Management

1. Careful observation postoperatively for improvement or progression of symptoms
2. Monitoring of visual acuity and extraocular motility
3. Follow-up imaging may be indicated in some situations

VI. Describe disease-related complications

A. Progression to orbital cellulitis/abscess

VII. Describe appropriate patient instructions

A. Adequate follow-up with other specialists: e.g., ear, nose, and throat (ENT) for contiguous infections/etiology, prevention and treatment

B. Monitoring of vision, symptoms and follow-up with ophthalmologist

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System,

2013-2014.

2. AAO, Focal Points: Preseptal and Orbital Cellulitis, Module #11, 2008.

Orbital cellulitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Routes of infection
 - a. Spread from adjacent structures
 - i. Sinus infection (especially ethmoiditis)
 - ii. Orbital fracture
 - iii. Dental abscess
 - iv. Periorbital skin infection
 - v. Intracranial infection/abscess
 - b. Spread from orbital structures
 - i. Panophthalmitis
 - ii. Dacryocystitis
 - iii. Dacryoadenitis
 - c. Post trauma
 - i. Retained foreign body
 - ii. Eyelid lacerations
 - iii. Orbital fractures
 - d. Postsurgical
 - i. Eye surgery
 - ii. Orbital surgery
 - iii. Lacrimal probing or surgery
 - iv. Sinus surgery
 - v. Dental procedures
 - e. Endogenous
 - i. Bacteremia
2. Anatomical factors
 - a. Unique structure of the lids
 - b. "Closed box" construction of the orbit
 - c. Proximity of paranasal sinuses

- d. Absence of lymphatic drainage within orbit
- e. Profuse venous connections among associated structures
- 3. Common organisms
 - a. *Staphylococcus* species
 - i. Consider MRSA in both indolent and aggressive infections
 - b. *Streptococcus* species
 - c. Gram negative rods (after trauma)
 - d. Fungal (mucor, aspergillus) especially in diabetic or immunocompromised patients
 - i. These may be particularly vision and life threatening
 - e. Anaerobes: peptococcus, peptostreptococcus, bacteroides

B. Define the relevant aspects of epidemiology of the disease

- 1. Can occur in all ages, all races
- 2. Age
 - a. Children
 - i. *Haemophilus* organism originating from ethmoid sinuses was prevalent in children, but has decreased due to childhood immunization
 - ii. Single organism infections (commonly *Staphylococcus* and *Streptococcus*) typical in younger children
 - iii. Polymicrobial disease more prevalent in older patients
 - b. Adults
 - i. More likely from *Staphylococcus* or *Streptococcus* species
 - ii. Polymicrobial disease

C. List the pertinent elements of the history

- 1. Age
- 2. Onset, duration
- 3. Symptoms
 - a. Malaise
 - b. Fever
 - c. Pain (especially with eye movements)
 - d. Vision loss
 - e. Diplopia
- 4. Recent infections
 - a. Upper respiratory infection

- b. Ear, nose, throat, facial or dental infection
 - c. Dacryocystitis
- 5. History of sinus infections or surgery
- 6. Trauma
- 7. Recent surgery
- 8. Mental status changes stiff neck, nausea, vomiting, headache
- 9. Past medical history
 - a. Diabetes mellitus (especially with ketoacidosis)
 - b. Immunocompromised state (history of cancer, renal disease, hyperalimentation) increases risk of fungal infection

D. Describe pertinent clinical features

- 1. Proptosis, restricted extraocular movement, decreased vision, afferent pupillary defect, reduced corneal sensation, eyelid edema, erythema, tenderness, warmth, conjunctival injection and chemosis, fever
- 2. Rule out headache, stiff neck, mental status changes
- 3. Fungal infections may present aggressively or as an indolent fashion
 - a. Most commonly in diabetic and immunocompromised hosts
 - b. Presentation includes features above plus characteristic signs of angio-invasion
 - i. Progressive cranial neuropathies
 - ii. Ischemic tissue on exam and at surgery
- 4. Eschar on roof of mouth or nose in diabetic patient is suggestive of mucormycosis
 - a. May progress to facial cellulitis (potentially necrotizing/ischemic), orbit apex syndrome, vision loss, other neurologic deficits, cavernous sinus thrombosis, carotid artery thrombosis
- 5. Orbital abscess may develop
 - a. Abscess may not be responsive to antibiotic therapy
 - b. Abscess may produce mass effect
 - i. May compress globe or optic nerve threatening vision
 - ii. May compress extraocular muscles producing diplopia
 - iii. Mass effect may cause increased pain
 - c. Subperiosteal abscess may develop adjacent to infected sinus
 - i. Typically extension of sinus infection through orbital wall causing subperiosteal orbital purulence without generalized orbital involvement
 - ii. May have a more benign course than other forms of orbital abscess, especially in children

- i) May respond to management of sinus disease, rather than surgical treatment specifically directed at orbit process

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Imaging of orbits - orbital fat stranding, orbital abscess (may be subperiosteal)
2. MRI may be useful in determining extent of tissue involvement, especially if fungal infection suspected.
3. Complete blood count (CBC)
4. Blood cultures (especially if febrile or systemic illness known)
5. Basic metabolic panel
6. Lumbar puncture (if suspect meningeal involvement)
7. Nasal culture swab (if sinusitis)
8. Conjunctival culture swab (if significant mucous discharge)
9. Tissue culture for fungus or atypical mycobacteria
10. Biopsy of nasal/sinus mucosa or necrotic skin lesion, look particularly for organisms or masquerade neoplasm

II. Define the risk factors

- A. Recent infection (sinus, upper respiratory, dacryocystitis, bacteremia), tooth abscess, trauma, or facial surgery
- B. Immunocompromised or diabetic patient (greater risk for mucormycosis and other fungi)

III. List the differential diagnosis

- A. Preseptal cellulitis
- B. Thyroid eye disease
- C. Orbital hemorrhage
- D. Benign or malignant primary tumors (lymphoproliferative disorders, lymphangioma/venous malformations, capillary hemangioma, ruptured dermoid cyst, meningioma, squamous cell carcinoma (SCC), basal cell carcinoma (BCCA), etc)
- E. Sinus tumor extension (fungal sinusitis, SCC)
- F. Metastatic tumor (breast, lung, prostate, leukemia, lymphoma)
- G. Rhabdomyosarcoma, neuroblastoma (especially children)
- H. Inflammatory lesions: Orbital inflammatory syndrome, Wegener's granulomatosis, sarcoidosis, systemic lupus erythematosus (SLE)
- I. Tolosa-Hunt syndrome
- J. Carotid-cavernous sinus fistula

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

A. Describe medical therapy options

1. Treat etiology
2. Infectious disease (ID) and/or ear, nose, and throat (ENT) consultation and co-management
3. Serial clinical exams and close observation: vital signs, visual acuity, pupillary light response, motility testing, mental status alteration
4. Beware that imaging may lag behind clinical improvement
5. IV antibiotics
6. Nasal/sinus irrigation, nasal decongestant
7. Antibiotic ointment, lubrication for exposure keratopathy.
8. Patients with fungal infections (e.g., Mucormycosis) require antifungal treatment and possibly local amphotericin irrigations (and likely surgery)
 - a. Lack of prompt treatment may lead to death and/or blindness within days
9. Orbital abscess will likely not respond to medical therapy alone
 - a. Exception is some subperiosteal abscess in children which may resolve with medical treatment of primary sinus disease

B. Describe surgical therapy options

1. Drain orbital or subperiosteal abscess
 - a. This may need to be undertaken urgently in cases of vision compromise
 - b. Drainage should be considered for subperiosteal abscess in adults or in children not improving with intravenous antibiotic therapy
2. Drain sinus using external or endoscopic approach
3. Obtain bacterial cultures
 - a. Adjust antibiotic coverage based on culture results and sensitivity
4. Treat any intracranial complication
5. Patients with fungal or fulminant infections (e.g., mucormycosis) may require repeated surgical debridement of necrotic tissue with local orbital irrigation with amphotericin B
 - a. Consider hyperbaric oxygen treatment

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

A. Complications

1. Medical
 - a. Complications of antibiotics
 - b. Complications of amphotericin B

- c. Need for surgery due to progression of disease
- 2. Surgical
 - a. Need for further surgery
 - b. Visual loss, scarring, motility disturbance
 - c. Hemorrhage
 - d. Ocular ischemia (central retinal artery occlusion (CRAO))
 - e. Spread of infection

B. Prevention

- 1. Serial clinical exams to monitor for deterioration
- 2. Adequate imaging to identify etiology
- 3. Aggressive medical treatment
 - a. Normalize underlying metabolic disturbance
- 4. Appropriate surgical approach and location of infection
- 5. Careful manipulation of globe and tissue at surgery
- 6. Control of hemostasis

C. Management

- 1. Careful observation postoperatively for improvement or progression of symptoms
- 2. Monitoring of visual acuity
- 3. Follow-up imaging as needed

VI. Describe disease-related complications

A. Progression to

- 1. Orbital abscess
- 2. Subperiosteal abscess
- 3. Cavernous sinus thrombosis
- 4. Meningitis
- 5. Brain abscess
- 6. Cranial neuropathies

B. Visual loss may result

C. Intracranial involvement, even death

D. Immunocompromised patients are at greater risk of more indolent fungal infection which may have a lethal course

VII. 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. Management of underlying disease processes to help restore immunocompetence and remove contributing factors**
- D. Monitoring of vision, symptoms and follow-up with ophthalmologist**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Preseptal and Orbital Cellulitis, Module #11, 2008.
3. Klok CE, Rubin PA. Role of inflammation in orbital cellulitis. *Int Ophthalmol Clin* 2006;46:57-68.

Thyroid eye disease

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease (also known as Graves ophthalmopathy or thyroid orbitopathy)

1. Autoimmune disorder: anti-thyrotropin-receptor antibodies (TSH-receptor antibodies) bind to TSH receptors on thyroid follicular cells and on orbital fibroblasts, resulting in stimulation of excessive thyroid hormone production.
2. Molecular mechanism: orbital fibroblasts are the target of the autoimmune process.
 - a. Subsets of fibroblasts differentiate into adipocytes when activated by TSH-receptor-antibodies
 - b. Adipocytes and fibroblasts produce interleukin-6 which increases B-cell maturation and increase orbital TSH receptor antibody production
3. Cytokine release, increased adipogenesis, and glycosaminoglycans (GAG) production lead to increased orbital fat, extraocular muscle volume and inflammatory response

B. Define the relevant aspects of epidemiology of the disease

1. Women to men: 5.5:1
2. Age 25 - 50 years
3. Smokers: highest external risk factor, 7.7 times increase to have disease

C. List the pertinent elements of the history

1. History of Graves hyperthyroidism or other thyroid abnormalities
2. Symptoms of systemic hyperthyroidism
 - a. Pretibial myxedema
 - b. Tremor
 - c. Weight loss
 - d. Goiter
3. Symptoms of the eye
 - a. Eye pain at rest and with movement
 - b. Conjunctival redness/swelling
 - c. Caruncle swelling
 - d. Eyelid redness and swelling
4. Family history
5. Smoking history
6. Myasthenia gravis symptoms

7. Vision loss
8. Rate of change of eye symptoms/time since onset

D. Describe pertinent clinical features

1. Lid retraction (most common)
2. Lid lag on downgaze
3. Periorbital edema
4. Proptosis
5. Conjunctival chemosis
6. Enlarged vessels over rectus muscle insertions
7. Corneal exposure, ulceration
8. Extraocular muscle (EOM) motility restriction
9. Elevated intraocular pressure (IOP) with attempted upgaze
10. Compressive optic neuropathy with decreased visual acuity and color vision, visual field defects, afferent pupil defect

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Thyroid function testing
 - a. TSH
 - b. Triiodothyronine (T3)
 - c. Levothyroxine (T4)
 - d. TSH-receptor antibodies
 - e. Thyroid-binding inhibitory immunoglobulins (TBII)
 - f. Thyroid-stimulating immunoglobulins (TSI)
 - g. Antithyroglobulin antibodies
 - h. Antithyroid peroxidase antibodies
2. Orbital computed tomography (CT) or magnetic resonance imaging (MRI) scan
 - a. Enlarged extraocular muscles seen with relative sparing of the tendons, and/or increased orbital fat volume
 - b. Orbital apex can be tight compressing optic nerve
 - c. Rare possible superior ophthalmic vein dilation/enlargement - beware of a vascular fistula and hemorrhage if patients are decompressed

F. List diagnostic criteria for thyroid eye disease

1. Eyelid retraction with
 - a. Thyroid dysfunction or abnormal regulation

- b. Exophthalmos
 - c. Optic nerve dysfunction or
 - d. EOM dysfunction
- 2. Or thyroid dysfunction/abnormal regulation with
 - a. Exophthalmos
 - b. Optic nerve dysfunction or
 - c. EOM involvement

II. Define the risk factors

- A. Smoking**
- B. Female**
- C. Family history**

III. List the differential diagnosis

- A. Orbital inflammatory diseases - specific and idiopathic**
- B. Orbital cellulitis**
- C. Carotid cavernous fistula**
- D. Orbital tumor**
- E. Orbital amyloidosis**
- F. Lid retraction alone**
 - 1. Contralateral ptosis
 - 2. Postsurgical
 - 3. Following vertical muscle surgery
 - 4. Posttraumatic
 - 5. Neurologic
 - a. Parinaud
 - b. Cranial Nerve (CN) III aberrant regeneration
 - 6. Silent sinus syndrome

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

- A. Describe medical therapy options**
 - 1. Supportive therapy

- a. Ocular lubricants
 - b. Elevate head of bed when sleeping
- 2. Corticosteroids
 - a. In setting of severe active inflammation, compressive optic neuropathy
- 3. Radiation therapy
 - a. Although anecdotal evidence suggests that orbital radiotherapy may be efficacious in patients with acute inflammation or optic neuropathy, two clinical trials failed to demonstrate measurable improvement in patients with moderate ophthalmopathy

B. Describe surgical therapy options

- 1. In active stage
 - a. Orbital decompression for compressive optic neuropathy
 - b. Tarsorrhaphy for severe corneal disease with proptosis
 - c. Early levator recession if corneal exposure cannot be managed other ways
- 2. In chronic stage
 - a. Staged surgical management once there is no change in signs/symptoms for 6 months
 - b. The logical stated sequence is important to avoid unnecessary surgery because the first procedure could influence the indications and findings to be addressed by the following procedures
 - i. Orbital decompression
 - ii. Strabismus surgery
 - iii. Eyelid retraction repair
 - iv. Blepharoplasty

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

A. Corticosteroids (intravenous or oral)

- 1. Corticosteroid dependence
- 2. Aseptic necrosis of the hip
- 3. Hyperglycemia
- 4. Increased infections
- 5. Hypertension
- 6. Reactivation of tuberculosis (TB)
- 7. Prevention
 - a. Keep dose and length of time on drug as minimal as possible in order to obtain desired effect

8. Management
 - a. Check blood pressure, purified protein derivative (PPD) when on chronic use

B. Radiation therapy

1. Radiation retinopathy
2. Prevention
 - a. Avoid radiation in diabetics, patients with vasculitis

C. Surgery

1. Orbital decompression
 - a. Blindness
 - b. Diplopia (strabismus surgery)
 - c. Sinus complications
 - d. Numbness
2. Strabismus surgery
 - a. Lower lid retraction (lid surgery)
 - b. Recurrent diplopia (wait until disease stable to reoperate)
3. Lid position surgery
 - a. Persistent lid retraction (reoperate)
 - b. Ptosis (ptosis repair)
4. Cosmetic lid surgery
 - a. Remove excess fatty tissue and skin

VI. 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 EOM restriction**

VII. Describe appropriate patient instructions

- A. Return if patient notices decrease in vision, decrease in color brightness, or significant exacerbation of inflammatory signs**
- B. For newly diagnosed patients, provide accurate counseling about:**
 1. Chronic nature of disease which can be frustrating for the patient
 2. Goals

- a. Preserve vision
- b. Provide comfort
- c. Surgical correction
 - i. Lid retraction
 - ii. Proptosis
 - iii. Corneal exposure
 - iv. Diplopia
 - v. Cosmesis

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 1: Update on General Medicine; Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 5: Neuro-Ophthalmology; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993.
3. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994.
4. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995.
5. AAO, Focal Points: Thyroid-Associated Orbitopathy, Module #1, 1997.
6. Dolman PJ, Rootman J. VISA Classification for Graves orbitopathy. *Ophthal Plast Reconstr Surg* 2006;22:319-24.
7. Garrity JA, Bahn RS. Pathogenesis of graves ophthalmopathy: implications for prediction, prevention, and treatment. *Am J Ophthalmol* 2006;142:147-153.
8. Krassas GE, Boboridis K. Recent developments in the medical treatment of thyroid eye disease. *Orbit* 2006;25:117-22.
9. Hatton MP, Rubin PA. Controversies in thyroid-related orbitopathy: radiation and decompression. *Int Ophthalmol Clin* 2005;45:1-14.
10. Abraham P, Avenell A, Watson WA, et al. Antithyroid drug regimen for treating Graves' hyperthyroidism. *Cochrane Database Syst Rev* 2005:CD003420.
11. AAO, Focal Points: Botulinum Toxin in Ophthalmology, Module #3, 2007.
12. Bahn RS. Graves' ophthalmopathy. *N Engl J Med* 2010 Feb 25;362(8):726-38.

Idiopathic orbital inflammatory syndrome (nonspecific orbital inflammation, orbital pseudotumor)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Idiopathic, sometimes occurring after upper respiratory infection (URI), especially in children

B. Define the relevant aspects of epidemiology of the disease

1. All ages, men and women

C. List the pertinent elements of the history

1. Pain is often the presenting feature
2. Possible decreased vision, double vision, depending on location of inflammation

D. Describe pertinent clinical features

1. Anterior orbital inflammation/diffuse inflammation
 - a. Pain
 - b. Erythema and edema of lids
 - c. Chemosis
 - d. Decreased vision
 - e. Decreased extraocular motility
 - f. Uveitis/scleritis
2. Apical inflammation
 - a. Pain
 - b. Decreased vision
 - c. Decreased extraocular motility
3. Myositic inflammation
 - a. Pain with eye movement
 - b. Decreased extraocular motility
 - c. Localized chemosis with conjunctival injection
4. Lacrimal inflammation - dacryoadenitis
 - a. Pain
 - b. S-shaped deformity of lid

- c. Localized chemosis, conjunctival injection
- 5. Sclerosing pseudotumor
 - a. Painless loss of orbital function
- E. Describe appropriate testing and evaluation for establishing the diagnosis**
 - 1. Computed tomography (CT)/Magnetic resonance imaging (MRI) scan
 - a. Anterior, diffuse, apical: enhancing infiltrative mass with irregular margins
 - b. Myositic
 - i. Fusiform enlargement of extraocular muscle involving the tendons of insertion
 - c. Lacrimal
 - i. Enlargement of the lacrimal gland with some hazy involvement of adjacent fat, bone intact
 - 2. Laboratory evaluation (See also Section II)
 - a. WBC with differential (eosinophilia in NSOI)
 - b. Erythrocyte sedimentation rate (elevated in NSOI)
 - c. Anti-nuclear antibody (positive in NSOI)
 - d. Cerebral spinal fluid pleocytosis (mild elevation in NSOI)
 - e. Rheumatoid factor (auto-immune process)
 - f. Angiotensin converting enzyme (sarcoidosis)
 - g. Lysozyme (sarcoidosis)
 - h. Antineutrophil cytoplasmic antibody (p-ANCA, c-ANCA) (Wegener)

II. List the differential diagnosis

A. Adults

- 1. Orbital cellulitis
- 2. Orbital inflammation of specific etiology
 - a. Sarcoidosis
 - i. Uveitis
 - ii. Dacryoadenitis
 - iii. Rare orbital lesions
 - iv. Tests
 - i) Angiotensin-converting enzyme
 - ii) Lysozyme

- iii) Serum calcium
 - iv) Chest x-ray (hilar adenopathy)
 - v) Positive gallium scan
- b. Wegener granulomatosis
 - i. Upper and lower respiratory tract lesions
 - ii. Vasculitis
 - iii. Nephritis
 - iv. Uveitis
 - v. Orbital inflammation
 - vi. Tests
 - i) Antineutrophil cytoplasmic antibodies (p-ANCA, c-ANCA)
 - ii) Abnormal urinary sediment
 - iii) Abnormal chest x-ray (CXR)
- c. Sjögren syndrome
 - i. Dry eyes and mouth
 - ii. Other autoimmune diseases
 - iii. Enlarged lacrimal glands
 - iv. Tests
 - i) Antinuclear antibody (ANA)
 - ii) Rheumatoid factor (RF)
 - iii) Sjögren antibodies
- d. Ruptured dermoid cyst
- e. Arteriovenous fistula
- f. Orbital metastasis
- g. Thyroid orbitopathy
 - i. Thyroid function testing
 - i) Thyroid stimulating hormone (TSH)
 - ii) Triiodothyronine (T3)
 - iii) Levothyroxine (T4)
 - iv) Thyroid antibodies - thyroid-binding inhibitory immunoglobulins (TBI), thyroid-stimulating immunoglobulins (TSI)
 - ii. Orbital CT or MRI scan
 - i) Enlarged extraocular muscles with sparing of the tendons, and/or increased

B. Children

1. Orbital cellulitis
2. Ruptured dermoid cyst
3. Rhabdomyosarcoma
4. Metastatic neuroblastoma
5. Leukemic orbital infiltrate

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

A. Describe medical therapy options

1. Oral prednisone with taper over weeks to several months; consider pulse-dosed intravenous dexamethasone prior to oral corticosteroids
2. Empiric therapy if confident of idiopathic diagnosis
3. Biopsy if atypical
4. Adjunctive non-steroidal anti-inflammatory drugs (NSAID) may be helpful
5. Many surgeons choose to biopsy all infiltrative orbital lesions prior to subjecting a patient to long-term corticosteroid therapy
 - a. Exceptions to this include presumed orbital myositis or orbital apex lesions
6. If no improvement or if recurrence, perform orbital biopsy
7. Orbital radiation or steroid sparing agents for resistant cases
8. Cytotoxic agents (methotrexate, cyclosporine, cyclophosphamide) for sclerosing pseudotumor

B. Describe surgical therapy options

1. Biopsy for diagnostic purposes only

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

A. Prednisone

1. Inability to taper drug without flare of disease
2. Hyperglycemia
3. Aseptic necrosis of the hip
4. Weight gain
5. Cushingoid facies
6. Depression
7. Prevention

- a. Use prednisone at minimally effective dose
- b. Limit long term use
- c. Discuss side effects with patient before use

B. Complications related to radiation therapy or cytotoxic agents

V. Describe disease-related complications

A. Loss of vision

B. Diplopia

C. Persistent or recurrent episodes of inflammation. Need to rule out:

1. Abscess/sequestrum
2. Unusual organism
3. Leaking dermoid
4. Hematological malignancy
5. Necrosis of solid tumor
6. Non-infective vasculitis

VI. Describe appropriate patient instructions

A. Take medications as instructed

1. Do not stop prednisone on own

B. Return immediately with worsening of signs/symptoms on prednisone

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. Zborowska B, Ghabrial R, Selva D, et al. Idiopathic orbital inflammation with extraorbital extension: case series and review. *Eye* 2006;20:107-13.

Sarcoidosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Multisystem disease of unknown origin

B. Define the relevant aspects of epidemiology of this disease

1. More common between ages 20-50 years
2. Reported in all racial groups, more prevalent among African-Americans and Scandinavians
3. Most common sites of involvement in systemic sarcoidosis
 - a. Chest with pulmonary infiltration or hilar adenopathy - very common
 - b. Skin lesions in some patients
4. Ocular manifestations in patients
 - a. Uveitis
 - i. May be a presenting sign of sarcoid
 - ii. Patients may develop visual loss
5. Lacrimal gland involvement most common orbital manifestation
 - a. Typically bilateral
 - b. Clinical enlargement of lacrimal gland is rare
6. Uncommon in children

C. Describe the pathology

1. Multisystem disease characterized by granulomatous inflammation
2. Hallmark is a noncaseating granuloma containing epithelioid cells, multinucleated giant cells (Langhans giant cell with nuclei at the periphery of the cell) and a rim of lymphocytes

D. Describe pertinent clinical features

1. Systemic
 - a. Pulmonary symptoms
 - b. Lymphadenopathy
 - c. Cutaneous
 - i. Occasional eyelid skin granulomas
 - d. CNS granulomatous disease
 - e. Arthritis
2. Ocular/periocular

- a. Acute or chronic anterior uveitis
- b. Orbital, lacrimal gland, lacrimal sac involvement
- c. Conjunctival granulomas
- d. Nummular corneal infiltrates
- e. Band keratopathy
- f. Mutton fat keratic precipitates especially involving the anterior chamber angle
- g. Koeppe and Busacca iris nodules
- h. Posterior synechiae, peripheral anterior synechiae
- i. Vitreitis - snowballs, "string of pearls"
- j. Occlusive retinal vascular disease
- k. Cystoid macular edema
- l. Optic nerve edema may be secondary to uveitis or CNS disease
 - i. Optic nerve granulomatous invasion is direct involvement of the optic nerve by sarcoidosis

E. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Chest X-ray, pulmonary function tests
- 2. Chest computed tomography of thorax with contrast
- 3. Specific neuro or body imaging of affected sites
- 4. Serum angiotensin converting enzyme, lysozyme
- 5. Directed biopsy
 - a. Conjunctiva
 - b. Lacrimal gland
 - c. Lymph node
 - d. Pulmonary-Transbronchial lung biopsy
 - e. Skin rash

II. List the differential diagnosis (site specific)

A. Orbital/lacrimal gland

- 1. Lymphoproliferative disorder (See Lymphoid hyperplasia and lymphoma)
- 2. Other orbital tumor (pleomorphic adenoma, epithelial choristoma, other)
- 3. Sjögren syndrome
- 4. Idiopathic orbital inflammation (orbital pseudotumor) (See Idiopathic orbital inflammatory syndrome (nonspecific orbital inflammation, orbital pseudotumor))

5. Specific inflammations (systemic lupus erythematosus, Wegener granulomatosis)
6. Infectious dacryoadenitis (bacterial, viral)

B. Ocular

1. Syphilis
2. Lyme disease
3. Tuberculosis
4. Familial juvenile systemic granulomatosis
5. Vogt Koyanagi Harada syndrome
6. Behçet disease
7. Sympathetic ophthalmia
8. Multifocal choroiditis with panuveitis

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

- A. Topical, periocular and systemic corticosteroids are tailored to the location and severity of inflammation
- B. Cycloplegia for uveitis
- C. Immunosuppressant agents (methotrexate, cyclosporine, etc.) usually reserved for corticosteroid resistant or intolerant patients

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

- A. Corticosteroid induced side effects
- B. Complications of immunosuppressants

V. Describe disease-related complications

A. Ocular

1. Dry eyes
2. Cataract
3. Band keratopathy
4. Glaucoma
5. Macular edema
6. Retinal vascular disease
7. Optic atrophy

B. Systemic

1. Pulmonary disease with decreased pulmonary function
2. Organomegaly: spleen, liver
3. Cardiac involvement
4. CNS defects

VI. Describe appropriate patient instructions

A. Call to report

1. Redness, pain, photophobia, blurred vision or decreased vision
2. New neurologic or ophthalmologic symptoms
3. New systemic complications

B. Follow the doctor's instructions regarding medication

C. Chronic disease with variable course

D. Side effects of medications must be understood

1. Do not suddenly stop systemic corticosteroids

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-ophthalmology, 2013-2014.
2. AAO, Basic and Clinical Science Course. Section 7: Eyelid, Orbit and Lacrimal System, 2013-2014.
3. AAO, Basic and Clinical Science Course. Section 9: Intraocular Inflammation and Uveitis, 2013-2014.
4. Bonfioli AA, Orefice F. Sarcoidosis. *Semin Ophthalmol* 2005;20:177-82.
5. Power WJ, Neves RA, Rodriguez A, et al. The value of combined serum angiotensin-converting enzyme and gallium scan in diagnosing ocular sarcoidosis. *Ophthalmology* 1995;102: 2007-2011.

Giant cell arteritis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Unknown cause of systemic granulomatosis arteritis

B. Define the relevant aspects of epidemiology of the disease

1. Elderly
2. Female: male 3:1

C. List the pertinent elements of the history

1. Headache
2. Jaw claudication
3. Scalp tenderness
4. Systemic weakness, malaise, weight loss, fever
5. Shoulder/muscle pain (polymyalgia rheumatica)
6. Loss of vision
7. Diplopia

D. Describe pertinent clinical features

1. Decreased vision or visual field associated with ischemic optic neuropathy - swollen or normal optic nerve head
2. Decreased vision associated with central retinal artery occlusion
 - a. Cherry red spot
3. Extraocular muscle (EOM) dysfunction associated with cranial nerve palsy

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Erythrocyte sedimentation rate (ESR)
 - a. Usually elevated
2. C-reactive protein (CRP)
 - a. Usually elevated
3. Temporal artery biopsy
 - a. Inflammation with or without giant cells
 - b. Disruption of internal elastic lamina
 - c. May have skip areas
 - d. May consider bilateral biopsies

II. Define the risk factors

- A. Elderly**
- B. Female**

III. List the differential diagnosis

- A. Nonarteritic ischemic optic neuropathy**
- B. Other optic neuropathies (infectious, inflammatory, infiltrative, compressive, toxic)**
- C. Nonarteritic central artery occlusion**
- D. Central retinal vein occlusion**

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

A. Describe medical therapy options

1. Systemic corticosteroids - IV for severe vision loss (methylprednisolone 250mg IV Q6 h for 3 days then oral prednisone 80-100mg/day), oral prednisone otherwise (1.0 to 1.5 mg/kg/day)
2. Initiate steroids while awaiting biopsy results as a delay in treatment can be vision threatening
3. Follow ESR and clinical symptoms
 - a. Decision to treat or stop treatment should be based on the complete clinical picture
4. Taper corticosteroids slowly
5. 65% of untreated patients may develop bilateral ischemic optic neuropathy

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

A. Prednisone

1. Inability to taper drug without flare of disease
2. Hyperglycemia
3. Aseptic necrosis of the hip
4. Weight gain
5. Cushingoid facies
6. Depression
7. Prevention
 - a. Limit long term use
 - b. Discuss side effects with patient before use

VI. Describe disease-related complications

- A. Loss of vision**
- B. Diplopia**
- C. Other organs such as the brain and heart may also suffer from ischemia**

VII. Describe appropriate patient instructions

- A. Take medications as instructed**
 - 1. Do not stop prednisone on own
- B. Return immediately if worsening of signs/symptoms on prednisone**

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 1: Update on General Medicine; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
- 2. Rahman W, Rahman FZ. Giant cell (temporal) arteritis: an overview and update. *Surv Ophthalmol* 2005;50:415-28.
- 3. AAO, Focal Points: Giant Cell Arteritis, Module #6, 2005.

Wegener granulomatosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease.

1. Necrotizing small vessel granulomatous vasculitis of unknown etiology
2. Classically affecting
 - a. Upper and lower respiratory system
 - b. Kidneys

B. Define the relevant aspects of epidemiology of the disease

1. Fifth decade
2. Males: females 2:1

C. List the pertinent elements of the history

1. Sinus symptoms
 - a. Rhinitis
 - b. Chronic sinusitis
 - c. Otitis
 - d. Epistaxis
 - e. Ulcerations of nasal mucosa
2. Respiratory symptoms
 - a. Cough
 - b. Hemoptysis
 - c. Dyspnea
 - d. Pleurisy
 - e. Cavitory pneumonitis
3. Arthralgia
 - a. Fever
 - b. Malaise
 - c. Weight loss
4. Ophthalmic symptoms
 - a. Ocular irritation
 - b. Proptosis
 - c. Diplopia

5. Renal symptoms
 - a. Often asymptomatic
 - b. Hematuria is usually microscopic (glomerulonephritis)

D. Describe pertinent clinical features

1. Non-ophthalmic
 - a. Chronic sinusitis
 - b. Mucosal ulcerations of nasopharynx
 - c. Saddle nose deformity and fistula formation
 - d. Lower respiratory cough and hemoptysis
 - e. Glomerulonephritis
 - f. Arthralgia
 - g. Rash
 - h. Less common organ system manifestations include
 - i. Gastrointestinal
 - ii. Cardiac
 - iii. Nervous
2. Limited form
 - a. Upper and lower respiratory tract, no kidney involvement
3. Ophthalmic
 - a. Episcleritis, scleritis
 - b. Marginal ulcerative keratitis
 - c. Retinal vasculitis
 - d. Orbital disease (20-45%)
 - i. More advanced cases can present with:
 - i) Proptosis
 - ii) Inflammation
 - iii) Chemosis
 - iv) Limitation of motility
 - v) Orbital congestion
 - vi) Visual loss
 - e. Dacryoadenitis, nasolacrimal obstruction, nasocutaneous fistula formation
 - f. Eyelid

- i. Periorbital edema, ptosis, lid retraction, or lid nodules

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography)/magnetic resonance imaging scan
 - a. Look for sinus disease, bony destruction, orbital infiltration
 - b. Baseline imaging is important to monitor subsequent recurrence of disease
2. Antineutrophil cytoplasmic antibodies (c-ANCA)
 - a. Most sensitive and specific
3. Perinuclear ANCA (p-ANCA)
 - a. Less sensitive
4. Complete blood count
5. Erythrocyte sedimentation rate
6. Rheumatoid factor
 - a. May be abnormal
7. Serum globulins
 - a. May be elevated
8. Chest x-ray
 - a. May reveal infiltrates or cavitary lesions
9. Blood urea nitrogen (BUN) and creatinine
 - a. Elevation with hematuria and proteinuria
10. The gold standard remains biopsy of the involved tissue

II. Define the risk factors

- A. Fifth decade, male**
- B. Renal failure from glomerular sclerosis is a major cause of death**

III. List the differential diagnosis

- A. Idiopathic orbital inflammation**
- B. Specific causes of orbital inflammation**
 1. Sarcoid
 2. Thyroid eye disease (thyroid orbitopathy)
- C. Fungal infection**
- D. Destructive lesions of the midface**

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

A. Describe medical therapy options

1. Cytotoxic or immunosuppressive agents, usually cyclophosphamide and prednisone
2. Long-term trimethoprim-sulfamethoxazole may suppress disease activity
3. Follow up frequently until disease stabilizes
4. Ensure that non-ophthalmic disease conditions are under treatment

B. Describe surgical therapy options

1. This is a medically treated systemic disorder
2. Complications of the disease, such as lacrimal obstructions, are treated surgically
 - a. Delay lacrimal drainage surgery until disease is under very good control
 - b. Risk of necrosis and fistula formation are high if the disease process is active
 - c. Following cytoplasmic ANCA is a useful way to monitor activity of the disease

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

A. Complications associated with medical therapies vary depending on drug used

VI. Describe disease-related complications

- A. Vision loss**
- B. Diplopia**
- C. Proptosis**
- D. Death**

VII. Describe appropriate patient instructions

- A. Take medications as prescribed**
- B. Return with worsening of symptoms**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 1: Update on General Medicine; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.70, 120-121.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.17.
4. Bhatia A, Yadava U, Goyal JL, et al. Limited Wegener's granulomatosis of the orbit: a case study

and review of literature. *Eye* 2005;19:102-4.

Oculoplastic manifestations of AIDS

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Mechanism of HIV-1 (US) and HIV-2 (Africa)
 - a. Retrovirus
 - b. RNA to DNA by reverse transcriptase
 - c. Infects T cells (CD4+)
 - d. Hallmark of disease is depletion of CD4+ cells
 - e. Leads to profound immunodeficiency
 - f. Causes life-threatening opportunistic infections/tumors (AIDS)
 - g. Clinical syndrome of AIDS
 - i. Recurrent opportunistic infections
 - ii. Unusual neoplasms

B. Define the relevant aspects of epidemiology of the disease

1. HIV to AIDS
 - a. No treatment
 - i. 100% conversion
 - b. Highly active antiretroviral therapy (HAART) for immune reconstitution
 - i. % of conversion has gradually fallen
 - ii. Corresponding decrease in opportunistic infection incidence
 - iii. Median incubation period from HIV to AIDS increased to over 10 years
 - iv. HAART has reduced deaths by 70%

C. List the pertinent elements of the history

1. No documented cases by casual contact
2. Homosexuality-males
3. High risk sexual activity
4. Intravenous drug abuse
5. Blood-borne exposure
6. HIV+ pregnant women

D. Describe pertinent oculoplastic clinical features

1. Infections

- a. Herpes Zoster
 - i. Occurrence in young adults should elevate suspicion of AIDS
 - ii. Treatment
 - i) Immune reconstitution (HAART)
 - ii) IV antivirals
 - iii) Topical antivirals
- b. Molluscum contagiosum
 - i. Etiology
 - i) Viral
 - ii. Appearance
 - i) Small elevation with central umbilication
 - iii. Clinical difference
 - i) Healthy host
 - (i) Variable number of lesions
 - (ii) Unilateral
 - (iii) Involve eyelid
 - (iv) Follicular conjunctivitis
 - ii) AIDS host
 - (i) Numerous lesions
 - (ii) Bilateral
 - (iii) Conjunctivitis may vary with immune status

2. Tumors

- a. Kaposi sarcoma
 - i. Epidemiology
 - i) Occurs in 30% of all AIDS patients
 - ii) 20% of these patients can have aggressive systemic dissemination
 - ii. Etiology
 - i) Herpes virus
 - ii) HIV may play a role
- b. Lymphoma
 - i. Mostly large B cell
 - ii. Usually involve lymph nodes, central nervous system, and lungs

- i) Most common tumor to effect CNS in AIDS
 - iii. Findings
 - i) Strabismus/diplopia
 - ii) Disc swelling/vision loss
 - iv. Diagnosis
 - i) Lymphomatous cells present
 - (i) Spinal fluid by lumbar puncture
 - (ii) Tissue biopsy
 - v. Treatment with multidrug chemotherapy and regional radiotherapy
 - vi. Prognosis
 - i) Poor, usually succumb to disease in 3 months
- c. Conjunctival squamous cell carcinoma
- d. Skin cancer
 - i. Biopsy suspicious lesions
 - i) Basal cell
 - (i) Increased frequency
 - (ii) Similar behavior as normal hosts
 - ii) Squamous cell
 - (i) Increased frequency
 - (ii) More aggressive
 - iii) Melanoma
 - (i) Increased frequency
 - (ii) More aggressive
- 3. Hypertrichosis
- 4. Ptosis
- 5. Facial lipoatrophy

E. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Demonstrate viral specific antibodies
 - a. Enzyme-linked immunosorbent assay (ELISA)
 - b. Western blot

II. Define the risk factors

- A. Unsafe sex-leading cause
- B. Intravenous drug abuse
- C. Blood transfusion
- D. Perinatal transmission from infected mother to child-about

III. List the differential diagnosis

- A. HIV prior to onset of AIDS
- B. Organ transplantation
- C. Chemotherapy for all other oncologic disease
- D. Congenital Immune deficiency

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

A. Describe the natural history, outcome and prognosis

1. Variable predictable stages ultimately lead to death
2. Three stages
 - a. Acute HIV Infection
 - i. Last 1-2 weeks
 - ii. Non specific viral illness
 - b. Asymptomatic with generalized lymphadenopathy
 - i. 2 to more than 10 years
 - ii. CD4 count declines
 - c. Symptomatic disease
 - i. CD4 count decline
 - ii. Opportunistic infections or malignancies or both

B. Describe medical therapy options

1. Immune reconstitution is the goal
2. All anti HIV drugs work through inhibition of reverse transcriptase enzyme
3. Combined use of three or more of these agents is referred to as highly active antiretroviral therapy (HAART)
4. All are associated with toxic side effects
 - a. Bone marrow suppression
 - b. Peripheral neuropathy
 - c. Gastrointestinal irritation

C. Describe surgical therapy options

1. There are no specific surgical treatments for AIDS
2. Surgery for opportunistic infections/neoplasms related to oculoplastic disease
 - a. Functional endoscopic sinus surgery/orbitotomy for sino-orbital fungus
 - b. Skin cancer excision with margins
 - c. Conjunctival squamous cell cancer excision with margins
 - d. Kaposi sarcoma
 - i. Treatment
 - i) Eyelid
 - (i) Radiation
 - (a) Lower recurrence rate
 - (ii) Cryotherapy
 - (iii) Excision
 - (iv) Intralesional chemotherapeutic injections
 - ii) Conjunctiva
 - (i) Surgically excise
 - (ii) Delineate surgical margin
 - e. Molluscum contagiosum excision/cryotherapy
3. Surgical Injury to medical personnel
 - a. Use of universal precautions
 - b. Institutional guidelines to prevent and treat needle sticks

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

- A. Immune reconstitution effects
- B. Recurrence
- C. Blindness
- D. Death
- E. Potential complications from long term HAART treatment

VI. Describe disease-related complications

- A. Opportunistic infection/neoplasm occurrence despite optimal immune reconstitution
- B. Facial lipoatrophy
- C. Blindness

- D. **Death**

VII. Describe appropriate patient instructions

- A. **Continual routine care to monitor immune status**
- B. **Optimize immune reconstitution**
- C. **Safe sex**
- D. **Sun screen/avoid sun**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. Cutaneous malignancy and human immunodeficiency virus disease. Wilkins K, Turner R, Dolev JC, et al., J Am Acad Dermatol, 2006: 54:189-206

Dermoid cysts

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Congenital choristoma - ectodermal rests pinched off at suture lines

B. Define the relevant aspects of epidemiology of the disease

1. Usually recognized in childhood

C. List the pertinent elements of the history

1. Most commonly presenting as a slowly progressive mass in superotemporal or superomedial quadrant of orbit
2. Present at or near birth

D. Describe pertinent clinical features

1. Nontender, well-circumscribed, firm mass
2. Most commonly seen at the lateral orbital rim (frontozygomatic suture)
3. Less commonly occurs at the suture between the frontal bone and the frontal process of the maxilla
4. Occasional presentation with rupture and acute inflammation
5. Rarely a deep dermoid situated at deeper orbital sutures may present later in life. Usual presentation is globe displacement due to a large mass

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan
 - a. Round, smooth borders, well-circumscribed mass
 - b. Bony indentation, occasional dumbbell extension to other side of bone
 - c. Lucent center due to fat content. Layering of contents can occur
2. Magnetic resonance imaging (MRI) can be useful in unusual presentations

II. Define the risk factors

- A. Usually seen in children as an anterior mass. Deeper masses tend to present later

III. List the differential diagnosis

A. Lacrimal gland cyst

B. Meningoencephalocele (medial)

C. Hemangioma (medial)

- D. Neoplasm
- E. Foreign body
- F. Mucocele
- G. Frontal osteoma

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

A. Describe surgical therapy options

1. Surgical resection
 - a. Age 1-5 years to avoid traumatic rupture
 - b. Lesion must be kept intact to avoid spillage of contents into orbit (which may cause inflammation) and increase the chance of recurrence. Controlled decompression may be helpful for larger lesions but important to remove all of the cyst
 - c. Usually adherent to frontozygomatic suture line. Less frequently adherent to the suture line between the frontal bone and the frontal process of the maxillary bone
 - d. No need for ongoing follow-up after complete resection

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

A. Cyst rupture with orbital inflammation. If occurs, copious irrigation at time of surgery

1. Consider postoperative anti-inflammatory medications

VI. Describe disease-related complications

A. Traumatic rupture leading to chronic inflammation

VII. Describe appropriate patient instructions

A. Surgical resection recommended because of continued growth and risk of rupture

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal system; Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Orbital Tumors in Children, Module #9, 1990, p.2.
3. Ahuja R, Azar NF. Orbital dermoids in children. *Semin Ophthalmol* 2006;21:207-11.

Infantile hemangioma (capillary hemangioma)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Abnormal growth of blood vessels with endothelial proliferation, etiology unknown

B. Define the relevant aspects of epidemiology of the disease

1. Infantile, female predominance

C. List the pertinent elements of the history

1. Onset within first months of life, rapid expansion over weeks to months, involution over months to years

D. Describe pertinent clinical features

1. Superficial strawberry surface lesion or deep bluish orbital lesion

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. No imaging for obvious superficial lesion
2. Magnetic resonance imaging (MRI) for deep orbital imaging (extensive lesion rare)
3. Rule out Kasabach-Merritt syndrome: capillary hemangioma associated with thrombocytopenia

II. Define the risk factors

A. Child

1. Not present at birth
2. Usually appear during first weeks to months after birth

B. Grow during first year of life then tend to regress over next 4-5 years

III. List the differential diagnosis

A. Lymphangioma

B. Sturge-Weber syndrome

C. Meningoencephalocele, when mass is medial

D. Rhabdomyosarcoma for lesion with orbital involvement

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

A. Describe medical therapy options

1. Observe for involution unless
 - a. Amblyopia
 - b. Anisometropia
 - c. Strabismus
 - d. Cosmesis may be an indication for treatment, although this must be considered in relation to potential complications
2. Corticosteroids
 - a. Intralesional - risk of corticosteroid particle emboli, eyelid necrosis
 - b. Topical - topical clobetasol propionate has been used to reduce capillary hemangiomas threatening the visual axis with less risk of adrenal suppression
 - c. Systemic - in conjunction with pediatrician
3. Systemic Propranolol - in conjunction with pediatrician

B. Describe surgical therapy options

1. Possible for well-circumscribed orbital lesions

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

- A. Corticosteroid intralesional injection - emboli to retina, central retinal artery, skin necrosis, hypopigmentation, fat atrophy
- B. Corticosteroids have been reported to cause growth retardation and adrenal suppression
- C. Propranolol can cause bradycardia and hypotension, and requires monitoring when initiating treatment

VI. Describe disease-related complications

- A. Amblyopia, strabismus

VII. Describe appropriate patient instructions

- A. Follow corticosteroid instructions as directed. Importance of follow-up visits

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.118-119.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.253-255.
4. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.18.

5. Cruz OA, Zarnegar SR, Myers SE. Treatment of periocular capillary hemangioma with topical clobetasol propionate. *Ophthalmol* 1995;102:2012-5.
6. Léauté-Labrèze C et al. Propranolol for severe hemangiomas of infancy. *N Engl J Med* 2008; 358:2649-2651

Cavernous hemangioma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Benign, encapsulated, lesion with large, dilated vascular spaces filled with red blood cells of unknown cause

B. Define the relevant aspects of epidemiology of the disease

1. Middle aged women
2. Most common benign primary orbital tumor of adults

C. List the pertinent elements of the history

1. Gradual proptosis, mass
2. Gradual vision loss or induced hyperopia
3. Incidental finding on imaging study

D. Describe pertinent clinical features

1. Proptosis
2. Indentation of back of globe
3. Hyperopia
4. Visual field loss
5. Vision loss
6. Diplopia
7. No symptoms

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT)/magnetic resonance imaging (MRI) scan
 - a. Well circumscribed, often intraconal mass with contrast enhancement (mass may extend into extraconal space)

II. Define the risk factors

A. Middle aged women

III. List the differential diagnosis

A. Hemangiopericytoma

B. Schwannoma

- C. Lymphoma
- D. Fibrous histiocytoma
- E. Varix
- F. Deep dermoid cyst
- G. Optic nerve glioma
- H. Optic nerve sheath meningioma
- I. Metastatic tumors and spread from adjacent structures

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

- A. Describe medical therapy options
 - 1. Observation with serial examinations
- B. Describe surgical therapy options
 - 1. Surgical resection via orbitotomy

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

- A. Complications of surgical resection
 - 1. Vision loss
 - 2. Bleeding
 - 3. Infection
 - 4. Ptosis
 - 5. Strabismus
 - 6. Anisocoria
 - 7. Diplopia

VI. Describe disease-related complications

- A. Vision loss from choroidal folds or optic nerve compression is rare
- B. Diplopia with large lesion

VII. Describe appropriate patient instructions

- A. Return prior to surgery if vision loss experienced

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular

Tumors; Section 7: Orbit, Eyelids, and Lacrimal System; Section 12: Retina and Vitreous, 2013-2014.

Rhabdomyosarcoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Unknown

B. Define the relevant aspects of epidemiology of the disease

1. Most common primary orbital malignancy of childhood
2. Onset: age 7-8 years typical, broad range

C. List the pertinent elements of the history

1. Rapid onset of unilateral proptosis
2. Eyelid swelling, conjunctival congestion
3. Pain and decreased vision are uncommon

D. Describe pertinent clinical features

1. Proptosis, often downward and lateral displacement of globe
2. Palpable mass
3. Ptosis
4. Lid edema and erythema, ecchymosis
5. Conjunctival chemosis

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

1. Computed tomography (CT) scan
 - a. Can have bony destruction, circumscribed or infiltrative lesion
 - b. Rapid growth of lesion can indent the globe
2. Magnetic resonance imaging (MRI)
 - a. Relative to the muscle, the mass is isointense on T-1 weighted image and slightly hyperintense on T-2 weighted image
 - b. Densely and homogeneously enhancing after administration of gadolinium
3. Urgent biopsy of lesion
 - a. Sheets and fascicles of rounded and spindled cells with alternating cellular and myxoid regions
4. Metastatic workup
 - a. Chest x-ray (CXR)
 - b. Bone marrow biopsy

- c. Lumbar puncture
- d. Lung parenchyma is the most common site, followed by bone marrow, bone, and locoregional lymph nodes

II. List the differential diagnosis of an orbital mass in a young patient

- A. Orbital cellulitis**
- B. Idiopathic orbital inflammation**
- C. Neuroblastoma**
 - 1. Check vanillylmandelic acid (VMA)
- D. Lymphangioma with hemorrhage**
- E. Ruptured dermoid cyst**
- F. Histiocytic Tumors (Eosinophilic granuloma, Langerhans cell histiocytosis, Letterer-Siwe, Hand-Schüller-Christian, Juvenile xanthogranuloma)**
- G. Sarcoma**

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

- A. Describe medical therapy options**
 - 1. Chemotherapy and radiation therapy after diagnosis is established
- B. Describe surgical therapy options**
 - 1. After surgical biopsy and metastatic workup to stage disease, mainstay of treatment is nonsurgical (chemotherapy and radiation)
 - 2. Under certain treatment protocols, surgical debulking of lesion may be performed after chemotherapy
- C. Combinations of surgery, XRT, and chemotherapy are used fairly successfully in the management of this disease. Patients should be referred to a center engaged in protocol management of these cases such that eligibility can be appropriately determined**

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

- A. Chemotherapeutic side effects**
 - 1. Depends on agents used
- B. Bleeding, vision loss after biopsy**
- C. Radiation**
 - 1. Developmental delay
 - 2. Second tumor

V. Describe disease-related complications

- A. Vision loss**
- B. Death**

VI. Describe appropriate patient instructions

- A. Serious condition which needs to be managed urgently**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.75.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.113.
4. Friedmann AM, Tarbell NJ, Schaefer PW, et al. Case 4-2004: A nine-month-old boy with an orbital rhabdomyosarcoma. *N Engl J Med.* 2004;350:494-502.

Lymphoid hyperplasia and lymphoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Unknown

B. Define the relevant aspects of epidemiology of the disease

1. Fifth or seventh decades

C. List the pertinent elements of the history

1. Progressive, painless proptosis
2. Motility disturbances
3. Firm nodular anterior orbital mass
4. Lacrimal gland mass
5. Subconjunctival salmon patch

D. Describe pertinent clinical features

1. See above
2. Spectrum from benign/reactive (polyclonal) to atypical to malignant (lymphoma)
3. May have associated systemic disease
 - a. Atypical lymphoma hyperplasia - 40% chance at 5 years, and lymphoma - 60% chance at 5 years
4. Mucosa-associated lymphoid tissue (MALT) lymphomas
 - a. Occur in a variety of organs, including the orbit, conjunctiva, salivary glands, skin, thyroid gland, lungs, stomach, and intestine
 - b. Often localized and of indolent clinical behavior.
 - c. The conjunctival form typically presents as a salmon patch
5. Less common forms of orbital lymphoma
 - a. Diffuse large cell
 - b. T-cell lymphoma

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. CT and/or MRI of orbit
 - a. Homogeneous lesions that mold to orbital tissues without displacing them
 - b. No bony involvement
 - c. Can also be infiltrative in appearance

2. Biopsy of lesion
 - a. Fresh tissue should be sent for flow cytometry to look for polyclonality (benign lymphoid hyperplasia) or monoclonality (lymphoma)
 - b. Immunohistochemical studies
 - c. Other cytogenetic/gene rearrangement studies
3. Systemic workup for lymphoma
 - a. Physical exam
 - b. Complete blood count (CBC)
 - c. Chest x-ray (CXR)
 - d. Bone marrow biopsy
 - e. Bone scan
 - f. Computed tomography (CT) of chest, abdomen, pelvis
 - g. Serum electrophoresis

II. Define the risk factors

- A. Fifth or seventh decades

III. List the differential diagnosis

- A. Well-circumscribed orbital lesions
 1. Cavernous hemangioma
 2. Schwannoma
 3. Hemangiopericytoma
 4. Fibrous histiocytoma
- B. Lacrimal gland tumors, inflammation
- C. Plasma cell tumor

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

- A. Describe medical therapy options
 1. Some patients may benefit from specific monoclonal antibody therapy (e.g. rituximab)
 2. Chemotherapy for disseminated systemic lymphoma
- B. Describe surgical and other therapy options
 1. Not treated by surgical excision

2. If systemic workup is negative, most lesions are usually radiated (gold-standard)
 - a. Dose depending on exact diagnosis
 - b. MALT tumors respond very well to radiation therapy
3. Long-term follow-up
 - a. Resolution
 - b. Local or systemic disease
 - c. Radiation-related complications

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

A. Most orbital complications are radiation related

1. Radiation retinopathy or neuritis
 - a. Unusual
 - i. Low doses given for most lymphoid lesions
 - ii. Shielding of globe and nerve
 - b. Occur after one year
2. Exposure keratopathy
 - a. Managed with ocular lubricants
3. Irritated skin
 - a. Usually self- limited

VI. Describe disease-related complications

A. Death from untreated systemic lymphoma

VII. Describe appropriate patient instructions

- A. Ongoing follow-up necessary because of potential for systemic disease**
- B. The most challenging task in managing patients with lymphoproliferative tumors is determining which tumor will remain confined locally and which tumor will have concurrent or eventual development of systemic lymphoma**
- C. Using histology, immunophenotype, location and bilaterality of the tumors as criteria, the following correlations are apparent:**
 1. Histology
 - a. Those tumors with the most benign pathologic features may still be associated with systemic lymphoma
 - b. High grade, large, cleaved-cell lesions have the highest percentage of systemic disease

2. Immunophenotype
 - a. Immunophenotype analysis of the constituted cell populations does not contribute to patient management
 - b. Polyclonal lesions are not necessarily restricted to the ocular adnexa, whereas most monoclonal lesions remain localized
 - c. The concept that polyclonality denotes benign behavior and monoclonality signifies malignancy does not apply rigidly to ocular adnexal lymphoid lesions
3. Currently, no single histologic, immunologic, or molecular genetic parameter can predict with accuracy which lesions are localized or are part of a systemic disease
4. Location
 - a. Proliferation within the substantia propria are less likely to be associated with systemic disease
 - b. Lymphoid tumors that originate exclusively from the preseptal skin without orbital involvement confer a poor prognosis or development of systemic lymphoma
5. Bilaterality
 - a. Patients with bilateral orbital or lacrimal gland lesions do not have a significantly increased incidence of lymphoma in comparison with those who present with strictly unilateral disease

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. Albert DM, Jakobiec FA (eds): Principles and Practice of Ophthalmology. Philadelphia, Saunders WB 2000 ed 2 (vol 4).

Pleomorphic adenoma (benign mixed tumor) of the lacrimal gland

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Unknown

B. Define the relevant aspects of epidemiology of the disease

1. Fourth to fifth decade

C. List the pertinent elements of the history

1. Slowly developing (>1 year) proptosis or globe displacement
2. Nonpainful

D. Describe pertinent clinical features

1. Globe displaced inferiorly and medially

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan
 - a. Well-circumscribed mass with pressure remodeling of lacrimal fossa may indent globe

II. Define the risk factors

A. Fourth to fifth decade

III. List the differential diagnosis

A. Inflammatory lesions of lacrimal gland

1. Idiopathic orbital inflammation
2. Sjögren syndrome
3. Sarcoid
4. Mumps
5. Tuberculosis (TB)

B. Lymphoid lesions

1. Benign reactive lymphoid hyperplasia
2. Atypical lymphoid hyperplasia
3. Lymphoma

C. Epithelial tumors

1. Malignant mixed
2. Adenoid cystic carcinoma

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

A. Describe surgical therapy options (See Surgical approaches to orbital tumors)

1. Complete excision without a preliminary biopsy to avoid tumor spill and potential for recurrence
2. Although controversial, some surgeons may perform small, limited preliminary biopsy prior to excision to confirm diagnosis
3. Follow up on ongoing basis to monitor for recurrence if biopsy performed
 - a. Tumor may undergo malignant transformation

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

A. Recurrence with incomplete tumor excision or tumor spillage at time of surgery

1. Monitor for recurrence, malignant transformation of tumor if recurs

B. Dry eye

VI. Describe disease-related complications

A. Diplopia

B. Ptosis

C. Vision loss

D. Induced refractive error due to compression of the globe

VII. Describe appropriate patient instructions

A. Follow up as recommended

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Adenoid cystic carcinoma of the lacrimal gland

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Unknown

B. Define the relevant aspects of epidemiology of the disease

1. Occurs predominately in the second to fourth decades
2. Less malignant course in children
3. No apparent sex predilection

C. List the pertinent elements of the history. (The historical factors of significance help to differentiate adenoid cystic carcinoma from benign mixed tumor)

1. Pain may be present
2. Progressive (signs and symptoms present < 1 year)
3. Paresthesia (perineural invasion)
4. Ptosis, especially temporally

D. Describe pertinent clinical features

1. Proptosis, axial or inferior and medial globe displacement or globe ptosis
2. Motility disturbance

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan
 - a. Lacrimal fossa mass with bony erosion
 - i. Typically irregular bony destruction
 - b. Possible apical extension into the superior orbital tissue
 - c. Less well circumscribed lesion than benign mixed tumor
2. Magnetic resonance imaging (MRI) scan is useful to define posterior extension of the lesion further

F. Describe the biological behavior of the tumor

1. The neoplasm tends to invade nerves and lymphatic channels resulting in microscopic spread
2. Local recurrence is common
3. A propensity for intracranial extension via the lacrimal nerve through the superior orbital fissure
4. Intracranial involvement is the principal cause of death
5. Bone and lung are potential sites of distant metastases

II. List the differential diagnosis - other causes of lacrimal gland enlargement

A. Inflammatory lesions of lacrimal gland

1. Idiopathic orbital inflammation
2. Sjögren syndrome
3. Sarcoid
4. Infectious dacryoadenitis

B. Lymphoid lesions

1. Benign reactive lymphoid hyperplasia
2. Atypical lymphoid hyperplasia
3. Lymphoma

C. Epithelial tumors

1. Malignant mixed tumor
2. Benign mixed tumor

D. Bony destructive lesions

1. Langerhans cell histiocytosis

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

A. Describe the natural course of the disease and prognosis

1. An actuarial survival rate of less than 50% at 5 years and 20% at 10 years, regardless of treatment regimens

B. Describe surgical therapy options

1. Due to rarity of this tumor, no controlled studies have been performed that prove one treatment plan over another
2. Several principles of treatment are commonly accepted and used.
 - a. Incisional biopsy with examination of permanent sections
 - b. Orbital exenteration with bone removal, after confirmation of specimen
 - i. Although this is the usual treatment, there are no controlled studies to show that radical surgery prolongs life
3. Other considerations
 - a. In children with tumors that lack basaloid features or neural invasion on histopathological examination, a globe-sparing procedure augmented with orbital plaque brachytherapy has been used
 - b. Other protocols

- i. Radiotherapy is usually added to surgical resection
- ii. Cytoreductive intra-arterial chemotherapy in conjunction with exenteration and adjuvant IV chemotherapy and radiation therapy (not to use alone) has been advocated
- iii. Follow up for local recurrence or distant metastasis

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

- A. Deformity associated with exenteration
- B. Local recurrence or intracranial spread
- C. Metastatic spread
 1. Usually bone and lung may occur late in course

V. Describe disease-related complications

- A. Loss of vision
- B. Pain
- C. Death

VI. Describe appropriate patient instructions

- A. Follow up long term for recurrence

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Optic nerve sheath meningioma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Arises in the arachnoid of the optic nerve sheath
2. Enlarge within the subarachnoid space, compressing the optic nerve

B. Define the relevant aspects of epidemiology of the disease

1. Most commonly occur in women (3:2) in the 3rd and 4th decades of life
2. Account for 2% of all orbital tumors and 1% of all intracranial meningiomas
3. More aggressive in younger patients with higher rates of intracranial extension
4. Most are benign

C. List the pertinent elements of the history

1. Typically occurs in middle-aged individuals
2. Onset of symptoms is typically very slow

D. Describe the pertinent clinical features

1. Gradual, painless, unilateral loss of vision
2. Axial proptosis but not usually severe
3. Decreased vision and APD
4. May cause a classic triad of visual loss, optic nerve atrophy and optociliary shunt vessels (30% of cases) or may also cause optic nerve swelling
5. Occasionally bilateral associated with NF
6. Pain and diplopia unusual

E. Describe appropriate testing and evaluation to determine level of functional impairment

1. Neuroimaging (MRI or CT) usually obviates the need for biopsy
2. Enlargement of the optic nerve with perineural enhancement results in the classic tram-track sign
3. Incisional biopsy may be warranted in atypical cases
4. Automated perimetry

II. Define the risk factors

A. Neurofibromatosis-2

B. Sex

C. Age

- D. **Pregnancy may accelerate growth**

III. List the differential diagnosis

- A. **May mimic optic nerve gliomas, inflammatory (sarcoid, Wegener) or neoplastic (lymphoma, metastatic) infiltration of the optic nerve**

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

A. Describe medical therapy options

1. Observation is appropriate in many cases since systemic morbidity and mortality is essentially nil and while most patients experience a progressive loss of vision, the rate and course of visual loss is variable

B. Describe radiation therapy options

1. Stereotactic fractionated radiotherapy has been shown to stabilize or improve vision in more than ½ of patients and is probably the best option for progressive or advanced disease

C. Describe surgical therapy options

1. Surgery is associated with a high rate of visual loss and recurrence and is only reserved for aggressive tumors with intracranial extension
2. Tumors with intracranial extension require a combined approach with neurosurgery

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

- A. **Complication is vision loss with either observation or surgery**
- B. **Radiation optic neuropathy is dose related and overall complication rate, including dry eye, cataracts, radiation retinopathy, etc is at least 10%**
- C. **Complications of intracranial surgery**

VI. Describe disease-related complications

- A. **Tumor growth may lead to continued loss of vision. Tumors arising from intracranial extension are more likely to affect the chiasm and both optic nerves**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. Dutton JJ. Optic nerve sheath meningiomas. *Surv Ophthalmol* 1992;37:167-83.
3. Miller NR. New Concepts in the Diagnosis and Management of Optic Nerve Sheath Meningioma. *J Neuro-Ophthalmol* 2006;26:200-9.

4. Kim, JW et al. Controversies in the Management of Optic Nerve Sheath Meningiomas. *Int Ophthalmol Clin* 2005;45(4):15-23.
5. Berman D, Miller NR: New concepts in the management of optic nerve sheath meningiomas. *Ann Acad Med Singapore* 35:168-174, 2006
6. Carrasco JR, Penne RB: Optic nerve sheath meningiomas and advanced treatment options. *Curr Opin Ophthalmol* 15:406-410, 2004

Sphenoid wing meningioma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. A tumor that arises from the meninges
2. Thought to arise from arachnoid villi cap cells
3. Originate intracranially and extend into the orbit through the bone, superior orbital fissure, or optic canal

B. Define the relevant aspects of epidemiology of the disease

1. Slow growing
2. Account for 20% of all intracranial neoplasms
3. Occur twice as frequently in women as men
4. Incidence increases with age
5. Most are benign

C. List the pertinent elements of the history

1. Typically occurs in middle-aged individuals
2. Slowly progressive, painless, unilateral exophthalmos

D. Describe the pertinent clinical features

1. Proptosis or globe dystopia, temporal fossa mass, eyelid edema, chemosis, diplopia, decreased vision
2. With a significant intracranial component, patients can have headache, swollen optic nerve head, decreased sensation of trigeminal nerve, seizure
3. Reactive hyperostosis of the involved bone and hyperplasia of the associated soft tissues

E. Describe appropriate testing and evaluation to determine level of functional impairment

1. Orbital imaging by CT. Tumors may cause hyperostosis and contain calcifications
2. Orbital imaging by MRI better delineates the dural extension of the tumor. Tumors enhance homogenously with gadolinium and the presence of dural enhancement ("dural tail") helps distinguish meningioma from fibrous dysplasia

II. Define the risk factors

A. Neurofibromatosis-2

B. Sex

C. Age

D. Ionizing radiation

- E. Pregnancy may accelerate growth**

III. List the differential diagnosis

- A. Orbital tumors can radiologically mimic hemangioma, neurofibroma, schwannoma, fibrous histiocytoma, hemangiopericytoma, or lymphoma**

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

A. Describe medical therapy options

- 1. Usually observed until they produce significant proptosis, compressive optic neuropathy, motility impairment, or cerebral edema

B. Describe surgical therapy options

- 1. Subtotal resection through combined intracranial (skull base resection) and orbital approaches to improve the volume-induced compressive effects of the lesion
- 2. Complete resection usually impossible

C. Describe radiotherapy options

- 1. Postoperative radiotherapy to reduce risk of recurrence of residual tumor in cases of subtotal resection
- 2. Definitive stereotactic radiosurgery for meningiomas that cannot be safely removed

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

A. Complications

- 1. Radiation may cause dry eye, optic neuritis, cataract, iritis and radiation retinopathy

B. Prevention

- 1. Low dose radiation is associated with fewer complications

VI. Describe disease-related complications

- A. Tumor growth may lead to continued loss of vision and increased symptoms from mass effect**

VII. Describe appropriate patient instructions

- A. Meningiomas are often slow growing tumors that can be observed until they produce functional consequences**
- B. Continued follow up including repeat imaging and automated perimetry will help guide management decisions**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. Heufelder MJ et al. Reconstructive and ophthalmologic outcomes following resection of sphenoorbital meningiomas. *Ophthal Plast Reconstr Surg*. 2009; 25:223-6.
3. Saeed P et al. Surgical treatment of sphenoorbital meningiomas. *Br J Ophthalmol* 2011; 95:996-1000.

Systemic corticosteroid therapy for orbital disorders

I. List the indications/contraindications

A. Indications

1. Orbital inflammatory disorders
 - a. Specific inflammatory disorders
 - i. Thyroid eye disease (Graves ophthalmopathy)
 - ii. Sarcoidosis
 - iii. Wegener granulomatosis (in conjunction with other agents)
 - iv. Giant cell arteritis
 - v. Connective tissue disorders (systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), dermatomyositis)
 - vi. Some cases of thrombophlebitis
 - vii. Orbital abnormalities associated with allergic aspergillus-related rhinosinusitis
 - b. Nonspecific inflammatory disorders: idiopathic orbital inflammation
2. Lymphoproliferative disorders (corticosteroid therapy not considered definitive but may be adjunctive to radiation therapy/systemic chemotherapy)
 - a. Reactive lymphoid hyperplasia
 - b. Lymphoma
3. Non-lymphoid orbital neoplasms
 - a. Capillary hemangioma - oral or intralesional injection
4. Orbital cellulitis in conjunction with appropriate antibiotics
5. Intra-operatively and post-operatively for orbital surgery

B. Contraindications

1. Relative/absolute contraindications may include medical/ophthalmic disorders potentially complicated by corticosteroid therapy (see below)
2. No definitive therapy for Wegener granulomatosis, lymphoproliferative disorders

II. Describe the pre-procedure/therapy evaluation

A. Diagnosis of underlying disorder for which corticosteroid therapy is recommended

B. Complete ophthalmic history with attention to history of glaucoma/glaucoma suspect status

C. Complete medical history with attention to:

1. High blood pressure (HBP)
2. Diabetes mellitus (DM)
3. Tuberculosis (TB) exposure/active TB (consider TB/anergy screen skin testing)
4. Musculoskeletal disease
5. Neuropsychiatric disease
6. Gastritis/peptic ulcer disease (PUD)/gastroesophageal reflux disease (GERD)
7. Immunodeficiency

D. Comprehensive eye exam

III. List the alternatives to this procedure/therapy (vary with disorder)

- A. Non-steroidal anti-inflammatory drug (NSAID) therapy**
- B. Topical corticosteroids**
- C. Intralesional/local corticosteroid injection**
- D. Radiation therapy**
- E. Corticosteroid-sparing chemotherapy agents (e.g., cyclophosphamide, methotrexate, azathioprine, cyclosporine, mycophenolate)**

IV. Describe treatment options

- A. Frequency of administration**
 1. Daily
 2. Split-dose (twice a day (BID))
 3. Alternate day
- B. Duration of therapy**
 1. Variable
 2. If response to therapy, taper course of treatment as opposed to sudden discontinuation
- C. Concomitant gastroprotective therapy (antacids, H₂ blockers)**
- D. Consider management in conjunction with internist, pediatrician, rheumatologist, oncologist as appropriate**

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

- A. Development/exacerbation of HBP**
- B. Development/exacerbation of hyperglycemia/DM**
- C. Weight gain/Cushingoid habitus**

- D. Gastrointestinal (GI)**
- E. Gastritis**
- F. PUD**
- G. Musculoskeletal**
 - 1. Steroid myopathy
 - 2. Avascular necrosis of femoral head
 - 3. Osteopenia/compression fractures
- H. Neuropsychiatric**
- I. Insomnia**
- J. Anxiety/depression**
- K. Immunosuppression (including re-activation of TB)**
- L. Adrenal suppression**
- M. Cutaneous fragility, easy bruising**
- N. Urinary tract infection**
- O. Thrush**
- P. Pulmonary embolism**
- Q. Deep vein thrombosis**
- R. Steroids for incorrect diagnosis (i.e. tumor, viral or fungal process) can delay proper treatment or exacerbate the true pathologic process**
- S. Potential elevation of intraocular pressure**

VI. Describe the follow-up care

- A. Monitoring of underlying disorder**
- B. Monitoring of medical parameters noted above (possibly in conjunction with internist)**

VII. Describe appropriate patient instructions

- A. Patient self-monitoring regarding symptoms of underlying disorder**
- B. Patient self-monitoring regarding above medical parameters, such as increased urination frequency and thirst, melanotic stool, lack of energy, and morning dizziness and sweating**
- C. Stress importance of not stopping suddenly stopping corticosteroids**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology, 2013-2014.

Surgical approaches to orbital tumors

I. List the indications/contraindications

A. Indications

1. Presence of orbital tumor with associated pain, visual symptoms, proptosis/globe displacement
2. Surgical goals may be diagnostic or therapeutic in nature

B. Contraindications

1. Medical contraindications to surgery
2. Anticoagulation should be discontinued preoperatively, unless emergent

II. Describe the pre-procedure/therapy evaluation

A. Complete ophthalmic/medical history with attention to history of:

1. Malignancy
2. Thyroid disease
3. Aspirin, anti-platelet, or anticoagulant therapy
4. Over-the-counter medications with anti-platelet or anticoagulant effects

B. Comprehensive eye examination

1. Hertel exophthalmometry
2. Globe malposition
3. Afferent pupillary defect
4. Color vision
5. Visual acuity
6. Visual field
7. Extraocular muscle (EOM) motility

C. Potential imaging options

1. Orbit/sinus computed tomography (CT) scan
2. Orbit/brain magnetic resonance imaging (MRI) scan
3. Ultrasound
4. Other studies (e.g., arteriography, venography)

D. Careful informed consent is necessary given potential serious complications from these procedures

III. List the alternatives to this procedure/therapy

- A. Observation**
- B. Diagnostic/therapeutic corticosteroid trial (suspected idiopathic orbital inflammation)**
- C. Fine-needle aspiration biopsy (selected cases)**
- D. Empiric radiation therapy or chemotherapy (in selected patients with disseminated lymphoma, metastatic carcinoma, etc.)**

IV. Describe the instrumentation, anesthesia and technique

A. Surgical spaces of the orbit

- 1. Subperiosteal space
 - a. Between bone and periorbita
- 2. Extraconal space
 - a. Between periorbita and muscle cone
- 3. Intraconal space
 - a. Within muscle cone
- 4. Episcleral space
 - a. Between Tenons capsule and globe

B. Anesthesia options

- 1. Local anesthesia +/- sedation (anterior lesions)
- 2. General anesthesia (generally required for lateral orbitotomy, deeper orbital lesions, orbital surgery in children/uncooperative adults)
- 3. Deep extubation to avoid postoperative hemorrhage

C. Possible use of perioperative antibiotics and systemic steroids

D. Instrumentation

- 1. Standard eyelid surgery instrumentation
- 2. Malleable retractors, neurosurgical instruments (pledgets, dissectors, nerve hooks, scissors)
- 3. Operating microscope +/- surgical loupes/headlight
- 4. Cryoprobe for encapsulated lesions, Allis forceps
- 5. Consider placing drain

E. Specific surgical approaches and surgical considerations

- 1. Anterior approach to superior orbit
 - a. Indicated for tumors in anterior, superior portion of orbit
 - b. Transcutaneous routes

- i. Upper lid crease or, occasionally, coronal incisions
 - ii. Extraperiosteal route
 - i) Dissection anterior to septum to superior orbital rim
 - ii) Incision of periosteum at arcus marginalis provides access to superior orbital rim, anterior portion of orbital roof, and subperiosteal space
 - c. Transconjunctival route
 - i. Access to episcleral, central, peripheral surgical spaces
 - d. Vertical upper eyelid split
 - i. Access to superomedial, extraconal or intraconal tumors
- 2. Anterior approach to inferior orbit
 - a. Indicated for tumors in anterior, inferior portion of orbit
 - b. Transcutaneous routes
 - i. Subciliary incision
 - ii. Extraperiosteal route
 - i) Dissection anterior to septum to inferior orbital rim
 - ii) Incision of periosteum at arcus marginalis provides access to inferior orbital rim, anterior portion of orbital floor, and subperiosteal space
 - iii. Can be combined with lateral canthal incision for swinging eyelid technique
 - c. Transconjunctival route
 - i. Dissection anterior or posterior to orbital septum
 - ii. Access to subperiosteal, extraconal, intraconal, and episcleral spaces
 - iii. Can be combined with lateral canthal incision for swinging eyelid technique
- 3. Anterior approach to medial orbit
 - a. Indicated for masses in medial subperiosteal, extraconal, intraconal, and episcleral spaces
 - b. Transcutaneous route
 - i. Lynch frontoethmoidal incision
 - i) Visible scar
 - ii) Not frequently used anymore
 - c. Transconjunctival route
 - i. Access to subperiosteal, extraconal, intraconal, episcleral spaces
 - ii. Transconjunctival peritomy incision
 - iii. Transcaruncular incision
 - iv. With medial rectus disinsertion +/- lateral wall removal, access to central intraconal

space including optic nerve

- v. May be combined with lateral canthotomy and inferior fornix transconjunctival incision

4. Lateral approach

- a. Indicated for masses in the intraconal, extraconal (lateral to the muscles), and lacrimal gland fossa spaces
- b. Transcutaneous route
 - i. Eyelid crease
 - ii. Lateral canthotomy
 - iii. With or without lateral orbital bone removal
 - iv. With or without lateral rectus traction suture
 - v. Suture +/- plate fixation lateral orbital rim
 - vi. May be combined with transconjunctival/ transcaruncular incision for medial orbital lesions

5. Endoscopic approach

- a. May be useful for inferior, medial, and lateral orbital masses in conjunctions with another surgical approach

6. Transcranial approach

- a. Typically indicated for orbital lesions superior to the optic nerve or those involving orbit, sinuses, and skull base
- b. Typically in conjunction with neurosurgery and/or ENT

F. Structures at risk

1. Superior orbit

- a. Levator - superior rectus complex
- b. Superior oblique
- c. Supraorbital/supratrochlear nerves and vessels
- d. Trochlea
- e. Lacrimal gland

2. Inferior orbit

- a. Inferior oblique
- b. Inferior rectus
- c. Nerve to inferior oblique

3. Medial orbit

- a. Canaliculi
- b. Lacrimal sac

- c. Anterior and posterior ethmoidal nerves and vessels
- d. Supratrochlear and infratrochlear nerves and vessels
- e. Medial canthal tendon
- f. Superior oblique and trochlea
- g. Inferior oblique
- 4. Lateral orbit
 - a. Lacrimal gland
 - b. Temporalis muscle
 - c. Lateral rectus muscle
 - d. Zygomatico temporal and facial sensory nerves

V. List the complications of the procedure/therapy, their prevention and management

A. Visual loss

- 1. Etiology
 - a. Hemorrhage with globe/optic nerve compression
 - b. Direct optic nerve/globe injury
 - c. Interruption of vascular supply to globe or optic nerve (e.g., central retinal artery or posterior ciliary vessel disruption)
- 2. Management directed towards underlying etiology

B. Orbital hemorrhage

- 1. Signs/symptoms
 - a. Severe pain
 - b. Reduced vision, proptosis, increased ecchymosis, increased intraocular pressure, relative afferent pupillary defect, funduscopy evidence of retinal vascular occlusion
- 2. Management
 - a. High dose corticosteroid therapy
 - b. Immediate re-opening of wound with evacuation of hematoma
 - c. If no visual loss and pain is not severe, hemorrhage can be observed

C. Diplopia

- 1. Etiology
 - a. Orbital edema/hemorrhage
 - b. Damage to EOM
 - c. Damage to nerve supply to EOM

- d. Entrapment in procedures where bone removal was performed

2. Management

- a. Management of underlying etiology
 - i. Prism
 - ii. Ocular occlusion
 - iii. Strabismus consultation with possible botulinum toxin therapy, strabismus surgery

D. Ptosis

1. Etiology

- a. Orbital edema/hemorrhage
- b. Mechanical displacement of levator (e.g., after orbital roof removal)
- c. Damage to levator
- d. Damage to superior division of cranial nerve III
- e. Damage to Muller muscle/sympathetic nerve supply

2. Management

- a. Directed towards underlying etiology
- b. May include observation and surgical repair

E. Neurotrophic keratopathy

F. Pupillary abnormalities

- 1. Efferent pupillary defect due to ciliary ganglion trauma
- 2. Anisocoria

G. Intraocular injury

- 1. Hyphema
- 2. Vitreous hemorrhage
- 3. Ruptured globe
- 4. Retinal detachment

H. Dry eye

I. Facial sensory loss

J. Cerebrospinal fluid leak

K. Infection

L. Loss of accommodation

VI. Describe the follow-up care

A. Office examination regarding presence of above complications

- B. Suture removal, if necessary**

VII. Describe appropriate patient instructions

- A. Antibiotic ointment on incision, possible oral antibiotics**
- B. Postoperative steroids**
- C. No/limited nose-blowing for surgery involving entry into sinuses**
- D. Activity limitations**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Zygomatic fractures

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Trauma

B. Define the relevant aspects of epidemiology of the disease

1. Male > female, young > old

C. List the pertinent elements of the history

1. Traumatic incident
2. Pain
3. Periorbital ecchymosis
4. Facial numbness
5. Trismus

D. Describe pertinent clinical features

1. Inferior displacement of lateral canthus
2. Malar flattening
3. Trismus
 - a. Due to coronoid process of mandible articulating with the displaced zygoma upon mouth opening
4. Palpable orbital rim step-off
5. Periorbital ecchymosis
6. Possible infraorbital nerve hypesthesia and other signs of floor fracture
7. Possible malocclusion and other signs of LeFort fracture

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Rule out ruptured globe, traumatic optic neuropathy
2. Computed tomography (CT) scan of orbit and paranasal sinuses with axial and coronal views demonstrating fracture, degree of displacement of bone

II. List the differential diagnosis

- A. Other type of facial fracture (Nasoorbital ethmoid (NOE) fractures, mandibular fractures, mid-facial fractures of the maxilla and pterygoids - LeFort I-III)

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

1. Describe medical therapy options
2. Observe if non-symptomatic, minimally displaced fracture
3. Soft diet

B. Describe surgical therapy options

1. Repair fracture if symptoms do not improve with observation or significant displacement
2. Examine patient and CT scan to diagnose affected buttresses of which up to 5 can be fractured
3. Plan surgical exposure based upon buttresses that need to be exposed
 - a. Upper gingival buccal incision for arch and/or lateral or medial facial buttresses
 - b. Temporal closed reduction approach for zygomatic arch
 - c. Lower eyelid for orbital floor, inferior and/or lateral orbital rim
 - d. Extended lateral canthotomy and/or lateral upper eyelid crease for frontozygomatic suture
4. Exposure, elevation, and repositioning of soft tissue
5. Realignment/reduction of displaced fragments while examining all the buttresses affected
6. Careful alignment of the zygomatic/sphenoid fracture (lateral wall) and the zygomatic/maxillary fracture should be given the highest priority in order to reposition the zygoma with appropriate three dimensional orientation
7. Rigid fixation of fragments working from more stable to less stable
8. Repair of any internal orbital fractures
9. Resuspension of soft tissue and closure of incisions

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

A. Postoperative orbital hemorrhage

1. Remove internal orbital plates, surgically drain if compromising vision

B. Postoperative loss of vision

1. Treatment depends on etiology
 - a. Optic nerve trauma (remove internal orbital plates, high dose corticosteroids)
 - b. Orbital hemorrhage (see above)

C. Infection

1. Remove implants

D. Implant extrusion or migration

1. Reposition, remove, or replace

E. Hypesthesia

F. Eyelid retraction

- G. Ectropion**
- H. Persistent restriction or rectus muscle dysfunction**
- I. Enophthalmos**

V. Describe disease-related complications

- A. Asymmetry of facial bones**
- B. Malar flattening**
- C. Malocclusion and trismus**
- D. Infraorbital nerve hypesthesia**
- E. Sinusitis**

VI. Describe appropriate patient instructions

- A. Wear appropriate face protection for contact sports**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.6.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.171, 184-190, 230-233.
4. AAO, Focal Points: Management of Orbital Trauma, Module #7, 1994, p.2.

Medial orbital wall fractures

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Trauma

B. Define the relevant aspects of epidemiology of the disease

1. Male > female, young > old

C. List the pertinent elements of the history

1. Traumatic event, but may be seemingly incidental in children
2. Pain
3. Periorbital edema, ecchymosis
4. Diplopia
5. Periorbital crepitus
6. Decreased vision
7. Post functional endoscopic sinus surgery (FESS) in which the lamina papyracea was breached

D. Describe pertinent clinical features

1. Periorbital edema, ecchymosis, subcutaneous emphysema
2. Enophthalmos or proptosis
3. Restriction of eye movements
4. Traumatic optic neuropathy
5. Intraocular injuries - traumatic iritis, lens subluxation, angle recession, commotio retinae etc
6. In children with a "trap-door" fracture
 - a. Minimal signs of external trauma
 - b. Limitation of adduction or abduction on fractured side
 - c. Horizontal ductions can elicit nausea, vomiting, and bradycardia due to the oculocardiac reflex

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan (axial and coronal views, bone and soft tissue windows)
 - a. Medial wall fracture
 - b. Opacification of ethmoid sinus
 - c. Possible entrapment of orbital tissue within fracture site
 - d. Rule out medial rectus muscle entrapment

- i. In children with "trap-door" fracture of medial wall with apparently unfractured or minimally displaced medial wall
2. Rule out ruptured globe, traumatic optic neuropathy
3. Can be accompanied by floor fracture

II. List the differential diagnosis

- A. Other facial fractures**
- B. Direct damage to medial rectus muscle and/or its innervation**
- C. Post traumatic fourth or sixth nerve palsy**

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

A. Describe medical therapy options

1. Adults
 - a. May observe for 7-10 days for resolution of motility disturbance resulting from edema, hemorrhage, muscle contusion
 - b. No observation may be appropriate if the fracture is large and associated with enophthalmos
2. Consider antibiotics, nasal decongestants
3. Tell patient to avoid nose blowing

B. Describe surgical therapy options

1. Indications for repair
 - a. Children with "trap-door" medial wall fracture may have severe restriction, which can cause pain and vagal symptoms of nausea, vomiting and bradycardia
 - i. Urgent surgical intervention to release incarcerated and potentially ischemia tissues
 - b. Adults
 - i. Unresolved diplopia in useful fields of gaze suspected to be restrictive in nature or secondary to globe malposition
 - ii. Enophthalmos, medial displacement of globe
 - iii. Large fracture on initial exam indicating that enophthalmos is likely to occur
 - iv. Appropriate timing is based on the clinical scenario, exam findings, and imaging
2. Approach
 - a. Transcaruncular (transconjunctival)
 - i. Careful release of entrapped tissue
 - b. Placement of implant to reproduce the anatomic position of the medial orbital wall

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

A. Postoperative orbital hemorrhage

1. Remove implant, surgically drain if compromising vision

B. Postoperative loss of vision

1. Treatment depends on etiology
 - a. Optic nerve trauma (remove implant, corticosteroids)
 - b. Orbital hemorrhage (see Orbital hemorrhage)

C. Infection

1. Treat with appropriate antibiotics
2. Consider removal of implant

D. Implant extrusion

1. Remove/reposition/replace implant

E. Damage to lacrimal drainage system or medial canthal ligament

1. Stay posterior to posterior lacrimal crest

F. Persistent restriction

1. Treatment depends on etiology
2. Follow-up imaging may help confirm implant position and differentiate continued tissue entrapment from intrinsic muscle dysfunction

G. Enophthalmos

1. Evaluation of implant position by CT scan
2. Consider implant replacement or augmentation of orbital volume

V. Describe disease-related complications

- A. Orbital emphysema and/or hemorrhage with potential for increasing intraorbital pressure and diminishing optic nerve and/or globe perfusion**
- B. Permanent diplopia**
- C. Enophthalmos**
- D. Orbital cellulitis**

VI. Describe appropriate patient instructions

- A. Avoid nose blowing**
- B. Follow-up when instructed**
- C. Avoid swimming or scuba diving or other activities associated with substantial pressure changes**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.198, 232-233.

Orbital floor fractures

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Trauma

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

1. Male > female, young > old

C. List the pertinent elements of the history

1. Traumatic event, but may be seemingly incidental in children
2. Pain, can be worse in upgaze if orbital tissue and/or inferior rectus muscle entrapped
3. Periorbital edema, ecchymosis
4. Diplopia
5. Periorbital crepitus
6. Decreased vision

D. Describe pertinent clinical features

1. Periorbital edema, ecchymosis, subcutaneous emphysema
2. Enophthalmos or proptosis
3. Infraorbital nerve hypesthesia
4. Restriction of eye movements
5. Traumatic optic neuropathy
6. Intraocular injuries - traumatic iritis, lens subluxation, angle recession, commotio retinae etc.
7. In children and young adults with a "trap-door" fracture
 - a. Minimal signs of external trauma
 - b. Double vision on upgaze and less commonly on downgaze
 - c. Supraduction can illicit nausea, vomiting, and bradycardia because of the oculocardiac reflex

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan (axial and coronal views, bone and soft tissue windows)
 - a. Fracture in floor of orbit involving or not involving the inferior orbital rim (blowout fracture)
 - b. Opacification or air/fluid level in maxillary sinus
 - c. Rule out zygomatic fracture
 - d. Rule out inferior rectus muscle entrapment

- i. In children within "trap-door" fracture of floor with apparently unfractured or minimally displaced orbital floor fracture
2. Rule out ruptured globe, traumatic optic neuropathy

II. List the differential diagnosis

- A. Other facial fracture - zygomatic fracture, Le Fort I and II fractures**
- B. Direct damage to the inferior rectus muscle and/or its innervation**
- C. Traumatic superior oblique palsy**

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

A. Describe medical therapy options

1. Adults
 - a. May observe for 7-10 days for resolution of motility disturbance resulting from edema, hemorrhage, rectus muscle contusion
 - b. No observation may be appropriate if the fracture is large and associated with early enophthalmos
2. Consider antibiotics, nasal decongestants
3. Tell patient to avoid nose blowing

B. Describe surgical therapy options

1. Indications for repair
 - a. Children with "trap-door" floor fractures have severe restriction, which can cause pain and vagal symptoms of nausea, vomiting and bradycardia
 - i. urgent surgical intervention is indicated to release entrapped and potentially ischemic tissue
 - b. Adults
 - i. Unresolved diplopia in useful fields of gaze suspected to be restrictive in nature or secondary to globe malposition
 - ii. Enophthalmos >2 mm
 - iii. Large fracture on initial exam (>50% of orbital floor on CT scan) indicating that enophthalmos is likely to occur
 - iv. Concomitant medial orbital wall fracture may increase risk of progressive enophthalmos. Orbital floor fracture repair may be indicated in this setting for small or medium sized defects.
 - v. Appropriate timing is based on the clinical scenario, exam, and imaging
2. Approach

- a. Transconjunctival
 - b. Transcutaneous - less often used
 - c. Without or with lateral canthotomy ("swinging eyelid") for better exposure
- 3. Procedure
 - a. Careful release of entrapped tissue
 - b. Placement of implant to reproduce the anatomic position of the orbital floor
 - c. Posterior aspect of the implant must rest on the posterior ledge

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

A. Postoperative orbital hemorrhage

- 1. Remove implant, surgically drain if compromising vision

B. Postoperative loss of vision

- 1. Treatment depends on etiology
 - a. Optic nerve trauma (remove implant, corticosteroids)
 - b. Orbital hemorrhage (See Orbital hemorrhage)

C. Infection

- 1. Treat with appropriate antibiotics
- 2. Consider removal of implant

D. Implant migration or extrusion

- 1. Remove/reposition/replace implant
- 2. Hypesthesia
- 3. May take up to 6 months to resolve following injury or surgery
- 4. Post-op CT scan to evaluate implant and/or screw placement if hypesthesia not resolving as anticipated
- 5. Eyelid retraction or ectropion
- 6. Avoid excessive traction and cautery of the lower eyelid
- 7. Treatment depends on location of eyelid scarring

E. Persistent restriction

- 1. Treatment depends on etiology
- 2. Consider follow-up imaging studies to evaluate implant position and possibility of residual tissue incarceration.

F. Enophthalmos

- 1. Evaluate for incorrect implant placement with follow-up CT scan

2. Consider implant replacement or augmentation of orbital volume

V. Describe disease related complications

- A. Orbital emphysema, edema, or hemorrhage with potential for increasing intraorbital pressure and diminishing blood flow to the optic nerve or globe.**
- B. Permanent diplopia**
- C. Enophthalmos**
- D. Orbital cellulitis**
- E. Sinusitis**

VI. Describe appropriate patient instructions

- A. Avoid nose blowing**
- B. Follow-up when instructed**
- C. Avoid swimming or scuba diving or other activities associated with substantial changes in pressure**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.52, 80.

Orbital roof fractures

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. High-energy blunt trauma
 - a. Motor vehicle accident, particularly when occupant unrestrained
 - b. Fall
 - c. Assault
 - d. Sports-related injury
2. Relatively low-energy blunt trauma can be seen in young children
 - a. Frontal sinus not yet developed so there is nothing to absorb energy as it extends along roof
 - b. May develop non-displaced linear roof fractures even with minor trauma
 - c. Cribriform plate may be involved
 - d. May present with delayed upper eyelid hematoma
3. Penetrating trauma

B. List the pertinent elements of the history

1. Mechanism of injury
2. Loss of consciousness - may indicate intracranial injury
3. Developing periorbital edema and ecchymosis
4. Change in vision
5. Diplopia
6. Supraorbital nerve hypesthesia
7. Pain

C. Describe pertinent clinical features

1. Decreased visual function (e.g., traumatic optic neuropathy)
 - a. Decreasing visual acuity
 - b. Altered color vision
 - c. Afferent pupillary defect (APD)
2. Decreased ocular motility
3. Blepharoptosis
4. Periorbital edema and ecchymosis

5. Developing upper eyelid hematoma - may indicate intracranial bleeding decompressing into orbit
6. Lacerations of brow or upper eyelid
7. Proptosis
8. Superior orbital rim step-off or frontal bone deformity
9. Cerebrospinal fluid (CSF) rhinorrhea
10. Intraocular injuries
11. Altered mental status or other neurologic deficits

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Visual function
 - a. Visual acuity
 - b. Color vision
 - c. Pupil exam
 - d. Visual fields
 - e. Dilated fundus examination
2. Ocular motility
3. Ptosis
 - a. Mechanical - superior orbital edema/hematoma, CSF accumulation, bone fragments
 - b. Paralytic - oculomotor (III) nerve palsy
 - c. Traumatic - direct damage to levator-superior rectus muscle complex
4. V1 function (relative numbness)
5. Globe displacement (ocular dystopia)
 - a. Exophthalmometry (assess for proptosis)
 - b. Resistance to retropulsion
 - c. Hypoglobus (inferior globe displacement)
6. Computed tomography (CT) scan
 - a. Axial and direct coronal views
 - b. Frontal sinus opacification
 - c. Presence of optic canal or skull base fractures
 - d. Intracranial injury - pneumocephalus, brain laceration, subdural or epidural hematomas
 - e. Orbital hemorrhage (See Orbital hemorrhage)
 - f. Globe injury

II. List the differential diagnosis

- A. Orbital hemorrhage (See Orbital hemorrhage)**
- B. Frontal sinus fracture**
- C. Oculomotor (III) nerve palsy (See Oculomotor (cranial nerve III) palsy)**

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

A. Describe the natural history, outcome and prognosis

1. Many roof fractures do not need repair and do well with few long-term sequelae
2. Traumatic ptosis and extraocular motility disturbance should be observed for at least 6 months for spontaneous recovery
3. Delayed development of an upper eyelid hematoma is an ominous sign indicating possible intracranial bleeding and requires immediate attention

B. Describe medical therapy options

1. Requires multidisciplinary team approach
2. High-dose corticosteroids if vision compromised

C. Describe surgical therapy options

1. The decision as to whether and when to proceed with surgical repair should be made with neurosurgery because many roof fractures involve herniated brain tissue and CSF leaks and a craniotomy may be needed
2. Alternative surgical approach through an existing laceration, brow incision, upper eyelid crease incision, or coronal flap may be used
3. Comminuted roof fractures often require reconstruction using cranial bone grafts or an alloplastic implant
4. Reduction and microplate fixation of supraorbital rim fractures
5. If there is compromise of the nasofrontal duct in a frontal sinus fracture the sinus should be obliterated

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

- A. Complications from use of high-dose corticosteroids - requires multi-disciplinary team approach**
- B. Intracranial bleeding - team approach with neurosurgeon**
- C. Continued CSF leaks**
- D. Persistent ptosis or diplopia - care not to damage levator-superior rectus muscle complex**
- E. Persistent diplopia**
- F. Supraorbital hypesthesia**
- G. Pulsatile proptosis - need proper reconstruction of orbital roof and repair of dural tears**

- H. Frontal sinusitis or mucocele formation - team approach with head and neck surgeon

V. Describe disease-related complications

- A. Intracranial injuries
- B. Pneumocephalus
- C. CSF rhinorrhea
- D. Orbital hemorrhage
- E. Pulsating proptosis
- F. Epistaxis
- G. Blepharoptosis
- H. Extraocular dysmotility and diplopia
- I. Superior orbital fissure or orbital apex syndrome
- J. Traumatic optic neuropathy

VI. Describe appropriate patient instructions

- A. Follow-up as instructed
- B. Report any decrease in vision
- C. Avoid nose blowing
- D. Avoid swimming
- E. Avoid contact sports

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monograph 8. Surgery of the Eyelid, Orbit, and Lacrimal System. Vol. 3, 1995, p.190-194.
3. AAO, Focal Points: Management of Orbital Trauma: Fractures, Hemorrhage, and Traumatic Optic Neuropathy. Vol. 12, No. 7, 1994, p.6-11.
4. Greenwald MJ, Boston D, Pensler JM, Radkowski MA. Orbital roof fractures in childhood. *Ophthalmology* 1989;96:491-496.
5. Hardy SP, Morales L Jr. Orbital roof and supraorbital ridge fractures. In: Holck DEE, Ng JD, eds. *Evaluation and Treatment of Orbital Fractures: A Multidisciplinary Approach*. Philadelphia: Elsevier Saunders; 2006:193-199.

Orbital hemorrhage

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Trauma
2. Iatrogenic - retrobulbar injection
3. Post-surgical eyelid, orbit
4. Systemic coagulopathy
5. Spontaneous bleed from vascular malformation

B. List the pertinent elements of the history

1. Usually acute onset proptosis, eyelid ecchymosis, pain
2. Decreased vision, double vision
3. Precipitating event common, spontaneous hemorrhage can occur

C. Describe pertinent clinical features

1. Proptosis, pain
2. Upper and lower eyelid ecchymosis
3. Hemorrhagic chemosis
4. Decreased ocular motility
5. Possible increased orbital tension, decreased vision, afferent pupil defect (APD), increased intraocular pressure (IOP), central retinal artery occlusion

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Blood workup for patient with no precipitating factor
2. Orbital imaging may be useful in evaluating cause of hemorrhage.

II. Define the risk factors

- A. Trauma, retrobulbar injection, recent surgery, systemic coagulopathy, orbital venous malformations (varix or lymphangioma)

III. List the differential diagnosis

- A. Rhabdomyosarcoma
- B. Orbital cellulitis
- C. Arteriovenous fistula

D. Lymphangioma

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

A. Describe surgical therapy options: emergent surgical intervention may save vision

1. If hemorrhage is postsurgical
 - a. Open incisions and evacuate clot
2. If vision threatening
 - a. Perform lateral canthotomy and lysis of superior and inferior limbs of the lateral canthal tendon
3. If IOP remains elevated and central retinal artery is occluded or pulsating
 - a. Consider orbital decompression
4. Follow very closely until stable

B. Describe medical therapy; primary treatment usually surgical, following medical options may be beneficial

1. Lower IOP
 - a. Acetazolamide
 - b. Mannitol
 - c. Timolol
 - d. Apraclonidine
2. Consider corticosteroids to reduce inflammation
3. Normalize systemic blood pressure
4. Consider discontinuation of anticoagulants

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

A. New incisions, or open incisions, as a result of surgical management of problem

1. Drain placement may be considered
2. Resuture after acute episode has clearly passed

VI. Describe disease-related complications

A. Vision loss

VII. Describe appropriate patient instructions

A. Report any further loss in vision immediately

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.194.
3. AAO, Focal Points: Management of Orbital Trauma, Module #7, 1994, p.6-8.

Dural cavernous fistula and traumatic carotid cavernous fistula

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. An abnormal communication between previously normal arterial and venous blood flow
2. Indirect
 - a. Small meningeal arterial branches communicate with the venous drainage
 - b. Produces low-flow dural cavernous fistula
 - c. Degenerative process in older patient with systemic hypertension and atherosclerosis
 - d. Spontaneous, onset can be insidious
3. Direct
 - a. Typically occurs after basal skull fracture or rupture of carotid cavernous aneurysm
 - b. High flow, carotid cavernous fistula (CCF)
 - c. Diversion of arterialized blood into the venous system causes venous outflow obstruction, which leads to elevated IOP, choroidal effusions, blood in Schlemm canal

B. Define the relevant aspects of epidemiology of the disease

1. Majority of cavernous sinus fistulas are indirect
2. 80% of direct carotid cavernous fistula from trauma

C. List the pertinent elements of the history

1. Headache, orbital pain
2. Orbital bruit
3. Proptosis
4. Red eye
5. Decreased vision, diplopia, ophthalmoplegia
6. Facial pain
7. Transient VIIth nerve palsy

D. Describe pertinent clinical findings

1. Ocular presentation is common
 - a. Proptosis
 - b. Eyelid edema
 - c. Pulsating exophthalmos

- d. Bruit
 - e. Conjunctival/episcleral "corkscrew" blood vessels extending to limbus
 - f. Chemosis
 - g. Exposure keratopathy
 - h. Elevated intraocular pressure
 - i. Blood in Schlemm canal
 - j. Diplopia
 - k. Visual loss - ischemic optic neuropathy (ION), central retinal vein occlusion (CRVO), choroidal effusion or detachment, retinal detachment, optic nerve compression from aneurysmal dilation of cavernous sinus
 - l. Dilation of retinal veins
 - m. Intraretinal/vitreous hemorrhage
 - n. Optic disc swelling
 - o. Angle closure glaucoma - iris/choroid congestion, forward displacement of iris-lens diaphragm
2. Facial pain - Numbness due to compression of gasserian ganglion or compression of the fifth nerve branches
 3. Bleeding
 - a. Fatal intracerebral hemorrhage
 - b. Severe epistaxis

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Can be missed even on good head computed tomography (CT) or magnetic resonance imaging (MRI)
2. Enlarged superior ophthalmic vein on orbital CT or MRI
 - a. Muscles may also be enlarged
 - b. Flow void
3. Ultrasound with Doppler can show reversed, arterialized blood flow in superior ophthalmic vein
4. Computed tomographic angiography and magnetic resonance angiography (MRA) can miss it. Source images of MRA often useful
5. Conventional angiography is gold standard. Done if clinical suspicion high and CT and MRI negative. Otherwise done only when treatment is planned.
6. Can assess muscle size and superior ophthalmic vein size by ultrasonography

II. Define the risk factors

- A. Trauma
- B. Old age or involutional changes
- C. Pregnancy
- D. Hypertension
- E. Elastic tissue diseases
- F. Cavernous sinus aneurysm

III. List the differential diagnosis

- A. Orbital process
 - 1. Thyroid eye disease
 - 2. Myositis
 - 3. Tumor
- B. Cavernous sinus thrombosis or cavernous sinus infiltrative process / neoplasm / infection
- C. Scleritis
- D. Chronic blepharoconjunctivitis

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

- A. Describe medical therapy options
 - 1. Treat corneal exposure
 - 2. Treat elevated intraocular pressure
 - 3. Correct diplopia
 - a. Occlusion
 - b. Prism
 - c. Muscle surgery when stable
- B. Describe surgical therapy options
 - 1. Endovascular treatment of the CCF
 - 2. Vascular access via transcutaneous canalization of the superior ophthalmic vein may be considered
 - 3. Surgical treatment done only if endovascular treatment fails
- C. Indications for treatment
 - 1. Absolute
 - a. Cortical venous drainage
 - b. Progressive optic neuropathy
 - 2. Relative

- a. Intractable ophthalmoplegia
- b. Intractable chemosis/exposure keratopathy

D. In some patients observation is appropriate

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

A. Aneurysm, stroke, recurrence, cranial nerve palsy with diplopia

VI. Describe disease-related complications

- A. Visual loss (corneal exposure, corneal edema, glaucoma, venous stasis retinopathy, vitreous hemorrhage, proliferative retinopathy, optic neuropathy, exudative retinal detachment, choroidal effusion)**
- B. Diplopia**
- C. Intractable pain**
- D. Intracranial hemorrhage**
- E. Severe epistaxis due to engorgement of nasal mucosa or erosion of cavernous sinus into sphenoid sinus**
- F. Death from progressive intracranial arterial insufficiency**
- G. High output cardiac failure**

VII. Describe appropriate patient instructions

- A. Refer to an interventional neuroradiologist or neurosurgeon for diagnosis and treatment**
- B. Ophthalmologic follow-up to manage ocular complications**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-ophthalmology, 2013-2014.
2. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Enucleation

I. List the indications

- A. Primary intraocular malignancies not amenable to alternative treatment**
- B. Selected cases of ocular trauma**
- C. Blind painful eye**
- D. Severe infection - corneal ulceration/perforation/panophthalmitis**

II. List the alternatives to this procedure/therapy

- A. Observation with medical therapy (for blind painful eye without neoplasm)**
- B. Other management options including below may be considered for individual patients on a case-by-case basis**
 - 1. Evisceration
 - 2. Fitting of a cosmetic scleral shell prosthesis
 - 3. Medical therapy: retrobulbar alcohol injection, retrobulbar chlorpromazine, atropine/prednisolone ophthalmic drops

III. Describe the pre-procedure/therapy evaluation

- A. Complete ophthalmic/medical history with attention to history of acetylsalicylic acid (ASA)/anti-platelet/anti-coagulant/anti-inflammatory therapy**
- B. Comprehensive eye evaluation**
- C. Possible second opinion to confirm need for enucleation**
- D. B-scan ultrasonography if media opaque to help**
- E. rule out intraocular malignancy**
- F. Potential additional imaging options (on case-by-case basis)**
 - 1. Orbit computed tomography (CT)/ magnetic resonance imaging (MRI) scan

IV. Describe the instrumentation, anesthesia and technique

- A. Anesthesia options**
 - 1. Monitored anesthesia care (MAC) anesthesia: retrobulbar anesthesia with sedation
 - 2. General anesthesia
 - 3. Consider retrobulbar bupivacaine injection prior to and at end of the procedure
- B. Technique options**

1. May want to consider lateral canthoplasty in certain cases (such as retinoblastoma in which a long segment of optic nerve to increase complete resection).
2. 360 degree conjunctival limbal peritomy
3. Isolation of extraocular muscles (EOMs) with placement on sutures/muscle transection (depending on technique)
4. Optic nerve transection
5. Hemostasis with cautery/clamping
6. Orbital implant type with or without wrapping material
 - a. Implant material: solid and porous
 - b. Style, shape, size; spheres, tunnels etc.
7. Attachment of EOM to implant
8. Tenon's capsule closure (one or two layers)
9. Conjunctival closure without tension
10. Consider conformer placement, appropriate size important
11. Possible temporary intermarginal suture placement
12. Dressing application (tight patch)
13. Consideration for orbital implant, peg or other coupling device

V. List the complications of the procedure/therapy, their prevention and management

A. Intraoperative complications

1. Removal of wrong eye
 - a. Confirm operative site by discussion with patient in preoperative area, marking of skin around eye, examination of eye, review of chart, and ophthalmoscopic examination of eye in operating room (OR), if appropriate
 - b. Adhere to the operating room "time-out" protocol
2. Intraoperative hemorrhage
3. EOM injury
4. Levator injury

B. Postoperative complications and management

1. Deep superior sulcus
 - a. Etiology - inadequate orbital volume or implant migration
 - i. Replacement volume of implant/prosthesis less than that of globe
 - ii. Orbital fat atrophy

- b. Management
 - i. Horizontal lid shortening if excess lower lid laxity
 - ii. Subperiosteal implant placement, orbital volume augmentation ("wedge" implant)
 - iii. Orbital implant replacement
 - iv. Volume augmentation (i.e. fat or filler)
- 2. Fornix contracture
 - a. Prevention
 - i. Preservation of conjunctiva during surgery
 - ii. Limited conjunctival dissection
 - iii. Conformer use
 - b. Management
 - i. Prosthesis modification
 - ii. Topical anti-inflammatory therapy
 - iii. Fornix/socket reconstruction with mucous membrane/amniotic membrane grafts
- 3. Socket contracture
 - a. Etiology
 - i. Implant extrusion
 - ii. Accidental trauma
 - iii. Surgical trauma
 - i) Prior to enucleation
 - ii) During enucleation
 - iii) Socket procedures
 - iv. Chemical burns
 - v. Prior radiation therapy
 - b. Management
 - i. Prosthesis modification
 - ii. Topical anti-inflammatory therapy
 - iii. Socket reconstruction
 - i) Dermis-fat graft
 - ii) Mucous membrane graft
- 4. Orbital implant exposure/extrusion
 - a. Etiology

- i. Implant size, shape and material may increase risk of exposure/extrusion
 - ii. Anterior placement of implant
 - iii. Inadequate Tenon's capsule closure
 - iv. Closure of conjunctiva under tension
 - v. Poor wound healing
 - vi. Infection
 - vii. Poor conformer/prosthesis fit
 - b. Management
 - i. Observation/topical antibiotics/spontaneous closure (small defects)
 - ii. Tissue grafts placement (donor sclera, allogenic dermis, autogenous grafts (hard palate, dermis fat, temporalis fascia, tarsoconjunctival grafts or flaps)
 - iii. Implant removal/replacement
5. Ectropion
- a. Due to increased lid laxity, skin cicatrix formation
 - b. Management
 - i. Medial +/- lateral canthal tendon tightening
 - ii. Skin graft placement
6. Entropion
- a. Due to socket/fornix contracture
 - b. Management
 - i. Prosthesis modification
 - ii. Marginal rotation surgery
 - iii. Fornix/socket reconstruction, mucous membrane grafting
7. Ptosis
- a. Etiology
 - i. Damage to the levator muscle or nerve supply
 - ii. Levator aponeurosis disinsertion
 - iii. Superotemporal implant migration
 - iv. Superior fornix scarring
 - b. Management
 - i. Prosthesis modification
 - ii. Levator surgery
 - iii. Frontalis suspension

8. Poor prosthesis motility
 - a. Etiology
 - i. Lost, restricted or paretic muscles
 - ii. Inadequate conjunctiva
 - iii. Poor prosthesis fit
 - iv. Inadequate implant volume or implant migration
 - b. Management
 - i. Reattachment of muscles
 - ii. Implant exchange or orbital volume enhancement
 - iii. Conjunctival augmentation or restore fornices
 - iv. New prosthesis
 - v. Peg placement
9. Giant papillary conjunctivitis-chronic mucous discharge
 - a. Prosthesis hygiene
 - b. Topical corticosteroids, nonsteroidal anti-inflammatory agents and mast cell stabilizers
 - c. Lubrication
 - d. New prosthesis if other medical measures fail
10. Infection

VI. Describe the follow-up care

- A. Dressing/tarsorrhaphy suture removal**
- B. Prosthesis fitting**
- C. Polycarbonate lens/safety frame prescription for normal eye**

VII. Describe appropriate patient instructions

- A. Prosthesis hygiene**
- B. Topical lubricants**
- C. Periodic prosthesis inspection/polishing/replacement at ocularist**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995.

3. AAO, Focal Points: The Anophthalmic Socket, Module #9, 1992.
4. Jordan DR, Klapper SR. Surgical techniques in enucleation: the role of various types of implants and the efficacy of pegged and nonpegged approaches. *Int Ophthalmol Clin* 2006;46:109-32.
5. Moshfeghi DM, Moshfeghi AA, Finger PT. Enucleation. *Surv Ophthalmol* 2000;44:277-301.
6. Rubin PA. Enucleation, evisceration, and exenteration. *Curr Opin Ophthalmol* 1993;4:39-48.
7. AAO, Focal Points: Choroidal Melanoma Update, Module #4, 2005.

Evisceration

I. List the indications/contraindications

A. Indications

1. Blind painful eyes in which intraocular malignancy has been excluded
2. May be preferable to enucleation in patients in whom
 - a. General anesthesia is contraindicated and/or shorter, technically simpler procedure preferred
 - b. Bleeding diathesis is present
 - c. Maximal cosmesis is a priority
 - d. Conjunctival scarring is present to decrease risk of further socket contracture

B. Contraindications

1. Possible presence of intraocular malignancy
2. Severe phthisis bulbi
3. Patient concern re: risk of sympathetic ophthalmia
4. Other management options including below may be considered for individual patients on a case-by-case basis:
 - a. Enucleation
 - b. Retrobulbar alcohol/chlorpromazine (Thorazine®) injection, topical therapy (atropine/prednisolone)
 - c. Fitting of a cosmetic scleral shell prosthesis
5. Physician concern regarding patient's immunosuppression or medical status that would preclude the use of standard immunomodulatory therapy for sympathetic ophthalmia in the unlikely event it would occur

II. Describe the pre-procedure/therapy evaluation

- A. Complete ophthalmic/medical history with attention to history of acetylsalicylic acid (ASA)/anti-platelet/anti-coagulant/anti-inflammatory therapy, repaired ruptured globe or intraocular procedures
- B. Comprehensive eye evaluation
- C. Possible second opinion to confirm need for evisceration
- D. B-scan ultrasonography if media opaque to help rule out intraocular malignancy

III. List the alternatives to this procedure/therapy

- A. Observation with medical therapy (for blind painful eye without neoplasm)**
- B. Retrobulbar alcohol injection**
- C. Enucleation**
- D. Retrobulbar chlorpromazine (Thorazine®) or atropine/prednisolone ophthalmic drops**
- E. Conjunctival flaps**
- F. Scleral shell prosthesis**

IV. Describe the instrumentation, anesthesia and technique

A. Anesthesia options

- 1. Monitored anesthesia care (MAC) anesthesia: retrobulbar anesthesia with sedation
- 2. General anesthesia
- 3. Consider retrobulbar bupivacaine injection prior to and at end of procedure

B. Technique options

- 1. 360 degree peritomy for 5 mm from limbus
- 2. Limbal incision
 - a. Corneal excision +/- excision of scleral triangles (3 o'clock and 9 o'clock) to facilitate closure. Alternatively, scleral incision may be made under the superior rectus and the cornea can be preserved
 - b. If the cornea is preserved, the corneal epithelium and endothelium should be removed
- 3. Efficient removal of all uveal content; alcohol swab of scleral shell; cautery hemostasis
- 4. Placement of implant within scleral envelope, posterior sclerotomy helpful (alternatively, some surgeons place implant posterior to scleral flaps following posterior scleral incision placement)
- 5. Scleral and conjunctival layered wound closure without tension
- 6. Conformer placement, correct sizing important
- 7. Temporary intermarginal tarsorrhaphy suture
- 8. Dressing placement (tight patch)

V. List the complications of the procedure/therapy, their prevention and management

A. Intraoperative complications

- 1. Evisceration of wrong eye
 - a. Confirm operative site by discussion with patient in preoperative area, marking of skin around eye, examination of eye, review of chart, and ophthalmoscopic examination of eye in operating room (OR), if appropriate
 - b. Intraoperative hemorrhage

B. Postoperative complications and management

1. Deep superior sulcus
 - a. Etiology - inadequate orbital volume
 - i. Replacement volume of implant/prosthesis less than that of globe
 - ii. Orbital fat atrophy
 - b. Management
 - i. Horizontal lid shortening if excess lower lid laxity
 - ii. Subperiosteal implant placement ("wedge" implant)
 - iii. Orbital implant replacement
2. Fornix contracture
 - a. Prevention
 - i. Preservation of conjunctiva during surgery
 - ii. Limited conjunctival dissection
 - iii. Conformer use
 - b. Management
 - i. Prosthesis modification
 - ii. Topical anti-inflammatory therapy
 - iii. Fornix/socket reconstruction with mucous membrane/amniotic membrane grafts
3. Socket contracture
 - a. Etiology
 - i. Implant extrusion
 - ii. Accidental trauma
 - iii. Surgical trauma
 - i) Prior to evisceration
 - ii) During evisceration
 - iii) Socket procedures
 - iv. Chemical burns
 - v. Prior radiation therapy
 - b. Management
 - i. Prosthesis modification
 - ii. Topical anti-inflammatory therapy
 - iii. Socket reconstruction

- i) Dermis-fat graft
 - ii) Mucous membrane graft
- 4. Orbital implant exposure/extrusion
 - a. Etiology
 - i. Placement of excessively large implant
 - ii. Anterior placement of implant
 - iii. Inadequate Tenon's capsule closure; wound closed under tension
 - iv. Poor wound healing
 - v. Infection
 - vi. Poor conformer/prosthesis fit
 - b. Management
 - i. Observation/topical antibiotics/spontaneous closure (small defects)
 - ii. Tissue grafts placement (donor sclera, allogenic dermis, autogenous grafts (hard palate, dermis fat, temporalis fascia, tarsoconjunctival grafts))
 - iii. Implant removal/replacement
- 5. Ectropion
 - a. Due to increased lid laxity, skin cicatrix formation
 - b. Management
 - i. Medial +/- lateral canthal tendon tightening
 - ii. Skin graft placement
- 6. Entropion
 - a. Due to socket/fornix contracture
 - b. Management
 - i. Prosthesis modification
 - ii. Marginal rotation surgery
 - iii. Fornix/socket reconstruction, mucous membrane grafting
- 7. Ptosis
 - a. Etiology
 - i. Damage to the levator muscle or nerve supply
 - ii. Levator aponeurosis disinsertion
 - iii. Superotemporal implant migration
 - iv. Superior fornix scarring
 - b. Management

- i. Prosthesis modification
 - ii. Levator surgery
 - iii. Frontalis suspension
- 8. Sympathetic ophthalmia
 - a. Extremely low risk after evisceration
 - b. Immunomodulatory therapy; removal of inciting eye may/may not be helpful

VI. Describe the follow-up care

- A. Dressing removal**
- B. Prosthesis fitting**
- C. Polycarbonate lens/safety frame prescription for normal eye**

VII. Describe appropriate patient instructions

- A. Prosthesis hygiene**
- B. Topical lubricants**
- C. Periodic prosthesis inspection/polishing/replacement at ocularist**

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. 3, 1995, p. 99-107, 148, 150.
- 3. Rubin PA. Enucleation, evisceration, and exenteration. *Curr Opin Ophthalmol* 1993;4:39-48.

Exenteration

I. List the indications

- A. Selected orbital malignancies (in absence of distant metastatic disease or for palliation)**
 - 1. Malignant epithelial lacrimal gland tumors
 - 2. Malignancies arising from sinuses, face, eyelids, conjunctiva, intracranial space
 - 3. Sarcomas unresponsive to alternative treatment
- B. Selected orbital infections**
 - 1. Phycomycosis
 - 2. Rare severe bacterial infection (often in immunocompromised patient) unresponsive to antibiotic therapy/less aggressive surgical resection
- C. Rarely severe orbital inflammation with visual loss and severe pain unresponsive to alternative therapy**
- D. Rare severe cases of socket contraction precluding prosthesis use to allow fabrication of orbital prosthesis**

II. List the alternatives to this procedure/therapy

- A. Medical therapy (depending on etiology)**
 - 1. Antimicrobial therapy
 - 2. Antineoplastic therapy
- B. Radiation therapy**
- C. More limited surgical therapy**

III. Describe the pre-procedure/therapy evaluation

- A. Complete ophthalmic/medical history with attention to history of acetylsalicylic acid (ASA)/anti-platelet/anti-coagulant/anti-inflammatory therapy**
- B. Comprehensive evaluation of the fellow eye**
- C. Second opinion to confirm need for exenteration**
- D. Imaging study options (orbit, head computed tomography scan (CT) +/- magnetic resonance imaging (MRI) scan**
- E. Metastatic disease evaluation**
- F. Possible consultations (internal medicine, hematology-oncology, infectious disease, radiation oncology)**
- G. Consultation with ear, nose and throat (ENT) service for a thorough head and neck evaluation for tumors with potential for regional metastases**

IV. Describe the instrumentation, anesthesia and technique

A. Anesthesia options: general anesthesia

B. Technique options

1. Extent of resection
 - a. Subtotal exenteration - periorbital and some or all of eyelids remain
 - b. Total exenteration - all intraorbital soft tissues with or without eyelids
 - c. Extended exenteration - all intraorbital soft tissues with orbital bone and sinus resection
2. Reconstructive options
 - a. Granulation
 - b. Split-thickness skin graft
 - c. Temporalis muscle transfer +/- skin graft
 - d. Pericranial flap +/- skin graft
 - e. Microvascular free flap
3. Rehabilitation options
 - a. Black patch
 - b. Adhesive mounted orbital prosthesis
 - c. Eyeglass mounted orbital prosthesis
 - d. Osseo-integration with magnetically attached orbital prosthesis
 - e. In selected patients undergoing subtotal exenteration, ocular prosthesis fitting

V. List the complications of the procedure/therapy, their prevention and management

A. Intraoperative complications

1. Removal of wrong eye
 - a. Prevention
 - i. Confirm operative site by discussion with patient in preoperative area
 - ii. Marking of skin around eye
 - iii. Examination of eye, review of chart, and ophthalmoscopic examination of eye in operating room (OR), if appropriate
2. Intraoperative hemorrhage
3. Entry into sinuses or intracranial space
4. Cerebrospinal fluid (CSF) leak

B. Postoperative complications and management

1. Delayed/incomplete socket healing
 - a. Etiology
 - i. Unknown
 - ii. Prior radiation therapy/chemotherapy
 - iii. Diabetes mellitus
 - iv. Persistent tumor/infection
 - b. Management
 - i. Medical therapy
 - ii. Surgical debridement
 - iii. Graft/flap placement
 - iv. Possible hyperbaric oxygen
2. Tumor/infection persistence/recurrence
 - a. Etiology
 - i. Unknown
 - ii. Persistent immunosuppression/metabolic acidosis
 - iii. Large reconstructive flaps may mask tumor recurrence and delay diagnosis of recurrence
 - b. Management
 - i. Correction of underlying medical abnormalities
 - ii. Medical treatment
 - iii. Possible radiation therapy
3. Osteomyelitis
 - a. Etiology
 - i. Unknown
 - ii. Persistent bone exposure due to delayed healing
 - b. Management
 - i. Medical therapy
 - ii. Surgical debridement
 - iii. Graft/flap placement
 - iv. Possible hyperbaric oxygen
4. Sino-orbital fistula
 - a. Etiology

- i. Pre-existing thin/dehiscent bone
 - ii. Surgical trauma
 - b. Management
 - i. Observation if small/asymptomatic, ENT consultation
 - ii. Surgical closure if indicated
- 5. CSF leak
 - a. Etiology
 - i. Pre-existing thin/dehiscent bone
 - ii. Surgical trauma
 - b. Management
 - i. Antibiotic coverage
 - ii. Neurosurgical consultation
 - iii. Surgical closure
- 6. Donor site morbidity
 - a. Etiology
 - i. Skin donor site infection

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit, and Lacrimal System, Vol. 1, 1993.
3. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995.
4. Moshfeghi DM, Moshfeghi AA, Finger PT. Enucleation. *Surv Ophthalmol* 2000;44:277-301.
5. Rubin PA. Enucleation, evisceration, and exenteration. *Curr Opin Ophthalmol* 1993;4:39-48.

Orbital implant options

I. List the indications/contraindications

A. Indications

1. Volume replacement after enucleation/evisceration
2. Facilitate prosthesis fitting after enucleation/evisceration
3. Facilitate prosthesis movement after enucleation/evisceration
4. Facilitate bony orbital growth (following enucleation in childhood)

B. Contraindications

1. Porous implants may be contraindicated in setting of infection or prior/planned radiation therapy
2. Placement of satisfactory alloplastic implant may be difficult in setting of small pediatric orbit; dermis-fat graft may be preferable in this setting
3. Implant placement may not be necessary and may lead to additional potential complications when performed in patients in whom prosthesis use is not contemplated

II. Implant options, advantages, and limitations

A. Alloplastic implants

1. Non-porous implants
 - a. Advantages
 - i. Potentially lower exposure/extrusion rates
 - ii. Less expensive
 - b. Disadvantage
 - i. May provide less motility than porous implants, particularly if drilling and peg placement performed
 - ii. Migration may be more likely
2. Porous implants
 - a. Advantages
 - i. May increase implant stability with successful fibrovascular ingrowth
 - ii. May allow peg placement to enhance motility
 - b. Disadvantages
 - i. May be associated with higher exposure/extrusion rate (varies with implant, wrapping material, and surgical technique)
 - ii. Need for implant wraps with some porous implants

- iii. Expense
- iv. Peg-related complications
 - i) Peg migration/prolapse/poor coupling to prosthesis
 - ii) Granuloma formation
 - iii) Infection
 - iv) Implant exposure, extrusion
 - v) Peg placement not indicated in patients that are blind in both eyes.
 - vi) Nystagmus may be a contraindication to peg

B. Autogenous implant (dermis-fat graft)

1. Advantages
 - a. No extrusion
 - b. May allow augmentation of mucosal lining of socket
 - c. May grow in pediatric patients with stimulation of bony orbital growth
2. Disadvantages
 - a. Second surgical site
 - b. Fat atrophy with graft shrinkage in adult patients
 - c. Graft hypertrophy possibly necessitating graft debunking in pediatric patients
 - d. Possible retention of hair bearing tissue
 - e. Conjunctival cysts
3. Surgical Technique
 - a. Selection of donor site
 - i. Periumbilical
 - ii. Left lower quadrant
 - iii. Superior buttock
 - b. Removal of epidermis
 - c. Incision of dermis
 - d. Excision of attached fat
 - e. Hemostasis
 - f. Placed into socket fat side deep
 - g. Secure recti muscles to graft periphery
 - h. Secure conjunctiva to dermis
 - i. Conformer or symblepharon ring placed +/- lid tarsorrhaphy sutures

- j. Harvest site closed with deep and skin sutures

III. Prosthesis considerations

A. Roles of prosthesis

1. Cosmesis
2. Preservation of conjunctival fornices (avoid removal of conformer/prosthesis for > 24 hours)

B. Prosthesis fitting generally 4-8 weeks after enucleation/evisceration

C. Prosthesis related inflammation

1. Etiology
 - a. Mechanical irritation of socket tissues
 - b. Poor prosthesis fit
 - c. Dry anophthalmic socket
 - d. Poor socket/prosthesis hygiene
 - e. Giant papillary conjunctivitis (GPC)
2. Management
 - a. Prosthesis hygiene (cleansing with dilute baby shampoo, contact lens solution, dish detergent, enzymatic solutions or ultrasonic cleaner may also be used to reduce protein deposits)
 - b. Topical lubricants
 - c. Mast cell inhibitor treatment; limited topical corticosteroid treatment
 - d. Prosthesis polishing/modification/replacement

D. Prosthesis related eyelid problems including increased lid laxity and/or ectropion leading to prosthesis instability

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.34.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p. 86-87, 90-99, 102-104, 106-107, 119-129, 149-154, 196, 201, 217, 221, 233-244.
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6. Chalasani R, Poole-Warren L, Conway RM, et al. Porous orbital implants in enucleation: a systematic review. *Surv Ophthalmol* 2007;52:145-55.

7. Custer PL, Trinkaus KM. Porous implant exposure: Incidence, management, and morbidity. *Ophthal Plast Reconstr Surg* 2007;23:1-7.
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9. Jordan DR, Klapper SR. Surgical techniques in enucleation: the role of various types of implants and the efficacy of pegged and nonpegged approaches. *Int Ophthalmol Clin* 2006;46:109-32.
10. Heher KL, Katowitz JA, Low JE. Unilateral Dermis-fat graft implantation in the pediatric orbit. *Ophthal Plast Reconstr Surg* 1998;14:81-88.
11. Migliori ME, Putterman AM. The domed dermis-fat graft orbital implant. *Ophthal Plast Reconstr Surg* 1991;7:23-30.

Socket contracture

I. List the indications and contraindications for surgical treatment

- A. Indications for socket contracture include inability to retain a prosthesis**
- B. Contraindications can include history of poor wound healing, burns, severe vascular disease**

II. Describe the pre-procedure evaluation

- A. Medical history of radiation**
- B. History of previous inability to wear prosthesis or extrusion of implant**
- C. Number of previous socket or lid surgeries**

III. List the alternatives to this procedure

- A. Prevent contraction by preserving conjunctiva and limiting dissection in the fornix which helps to avoid the need for reconstruction**
- B. Continuous wear of a conformer or prosthesis helps to prevent contraction**
- C. Amniotic membrane grafting is an alternative to mucous membrane grafting**
- D. Dermis fat grafting is an adjunct to socket reconstruction**
- E. Facial prosthesis**
- F. Black patch or opaque lens in spectacles**
- G. Vascularized free flaps**
 - 1. Radial-free forearm grafts**
 - 2. Thoracodorsal artery perforator adipose flap**
- H. Bone expansion osteotomy**
- I. Serial expanders such as hydrogel expanders or inflatable expanders**

IV. Describe the technique

- A. Technique**
 - 1. Excision scar tissue**
 - 2. Determine tissue deficit**
 - 3. Mucous membrane graft**
 - 4. Conformer may need to be sutured in place or temporary tarsorrhaphy placed over conformer**
 - 5. Fornix sutures placed though lids to maintain fornix**

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

A. Intraoperative

1. Avoid Stensen duct (adjacent to upper 2nd molar tooth) when harvesting buccal mucosa

B. Postoperative

1. Maintain fornix with conformer - may need to leave sutured in place for weeks

C. Prevention of complications

1. Avoid overly aggressive socket dissection
2. Avoid sacrificing conjunctiva
3. Stress need to comply with conformer and prosthesis wear

D. Management of complications

1. Topical, oral, or systemic antibiotics for infection
2. Repair of wound dehiscence
3. Replacement of lost conformer
4. Pressure patch any bleeding

VI. Describe the follow-up care

A. Evaluate wounds 7-10 days

VII. Describe appropriate patient instructions (postoperative care)

A. Leave conformer in place prescribed length of time

B. Remove pressure patch as instructed

C. Topical or systemic antibiotics as instructed

D. Return for follow up as instructed

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Congenital socket malformations

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Anophthalmos results when the primary optic vesicle fails to grow out from the cerebral vesicle (rare)
2. A microphthalmic globe results when the eye is abnormally small
3. Microphthalmia with cyst results when the embryonic choroidal fissure fails to close

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

1. Generally sporadic but various inherited patterns may exist
2. List the pertinent elements of the history
3. Defect present at birth
4. An associated cyst may enlarge with time

C. Describe pertinent clinical features

1. Isolated ocular defect or associated with intracranial or systemic defects
2. Generally unilateral but may be bilateral
3. The microphthalmic globe often has colobomatous defects
4. The opposite globe may have colobomatous defects
5. Cyst of rudimentary sclera lined rudimentary neuroglia which communicates through channel into vitreous cavity

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. External ocular examination may be characteristic
2. Imaging with ultrasound, computed tomography (CT) or magnetic resonance imaging (MRI) will demonstrate characteristic findings
3. All children with microphthalmia have hypoplastic orbits.

II. Define the risk factors

A. Generally sporadic

B. Genetic associations with microphthalmos include trisomy (usually 13),

C. Microphthalmia may occur in association with Goldenhar and other syndromes,

D. Maternal infection, radiation or exposure to drugs such as retinoic acid may be associated with microphthalmia

III. List the differential diagnosis

- A. Other cystic lesion of orbit presenting at birth**
 - 1. Teratoma
 - 2. Lymphangioma
 - 3. Encephalocele
 - 4. Epithelial choristoma (dermoid or epidermoid cyst)
 - 5. Orbital lipodermoid

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

- A. Observation appropriate with or without scleral shell**
- B. Treatment of microphthalmic orbit begins shortly after birth and consists of socket expansion with progressively larger custom conformers/prosthesis. In severe bony asymmetry intraorbital tissue expanders may be progressively inflated to enlarge the hypoplastic orbit.**
- C. Simple drainage of cyst sometimes effective in addressing orbital complications resulting from excess cyst volume**
- D. Apparent deformity may necessitate removal of the cyst with or without removal of the microphthalmic eye**
- E. If globe removed appropriate socket management to stimulate orbit growth needs to be considered**

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

- A. Recurrence generally seen with simple drainage**
- B. Removal of the cyst with or without enucleation of attached globe may diminish orbital and eyelid growth, necessitating additional surgery to expand eyelids and socket**

VI. Describe disease-related complications

- A. Microphthalmic eye generally non-functional**
- B. Systemic associations may be associated with systemic morbidity.**

VII. Describe appropriate patient instructions

- A. Karyotype and genetic counseling indicated if additional abnormalities.**

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology, 2013-2014.

2. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2013-2014.
3. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System, 2013-2014.

Facial anatomy

I. Introduction

- A. The face is composed of functional and aesthetic units**
 - 1. Forehead
 - 2. Eyelids
 - 3. Cheek
 - 4. Nose
 - 5. Lips
 - 6. Neck
- B. Effective treatment of cosmetic and reconstructive upper eyelid problems must include consideration of eyebrow and forehead anatomy and surgery**
- C. Effective lower eyelid cosmetic and reconstructive surgery must include consideration of the midface and cheek**
- D. The lower face and neck are important anatomic areas that are relevant to the aesthetic and functional outcomes of midface, cheek, and forehead pathology and repair**
- E. Symmetry and aesthetic harmony of the various facial units are extremely important**
- F. Only soft tissue anatomy is discussed in this section**
- G. Structural planes of the face include: skin; subcutaneous tissues; the superficial musculoaponeurotic system (SMAS) and mimetic muscles; the deep facial fascia; and the plane containing the facial nerve, parotid duct and buccal fat pad.**

II. Skin

- A. Describe the relevant aspects of the anatomy**
 - 1. Skin consists of 3 layers
 - a. Epidermis (from deeper to superficial)
 - i. Basal layer
 - i) Main proliferative layer
 - ii. Spinous layer
 - i) Spiny projections = desmosomes
 - iii. Granular layer
 - i) Participates in cornification process
 - iv. Cornified layer (stratum corneum)
 - i) Desquamation occurs here

- ii) Thinnest on eyelids and genitalia
- b. Dermis
 - i. Primarily composed of collagen
 - i) Type I (80%) and Type III in humans
 - ii. Elastin fibers 3% of dermis
 - iii. Superficial papillary dermis
 - i) Thin, randomly arranged primarily Type III collagen
 - iv. Deeper reticular dermis
 - i) Course collagen, parallel to surface of skin
 - v. Contains blood vessels, lymphatics, nerves, epithelial adnexa
- c. Subcutaneous fat
 - i. Variable thickness
- d. Melanocytes
 - i. Produce melanin that is passed to lower layers of epidermis

B. Clinical correlations

1. Skin is largest organ in the body
2. Collagen, elastin, and gravitational changes are the cause of most cutaneous aging characteristics
3. Resurfacing techniques may remove tissue to reticular dermis and allow collagen genesis
4. Adnexal structures help reepithelialization
5. Skin is the source of many lesions, benign and malignant, treated by ophthalmologists
6. Adnexal structures, such as hair follicles and cutaneous glands, can develop malignant changes

III. Upper facial anatomy (scalp and frontal region)

A. Describe the relevant aspects of the anatomy

1. Layers of the Scalp (S.C.A.L.P.)
 - a. Skin
 - i. See section above
 - ii. Tends to be thick in scalp
 - b. SubCutaneous tissue
 - i. Fibro-fatty layer adherent to the skin and underlying muscle and its aponeurosis
 - c. Epicranial Aponeurosis (Galea Aponeurotica)
 - i. Galea represents connective tissue layer over cranium

- ii. Galea splits to form sheath around muscles anteriorly (frontalis) and posteriorly (occipitalis)
 - iii. Continuous with superficial musculoaponeurotic system (SMAS) in lower face
 - d. **Loose areolar tissue**
 - i. Occupies sub-aponeurotic space
 - ii. Loosely connects the aponeurosis to periosteum of skull
 - e. **Periosteum of skull or pericranium**
2. **Muscles**
- a. **Occipitofrontalis muscle**
 - i. Consists of 4 bellies
 - i) 2 occipitalis
 - ii) 2 frontalis
 - ii. Connected by galea aponeurotica
 - iii. Occipitalis arises from the highest nuchal line on the occipital bone
 - iv. Occipitalis inserts on the galea
 - v. Frontalis arises from the galea and inserts in the brow (no bony attachment)
 - vi. Occipitofrontalis allows the scalp to move anteriorly and posteriorly, raising brows and wrinkling forehead as in surprise
 - vii. Frontal bellies elevate the brows
 - viii. Occipitalis innervated by posterior auricular branch, facial nerve
 - ix. Frontalis innervated by the temporal branch of the facial nerve
3. **Nerves**
- a. Motor function: see above
 - b. Sensory
 - i. Ophthalmic nerve (first division of trigeminal nerve)
4. **Lymphatic drainage**
- a. Scalp: frontal scalp: anterior auricular and parotid nodes; temporal and parietal scalp: parotid and retroauricular nodes; occipital scalp: occipital and deep cervical nodes

B. Clinical correlations

- 1. The loose areolar tissue layer is avascular and a good plane in which to elevate a frontal flap for brow lift or reconstruction
- 2. Anteriorly, the subperiosteal plane is elevated for the endoscopic brow lift
- 3. Horizontal frontal rhytides are created by contraction of the frontalis muscle
 - a. These can be relaxed with botulinum toxin or a forehead lift

4. Note that lymphatic drainage is to deep facial and cervical nodes with respect to infection and tumor spread

IV. Upper facial anatomy (eyebrows and eyelids)

A. Describe the relevant aspects of the anatomy

1. The eyebrow is the junction of many anatomic structures of the upper and mid face and forms the functional foundation for this region
 - a. In the region of the eyebrow, the muscle plane is firmly fixed to the skin
 - b. The frontalis muscle inserts into the eyebrow, where it interdigitates with the orbicularis oculi and corrugator supercilii muscles
 - c. Laterally, the muscles are attached at the lateral raphe
 - d. The corrugator supercilii inserts into the medial cutaneous portion of the brow
 - e. A fat layer is posterior to the brow muscles
 - i. This is brow fat or retro-orbicularis oculi fat (ROOF)
 - ii. This is continuous with the posterior orbicularis fascia of the upper lid in Caucasians and with the preseptal eyelid fat in Asian lid configurations
 - iii. The fat allows the brow to slide over the frontal bone
 - f. The posterior boundary of the eyebrow fat pad continues into the eyelid as the superior orbital septum
 - g. Deep attachments from the brow musculature extend through the fat pad to insert in the periosteum
 - i. These attachments are weakest laterally
 - ii. These form the structural support for the brow
2. Muscles
 - a. Orbicularis oculi
 - i. Three parts: orbital, preseptal, pretarsal
 - ii. Sphincter muscle that closes the eye, contributes to the lacrimal pump
 - iii. Superior portion supplied by the temporal branch, facial nerve
 - iv. Inferior portion supplied by the zygomatic branch, facial nerve
 - b. Corrugator supercilii
 - i. Origin from the periosteum of the nasal process, frontal bone
 - ii. Fibers insert into medial aspect of brow
 - iii. Moves brows medially
 - iv. Innervated by temporal branch, facial nerve

- c. Procerus
 - i. Continuous with the inferior medial frontalis
 - ii. Origin is from the lower portion of nasal bone
 - iii. "Inserts" into inferior frontalis
 - iv. Moves medial brow inferiorly
 - v. Innervated by the buccal branch, frontal nerve
- 3. Nerves
 - a. Facial nerve (CN VII)
 - i. Innervates the majority of facial musculature
 - ii. Emerges from the stylomastoid foramen, enters parotid gland
 - iii. Divides into 5 terminal branches:
 - i) Temporal
 - ii) Zygomatic
 - iii) Buccal
 - iv) Mandibular
 - v) Cervical
 - iv. The temporal branch lies anterior to the zygoma, crossing through a point 1.5 cm lateral to the eyebrow
 - v. The temporal branch runs in the plane of the galea aponeurotica and the superficial temporal fascia
 - vi. No significant branches of facial nerve posterior to hairline
 - b. Sensory innervation to the brow
 - i. Supraorbital and supratrochlear nerves of the ophthalmic division of trigeminal nerve (CN V)
- 4. Vascular supply and lymphatic drainage of the eyelid
 - a. Arterial supply comes from the internal carotid artery and the external carotid artery
 - i. Collateral circulation between the two systems is extensive
 - b. Eyelid venous drainage can be divided into pretarsal and posttarsal
 - i. Pretarsal tissues drain to the angular vein medially and the superficial temporal vein laterally
 - ii. Posttarsal tissues drain into the orbital veins and the deeper branches of the anterior facial vein and the pterygoid plexus
 - c. Lymphatic vessels serving the medial portion of the eyelids drain to the submandibular nodes, and vessels serving the lateral portion drain to the superficial preauricular nodes then into the deeper cervical nodes

B. Describe clinical correlates

1. The extensive vascular supply to the eyelid makes infection uncommon and eyelid reconstruction highly successful
2. Avulsed portions of the eyelid should be sewn in place. Revascularization is sometimes possible
3. Vascular supply to the median forehead flap
4. Clinical correlations
 - a. The corrugator produces vertical glabellar creases
 - b. The procerus produces horizontal glabellar creases
 - c. Botulinum toxin and brow lift techniques can be used to treat these creases
 - d. Lateral brow ptosis occurs first due to lack of periosteal attachments
 - e. Can change dissection planes without facial nerve injury posterior to hairline
 - f. The extensive vascular supply to the eyelid makes infection uncommon and eyelid reconstruction highly successful
 - g. Avulsed portions of the eyelid should be sewn in place. Revascularization is sometimes possible
 - h. Vascular supply to the median forehead flap

VI. Temporal region anatomy

A. Describe relevant aspects of the anatomy

1. The temporal region is bounded laterally by the ear, superiorly and medially by the temporal line, and inferiorly by the zygomatic arch
2. Anatomic layers of the temporal region (from superficial to deep)
 - a. Skin, with temporal hairline
 - b. Subcutaneous fat
 - c. Superficial temporalis fascia
 - i. This layer is continuous with the galea aponeurotica
 - ii. Temporal branch of facial nerve runs approximately 1.5 cm from lateral brow
 - d. Subgaleal fascia
 - i. Very loose tissue
 - e. Temporalis fascia proper
 - f. Temporalis muscle
 - g. Approximately 2.5 cm above zygoma

- i. Temporalis fascia proper splits into the intermediate and deep temporalis fascia to envelop the intermediate temporal fat pad
 - i) Intermediate temporal fat pad extends to zygomatic arch and medially to the lateral orbital rim; continuous with buccal fat pad
- 3. The conjoined tendon lies along the temporal line
 - a. The conjoined tendon represents a fusion of the superficial temporalis fascia, temporalis fascia proper, galea aponeurotica, and periosteum
- 4. Muscles
 - a. Temporalis muscle
 - i. Arises from the temporalis (infratemporal) fossa along the temporal line
 - ii. Inserts on coronoid process and anterior border of ramus of mandible
 - iii. Closes jaw, posterior portion retracts jaw
 - iv. Innervated by mandibular division, motor trigeminal nerve
- 5. Nerves
 - a. Temporal branch, facial nerve runs in this region
 - b. See relevant aspects of the anatomy (VI.A.2.c.ii.) above and clinical correlations below
 - c. Sensation provided by the mandibular nerve (third division of trigeminal nerve)
- 6. Lymphatics
 - a. Drains to deep facial and deep cervical nodes

B. Clinical correlations

- 1. As the temporal branch of the facial nerve crosses the zygomatic arch anteriorly, it is unsafe to dissect from the upper face to lower face across this region
 - a. A safe approach to the medial zygomatic arch may be carried out in a dissection plane just deep to the intermediate temporal fat pad
- 2. As the temporal branch of the facial nerve lies in layers anterior to the temporalis fascia proper, this should be the dissection plane over the temporalis muscle
 - a. The dissection plane for a coronal or endoscopic brow lift should be just anterior to the temporalis fascia proper
 - b. The temporal optical space should be joined to the central subperiosteal space through the conjoined tendon from lateral to medial
- 3. The conjoined tendon is quite strong. Sharp dissection may be required to open it and care must be taken to maintain a safe plane, deep to the galea medially and superficial temporal fascia laterally
- 4. Note that lymphatic drainage is to deep facial and cervical nodes with respect to infection and tumor spread

VII. Mid Face Anatomy

A. Describe the relevant aspects of the anatomy

1. Nose

- a. The external structure of the nose consists of skin, bone and cartilage. Most of the external shape of the nose is from the soft tissue structures rather than the underlying bones. The nose is made of 3 distinct portions with the upper 1/3 rigidly fixed into position, the middle third semi mobile and the inferior third mobile
 - i. The nose profile
 - i) Root, dorsum, tip and columella
 - ii) The coronal plane
 - (i) Ala, alar sulcus and nostrils
 - ii. The soft tissues are supported by the nasal bones and the frontal processes of the maxilla. The nasal cartilage is supported by the nasal bones, maxillae, vomer and ethmoid bones
 - iii. The external cartilages
 - i) The triangular shaped lateral cartilage
 - ii) The bilateral alar cartilage
 - (i) Forms the columella
 - (ii) Nasal tip
 - (iii) Outer nostril walls
 - (iv) The alar base and the external naris vary widely among individuals and ethnic types with variable configuration and distribution of fibro-fatty connective tissue
- b. Skin
 - i. Thickness of the nasal skin
 - i) Varies from dorsum to tip
 - (i) Upper 1/3 thick
 - (ii) Middle 1/3 thin
 - (iii) Inferior 1/3 thick with sebaceous glands in nasal tip
 - ii. Varies among ethnic backgrounds with some Caucasian individuals having thin tissue paper like skin over the nose and other ethnic backgrounds may have thick oily skin which may be difficult to advance or re-drape
- c. SMAS
 - i. Nasal SMAS is continuous with facial SMAS
 - ii. Invests and connects the nasal musculature

- iii. Sling for mimetic muscles
 - d. Muscles
 - i. The nasal muscles animate the nose during facial expression and regulate airflow. The muscle fibers intertwine with each other
- 2. The Midface
 - a. The midface extends from the inferior orbital rim to the mouth. The soft tissues of the cheek and midface descend and the nasolabial folds become more pronounced with age. Diffuse connective tissue attachments exist between the subcutaneous fat and the dermis. Areas with sparse fat including the pretragal region, zygomatic arch, orbicularis oculi, nasolabial fold and orbicularis oris have dense connective tissue attachments into the dermis. The SMAS has osseous fixation at several distinct points throughout the midface
 - b. Osseous Fixation of Midface SMAS
 - i. Zygomatic ligaments
 - i) Extensive complex from the zygoma to the SMAS
 - (i) Extends posteriorly and inferiorly around the origin of the zygomatic major muscle
 - (ii) Continuous with the masseteric cutaneous ligaments
 - ii. Orbitomalar ligament
 - c. A distinct bony attachment arising from the periosteum of the inferior orbital rim and inserting into the malar dermis cheek fat pads
 - i. Deep medial fat pad
 - ii. Deep lateral fat pad
 - d. Tear trough
 - i. Concave obliquely oriented groove medial aspect lower eyelid from medial cantus to midpupillary line. May be multifactorial
 - i) Descent of cheek
 - ii) Loss of facial volume
 - iii) Maxillary retrusion
 - iv) Change in subcutaneous fat
 - ii. Various classification methods
 - e. The remaining details of the midfacial anatomy are described below in the lower facial anatomy

B. Clinical correlation

1. Seventh nerve paralysis may cause nasal obstruction due to muscular hypofunction
2. Dissection under the nasal SMAS may enhance preservation of the vascularity of an overlying flap
3. Tear trough augmentation

4. Volume restoration of the deep medial cheek fat diminishes the nasolabial fold and improves cheek projection

VIII. Lower Facial Anatomy

A. Describe the relevant aspects of the anatomy

1. Lips
 - a. Vermillion border
 - i. Mucocutaneous junction
 - ii. Abrupt transition from keratinized to non-keratinized squamous epithelium
 - b. Cupid bow
2. Philtral columns
3. Muscles of mastication
 - a. All are innervated by mandibular division, motor trigeminal nerve
 - b. All are involved in biting and chewing
 - i. Temporalis: see above
 - ii. Masseter
 - i) Origin: zygomatic arch
 - ii) Insertion: ramus and condyloid process of mandible
 - iii) Closes jaw
 - iii. Medial and lateral pterygoid muscles
4. Muscles of facial expression (mimetic muscles)
 - a. Lie in the SMAS fascia (see below)
 - b. All arise from the fascia or bone
 - c. All insert in the skin
 - d. All innervated by facial nerve from posterior surface, except for buccinator, levator anguli oris, and mentalis, which are innervated from anterior surface
 - i. Anterior and posterior dilator nares: enlarge nares
 - ii. Depressor septi: constricts nares
 - iii. Nasalis: depresses nasal cartilage
 - iv. Levator labii superioris: raises upper lip
 - v. Levator labii superioris alaeque nasi: raises upper lip and dilates nares
 - vi. Zygomaticus minor: raises lip; works with levator labii superioris and levator labii superioris alaeque nasi to create nasolabial fold

- vii. Levator anguli oris: raises angle of mouth
- viii. Zygomaticus major: draws angle of mouth up and back
- ix. Risorius: retracts angle of mouth
- x. Depressor labii inferioris: draws lip down and back
- xi. Mentalis: raises and protrudes lower lip
- xii. Orbicularis oris: closes lips, protrudes lips, presses lips to teeth
- xiii. Buccinator: compresses cheek

5. Fascial planes of the lower face

- a. SMAS
 - i. Surrounds mimetic muscles
 - ii. Extension of superficial cervical fascia of neck
 - iii. Includes the suborbicularis oculi fat pad (SOOF) (see below)
 - iv. Continuous with superficial temporal fascia
 - v. Acts to distribute facial muscle contractions to the skin via ligamentous connections
 - vi. Fat lies anterior to SMAS, deep to skin
 - vii. Major vessels and nerves run deep to the SMAS
- b. SOOF
 - i. Supraperiosteal suborbicularis fat pad
 - ii. Present over the zygoma and maxilla
 - iii. Continuous superiorly with the ROOF pad deep to eyebrow
- c. Parotidomasseteric fascia
 - i. Overlies parotid gland and masseter muscle
 - ii. Continuation of deep cervical fascia of the neck
 - iii. Facial nerve is deep to this layer in lower face (see below)
 - iv. Continuous with deep temporalis fascia superior to zygomatic arch

6. Facial nerve

- a. Arises from the pons at cerebellopontine angle
- b. Travels with nervus intermedius
- c. Leaves cranial fossa with acoustic nerve via internal auditory meatus
- d. Enters facial canal; geniculate ganglion at acute bend of facial canal
- e. Exits at stylomastoid foramen
- f. Enters parotid gland and divides into branches

- g. Branches
 - i. Great petrosal nerve carries lacrimal secretory fibers
 - ii. Nerve to stapedius muscle
 - iii. Chorda tympani nerve: taste anterior 2/3 of tongue and innervation of submandibular and sublingual glands
 - iv. Muscular branches
 - i) Temporal
 - ii) Zygomatic
 - iii) Buccal
 - iv) Mandibular
 - v) Cervical

7. Sensory innervation

- a. Maxillary nerve (second division of trigeminal nerve)
 - i. Lower eyelid, cheek, side of nose, nasal vestibule, upper lip
 - ii. Infraorbital nerve is terminal nerve to cheek region
- b. Mandibular nerve (third division of trigeminal nerve)
 - i. Sensation to lower teeth, gingiva, mandible
 - ii. Sensation to temporal area (see above) and tympanic membrane
 - iii. Motor innervation to temporalis, masseter, medial and lateral pterygoids

8. Parotid gland and duct

- a. Lie deep to SMAS and deep parotidomasseteric fascia
- b. Anterior to masseter muscle
- c. After coursing over masseter and buccal fat pad, parotid duct pierces buccinator muscle and enters oral cavity at Stensen's duct

9. Lymphatic drainage

- a. Eyelids and conjunctiva: submandibular and parotid nodes
- b. Cheek: parotid and submandibular nodes
- c. Side of nose, upper lip, lateral lower lip: submandibular nodes
- d. Medial lower lip: submental nodes

B. Clinical correlations

- 1. SMAS dissection and shortening is one of the most common techniques in face lifting and is felt to have greater longevity than skin only face lift
- 2. Creation of skin flaps of the lower face (e.g. Mustarde flap) must take into consideration anatomic layers and position of important structures such as facial nerve and parotid duct

3. Note that lymphatic drainage is to deep facial and cervical nodes with respect to infection and tumor spread
4. The temporal and mandibular branches of the facial nerve are at greatest risk of surgical injury
5. Infraorbital nerve frequently injured in midface trauma (e.g., blow-out fractures) or midface surgery (e.g., SOOF lifts), causing midface anesthesia

IX. Neck Anatomy

A. Describe relevant aspects of the anatomy

1. Surface anatomy defining features
 - a. Thyroid cartilage
 - b. Anterior triangles
 - c. Posterior triangles
 - d. Sternocleidomastoid muscle
2. Muscles
 - a. Platysma
 - i. Retracts and depresses angle of mouth
 - ii. Arises from deltoid and pectoral fascia
 - iii. Inserts on the mandible and skin
 - iv. Innervated by facial nerve
 - b. Sternocleidomastoid
 - i. Bends head to same side, rotates head, raises chin to opposite side
 - ii. Arises from sternum, clavicle
 - iii. Inserts into the mastoid process
 - iv. Innervated by Accessory, C2, C3 nerves
 - c. Anterior belly digastric muscle
 - i. Opens jaw, draws hyoid bone forward
 - ii. Arises from lower border mandible
 - iii. Inserts on body and great cornu of hyoid
 - iv. Innervated by trigeminal nerve
 - d. Posterior belly digastric muscle
 - i. Draws hyoid back and raises hyoid
 - ii. Arises from mastoid notch in temporal bone
 - iii. Inserts on body and great cornu of hyoid

- iv. Innervated by facial nerve
- e. Stylohyoid
 - i. Draws hyoid up and back
 - ii. Arises from styloid process
 - iii. Inserts on body of hyoid
 - iv. Innervated by facial nerve
- f. Mylohyoid
 - i. Raises hyoid
 - ii. Arises from mylohyoid line of mandible
 - iii. Inserts on body of hyoid
 - i) Innervated by trigeminal nerve
- g. Various additional muscles depressing hyoid and thyroid cartilages
 - i. Include geniohyoid , sternohyoid , sternothyroid , thyrohyoid , omohyoid
- 3. Facial planes of the neck
 - a. Superficial cervical fascia of neck continuous with SMAS
 - b. Deep cervical fascia of the neck
 - i. Found on superficial surface of strap muscles superior to the hyoid bone
 - ii. Overlies mylohyoid muscle and extends superiorly over body of mandible
- 4. Lymphatics
 - a. Note afferent sources from scalp and face above

B. Clinical correlations

- 1. Note continuity in planes from lower face into neck
- 2. Dehiscence of midline interdigitation of platysma bands contributes to laxity of neck tissues
- 3. Neck lymph nodes are common site for distant spread of facial infection and malignancies
- 4. Subcutaneous fat commonly removed in subcutaneous fat

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology, 2013-2014.
- 2. Owsley JQ: *Aesthetic Facial Surgery* WB Saunders, Philadelphia, 1994.
- 3. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.18-21.
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Eyelid skin and subcutaneous tissue

I. Describe relevant aspects of the anatomy

- A. The eyelid skin is the thinnest skin of the body
- B. Just beneath the skin and its thin layer of subcutaneous tissue lies the orbicularis oculi muscle
- C. As the skin extends superiorly to the location of the eyebrow, and inferiorly to the infraorbital rim, it becomes thicker
- D. The upper eyelid crease approximates the attachments of the levator aponeurosis to the pretarsal orbicularis bundles and skin. Racial variation can be noted in the location of the eyelid crease. The skin receives sensory innervation from branches of the trigeminal nerve
- E. The blood supply to the periocular soft tissues is by the internal carotid via the ophthalmic artery and its branches - the supraorbital and lacrimal, as well as the external carotid via the arteries of the face - the angular and temporal

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.9-11.

Protractors

I. Describe relevant aspects of the anatomy

A. Orbicularis oculi muscle

1. Innervated by cranial nerve (CN) VII
2. Narrows the palpebral fissure, closes the eye
3. A portion contributes to the lacrimal pump
4. Divisions are anatomic and physiologic
 - a. Pretarsal component arises from deep origins at the posterior lacrimal crest and superficial origins at the anterior limb of the medial canthal tendon
 - i. Upper and lower pretarsal segments fuse laterally to form the lateral canthal tendon
 - ii. Deep head of pretarsal portion medially encircles both canaliculi to aid tear drainage
 - b. Preseptal portion has deep origins from fascia around the lacrimal sac and posterior lacrimal crest, and superficial origins from anterior limb of medial canthal tendon
 - i. Laterally fuses to form the lateral palpebral raphe overlying the lateral orbital rim
 - c. Orbital portions arise from the anterior limb of the medial canthal tendon and surrounding periosteum
 - d. Palpebral portions (pretarsal and preseptal) are more involved in involuntary eyelid movement (blink), and the orbital portion is responsible for forceful eyelid closure (wink, blepharospasm). The orbital portion of the orbicularis is involved in both voluntary and forced eyelid closure. The preseptal fibers contribute to voluntary and involuntary eyelid closure. The pretarsal orbicularis primarily contribute to involuntary closure

B. Canthal tendons

1. Lateral canthal tendon
 - a. Attaches at the lateral orbital tubercle on the inner aspect of the lateral orbital rim
 - b. Splits into the superior and inferior crus, which attach to the respective tarsal plates
 - c. The lateral canthal tendon usually inserts 2 mm more superiorly than the medial canthal tendon
 - d. Horizontal eyelid instability is usually the result of lateral canthal lengthening, and surgical correction should be directed to shortening the lateral canthus
2. Medial canthal tendon
 - a. The anterior and posterior limbs of the tendon surround the lacrimal sac, attaching on the anterior and posterior lacrimal crests
 - b. The attachment to the periosteum of the anterior lacrimal crest is diffuse and strong
 - c. The attachment to the posterior lacrimal crest is more delicate, but important in maintaining

apposition of the eyelids to the globe

- d. The arrangement of the canthal tendon blending into the lacrimal sac fascia is the key anatomic factor in the "lacrimal pump"

II. Describe clinical correlates

- A. **Weakness of the protractors is responsible for the lagophthalmos, incomplete blink and ectropion seen in facial nerve palsy**
 - 1. Tearing and corneal exposure may result
- B. **Overactivity of the protractors is seen in disorders such as hemifacial spasm, essential blepharospasm and orbicularis myokymia**
- C. **Orbicularis weakness may be associated with tearing even without overt lagophthalmos or ectropion from disruption of the lacrimal pump mechanism**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
- 2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.88-89.
- 3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.169-171.
- 4. Reifler DM. Early description of Horner's muscle and the lacrimal pump. *Surv Ophthalmol*, 1996; 41:127034.
- 5. Muzaffar AR, Mendelson BC, Adams WP Jr. Surgical anatomy of the ligamentous attachments of the lower lid and lateral canthus. *Plast Reconstr Surg*. 2002;110(3):873-884; discussion 897-911.

Orbital septum

I. Describe relevant aspects of the anatomy

- A. Multi-layered sheet of thin fibrous tissue that arises from the periosteum of the orbital rims at the arcus marginalis
- B. Fuses with the levator aponeurosis in the upper eyelid, 2 to 5 mm above the superior border of the tarsal plate in non-Asians
- C. The fusion point of the septum with the levator is variable in the Asian eyelid, and may be above the superior border of the tarsal plate, or inferiorly over the anterior surface of the tarsal plate
- D. Fuses with the capsulopalpebral fascia at or just below the inferior border of the tarsal plate in the lower eyelid

II. Describe clinical correlates

- A. The septum attenuates with age in both the upper and lower eyelids, allowing anterior prolapse of the orbital fat
- B. The septum serves as a barrier between the eyelid and orbit, limiting spread of infection and hemorrhage

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.3-4, 16-17, 27-28.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.86, 90.
4. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.14, 18, 23, 84-85.

Orbital fat

I. Describe the relevant aspects of the anatomy

A. Enclosed within an intricate network of fibrous septae and blood vessels

1. Intermuscular septum divides intraconal from extraconal fat
2. Injury to fibrous septae can cause fibrosis
3. Relationship of fibrous septae and extraocular muscles - inferior rectus muscle entrapment in orbital floor fracture

B. Extraconal fat

1. Preaponeurotic fat
 - a. Extraconal
 - b. Intraconal
 - c. Fat exposure is a surgical landmark to identify penetration of septum and location of levator aponeurosis and muscle in ptosis surgery. Identification of the central fat pad is a helpful finding in eyelid trauma.
 - d. Preaponeurotic fat removed during blepharoplasty
2. Upper eyelid fat pads
 - a. Central - yellow color, can extend inferiorly to junction of orbital septum with levator aponeurosis
 - b. Medial - whiter color
 - c. Lateral area of preaponeurotic space contains lacrimal gland
 - d. Medial and central upper eyelid fat pads are separated by fascia connections continuous with the trochlea
3. Lower eyelid fat pads
 - a. Medial, central, lateral
 - b. Inferior oblique muscle divides medial and central fat pads

C. Intraconal orbital fat

1. Fat within intricate septa provides globe support, optic nerve protection, muscle support within the bony orbit
2. Involved in thyroid orbitopathy
 - a. Orbital fibroblast undergo adipogenesis
 - b. Orbital fat expands with accumulation of glycosaminoglycans

D. Suborbicularis oculi fat (SOOF): deep subcutaneous fat, posterior to the orbicularis oculi overlying the body of the zygoma, extends into midfacial soft tissues

E. Retro-orbital suborbicularis fat (ROOF)

1. May enlarge with thyroid eye disease

II. Describe clinical correlates

A. Excessive manipulation of orbital fat can result in:

1. Fibrosis and muscle restriction
2. Fat atrophy
3. Enophthalmos
4. Hemorrhage

B. Thinning of the orbital septum and loss of fibrous support within the orbit contributes to fat prolapse

1. Removal or repositioning of preaponeurotic fat is a major component of blepharoplasty
2. Complications of blepharoplasty include orbital hemorrhage
3. Reduction of proptosis in thyroid eye disease (thyroid orbitopathy) can be obtained by removal of bony walls of the orbit and/or removal of orbital fat
4. The fat component of a dermis fat graft can enlarge in children and atrophy in adults

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.87-88.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.16, 25-27, 84-85.

Eyelid retractors

I. Describe relevant aspects of the anatomy

A. The upper eyelid retractors are the levator muscle with its aponeurosis (innervated by cranial nerve (CN) III) and the superior tarsal muscle (Müller muscle, sympathetic innervation)

1. The levator muscle originates from the periosteum of the lesser wing of the sphenoid, above the annulus of Zinn
 - a. The muscular portion is approximately 40 mm in length
 - b. The aponeurotic portion measures 14 to 20 mm in length
 - c. The superior transverse ligament (Whitnall) is a condensation of elastic fibers of the anterior sheath of the levator found at the transition between the muscular and aponeurotic portion
 - i. Whitnall ligament functions as a suspensory ligament for the upper lid and superior orbital tissues
 - ii. Acts as a fulcrum for the levator
 - iii. Medially, Whitnall ligament attaches to connective tissue around the trochlea
 - iv. Laterally, it forms septa through the lacrimal gland, then attaches to the lateral orbital wall 10 mm above the orbital tubercle
 - d. The lateral horn of the levator aponeurosis inserts on the lateral orbital tubercle (after dividing the lacrimal gland into orbital and palpebral lobes), and the medial horn inserts onto the posterior aspect of the medial canthal tendon and the posterior lacrimal crest
 - e. The anterior portion of the distal aponeurosis sends fibers that insert between pretarsal orbicularis muscle bundles
 - i. The upper eyelid crease is formed by the most superior of these fibers
 - f. The posterior portion of the distal aponeurosis inserts on the anterior surface of the lower half of the tarsus, firmly attached approximately 3 mm above the lid margin and more loosely attached superiorly
2. Müller muscle originates at the undersurface of the levator at the level of Whitnall ligament, and extends inferiorly to insert on the superior aspect of the tarsal margin
 - a. Sympathetic innervation, provides about 2 mm of elevation
 - b. Firmly attached to the underlying conjunctiva
 - c. The peripheral arterial arcade is located between Müller and the levator aponeurosis just above the superior tarsal border

B. The lower eyelid retractors include the capsulopalpebral fascia and the inferior tarsal muscle

1. The capsulopalpebral fascia is analogous to the levator aponeurosis
 - a. Originates as the capsulopalpebral head from attachments to the inferior rectus muscle
 - b. The capsulopalpebral head courses anteriorly, divides and encircles the inferior oblique

muscle, and fuses with the sheath of the oblique

- c. The 2 portions of the capsulopalpebral head then join to form Lockwood's ligament anterior to the inferior oblique muscle
 - d. The capsulopalpebral fascia extends anteriorly from Lockwood ligament, sending strands to the inferior fornix
 - e. The capsulopalpebral fascia inserts on the inferior border of the inferior tarsal plate just after fusing with the orbital septum
2. The inferior tarsal muscle is analogous to Müller muscle in the upper eyelid
- a. Poorly developed smooth muscle
 - b. Runs posterior to the capsulopalpebral fascia, and is most abundant in the region of the inferior fornix

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 9. Surgical Anatomy of the Ocular Adnexa, 1996.
3. AAO, Focal Points: Botulinum Toxin in Ophthalmology, Module #3, 2007.

Tarsus

I. Describe relevant aspects of the anatomy

- A. Firm dense plates of connective tissue that function as the skeleton of the eyelids
- B. Upper eyelid tarsal plate measures 10 to 12 mm vertically in the center of the eyelid
- C. Lower eyelid tarsal plate measures 4 mm or less vertically
- D. Both tarsal plates measure 1 mm thick, tapering at the medial and lateral ends
- E. Attach to the periosteum medially and laterally via the medial and lateral canthal tendons
- F. Tarsal plates may displace horizontally with age as a result of stretching of the medial or lateral canthal tendons
- G. Meibomian glands, holocrine sebaceous glands, lie within tarsal plates
- H. Conjunctiva intimately attached to posterior surface

II. Describe clinical correlates

- A. A chalazion is a chronic granulomatous inflammation of a meibomian gland (See Chalazion)
- B. The conjunctiva and the tarsal plate form the posterior lamella of the eyelid
- C. The strength of a primary lid margin repair lies in lamellar sutures in the tarsus closure
- D. In eyelid reconstruction, tarsal replacement can be obtained using a free tarsal graft or a tarsoconjunctival flap (Hughes procedure)

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.85, 180-181.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.23, 48-49, 84-85, 139.

Conjunctiva

I. Describe relevant aspects of the anatomy

- A. The conjunctiva is the mucous membrane lining the posterior surface of the eyelids (palpebral conjunctiva), and covering the surface of the globe to the limbus (bulbar conjunctiva)**
1. Consists of stratified nonkeratinizing squamous epithelium with mucus-secreting goblet cells (most numerous in the fornices)
 2. The underlying connective tissue (substantia propria) contains scattered lymphocytes, plasma cells, and lymphoid follicles
 3. The accessory lacrimal glands of Krause, approximately 20 in number, are located along the conjunctival fornices
 4. The accessory lacrimal glands of Wolfring, less numerous, are found in the subconjunctival tissue near the superior edge of the upper tarsus and the inferior edge of the lower tarsus
 5. No innervation to the accessory lacrimal glands has been identified

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.95.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.87, 131, 154-157.

Suborbicularis fat pads

I. Describe relevant aspects of the anatomy

- A. Suborbicularis oculi fat (SOOF, malar fat pad) consists of the deep, nonseptate subcutaneous fat and connective tissue that lies beneath the orbicularis in the lower eyelid, and extends into the midface soft tissues
- B. The sub-brow fat pad (ROOF—retroorbicularis oculi fat) also undergoes gravitational descent, compounding redundant upper eyelid skin

II. Describe clinical correlates

- A. The SOOF descends with aging, contributing to the midface ptosis and resultant increase in the depth of the nasolabial folds
- B. Elevation of the SOOF to its normal position improves lower eyelid and midface contour
- C. Elevation of the ROOF accompanies brow plasty
- D. "Sculpting" of the ROOF can improve eyebrow contour

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Upper Eyelid Blepharoplasty, Module #8, 1995.
3. AAO, Focal Points: Aesthetic Surgery of the Lower Eyelid, Module #7, 1995.

Eyelid margin

I. Describe relevant aspects of the anatomy

- A. The eyelid margin has a well-defined anterior and posterior margin
- B. The punctum is present on the lacrimal papilla. The portion of the eyelid margin medial to the punctum contains the canaliculus and is thinner than the majority of the eyelid
- C. The gray line is formed by a specialized section of pretarsal orbicularis muscle (Riolan's) that is just anterior to the tarsus
- D. The mucocutaneous junction is found just posterior to the meibomian gland orifices
- E. Reapproximation of eyelid margin structures are imperative in eyelid reconstruction to avoid notching and margin irregularity

II. Describe clinical correlations

- A. Inversion of the eyelid margin is known as entropion
 - 1. The most common cause of entropion is involutional
 - 2. Marginal entropion is a subtle turning in of the eyelid margin as the result of mild posterior lamellar shortening
 - 3. Marginal entropion is a common cause of misdirected eyelashes
- B. Epiblepharon is a condition where an extra roll of eyelid skin displaces the lashes toward the cornea
 - 1. The position of the lid margin is normal
- C. Distichiasis is a condition where an extra row of eyelashes emerges from the meibomian gland orifices
- D. The mucocutaneous junction of the eyelid may not form normally after eyelid reconstruction
 - 1. An anterior position of the junction causes an erythematous lid margin
 - 2. A posterior position of the junction may bring skin and lanugo hairs against the eye

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
- 2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.26.
- 3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p. 84, 101, 108, 174-176.
- 4. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p. 130-131, 205, 209-217, 228-229, 232.
- 5. AAO, Focal Points: Management of Eyelid Trauma, Module #10, 1996, p. 3.

Eyelashes

I. Describe relevant aspects of the anatomy

- A. The eyelash roots are anterior to the tarsal plates. The eyelashes emerge from the lid margin anterior to the gray line**
 - 1. The eyelashes form 3 or 4 rows in the upper eyelid and 2 or 3 rows in the lower eyelid
 - 2. A few eyelashes may be found on the caruncle
- B. Eyelash abnormalities usually result from chronic inflammation, infection, congenital anomalies, tumor or trauma**
 - 1. Trichiasis - misdirected or turning in of lashes
 - 2. Distichiasis - an anomalous row of eyelashes emerging from the meibomian gland orifices
 - 3. Lash loss (madarosis)- secondary to tumor, chronic inflammation, alopecia, trichotillomania
 - 4. Eyelash abnormalities should be differentiated from eyelid malpositions, such as involutional entropion, marginal entropion or epiblepharon
 - 5. Eyelash ptosis seen in floppy eyelid syndrome
- C. Eyelash enhancement**

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
- 2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.8, 26.
- 3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.70, 75, 79, 101, 108, 128, 139.
- 4. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.131.
- 5. AAO, Focal Points: Management of Trichiasis, Module #4, 2001.

Meibomian glands

I. Describe relevant aspects of the anatomy

- A. **Specialized sebaceous glands originating in the tarsal plates, responsible for the production of the oily outer layer of tear film that prevents evaporation of the aqueous layer**
- B. **The upper eyelid contains approximately 40 glands, and the lower eyelid contains approximately 20 glands**
- C. **The eyelashes and Meibomian glands originate from a common pilosebaceous unit in the second month of gestation**
 - 1. Congenital distichiasis is characterized by an extra row of lashes emanating from the meibomian gland orifices from birth
 - 2. Trauma or chronic irritation may result in development of a ciliary follicle in place of a Meibomian gland, called acquired distichiasis

II. Meibomian gland disease

- A. **Dysfunction (posterior eyelid margin disease, blepharitis)**
- B. **Chalazia/Hordeola**
- C. **Sebaceous carcinoma**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
- 2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.25.
- 3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.84, 101.

Nerve supply of eyelids

I. Describe relevant aspects of the anatomy

A. Sensory innervation is provided by branches of the first (ophthalmic) and second (maxillary) divisions of cranial nerve (CN) V

1. The ophthalmic division (CN V₁) divides into 3 branches
 - a. Frontal
 - b. Lacrimal
 - c. Nasociliary
2. The frontal and lacrimal nerves enter the orbit through the superior orbital fissure, superior to the annulus of Zinn, and course anteriorly to innervate
 - a. Medial canthus (via the supratrochlear branch)
 - b. Upper eyelid (via the lacrimal and supratrochlear branches)
 - c. Forehead (via the supraorbital branch)
3. The nasociliary branch enters the orbit through the superior orbital fissure and the annulus of Zinn, and travels anteriorly to innervate the eye
4. Branches of the maxillary nerve (CN V₂) innervate the lower eyelid and cheek

B. Motor supply is provided by CN III (levator), CN VII (orbicularis), and sympathetics (Müller muscle)

II. Describe clinical correlates

A. Manifestations of sensory nerve weakness

1. Numbness in the distribution of the infraorbital nerve following trauma suggests an orbital floor fracture
2. Numbness in the distribution of the supraorbital nerve may follow brow or forehead lifting surgery
3. Numbness with a previous history of facial skin cancer suggests perineural cancer spread

B. Manifestations of motor nerve weakness

1. Ptosis due to CN III palsy
2. Ectropion, lagophthalmos and brow ptosis secondary to facial nerve paralysis
3. Frontal branch paresis after endoscopic or coronal forehead lifting

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.18-19, 30.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.36-38.

Chalazion

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Plugging of a meibomian or Zeis gland or glands results in trapping of sebaceous material
2. The trapped material elicits chronic granulomatous inflammation

B. Define the relevant aspects of epidemiology of this disease

1. All ages, men and women, no racial predilection

C. List the pertinent elements of the history

1. Generally, some degree of acute inflammation characterized by mild pain, and erythema precedes a chronic phase consisting of a painless well circumscribed mass within the pretarsal eyelid
 - a. Resolution may occur prior to the development of the chronic phase
2. May fluctuate in size
3. May spontaneously drain posteriorly or anteriorly
4. Blurred vision secondary to induced astigmatism
5. Topical or oral tetracyclines (such as doxycycline) may be appropriate for recurrent or multiple episodes

D. Describe pertinent clinical features

1. Nodule on the tarsal conjunctival surface, visible internally with eyelid eversion, and often visible externally if large enough in size
2. Overlying skin may have erythema
3. May localize anteriorly
4. Chronic lesions often appear cyst-like

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Usually diagnosed clinically
2. Histopathologic confirmation for atypical, persistent or recurrent lesions

II. Define the risk factors

A. Rosacea

B. Chronic posterior blepharitis (meibomian gland dysfunction)

III. List the differential diagnosis

A. Malignancy

1. Sebaceous gland carcinoma
2. Basal cell carcinoma
3. Squamous cell carcinoma

B. Granulomatous disorders

1. Sarcoidosis
2. Atypical mycobacterial infection
3. Wegener granulomatosis

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

A. Describe medical therapy options

1. Warm compresses
2. Eyelid hygiene/scrubs
3. Topical or systemic tetracyclines
4. Topical, oral, or intralesional/perilesional corticosteroid injection; small marginal lesions

B. Describe surgical therapy options

1. Surgical drainage via a transconjunctival or cutaneous route
 - a. Incision and curettage
 - b. Incision with excision of granulomatous tissue and cyst wall
2. Combined excision and corticosteroid injection
3. Biopsy for recurrent or atypical lesions

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

A. Eyelid margin scarring or notching

1. Use care when removing tissue at or near the lid margin as there is a risk for eyelid scarring or notching
2. Can revise severe scar or notch with full-thickness pentagonal wedge resection

B. Damage to surrounding meibomian glands

1. Prevent by making vertical cuts through tarsal conjunctival surface

C. Depigmentation of overlying skin with corticosteroid injection

1. Avoid use in patients with dark skin
2. Use a small, dilute dose combined with surgical treatment
3. Potential risk of visual loss due to intravascular injection which may, albeit rarely, lead to central retinal artery occlusion

D. Damage to punctum or canaliculus

1. Use caution when excising pericanalicular chalazia
2. If punctum or canaliculus is violated, repair with silicone stent intubation

E. Repeated episodes or surgical treatments can lead to posterior lamellar scarring and misdirected eyelashes or eyelash loss

VI. Describe disease-related complications

- A. Rarely eyelid scarring with or without trichiasis**
- B. Eyelid malposition (ex/ ptosis from longstanding lesion of upper eyelid)**
- C. Recurrent lesions**

VII. Describe appropriate patient instructions

- A. Prevention of new chalazia with warm compresses, eyelid hygiene, long-term systemic tetracyclines**
- B. Return if chalazion recurs**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.

Blepharitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Staphylococcal blepharitis
 - a. Bacterial infection and/or inflammatory reaction to bacterial antigens and toxins. Usually involving the eyelids and frequently the conjunctiva
2. Meibomian gland dysfunction
 - a. Altered meibomian gland secretions result in gland blockage and decreased lipid layer in tear film. Abnormal lipids contribute to ocular and eyelid inflammation and dry eyes
3. Seborrheic blepharitis
 - a. Scaling of eyelid margin associated with inflammatory changes of the eyelid

B. Define the relevant aspects of epidemiology of this disease

1. Staphylococcal disease usually occurs in younger individuals

C. List the pertinent elements of the history

1. Burning, itching, and foreign body sensation
2. Usually worse in the morning
3. Redness of the eyelids
4. Crusting of the eyelids
5. Filmy, blurred vision
6. Recurrent chalazia/hordeola

D. Describe pertinent clinical features

1. Staphylococcal
 - a. Hard scales and crusts around eyelid cilia
 - b. Injection and telangiectasis of eyelid margins
 - c. Poliosis
 - d. Madarosis
 - e. Trichiasis
 - f. Conjunctival hyperemia
 - g. Papillary reaction of tarsal conjunctiva
 - h. Corneal changes such as punctate epithelial keratopathy, marginal infiltrates, phlyctenules
2. Meibomian gland dysfunction

- a. Inflammation of posterior lid margin, conjunctiva, and cornea
 - b. Eyelid margin with irregularity and thickening.
 - c. Prominent telangiectasis
 - d. Pouting or plugged meibomian gland orifices
 - e. Turbid, thick secretions
 - f. Foamy tear meniscus
 - g. Conjunctival hyperemia, and often papillary reaction
 - h. Corneal changes such as punctate epithelial keratopathy, marginal infiltrates, pannus
3. Seborrheic
- a. Anterior eyelid margin inflammation with oily or greasy crusting
 - b. Crusting also on eyebrows and scalp
 - c. Punctate epithelial keratopathy
- E. Describe appropriate testing and evaluation for establishing the diagnosis**
- 1. Culture for cases not responding to empiric therapy, particularly those with chronic component of conjunctivitis
 - 2. Chronic unilateral blepharoconjunctivitis should prompt an eyelid margin biopsy

II. Define the risk factors

- A. Rosacea**
- B. Oral retinoid therapy**

III. List the differential diagnosis

- A. Sebaceous cell carcinoma (particularly if asymmetric and/or unilateral)**

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

- A. Describe medical therapy options**
 - 1. Lid hygiene consisting of warm compresses
 - 2. Eyelid scrubs with dilute baby shampoo or commercially available eyelid wipe once or twice daily depending on severity
 - 3. Ocular lubrication
 - 4. Erythromycin or bacitracin ophthalmic ointment applied to eyelids at bedtime
 - 5. Topical azithromycin may be an option
 - 6. Meibomian gland expression may be beneficial

7. Oral tetracycline, doxycycline, or minocycline for chronic cases
8. Short course of topical corticosteroids may be used when inflammation severe
9. Long term treatment and followup is required

B. Describe surgical therapy options

1. No specific surgical treatment exists
2. Anatomic changes resulting in trichiasis or entropion may require surgical correction

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

A. Topical corticosteroids

1. Increased intraocular pressure, cataract, infection
2. Prevent by using lowest strength and dose possible, for only brief period when inflammation severe

B. Systemic tetracyclines

1. Photosensitization, gastrointestinal (GI) upset, azotemia, candidiasis
2. Use is contraindicated in children and pregnant or nursing women
3. Taper based on clinical response and use the lowest dose possible

VI. Describe disease-related complications

A. Corneal neovascularization and scarring

B. Madarosis

C. Trichiasis, marginal entropion

D. Cicatricial entropion

E. Chalazion

F. Reflex epiphora

VII. Describe appropriate patient instructions

- A. Eyelid hygiene will likely need to be performed indefinitely**
- B. Intermittently may need more aggressive therapy with topical or systemic medications**
- C. Return for recurrence of symptoms**
- D. Follow-up in 3 to 4 weeks when symptoms severe and new medication started**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids and Lacrimal System, Section

8: External Disease and Cornea, 2013-2014.

2. AAO, Focal Points: Diagnosis and Treatment of Chronic Blepharitis, Module #10, 1989.
3. AAO, Preferred Practice Patterns Committee, Cornea and External Disease Panel. Blepharitis Preferred Practice Pattern, 2008.

Epidermal inclusion cysts

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Epidermis lodged in the dermis due to trauma or comedone
2. Cyst wall lined by normal epidermis that produces keratin

B. Define the relevant aspects of epidemiology of this disease

1. Any age
2. Male or female

C. List the pertinent elements of the history

1. Previous skin trauma
2. Slowly enlarging, smooth, round nodule
3. May have episodes of inflammation and tenderness

D. Describe pertinent clinical features

1. Freely mobile cyst or nodule (lesion nonmobile if at lid margin)
2. Often has central pore (remaining pilar duct)
3. Erythema, edema, and tenderness if inflamed or infected
4. Positive transillumination

E. Describe appropriate laboratory testing for establishing the diagnosis

1. Usually diagnosed clinically
2. If lesion is excised, submit for histopathologic examination

II. Define the risk factors

A. Previous skin trauma

III. List the differential diagnosis

A. Pilar cyst

B. Molluscum contagiosum

C. Eyelid malignancy-basal, squamous or sebaceous cell

D. Eccrine or apocrine hidrocystoma

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

A. Describe medical therapy options

1. Corticosteroid injection, perilesional if severely inflamed

B. Describe surgical therapy options

1. Complete excision including the cyst wall to prevent recurrence
2. Marsupialization for small cysts
3. Incision and curettage if inflamed and fluctuant

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

A. Corticosteroid injection

1. Localized atrophy
2. Hypopigmentation
3. Use lowest dose needed

B. Surgical excision

1. Scar
2. Infection
3. Recurrence
4. Use careful surgical technique to decrease incidence of complications

VI. Describe disease-related complications

A. Gardner syndrome

1. Multiple epidermal inclusion cysts associated with colon cancer
2. Autosomal dominant inheritance

B. Cysts may become inflamed or infected

VII. Describe appropriate patient instructions

A. Return if lesion becomes inflamed, tender, or enlarges

B. If surgically excised, return for recurrence if patient desires re-excision

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.

Actinic keratosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Chronic, cumulative sun exposure

B. Define the relevant aspects of epidemiology of this disease

1. More common in the elderly, particularly those with fair complexion and a history of chronic sun exposure

C. List the pertinent elements of the history

1. Chronic sun exposure
2. Age
3. History of previous skin cancer

D. Describe pertinent clinical features

1. Flat, scaly, erythematous, keratotic plaque
2. Found on sun-exposed skin (face, dorsum of hands, head, neck, forearms)
3. May be pigmented
4. Can form a keratin horn
5. Sandpapery texture on palpation
6. Lesion will vary in size and texture over periods of time depending on the amount of keratinization which can flake off and reaccumulate over time

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Biopsy lesions to obtain histopathologic confirmation of diagnosis

II. Define the risk factors

A. Cumulative ultraviolet radiation (sun exposure)

B. Fair complexion

C. Advanced age

D. History of previous skin cancer

E. Immunocompromised state

III. List the differential diagnosis

A. Squamous cell carcinoma

B. Seborrheic keratosis

C. Basal cell carcinoma

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

A. Define medical therapy options

1. Topical 5% fluorouracil or imiquimod therapy (particularly for extensive facial lesions)
2. Chemical peel
3. Topical retinoid therapy
4. Photodynamic therapy

B. Define surgical therapy options

1. Surgical excision
2. Cryosurgery

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

A. Topical 5% fluorouracil

1. Extreme photosensitivity expected and normal
2. Severe erythema and crusting expected and normal
3. Careful patient selection and counsel prior to use
4. Recurrence or progression to squamous cell carcinoma

B. Surgical excision

1. Scarring or tissue loss can result in
 - a. Eyelid malposition
 - b. Trichiasis
 - c. Mechanical impairment of lacrimal drainage
2. Recurrence or progression to squamous cell carcinoma
3. Infection - rare
4. Complications can generally be prevented by careful surgical technique

C. Cryosurgery

1. Hypopigmentation
2. Scarring may occur especially if the freeze is too deep
3. Since no excision occurs, free surgical margins cannot be checked
4. Recurrence or progression to squamous cell carcinoma

VI. Describe disease-related complications

A. Risk of malignant transformation

1. Risk of transformation to squamous cell carcinoma is possible, generally considered 10-15%
2. Squamous cell carcinoma that develops from an actinic keratosis is usually very indolent but may be invasive

VII. Describe appropriate patient instructions

- A. Follow-up with a dermatologist for routine skin surveys**
- B. Explain that most patients will develop new lesions in the future**
- C. Have any new growths or changing lesions evaluated**
- D. Use protective clothing and sunscreen**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.

Nevi

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Benign, melanocytic neoplasm of the skin, commonly occurring on the eyelids or conjunctiva

B. Define the relevant aspects of epidemiology of this disease

1. Affects males and females
2. May be congenital or acquired

C. List the pertinent elements of the history

1. Pigmented lesion present since birth or acquired
2. Recent changes in the appearance of the lesion
3. Primary acquired melanosis (PAM) of the conjunctiva

D. Describe pertinent clinical features

1. Congenital nevus
 - a. Usually larger than acquired nevi, and may be of substantial size
 - b. Usually deeply pigmented
 - c. Border often irregular
 - d. Frequently covered with hair
 - e. A chance of malignant degeneration exists proportional to the size of the lesion
 - f. Kissing nevi are congenital nevi appearing in a symmetrical fashion on adjacent portions of the upper and lower eyelids, and are formed from melanocytic migration into the lids prior to separation which usually occurs by the 26th week of gestation
2. Acquired nevus
 - a. Junctional nevus
 - i. A pigmented lesion, usually appearing in adolescence, small (less than 0.5 cm), round, brown macule
 - ii. Histopathologic examination shows nests of melanocytes present at the dermal-epidermal junction. Initially the growth is radial
 - b. Compound nevus
 - i. In older children and adults, the lesion increases in thickness and becomes a dome shaped papule
 - ii. Histologic examination shows nests of melanocytes extending into the superficial dermis
 - c. Intradermal nevus

- i. Later in life, the lesion may become more dome-shaped, papillomatous or pedunculated
 - ii. As the adult loses skin and hair pigment in general, the nevus often becomes depigmented or amelanotic
 - iii. Histopathologic examination shows nests of nevus cells confined entirely to the dermis
- 3. PAM of the Conjunctiva
 - a. Has 13% malignant degeneration rate to melanoma
 - b. Development of conjunctival pigmentation during adulthood should be followed closely

E. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Usually diagnosed based on clinical appearance
- 2. Biopsy is performed for any suspicious lesion to confirm diagnosis and rule out malignancy

II. Define the risk factors

- A. Sun exposure in early childhood is thought to increase the number of nevi that develop

III. List the differential diagnosis

- A. Freckle
- B. Lentigo simplex
- C. Solar lentigo
- D. Malignant melanoma
- E. Seborrheic keratosis
- F. Squamous papilloma

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

A. Describe medical therapy options

- 1. Observation
- 2. Sunscreen use for prevention in early childhood
- 3. Return for evaluation of lesions demonstrating changes
 - a. Growth
 - b. Bleeding
 - c. Ulceration
 - d. Irregular borders

- e. Uneven pigmentation
- f. Changing color
- 4. Congenital nevi require more frequent follow-up and close monitoring for any suspicious changes

B. Describe surgical therapy options

- 1. Biopsy any suspicious lesion (those with irregular border, growth, bleeding, ulceration, variable pigmentation)
- 2. Surgical excision may also be performed for cosmesis or mechanical irritation

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

A. Scarring

- 1. Prevent with careful surgical technique

B. Recurrence after excision

- 1. Prevent with complete excision

VI. Describe disease-related complications

A. Development of malignant melanoma

- 1. More common in congenital nevi
- 2. Risk is directly related to size of congenital nevi
- 3. Close serial follow-up or excision warranted for congenital nevi

VII. Describe appropriate patient instructions

A. Watch for suspicious changes including growth, bleeding, ulceration, uneven pigmentation or irregular borders

B. Suspicious or large lesions measuring more than 6 mm should be biopsied.

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Orbit, Eyelids and Lacrimal System; Section 2: Fundamentals and Principles of Ophthalmology; Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 6: Pediatric Ophthalmology and Strabismus 2013-2014.
- 2. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993.
- 3. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
- 4. Shields et al., Primary Acquired Melanosis of the Conjunctiva: Experience with 311 Eyes. Trans Am Ophthalmol Soc. 2007 Dec;105; 61-72.

Lentigo maligna (Hutchinson melanotic freckle)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Possibly actinic induced hyperplasia of hyperpigmented and pleomorphic melanocytes

B. Define the relevant aspects of epidemiology of the disease

1. Age greater than 50
2. Living in areas with high ultraviolet (UV) exposure

C. List the pertinent elements of the history

1. Previous history of actinic pre-cancerous or malignant skin lesions
2. Pigmented skin lesion in sun exposed area that has slowly increased in size or changed in color

D. Describe pertinent clinical features

1. The term "lentigo maligna" is still used today by pathologists to refer to a melanoma in situ that occurs on skin damaged severely by sunlight
 - a. As is true for all melanomas *in situ* on all anatomic sites, the lesions tend to be
 - i. Flat or very slightly elevated
 - ii. Asymmetrical
 - iii. Notched in outline
 - iv. Uneven in pigmentation with colors that range from light brown to black
 2. Macular pigmented skin lesion (flat, not elevated nodule)
 3. Asymmetric lesion with irregular borders
 4. Variation in pigment (light brown to black)
 5. Frequently located in malar area
 6. 30 - 50 % of lesions will progress to vertically invasive nodular melanoma
- ### E. Describe appropriate testing and evaluation for establishing the diagnosis
1. Pathological examination of the entire excised specimen
 2. Benign melanocytes in basal layer of the epidermis
 3. Signs of sun damage, namely, marked solar elastosis in the upper part of the dermis

II. Define the risk factors

A. Sun exposure

III. List the differential diagnosis

- A. Malignant melanoma**
- B. Benign freckle**
- C. Lentigo senilis, small 3-5 mm uniformly pigmented lesions in sun exposed areas**
- D. Benign nevi (1-3 mm), usually occur in childhood and increase at puberty**

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

- A. Describe medical therapy options**
 - 1. Consideration of topical chemotherapeutic agents
 - 2. Possible observation for small macular lesions
- B. Describe surgical therapy options**
 - 1. Total surgical excision utilizing permanent sections to determine complete removal
 - 2. Close observation for recurrence and onset of new lesions

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

- A. Compromise of the functional integrity of the eyelid (cicatricial ectropion, lagophthalmos, ocular exposure)**

VI. Describe disease-related complications

- A. Malignant transformation into melanoma with distant metastasis**

VII. Describe appropriate patient instructions

- A. Self examination for recurrence or new lesions**
- B. Limit sun exposure (hat, topical sun block)**
- C. Routine periodic follow up evaluations**

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
- 2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.

Basal cell carcinoma of the eyelid

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Neoplastic transformation of normal skin cells into a malignant cell line

B. Define the relevant aspects of epidemiology of the disease

1. More common with decreased racial skin pigmentation (red or blonde hair, blue eyes, fair skin)
2. Living in areas with high ultraviolet (UV) exposure
3. Basal cell nevus syndrome

C. List the pertinent elements of the history

1. Unprotected extensive sun exposure
2. Skin radiation for inflammatory (acne) or malignant skin conditions
3. Previous skin cancer
4. Non healing, ulcerated skin lesion

D. Describe pertinent clinical features

1. Often elevated, but may be flat
2. Central ulceration possible
3. Irregular margins
4. Often elevated pearly margins with telangiectasias
5. Non tender
6. Loss of normal skin or eyelid margin architecture (i.e., loss of lashes)
7. Occasionally pigmented or cystic
8. Morpheaform basal cell carcinoma is an uncommon variant
 - a. Tumor cells induce a proliferation of fibroblasts within the dermis
 - b. An increased collagen deposition (sclerosis) that clinically resembles a scar
 - c. The tumor appears as a white or yellow, waxy, sclerotic plaque that rarely ulcerates
 - d. Eyelid notching if along eyelid margin

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Incisional or excisional biopsy depending on clinical presentation

II. Define the risk factors

A. Sun exposure

- B. Previous skin cancer**
- C. Previous skin therapeutic radiation**
- D. Basal cell nevus syndrome**
 - 1. Autosomal dominant
 - 2. Multiple basal cell skin cancers - these tumors may have the appearance of nevi
 - 3. Other abnormalities including odontogenic jaw cysts, palmar and plantar pits, musculoskeletal anomalies
- E. Xeroderma pigmentosum**
- F. Skin type**
 - 1. Skin that easily sunburns
 - 2. Fair skin with blue eyes (though can occur in all skin types)
- G. Racial predilection (i.e. Irish descent), usually reflecting skin type**

III. List the differential diagnosis

- A. Squamous cell skin cancer**
- B. Sebaceous cell carcinoma**
- C. Other adenocarcinomas of dermal appendages**
- D. Actinic keratosis**
- E. Chronic inflammatory blepharitis**
- F. Keratoacanthoma**
- G. Papilloma**

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

- A. Describe medical therapy options**
 - 1. Incisional biopsy mandatory before consideration of non-surgical alternatives
 - 2. Cryotherapy for small lesions and in patients that are not surgical candidates
 - 3. Topical therapy (imiquimod, Aldara) in patients that are not good surgical candidates. May cause irritation with ocular exposure
 - 4. Radiation for palliative treatment or in patients who are not good surgical candidates
 - 5. Oral therapy with Hedgehog pathway inhibitors (e.g. vismodegib, (Erivedge)) may be an option in patients who are not candidates for surgery or radiation
 - 6. Photodynamic therapy indicated for basal cell nevus syndrome or an extensive lesion that had failed conventional therapies
- B. Describe surgical therapy options**

1. Incisional biopsy to confirm clinical diagnosis
2. Treatment of choice is total excision of the lesion with either intraoperative frozen section control of tumor margins or Mohs' micrographic surgery
3. Reconstruction to re-establish function and aesthetic appearance

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

A. Basal cell carcinoma (BCC) recurrence

1. Regular follow-up for early detection of any recurrence or new tumor

B. Eyelid/facial dysfunction with ocular surface compromise or other ocular/facial functional compromise (tearing, diplopia, ptosis), especially with large tumors

1. Appropriate reconstruction is directed to restore adequate function
2. Secondary procedures may be used to restore function
 - a. For example, loss of the canalicular system may be addressed with a Jones tube, but only when adequate follow-up insures that recurrence is unlikely
3. Exenteration can be considered if clinical orbital extension of tumor is confirmed histopathologically or radiologically

C. Unacceptable eyelid/facial aesthetic result

1. Reconstruction is designed to provide the best cosmetic result possible
 - a. Procedures such as local skin flaps, rather than skin grafts, are chosen, when possible, to provide the best aesthetic result
 - b. Additional procedures may be undertaken to correct functional or aesthetic issues

VI. Describe disease-related complications

- A. Direct tumor extension into orbit with functional compromise (vision, diplopia, pain)**
- B. Direct tumor extension in adjacent facial tissues (lacrimal drainage, perineural spread)**
- C. Rarely intracranial extension**

VII. Describe appropriate patient instructions

A. Protection from sun exposure

1. Physical barrier (hat, long sleeves, long pants)
2. Topical sun-block with UVA and UVB protection
 - a. Physical sun-block (zinc oxide or titanium dioxide)
 - b. Chemical sun-block

B. Appropriate medical evaluation of any non-healing skin lesions

C. Routine periodic follow up examination of all sun exposed skin areas

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.
3. Bernardini FP. Management of malignant and benign eyelid lesions. *Curr Opin Ophthalmol* 2006;17:480-4.
4. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
5. AAO, Focal Points: Periorbital Skin Cancers: The Dermatologist's Perspective, Module #1, 2006.

Squamous cell carcinoma of the eyelid

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Neoplastic transformation of normal epidermal skin cells into a malignant cell line

B. Define the relevant aspects of epidemiology of the disease

1. Decreased racial skin pigmentation (red or blonde hair, blue eyes, fair skin)
2. Living in areas with high ultraviolet (UV) exposure

C. List the pertinent elements of the history

1. Unprotected extensive sun exposure
2. Skin radiation for inflammatory (acne) or malignant skin conditions
3. Previous skin cancer
4. Non healing, hyperkeratotic skin lesion with inflammation and/or bleeding
5. Most cutaneous squamous cell carcinomas arise from pre-existing lesions such as actinic keratosis, Bowen disease, radiation dermatoses, burn scars, and inflammatory lesions

D. Describe pertinent clinical features

1. Hyperkeratotic macular (flat) or nodular (elevated) skin lesion
2. May have significant inflammatory component
3. Later phases may have ulceration
4. Associated sun damaged skin
5. Loss of normal eyelid architecture (i.e., loss of lashes, eyelid margin distortion)
6. Perineural extension - centripetal perineural spread
7. Regional lymph node spread

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Incisional or excisional biopsy depending on clinical presentation
2. Computed tomography (CT) or magnetic resonance imaging (MRI) with enhancement if intraorbital or intracranial spread is suspected

II. Define the risk factors

A. Sun exposure

B. Previous skin cancer

C. Previous skin therapeutic radiation

D. Psoralen therapy

III. List the differential diagnosis

- A. Basal cell carcinoma - nodular, morpheaform (sclerosing)**
- B. Actinic keratosis**
- C. Chronic inflammatory blepharitis**
- D. Keratoacanthoma (considered to be a low grade squamous cell carcinoma by some pathologists)**
- E. Papilloma**
- F. Sebaceous cell carcinoma**
- G. Nonspecific, chronic dermatitis**
- H. Inverted follicular keratosis**
- I. Pseudoepitheliomatous hyperplasia**

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

A. Describe medical therapy options

1. Incisional biopsy mandatory before consideration of non-surgical alternatives
2. Radiation for palliative treatment only, usually radio-resistant
3. Systemic chemotherapy for widespread unresectable, local disease or distant metastasis
4. Cryotherapy for individuals who refuse surgery or are poor surgical candidates

B. Describe surgical therapy options

1. Incisional biopsy
2. Treatment of choice is total excision of the lesion with either intraoperative frozen section tumor margin control or Mohs' micrographic surgery
3. Reconstruction to re-establish function and aesthetics

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

- A. Squamous cell carcinoma (SCC) recurrence**
- B. Eyelid/facial dysfunction with ocular surface compromise or other ocular/facial functional compromise (tearing, diplopia, ptosis, lagophthalmos, cicatricial eyelid ectropion/retraction)**
- C. Unacceptable eyelid/facial aesthetic result**

VI. Describe disease-related complications

- A. Direct tumor extension into orbit with functional compromise (vision loss, diplopia, pain)**
- B. Perineural spread - tic douloureux, ophthalmoplegia**
- C. Direct tumor extension in adjacent facial tissues (lacrimal drainage, perineural spread)**

D. Regional metastasis

VII. Describe appropriate patient instructions

- A. Avoid unprotected sun exposure (hat, topical sun block)**
- B. Appropriate medical evaluation of any non-healing skin lesions**
- C. Routine periodic follow up examination of all sun exposed skin areas**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993.
3. AAO, Focal Points: Periorbital Skin Cancers: The Dermatologist's Perspective, Module #1, 2006.

Sebaceous adenocarcinoma of the eyelid

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Neoplastic transformation of normal sebaceous cells (meibomian glands, glands of Zeis, caruncle, skin) into a malignant cell line
2. Rarely, it may be encountered in children who have received radiation therapy for retinoblastoma or cavernous hemangioma of the face

B. Define the relevant aspects of epidemiology of the disease

1. More common in older patient groups, but can be seen in any age group
2. More frequent in females
3. More frequent in the upper eyelid

C. List the pertinent elements of the history

1. Persistent nodular lesion of the eyelid meibomian glands masquerading as chalazion
2. Persistent blepharoconjunctivitis (masquerading as benign inflammatory blepharitis)

D. Describe pertinent clinical features

1. Unilateral chronic blepharitis that does not respond to medical management
2. Infiltrative destruction of the normal eyelid margin architecture
3. Loss of eyelashes
4. Yellow coloration of retained lipid material
5. Multicentric origin results in noncontiguous tumor
6. Intraepidermal extension results in widespread papillary elevation of tarsal conjunctiva (pagetoid spread)
7. Can mimic leukoplakia, ocular mucous membrane (cicatricial) pemphigoid, squamous cell carcinoma of the conjunctiva, and carcinoma *in situ*

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. A generous representative specimen is helpful in making the diagnosis
 - a. Full thickness eyelid biopsy can avoid sampling of superficial conjunctival reactive inflammation
 - b. Discussion with the pathologist regarding the suspicion of sebaceous adenocarcinoma may be helpful to diagnose this uncommon tumor
 - c. The most common site of sebaceous cell carcinoma on the body is the eyelid.
 - d. Special lipid stains may be helpful

II. List the differential diagnosis

- A. Chalazion (masquerade)**
- B. Blepharoconjunctivitis (masquerade)**
- C. Basal cell or squamous cell carcinoma (skin cancer)**
- D. Superior limbic keratoconjunctivitis**
- E. Ocular cicatricial pemphigoid**

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

A. Describe medical therapy options

- 1. Radio-resistant, palliation only

B. Describe surgical therapy options

- 1. Complete wide surgical excision with frozen section control or Mohs' micrographic surgery
- 2. Skip lesions possible, pagetoid spread, multicentric origination make total excision difficult
- 3. Careful, long term monitoring is needed
- 4. Sentinel lymph node biopsy may be helpful in identifying early, subclinical microscopic regional lymph node metastasis
- 5. Neck dissection for regional adenopathy
- 6. Orbital exenteration for widespread local disease

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

- A. Local tumor recurrence requiring repeat surgery (exenteration)**
- B. Regional tumor recurrence (chemotherapy and or radiotherapy)**
- C. Eyelid/facial dysfunction with ocular surface compromise or other ocular/facial functional compromise (tearing, diplopia, ptosis, lagophthalmos, cicatricial eyelid ectropion/retraction)**

V. Describe disease-related complications

- A. Direct tumor extension into orbit with functional compromise (vision, diplopia, pain)**
- B. Direct tumor extension in adjacent facial tissues (lacrimal drainage, perineural, lymphatic)**
- C. Regional metastasis**

VI. Describe appropriate patient instructions

- A. Prolonged follow up needed to monitor possible tumor recurrence**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Periorbital Skin Cancers: The Dermatologist's Perspective, Module #1, 2006.

Malignant melanoma of the eyelid

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Neoplastic transformation of normal skin melanocytes into a malignant cell line

B. Define the relevant aspects of epidemiology of the disease

1. Decreased racial skin pigmentation (blonde hair, blue eyes)
2. Living in areas with high ultraviolet (UV) exposure

C. List the pertinent elements of the history

1. Unprotected extensive sun exposure
2. Pre-existing acquired pigmented lesion (lentigo maligna or nevus)
3. May occur de novo
4. Historical change in pigment density, distribution, or color (surface bleeding, increase in size and thickness)
5. New nodule formation in a previous flat pigmented lesion

D. Describe pertinent clinical features

1. Variation in pigmentation density and/or color
2. Irregular, vague border with pigmentary infiltration into "normal" skin
3. Elevated pigmented nodule in a flat pigmented area
4. May present with multiple lesions near primary site

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Incisional biopsy incorporating the maximum thickness of the lesion (punch biopsy may be useful)

F. Preoperative metastatic work-up for tumors greater than 1.5 mm thick

1. Systemic evaluation for liver, lung and brain metastatic disease
2. Increased incidence of metastatic disease with increased melanoma thickness or deeper cutaneous invasion
 - a. Thin lesions <0.75 confer 98% 5 year survival
 - b. Thicker lesions >4mm with ulceration confer a survival rate of less than 50% at 5 years

II. Define the risk factors

A. Sun exposure

B. Decreased racial skin pigmentation

C. Genetic predisposition

- D. Environmental mutagens

III. List the differential diagnosis

- A. Lentigo maligna
- B. Pigmented seborrheic keratosis
- C. Benign nevus
- D. Lentigo senilis (multiple small 3-5 mm uniformly pigmented lesions)
- E. Pigmented basal cell carcinoma

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

- A. Describe medical therapy options
 - 1. Immunotherapy
- B. Describe surgical therapy options
 - 1. Possible sentinel lymph node mapping and biopsy
 - 2. Wide surgical removal with permanent histologic monitoring of resected margins

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

- A. Melanoma recurrence, local or metastatic
- B. Eyelid/facial dysfunction with ocular surface compromise or other ocular/facial functional compromise (tearing, diplopia, ptosis, lagophthalmos, cicatricial eyelid ectropion/retraction)

VI. Describe disease-related complications

- A. Direct tumor extension into orbit with functional compromise (vision, diplopia, pain)
- B. Direct tumor extension in adjacent facial tissues (lacrimal drainage, perineural)
- C. In-transit and regional metastasis
- D. Distant metastasis

VII. Describe appropriate patient instructions

- A. Avoid unprotected sun exposure (hat, topical sun block)
- B. Appropriate medical evaluation of any pigmented skin lesions that have a change in character (pigment density, color, size, thickness)
- C. Routine periodic follow up examination of all sun exposed skin areas
- D. Systemic evaluation including liver function tests, palpate lymph nodes

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.164.
3. Boulos PR, Rubin PA. Cutaneous melanomas of the eyelid. *Semin Ophthalmol* 2006;21:195-206.

Masquerading syndrome for eyelid tumors

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. A malignant neoplastic disease that superficially appears to be benign leading to a delay in the correct diagnosis

B. List the pertinent elements of the history

1. Suspicious characteristics
2. Non healing presumed benign lesion or inflammatory eyelid lesion
3. Presumed benign inflammation is limited to one eyelid or only one eyelid area
4. Multiple recurrent chalazia
5. Lack of clinical improvement despite maximal medical therapy

C. Describe pertinent clinical features

1. Loss of normal eyelid margin anatomical features, including loss of eyelashes
2. Atypical Meibomian gland changes with altered anatomic gland appearance
3. Cicatricial rotation of eyelid margin (entropion or ectropion)

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Biopsy with histologic examination
 - a. Consider conjunctival map biopsies
2. Full thickness eyelid biopsy is indicated to avoid superficial inflammatory change overlying a potential malignant lesion
 - a. Discussion with pathologist may help in identifying a rare eyelid tumor such as sebaceous cell carcinoma

II. List the differential diagnosis

A. Sebaceous adenocarcinoma

B. Blepharoconjunctivitis

C. Basal cell or squamous cell skin cancer

D. Chalazion

E. Superior limbic keratoconjunctivitis

F. Granulomatous inflammatory disorders: sarcoidosis, Wegener granulomatosis

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

A. Describe surgical therapy options

1. Full thickness eyelid biopsy
2. Definitive surgical resection of the eyelid malignancy utilizing either Mohs' micrographic surgery or intraoperative frozen sections
3. Reconstruction of eyelid/soft tissue defect

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

- A. Local tumor recurrence requiring repeat surgery**
- B. Distant tumor recurrence (chemotherapy might be needed)**
- C. Eyelid/facial dysfunction with ocular surface compromise or other ocular/facial functional compromise (tearing, diplopia, ptosis, lagophthalmos, cicatricial eyelid ectropion/retraction)**

V. Describe disease-related complications

- A. Direct tumor extension into orbit with functional compromise (vision, diplopia, pain)**
- B. Direct tumor extension in adjacent facial tissues (lacrimal drainage, perineural, lymphatic)**
- C. Distant metastasis**

VI. Describe appropriate patient instructions

- A. Prolonged follow up needed to monitor possible tumor recurrence**
- B. Self examination of regional lymph nodes**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Mohs micrographic surgery for malignant eyelid tumors

I. List the indications/contraindications

A. Relative indications

1. Basal cell or squamous cell carcinoma of eyelids or periorcular area

B. Stronger indications

1. Basal cell or squamous cell carcinoma that are recurrent, located in the medial canthus, supraorbital nerve region or arise in a previously irradiated area

C. Relative contraindications

1. Malignant melanoma and sebaceous adenocarcinoma may be cleared by "slow Mohs" which is a modified technique using permanent sections.

D. Summary of benefits of Mohs surgery

1. Potential tissue preservation
2. Decreased recurrence rate

II. Describe the pre-procedure evaluation

A. Patient history

1. Ocular symptoms of epiphora, decreased vision, diplopia, and pain will help determine tumor extent
2. Time course of tumor
3. Prior tumor excision history
 - a. Mohs excision or no Mohs excision
4. Incisional biopsy result
5. History of radiation and cryotherapy for skin cancer in the region

B. Clinical examination

1. Size and location of tumor will aid in determination of presumed tumor extent
2. Immobile lesion by palpation suggests deep tissue involvement
3. Medial canthal location frequently requiring deeper surgical resection
4. Orbital functional evaluation including
 - a. Vision
 - b. Pupillary exam

- c. Ocular motility
- d. Proptosis
- 5. Computed tomography (CT) or magnetic resonance imaging (MRI) if orbital involvement suspected
- 6. Lacrimal drainage assessment, if clinical involvement suspected

C. Preoperative assessment

- 1. Devise a tentative plan for soft tissue reconstruction following Mohs' surgery, may need to be revised
- 2. Aim for stable medical health, including normal coagulation studies, when appropriate
- 3. No aspirin compounds, must consider risk of discontinuing antiplatelet medications in patients with history of cardiac problems, stroke or venous thrombosis

III. List the alternatives to this procedure

- A. Primary nodular basal cell carcinoma (BCC) can be removed with standard tumor excision with frozen section control of resected margins
- B. Morpheaform and medial canthal lesions should be treated by Mohs' surgery
- C. Radiation and cryotherapy are not good alternatives to complete surgical excision
- D. Orbital exenteration may be indicated for orbital involvement

IV. Describe the instrumentation, anesthesia and technique

- A. Local IV sedation, or general anesthesia
- B. Standard surgical instruments for facial surgery
- C. Reconstructive techniques as needed

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

A. Intraoperative

- 1. Tumor extension beyond anticipated area requiring more extensive reconstruction than anticipated
- 2. Hemorrhage
- 3. Intracranial tumor extension
- 4. Technical map interpretation and topography problems resulting in false negative tissue report

B. Postoperative

- 1. Positive margins on permanent sections requiring additional soft tissue resection
- 2. Poor functional result following eyelid reconstruction with corneal exposure, epiphora, decreased vision, lagophthalmos, eyelid malposition

3. Soft tissue infection
4. Recurrence of skin cancer

VI. Describe the follow-up care

- A. Confirmation of eyelid reconstruction functional integrity with good globe protection**
- B. Review permanent histologic report to confirm tumor free status**
- C. Suture removal**
- D. Periodic patient evaluation for tumor recurrence**

VII. Describe appropriate patient instructions

- A. Tumor prevention by patient use of sun blocking topicals and protective clothing (hat)**
- B. Follow up with dermatologist**
- C. Discuss the warning signs of tumor recurrence**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 9. Surgical Anatomy of the Ocular Adnexa, 1996.
3. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993.
4. AAO, Focal Points: Periorbital Skin Cancers: The Dermatologist's Perspective, Module #1, 2006.

Blunt trauma to the eyelids

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Traumatic injury to eyelids, midface, orbit, or eye
2. Source of injury may be any event that results in the transfer of kinetic or thermal energy into soft tissue or bone

B. Define the relevant aspects of epidemiology of the disease

1. Participation in any activity or occupation with exposure to potential traumatic environment
 - a. Auto, bike, sports, fights, occupational hazards, etc.

C. List the pertinent elements of the history

1. Usually blunt trauma to the face from any source
2. Age of patient. Witnesses in child or infant
3. Circumstances surrounding the injury
4. Visual loss
5. Neurologic status. Loss of consciousness
6. Foreign bodies
7. Contact lens wearer
8. Animal bites
9. Human bites
10. Thermal or chemical burns
11. Tetanus immunization status

D. Describe pertinent clinical features

1. Patient's general condition is given priority
 - a. Adequate airway
 - b. Hemorrhage and shock
 - c. Neurological status
 - d. Extent of extraorbital injuries
2. Rule out ruptured globe or other intraocular injury - hyphema, subluxed lens, cataract, commotio retinae
3. Possible occult penetrating eyelid, lacrimal, ocular, or orbital injury, note if there is any laceration that extends through the orbital septum
4. Hemorrhagic eyelid edema

5. Possible facial or orbital fractures
6. Possible traumatic optic neuropathy with loss of vision

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Complete ocular and orbital evaluation to determine ocular or orbital injury
2. Axial and coronal computed tomography (CT) scan to evaluate possible facial and orbital fractures or hemorrhage, head or spinal injury, if clinically indicated
3. Magnetic resonance imaging (MRI) scan is generally not needed.
 - a. In unusual cases, such as suspected non metallic foreign body or intrasheath hematoma, MRI may be helpful

II. Define the risk factors

- A. Participation in high risk activities, but can occur at any time, anywhere**

III. List the differential diagnosis

- A. Occult malignancy**
- B. Kaposi sarcoma with hemorrhage**
- C. Soft tissue or blood malignancy with pathologic spontaneous hemorrhage**
- D. Other bleeding disorders**
- E. Non-accidental child trauma**
- F. The diagnosis of rhabdomyosarcoma may be somewhat delayed because of the appearance of possible blunt trauma to the periocular region**
- G. Lymphangioma with hemorrhage**
- H. Inflammation**
- I. Infection**

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

A. Describe medical therapy options

1. Observation only
2. Medical management of ocular injuries
3. Possible corticosteroid treatment of optic neuropathy
4. Ice packs, head of bed elevation
5. Possible oral antibiotics and avoidance of nose blowing if fractures are present
6. Avoid aspirin intake in the event surgery is needed

B. Describe surgical therapy options

1. Repair of ruptured globe, if indicated
2. Fracture repair, if indicated
3. Drainage of orbital hemorrhage, if indicated
4. Eyelid laceration repair
 - a. Careful layered closure to prevent eyelid malposition and deformity
 - b. Margin repair with sutures through gray line and lash line. Sutures may be used and secured and away from the globe to avoid corneal abrasions. Alternatively absorbable sutures can be considered, particularly in infants, children and uncooperative adults where suture removal may be challenging.
 - c. Tarsal plate closure with interrupted partial thickness absorbable sutures, not extending through the conjunctiva
 - d. Orbicularis muscle closure with absorbable sutures if desired
 - e. Skin closure with interrupted absorbable or permanent sutures
5. Repair of lacrimal system laceration with stent

V. Describe disease-related complications

A. Long term sequelae

1. Corneal abrasion from exposed conjunctival suture(s) in eyelid laceration repair
2. Eyelid margin distortion or notching from full-thickness laceration repair
3. Eyelid malposition (ptosis most common)
4. Ocular or orbital injuries

VI. Describe appropriate patient instructions

- ### **A. If acute, complete ocular or orbital evaluation difficult because of hemorrhagic edema, repeat evaluation indicated after edema decreases**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.
3. AAO, Focal Points: Management of Eyelid Trauma, Module #10, 1996.
4. AAO, Focal Points: Eyelid Trauma, Module #10, 1991.

Penetrating trauma to the eyelids

I. Describe the approach to establishing the diagnosis

A. Describe the mechanism of injury

1. Traumatic injury to the eyelids, midface, orbit, or eye
2. Determine the object causing the injury, may be any event with a skin laceration
3. Ascertain the circumstances of injury
4. Determine extent of injury, can include the eye, orbit, midface structures, or brain
5. Evaluate for injury of adjacent structures consider trajectory and kinetic force of the object

B. Define the relevant aspects of epidemiology of the disease

1. Participation in any activity or occupation with exposure to potential traumatic environment, i.e., auto, bike, sports, fights, occupational hazard - nail gun, power tools
2. Wearing of protective eyewear or other protective device , i.e., goggles, welding helmet

C. List the pertinent elements of the history

1. Sharp, penetrating injury may easily extend into globe or brain
2. History of vision loss, or neurologic deficit mandates further evaluation
3. Penetrating instrument, wood or pencil injury may leave foreign body (FB) fragments in orbit
4. Child's account of injury may be highly inaccurate

D. Describe pertinent clinical features

1. Complete ocular and orbital evaluation including
 - a. Visual function i.e., vision, pupils, color vision, visual field
 - b. Diplopia secondary to orbital cicatricial changes, EOM injury, or neurologic injury
 - c. Ocular motility
 - d. Eyelid ptosis
 - e. Dilated fundus examination
 - f. Neurologic examination
2. Evaluation of the penetration site
 - a. Explore the penetration site for retained foreign body or inoculation of debris
 - b. Orbital fat in wound implies penetration of the septum and orbital involvement
 - c. Inspect for violation of the orbital septum, which suggests orbital involvement
 - d. Penetration of the orbital septum by a small instrument (ice pick) may not show fat in the wound, but even a small laceration without fat in the wound does not negate deep penetration ("ice pick stab")

3. Examination of the penetrating object if available for broken pieces, debris, depth of injury
4. Determine the location of injury on eyelid which can predict injury to adjacent structures
 - a. Medial lid trauma may involve lacrimal system
 - b. Superior sulcus injury may involve the levator superior rectus complex
5. History of finger or object engaging lower eyelid with lateral traction and subsequent "laceration" in the medial canthal area is almost certainly an avulsive injury with canalicular involvement
 - a. Typical of dog bite injuries
6. Trivial appearing lacerations in the medial canthal area may be associated with canalicular lacerations
7. Alteration in mentation and cerebrospinal fluid (CSF) rhinorrhea
8. Eyelid or orbital emphysema indicates penetration of the sinuses

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan to evaluate possible facial and orbital fractures, if clinically indicated
 - a. Intracranial air implies intracranial penetration
2. Magnetic resonance imaging (MRI) scan to evaluate possible organic foreign body (after CT or plain films to rule out metallic foreign body)
3. Visual field test, if traumatic optic neuropathy suspected
4. Lacrimal evaluation to rule out canalicular laceration

II. Define the risk factors

- A. Participation in high risk activities, but can occur at any time, anywhere**
- B. Injury is often the result of eyewear or eye protection**
- C. Dog bite injury often causes an avulsion of the eyelid with canalicular tear**

III. List the differential diagnosis

- A. Self inflicted injury needs immediate psychiatric evaluation**

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

- A. Describe medical therapy options**
 1. Antibiotic prophylaxis especially for suspected intracranial penetration
 2. Tetanus toxoid status
- B. Describe surgical therapy options**
 1. Exploration of wound

2. Copious saline irrigation
3. Repair of levator laceration, without resecting or advancing the levator aponeurosis
4. Repair of extraocular muscle (EOM) lacerations
5. Repair of orbital septum not indicated
6. Anatomic repair of eyelid margin with sutures tied anteriorly and deep sutures in tarsal plate placed partial thickness without incorporating the conjunctiva
7. Canalicular repair with silicone intubation
8. Eyelid canthal repositioning with appropriate posterior attachment to posterior lacrimal crest (medial canthal tendon) and internal aspect of the lateral orbital rim (lateral canthal tendon)
9. Neuro-surgical consultation for intracranial injuries
10. Removal of an orbital foreign body
 - a. All organic foreign bodies should be removed
 - b. Large metallic foreign bodies should be removed
 - c. Small foreign bodies less than 1-2mm may not need to be removed depending upon position.

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

- A. Epiphora secondary to overlooked lacrimal injury**
- B. Traumatic ptosis**
 1. Observe since frequent complete resolution in 6 months
- C. Eyelid malposition (ectropion, entropion), frequently cicatricial type, associated with soft tissue loss, may require grafting or complex reconstruction to correct**
- D. Intracranial injury requires urgent neurological evaluation**

VI. Describe appropriate patient instructions

- A. If acute, complete ocular or orbital evaluation is difficult because of hemorrhagic edema, repeat evaluation indicated after edema decreases or as soon as possible**

Additional Resources

1. AAO, Basic and Clinical Sciences 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.

Eyelid defects not involving the eyelid margin

I. List the indications/contraindications

A. Indications

1. Soft tissue defects of anterior lamella (skin and orbicularis muscle) or posterior lamella (conjunctiva and tarsus)
2. Posttrauma, postsurgical, (tumor removal), or congenital

II. Describe the pre-procedure evaluation

A. The surgeon's choice of procedure will depend upon:

1. Age of the patient
2. Anatomic character (position, laxity) of the eyelids
3. Size and position of the defect
4. Personal experience and preference

B. Patient history

1. Exposure symptoms secondary to lagophthalmos or eyelid malposition
2. Tumor type with risk of possible recurrence

C. Clinical examination

1. Size and location of defect
2. Full thickness or partial thickness
3. Status of adjacent tissue; laxity, solar changes, etc.
4. Physical examination and possible computed tomography (CT) scan to evaluate eye and orbit for possible foreign bodies

D. Goals of treatment

1. Avoid distortion of major facial anatomical features: eyelid margin, eyebrow, canthi
2. Provide adequate vertical eyelid tissue
3. Provide for adequate eyelid closure
4. Provide a smooth, epithelialized, non-keratinizing internal eyelid surface
5. May reconstruct either the anterior or posterior eyelid lamella with a free graft, but not both
 - a. One of the layers must provide the blood supply
6. Minimize vertical tension to avoid eyelid retraction or ectropion

7. Avoidance of vertical tension may require a vertically oriented wound closure
8. Maintain sufficient and anatomic canthal fixation
9. Match like tissue to like tissue
 - a. If a skin graft is needed, utilize similar skin donor site (eyelid, retroauricular, preauricular)
10. Choose the simplest technique, consider
 - a. Primary closure
 - b. Primary closure with undermining
 - c. Mobilization of adjacent tissue using a local skin-muscle or skin flap
 - d. Full-thickness skin graft

III. List the alternatives to this procedure

- A. Healing by second intention, if defect is anterior lamella only and not near major facial anatomic feature: eyelid margin, eyebrow, or canthi

IV. Describe the instrumentation, anesthesia and technique

- A. Local, IV sedation, or general anesthesia
- B. Standard surgical instruments for eyelid and facial surgery
- C. Deep absorbing sutures as needed to support and fixate deep tissues
- D. Skin sutures to evert the wound margin

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

- A. Intraoperative
 1. Hemorrhage
 2. Injury to donor site, contralateral donor site eyelid malposition
- B. Postoperative
 1. Eyelid retraction with lagophthalmos and corneal exposure
 2. Eyelid ptosis with an adynamic thickened upper eyelid
 3. Epiphora secondary to lower eyelid ectropion with punctal malposition
 4. Poor aesthetic appearance with poor skin graft match to adjacent tissue
 5. Soft tissue infection or necrosis
 6. Corneal abrasion from exposed conjunctival sutures

VI. Describe the follow-up care

- A. Topical and oral antibiotics, if appropriate**
- B. Confirmation of eyelid functional integrity with adequate globe protection**
- C. Suture removal**

VII. Describe appropriate patient instructions

- A. Signs and symptoms of infection, post operative hemorrhage, wound dehiscence**
- B. Discussion of underlying disease process, i.e., tumor recurrence**
- C. Avoidance of trauma**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Eyelid Trauma, Module #10, 1991, p.2-3.
3. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.

Concepts of upper eyelid reconstruction

I. List the indications and contraindications

A. Indications

1. Full thickness eyelid defects following tumor resection or trauma
2. Congenital coloboma

B. Contraindications

1. Active soft tissue infection

II. Describe the pre-procedure evaluation

A. Patient history

1. Etiology of the defect
 - a. Other medical conditions
 - b. Diabetes mellitus
 - c. Post radiation
2. History of self-trauma
3. Medications
4. Monocular status
5. Sicca

B. Clinical examination

1. Ocular evaluation
2. Facial nerve function (orbicularis muscle function)
3. Size of defect
4. Laxity of adjacent tissue to determine ability to advance remaining eyelid tissues to close the defect
5. Corneal and tear film integrity
6. Involvement of lacrimal system
7. Verify integrity of lacrimal outflow system

C. Goals of surgery

1. Re-establish functional integrity of the eyelid
 - a. Good aesthetic result is desired, but secondary to eyelid function
 - b. Provide adequate eyelid closure

2. Avoid distortion of major facial anatomical features, if possible; eyelid margin, medial and lateral canthi
3. Provide adequate vertical eyelid tissue to protect cornea
4. Provide adequate horizontal eyelid tissue by release of the upper limb of the lateral canthal tendon to minimize defect size before planning flap size
5. Provide a smooth, epithelialized internal eyelid surface
6. Minimize vertical tension to avoid upper or lower eyelid retraction or ectropion with ocular exposure
7. Maintain sufficient and anatomic medial and lateral canthal fixation
8. Choose the simplest technique
 - a. Primary closure
 - b. Primary lid margin closure with canthotomy and cantholysis
 - c. Primary lid margin closure with canthotomy, cantholysis and Tenzel semicircular rotational flap to advance lateral portion of the eyelid and repair margin primarily
 - d. Free tarsal conjunctival composite graft from the contralateral upper eyelid with an overlying skin orbicularis muscle flap
 - e. Transposition (Cutler-Beard) full thickness eyelid flap from the lower to upper eyelid
9. Reconstruction of lacrimal system as necessary (See Lacrimal outflow system trauma)

III. List the alternatives to this procedure

- A. Temporary patching to protect the cornea**

IV. Describe the instrumentation, anesthesia and technique

- A. Standard surgical instruments for eyelid and facial surgery**
- B. Local, intravenous (IV) sedation, or general anesthesia**
- C. Determination of sufficient horizontal laxity for direct closures**
- D. If direct closure not possible flap or graft should be positioned with either permanent or absorbable sutures**
- E. Considerations**
 1. If standard Cutler-Beard flap will not make a wide enough flap the lateral canthal tendon may be released
 2. Divide Cutler-Beard flap 2-4 weeks after operation
- F. Re-approximation of the eyelid margin anatomical landmarks is essential**
- G. Eyelid margin closure with 2 or 3 sutures through the eyelash line and gray line (optional)**
 1. Sutures secured anteriorly will avoid corneal abrasions

2. Absorbable sutures can be considered in infants and children or when avoidance of suture removal is preferred

- H. Absorbable sutures to secure skin graft to underlying tarsal conjunctival flap**
- I. Tarsal plate suture closure with interrupted partial thickness absorbing sutures, not extending through the conjunctival surface**
- J. Orbicularis muscle closure with absorbing sutures**
- K. Skin closure with interrupted, absorbable or non-absorbable sutures**
- L. Lateral canthal attachment to the periorbita of the inner aspect of the lateral orbital rim**

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

A. Intraoperative

1. Hemorrhage
2. Inadequate tissue to close the defect, will need to perform lateral canthotomy and superior cantholysis

B. Postoperative

1. Infection
2. Poor tissue approximation with eyelid margin irregularity
3. Corneal surface problems related to irregular eyelid margin and loss of tear film integrity
4. Ptosis, usually secondary to postoperative edema and horizontal tightness and will often resolve
5. Corneal abrasion from lid margin sutures
6. Thickened, immobile upper eyelid
7. Canthal distortion due to inadequate fixation or wound dehiscence
8. Younger patients (< 7 years) may require alternate flap design to prevent occlusion amblyopia; free tarsal conjunctival graft with orbicularis muscle skin flap
9. Eyelid retraction with lagophthalmos and exposure keratitis

C. Prevention of complications

1. Intraoperative control of hemostasis
2. Diligent surgical technique with regard to wound tension and closure
3. Confirm that marginal sutures are rotated away from corneal surface

D. Management of complications

VI. Describe the follow-up care

- A. Ocular evaluation with corneal and tear film inspection**
- B. Suture removal**

- C. Topical antibiotics
- D. Monitor patient for amblyopia, if coloboma repair in a young patient

VII. Describe appropriate patient instructions (postoperative care)

- A. Avoid eyelid trauma to the reconstructed area (protective eye shields helpful)
- B. Observe for tumor recurrence if indicated

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.

Concepts of lower eyelid reconstruction

I. List the indications/contraindications

A. Indications

1. Full thickness eyelid defects following tumor resection or trauma
2. Congenital coloboma

B. Contraindications

1. Active soft tissue infection

II. Describe the pre-procedure evaluation

A. Patient history

1. Etiology of the defect
2. Medical conditions
 - a. Diabetes mellitus
 - b. Post radiation therapy
 - c. History of self-trauma
3. Medications

B. Clinical examination

1. Ocular evaluation
2. Facial nerve function (orbicularis muscle function)
3. Size of defect
4. Laxity of adjacent tissue to determine ability to advance remaining eyelid tissues to close the defect
5. Tarsus and conjunctival surface of the upper eyelid to determine suitable donor site for tarsal conjunctival transposition flap to the lower eyelid
6. Corneal and tear film integrity
7. Verify integrity of lacrimal outflow system

C. Goals of surgery

1. Re-establish functional integrity of the eyelid
 - a. Good aesthetic result is desired, but secondary to eyelid function
 - b. Provide adequate eyelid closure
2. Avoid distortion of major facial anatomical features, if possible; eyelid margin, medial canthus

3. Provide adequate vertical eyelid tissue to protect cornea
4. Provide adequate horizontal eyelid tissue by release of the lower limb of the lateral canthal tendon to minimize defect size before planning upper eyelid tarsal conjunctival flap size
5. Provide a smooth, epithelialized, non-keratinized internal eyelid surface
6. Minimize vertical tension to avoid lower eyelid retraction or ectropion with ocular exposure
7. Maintain sufficient and anatomic medial canthal fixation
8. Choose the simplest technique
 - a. Primary closure
 - b. Primary lid margin closure with canthotomy and cantholysis
 - c. Primary lid margin closure with canthotomy, cantholysis and Tenzel semicircular rotational flap to advance lateral portion of the eyelid and repair margin primarily
 - d. Free tarsal graft and skin muscle flap
 - e. Hughes tarsoconjunctival flap for posterior lamella, skin graft or skin muscle flap for anterior lamella
9. Reconstruction of lacrimal system as necessary (See Lacrimal outflow system trauma)

III. List the alternatives to this procedure

- A. Temporary patching to protect the cornea

IV. Describe the instrumentation, anesthesia and techniques

- A. Standard surgical instruments for eyelid and facial surgery
- B. Local IV sedation, or general anesthesia
- C. Determination of sufficient horizontal laxity for direct closure
- D. If direct closure not possible flap or graft should be positioned with either permanent or absorbable sutures
- E. Considerations
 1. If standard Hughes flap will not make a wide enough flap the lateral canthal tendon may be released
 2. If there is adequate anterior lamella, a free tarsal graft may be used to provide posterior lamella
 3. Free tarsal conjunctival composite graft from the contralateral upper eyelid can be considered. An overlying skin orbicularis advancement flap may be needed
 4. Divide conjunctival Hughes flap 2-4 weeks after operation
- F. Re-approximation of the eyelid margin anatomical landmarks is essential
- G. Eyelid margin closure with 2 or 3 sutures through eyelash line and gray line (optional). Sutures secured anteriorly will avoid corneal abrasions. Absorbable sutures can be considered in infants

and children or when avoidance of suture removal is preferred

- H. Skin graft sutures to secure skin graft to underlying tarsal conjunctival flap
- I. Tarsal plate suture closure with interrupted *partial* thickness absorbing sutures, not extending through the conjunctival surface
- J. Orbicularis muscle closure with absorbing sutures
- K. Skin closure with interrupted absorbing or permanent sutures
- L. Lateral canthal attachment to the lateral orbital rim periosteum

V. List the complications of the procedure/therapy, their prevention and management

A. Intraoperative

1. Hemorrhage
2. Inadequate tissue to close the defect, will need to perform a lateral canthotomy and inferior cantholysis or upper eyelid tarsal-conjunctival transposition flap or full thickness flap to the lower eyelid defect
3. Larger defects still may require a mid forehead or glabellar flap possibly combined with temporal transposition flap or cheek rotational flap

B. Postoperative

1. Infection
2. Poor tissue approximation with eyelid margin irregularity
3. Corneal surface problems related to irregular eyelid margin, lower eyelid retraction with loss of tear film integrity, or suture corneal abrasion
4. Thickened immobile lower eyelid
5. Upper eyelid retraction
6. Lateral canthal angle distortion (rounding) secondary to inadequate periosteal fixation at lateral canthal angle
7. Young patient (< 7 years) may require alternate flap design to avoid occlusion amblyopia; free tarsal conjunctival graft with skin-muscle advancement flap

VI. Describe the follow-up care

- A. Ocular evaluation with corneal and tear film inspection
- B. Suture removal
- C. Topical antibiotics
- D. Monitor patient for amblyopia, if coloboma repair in a young patient

VII. Describe appropriate patient instructions

A. Avoid eyelid trauma to the reconstructed area (protective eye shield may be helpful)

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Management of Malignant Eye Tumors, Module #6, 1989, p.6.

Lateral canthal defects

I. List the indications/contraindications

A. Indications

1. Skin, orbicularis muscle, and/or lateral canthal soft tissue defects post tumor resection, surgery, infection, or trauma

B. Contraindications

1. Active soft tissue infection

II. Describe the pre-procedure evaluation

A. Patient history

1. Etiology of the defect
2. Other medical conditions
 - a. Diabetes mellitus
 - b. Post radiation therapy
 - c. History of self trauma
3. Medications

B. Clinical examination

1. Ocular evaluation
2. Facial nerve function (orbicularis and frontalis muscle function)
3. Size of defect
4. Laxity of adjacent tissue to determine ability to advance remaining eyelid tissues to close the defect
5. Corneal and tear film integrity
6. Assess infection vs. wound dehiscence in post surgical/traumatic lateral canthal defect

C. Goals of surgery

1. Avoid distortion of major facial anatomical features; eyelid margin, eyebrow, palpebral fissure
2. Plan to provide adequate horizontal anterior lamellar eyelid tissue by skin/orbicularis muscle advancement flaps
3. Plan periosteal rotational flap from the lateral orbital rim based at the inner aspect of the lateral orbital rim to provide posterior lamella support to the remaining tarsus
4. Provide for adequate eyelid closure and opening
5. Provide a smooth, epithelialized non-keratinized conjunctival eyelid surface

6. Minimize vertical tension to avoid eyelid retraction or ectropion with lagophthalmos
7. Maintain adequate attachment of the reconstructed lateral canthal soft tissues to the lateral orbital rim to avoid lateral canthal displacement postoperatively
8. Choose the simplest technique
 - a. Lateral tarsal strip type of procedure
 - b. Posterior lamellar free tarsal graft with skin muscle flap
 - c. Periosteal strip with skin muscle flap
 - d. Posterior lamellar reconstruction using a laterally based Hughes tarsoconjunctival flap with a skin graft or skin muscle flap

III. List the alternatives to this procedure

- A. Temporary patching
- B. If small defect, consider wound healing by second intention without primary surgical intervention

IV. Describe the instrumentation, anesthesia and technique

- A. Standard surgical instruments for facial surgery
- B. Local IV sedation, or general anesthesia
- C. Both posterior lamellar support with lateral canthal reconstruction and anterior lamellar (skin and orbicularis muscle) reconstruction needed
 1. Posterior support
 - a. Lateral orbital rim periosteal flap combined with temporalis fascial rotation with the flap based at the internal aspect of the rim
 - b. Lateral based tarsal/conjunctival flap (Hughes') from the upper eyelid
 - c. Free tarsal graft
 2. Anterior soft tissue reconstruction from skin/muscle advancement flap from adjacent tissues or a free skin graft
 - a. Cannot use both a posterior and anterior lamellar free grafts
 - b. Tarsal plate suture closure with interrupted partial thickness absorbing sutures, not extending through the conjunctival surface
 - c. Orbicularis muscle closure with absorbing sutures
 - d. Skin closure with interrupted absorbable or non-absorbable sutures

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

- A. Intraoperative

1. Hemorrhage
2. Inadequate local soft tissues requires transposition flap from forehead or upper eyelid area

B. Postoperative

1. Infection
2. Poor tissue approximation with eyelid margin irregularity
3. Corneal surface problems related to irregular eyelid margin and lateral canthal soft tissues with loss of tear film integrity
4. Ptosis, probably secondary to postoperative edema and horizontal tightness and will typically resolve
5. Reestablishment of the mucocutaneous junction may be too anterior or posterior causing either an erythematous lid margin or cornea irritation with eyelid skin and eyelashes against the cornea

VI. Describe the follow-up care

- A. Ocular evaluation with corneal and tear film inspection
- B. Suture removal
- C. Topical antibiotics

VII. Describe appropriate patient instructions

- A. Avoid eyelid trauma to the reconstructed area (protective eye shield may be helpful)
- B. Watch for tumor recurrence

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.
3. AAO, Focal Points: Management of Eyelid Trauma, Module #10, 1996.

Medial canthal defects

I. List the indications/contraindications

A. Indications

1. Skin, orbicularis muscle, and/or medial canthal soft tissue defects post tumor resection or trauma

B. Contraindications

1. Active soft tissue infection

II. Describe the pre-procedure evaluation

A. Patient history

1. Etiology of the defect
2. Other medical conditions - diabetes, post radiation, history of self trauma

B. Medications

C. Clinical examination

1. Ocular evaluation
2. Lacrimal drainage function
3. Canalicular, lacrimal sac and lacrimal duct anatomy
4. Facial nerve function (orbicularis muscle function)
5. Size of defect
6. Laxity of adjacent tissue to determine ability to advance remaining eyelid tissues to close the defect
7. Status of the posterior and anterior limbs of the medial canthal tendon
8. Corneal tear film integrity

III. List the alternatives to this procedure

A. Temporary patching

- B. If small defect, consider secondary wound healing by second intention without primary surgical intervention. Defects of the lid margin usually do not heal well with granulation alone; however small defects of the medial canthal concavity may do well without repair.**

IV. Describe the instrumentation, anesthesia and technique

A. Standard surgical instruments for eyelid and facial surgery

B. Local IV sedation, or general anesthesia

C. Goals of surgery

1. Avoid distortion of major facial anatomical features: eyelid margin, eyebrow, palpebral fissure
2. Avoid distortion of the puncta, if the remaining lacrimal drainage system is intact
3. Plan skin grafting or local skin-muscle flaps for larger defects
4. Local flaps can be planned from the glabellar or mid-forehead area
5. Adequate eyelid closure (blinking dynamics)
6. Plan reconstruction of the lacrimal drainage system if canalicular or lacrimal sac defect can be primarily repaired with silicone intubation.
7. Defer dacryocystorhinostomy (DCR) with Jones tube if tumor recurrence is possible because of tumor recurrence with access into the nose following Jones tube placement
8. Minimize vertical tension to avoid eyelid retraction or ectropion with lagophthalmos
9. Avoid soft tissue flaps from the glabellar or forehead area if deep tumor recurrence is possible; thick tissue flaps may delay detection of deep tumor recurrence
10. Reattach the reconstructed medial canthal tendon (soft tissues) to the posterior crest of the lacrimal sac fossa to avoid anterior displacement from the ocular surface

D. Choose the simplest technique

1. If only a small portion of the canaliculus has been removed, primary closure with canalicular repair may be possible
2. For larger medial defects, consider primary closure of the defect reattaching the medial end of the tarsus to the posterior lacrimal crest
3. Canthotomy and cantholysis may be required, if lower eyelid is too tight
4. Consider free posterior lamellar (tarsal) graft covered with a skin muscle flap, although sufficient skin and muscle are usually not present with this type of defect
5. Medially based Hughes tarsoconjunctival graft with an overlying free skin graft
6. Canalicular repair can be accomplished with silicone stents using bicanalicular intubation into nose, monocanalicular intubation or blunt-tipped pigtail probe technique

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

A. Intraoperative

1. Hemorrhage
2. Inadequate local soft tissues requires transposition flaps from the forehead area
3. Inadequate attachment to posterior lacrimal crest causing anterior displacement of the eyelid from globe
4. Injury to the lacrimal collecting system
 - a. Reconstructed medial eyelid may not allow normal pump function

B. Postoperative

1. Infection
2. Epiphora
3. Poor tissue approximation with eyelid margin irregularity
4. Corneal surface problems related to irregular eyelid margin and medial canthal soft tissues with loss of tear film integrity
5. Ptosis of upper eyelid repair, probably secondary to postoperative edema and horizontal tightness and will typically resolve

VI. Describe the follow-up care

- A. Topical antibiotics**
- B. Ocular evaluation with corneal and tear film inspection**
- C. Suture removal**
- D. Regular dermatologic follow up**

VII. Describe appropriate patient instructions

- A. Avoid eyelid trauma to the reconstructed area (protective eye shield may be helpful)**
- B. Watch for tumor recurrence**

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. 1, 1993, p.182-185.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.236-240,274-276.
4. AAO, Focal Points: Management of Eyelid Trauma, Module #10, 1996.

Involucional ectropion

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Lower eyelid horizontal laxity, usually involving either the lateral or medial canthal tendons or both
2. Gravitational effects on the eyelid and midface lead to eversion of the eyelid margin and loss of eyelid apposition to the globe
3. Eyelid retractor dehiscence may have a role in the etiology of tarsal ectropion
4. Senescent reduced tone in the orbicularis oculi muscle (see E3 below)

B. Define the relevant aspects of epidemiology of the disease

1. Advanced age with involutional horizontal tendon laxity

C. List the pertinent elements of the history

1. Chronic ocular exposure symptoms; mucoid discharge, foreign body sensation
2. Epiphora, secondary to involvement of the punctum with poor lacrimal drainage, and lacrimal pump insufficiency

D. Describe pertinent clinical features

1. Lateral canthal tendon laxity
2. Medial canthal tendon laxity
3. Epiphora, secondary to reflex lacrimal secretion and/or punctal ectropion and secondary stenosis due to exposure
 - a. Other causes of epiphora must be ruled out or treated
4. Palpebral conjunctival hypertrophy and keratinization
5. Ocular exposure, secondary to lower eyelid malposition
6. Inferior corneal epithelial changes secondary to lower eyelid malposition
7. Chronic blepharitis/meibomian gland inflammation
8. Reduced orbicularis tone but otherwise normal facial nerve function
9. No anterior lamellar insufficiency

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Biopsy, if eyelid margin anatomy altered or loss of lashes
2. Distraction test-test for medial or lateral canthal tendon laxity
 - a. Eyelid laxity if >7-8 mm from globe
3. Snap back test
 - a. After distraction from the globe, the eyelid does not return to normal position without one or

more blinks

- b. Qualitatively measures orbicularis tone

II. Define the risk factors

- A. Advanced age
- B. Chronic eyelid rubbing
- C. Floppy eyelid syndrome

III. List the differential diagnosis

- A. Paralytic ectropion secondary to facial nerve dysfunction
- B. Cicatricial ectropion, secondary to inadequate vertical anterior lamella (skin or orbicularis muscle)
- C. Eyelid retraction, secondary to thyroid eye disease (Graves ophthalmopathy)
- D. Eyelid retraction, secondary to cicatricial inferior tethering following trauma or lower blepharoplasty
- E. Eyelid cancer (mechanical ectropion)

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

A. Describe medical therapy options

- 1. Topical lubrication
- 2. Eyelid hygiene, topical antibiotics, and occasional corticosteroids for blepharitis
- 3. Horizontal eyelid taping (temporary only)

B. Describe surgical therapy options

- 1. Medial ectropion only
 - a. Mild
 - i. Thermal cautery of posterior (conjunctival) surface inferior to the punctum
 - b. Mild to moderate
 - i. Posterior conjunctival resection with vertical closure rotating the punctum internally (medial spindle procedure)
 - ii. Medial canthal tendon plication/shortening
 - iii. Lateral canthal tightening
- 2. Ectropion of lower eyelid
 - a. Lateral canthal tightening procedure e.g. tarsal strip
 - i. May include horizontal tightening of the lateral canthal tendon

- ii. Re-approximation of the tendon to the inner aspect of the lateral orbital rim
- b. Total tarsus ectropion may require reinsertion of dehiscenced lower eyelid retractors to the inferior tarsal plate via a conjunctival approach combined with a lateral canthal tightening procedure
- c. In the presence of lateral canthal tendon laxity, avoid horizontal resection of the tarsal plate or full thickness eyelid resection
 - i. May horizontally shorten the palpebral fissure
 - ii. May contribute to blunting of the lateral canthal angle
 - iii. Does not address one aspect of the pathology, lateral canthal tendon laxity
- d. When severe
 - i. Often caused by a "combined mechanism" ectropion with vertical insufficiency of the anterior lamella
 - i) May require concomitant skin graft or other recruitment of anterior lamella (e.g. midface elevation) and horizontal tightening
 - (i) Medial canthal tendon plication/tightening
 - (ii) Lateral canthal tendon tightening/tarsal strip procedure
 - ii. May instead be caused by a "combined mechanism" of involutional ectropion and involutional eyelid retraction
 - i) Treatment may require medial and/or lateral canthal tendon tightening, release of lower eyelid retractors, and/or midface elevation or other augmentation of the anterior lamella

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

- A. Aesthetic compromise of the palpebral fissure by horizontally shortening the tarsal plate or eyelid within the palpebral fissure**
 - 1. Prevented by careful choice of procedure
- B. Recurrence of the ectropion**
- C. Trichiasis, secondary to palpebral conjunctival cicatricial changes**
 - 1. Managed by surgical relaxation of conjunctival cicatrix or by permanent epilation of offending lashes (q.v.)
- D. Mal-apposition or imbrication of the lateral upper and lower eyelids at the commissure**
 - 1. Prevented by reapproximation of the gray lines with buried absorbable suture
- E. Vertical dystopia of the lateral canthi relative to one another**
 - 1. Prevented by intraoperative assessment of symmetry
- F. Infection**
- G. Hematoma or hemorrhage**

VI. Describe disease-related complications

A. Chronic ocular exposure with

1. cicatricial palpebral conjunctival changes resulting in trichiasis of the eyelid margin after correction of the ectropion
2. superficial keratitis with or without symptoms of foreign body sensation

B. Ocular surface inflammation with pterygium formation

C. Conjunctival keratinization and lid imbrication

VII. Describe appropriate patient instructions

A. Avoid constant eyelid rubbing

B. Recommend blotting, rather than wiping, tears from medial canthal recess

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.

Paralytic ectropion

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. This form of ectropion occurs as a result of facial nerve (cranial nerve (CN) VII- zygomatic branch) paralysis that affects the inferior orbicularis oculi
 - a. Non-paralytic, age-related reduction in orbicularis tone is associated with involutional ectropion (q.v.)
2. Gravity, acting on a lax lower eyelid, without an active orbicularis causes the eyelid to lose its position against the globe and turns outward

B. Define the relevant aspects of epidemiology of the disease

1. Occurs in the presence of facial palsy
 - a. Etiologies include viral, post-surgical, traumatic, idiopathic, etc (see Facial nerve palsy)
2. Severity of clinical appearance correlates with aging changes of the eyelids and face (especially canthal tendon laxity, lower and midface descent)

C. List the pertinent elements of the history

1. Patients typically have a history of facial nerve paralysis and incomplete recovery
2. Epiphora variably related to tear pump insufficiency, punctal eversion outside the tear lake, punctal stenosis, and reflex lacrimation in response to corneal exposure
3. Complaints regarding ocular burning, foreign body sensation, tearing, or dryness (Corneal foreign body sensation)
4. Concerns regarding aesthetics of out-turned eyelid

D. Describe pertinent clinical features

1. An outward turning of the lower eyelid with evidence of an active or remote facial nerve palsy
2. Degree of ectropion dependant on amount of facial nerve function
3. Punctal stenosis
4. Elevated tear meniscus
5. Keratinization, erythema, and edema of the eyelid margin and palpebral conjunctiva
6. Exposure keratopathy
7. Often, but not always, seen with paralytic lagophthalmos
 - a. Evaluate quality of Bell's phenomenon
8. Evaluate for other cranial neuropathies
 - a. Traumatic, concomitant loss of corneal sensation and/or extraocular motility may result in neurotrophic keratitis

II. Define the risk factors

- A. Facial paralysis**

III. List the differential diagnosis

- A. Paralytic lower lid retraction, where the lid is inferiorly displaced without outward rotation**
 - 1. Paralytic ectropion and retraction may occur together
- B. Involutional ectropion**
- C. Cicatricial ectropion**
- D. Mechanical ectropion**

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

A. Describe medical therapy options

- 1. Medical therapy is directed at treating the ocular exposure caused by the ectropion using lubricants (artificial tears and ointments)
- 2. Mechanical therapy may include taping the lower eyelid upward

B. Describe surgical therapy options

- 1. Basic therapy includes horizontal tightening and elevation of the lid using lateral canthoplasty
- 2. More advanced treatments would include
 - a. Facial slings, Arion slings
 - b. Sub orbicularis oculi fat (SOOF) lift
 - c. Medial canthoplasty or canthopexy
 - d. Correction of lower eyelid retraction with or without posterior lamella spacer grafts (stiffer grafts including cartilage, hard palate grafts)
- 3. Associated treatments to increase facial tone include facial nerve and cross face nerve grafts
- 4. Temporary or permanent intermarginal tarsorrhaphy may be useful in cases where options above are not possible or appropriate

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

- A. Recurrence or persistence of the ectropion**
- B. Infection**
- C. Hematoma or hemorrhage**
- D. Aesthetic compromise of the palpebral fissure by horizontally shortening the tarsal plate or eyelid within the palpebral fissure**

1. Prevented by careful choice of procedure

E. Recurrence of the ectropion

F. Trichiasis, secondary to palpebral conjunctival cicatricial changes

1. Managed by surgical relaxation of conjunctival cicatrix or by permanent epilation of offending lashes (q.v.)

G. Mal-apposition of the lateral upper and lower eyelids at the commissure

1. Prevented by reapproximation of the gray lines with buried absorbable suture

H. Vertical dystopia of the lateral canthi relative to one another

1. Prevented by intraoperative assessment of symmetry

VI. Describe disease-related complications

A. Ocular exposure with corneal decompensation and foreign body sensation

B. Epiphora due to loss of lacrimal pump mechanism and/or reflex tearing from ocular exposure

VII. Describe appropriate patient instructions

A. Maintenance of ocular lubrication

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.70-74.
3. AAO, Focal Points: Ophthalmic Management of Facial Nerve Paralysis, Module #4, 2000, p.4-5.
4. AAO, Focal Points: Ectropion and Entropion, Module #10, 1994, p.3, 8.

Cicatricial ectropion

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Caused by vertical shortening of the anterior lamella of the lid
2. Much more common in the lower lid than upper lid
3. Horizontal laxity of the lid may contribute to severity

B. Define the relevant aspects of epidemiology of the disease

1. Thermal, radiation, or chemical burns
2. Mechanical trauma
3. Surgical trauma - tumor removal, blepharoplasty
4. Chronic actinic skin damage
5. Chronic inflammation of the eyelid
 - a. Rosacea
 - b. Atopic dermatitis
 - c. Eczematous dermatitis
 - d. Herpes zoster
 - e. Ichthyosis
 - f. Infection

C. List the pertinent elements of the history

1. Complaints regarding ocular burning, foreign body sensation, tearing, or dryness
2. Concerns regarding aesthetics of out-turned eyelid
3. Patient often working in a setting of chronic sun exposure
4. History of diseases and injuries as noted above

D. Describe pertinent clinical features

1. Outward rotation of eyelid margin and/or punctum, often with punctal stenosis
2. Vertical shortening of the anterior lamella of the eyelid such that digital elevation of the lower lid above the superior limbus (with the eye in primary position) is not possible (for upper lid, depression below inferior limbus)
3. Cutaneous scar or other manifestation of diseases noted above
4. Horizontal laxity of the eyelid (especially lower)
5. Keratinization of eyelid margin and palpebral conjunctiva

6. Signs of ocular exposure
7. When patient opens mouth, the lower lid retracts

II. List the differential diagnosis

- A. Involutional ectropion (lower lid)**
- B. Paralytic ectropion (lower lid)**
- C. Eyelid retraction**
- D. Mechanical ectropion (lower lid, caused by mass effect)**

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

A. Describe medical therapy options

1. Treat underlying conditions as outlined in I.B.
 - a. Corticosteroid ointment massage into affected area may release some contraction due to inflammatory conditions
2. Treat ocular exposure with artificial tears, lubricants, etc.

B. Describe surgical therapy options

1. Occasionally, horizontal tightening alone is adequate for lower lid correction if cicatricial changes are not severe, although often this is only a temporizing procedure because it does not address the shortening of the anterior lamella
2. Standard approach
 - a. Vertical cicatricial traction is surgically incised and relaxed
 - b. Eyelid is horizontally tightened as needed
 - c. Anterior lamella of eyelid is vertically lengthened, utilizing
 - i. full thickness skin graft
 - i) Upper eyelid skin
 - ii) Posterior auricular skin graft
 - iii) Preauricular skin graft
 - iv) Supraclavicular graft
 - ii. Skin-muscle flap
 - i) Advancement flap
 - ii) Rotational flap
 - iii) Transpositional flap
3. Some cases may be improved with suborbicularis oculi fat (SOOF) or other midface lift which may eliminate need for skin graft

4. Postoperative traction suture often critical to reduce risk of skin graft contracture and recurrent ectropion

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

A. Corticosteroid massage

1. May cause skin thinning and high recurrence rate

B. Surgical therapy

1. Standard surgical complications (e.g. infection, bleeding, etc)
2. Graft failure or flap ischemia/necrosis
 - a. Compression bolsters may reduce incidence
 - b. Cessation of smoking and other tobacco use may reduce incidence
3. Graft prominence
 - a. Incidence reduced with careful suturing technique and graft selection
 - b. Can reduce graft prominence with careful use of corticosteroids
4. Recurrence of cicatricial ectropion
 - a. Postoperative traction suture(s) may reduce risk of recurrent ectropion.

V. Describe disease-related complications

- A. Persistent ocular exposure, possible corneal complications**
- B. Punctal ectropion**
- C. Thickened, hypertrophic tarsal conjunctiva**
- D. Tearing**
- E. Aesthetic concerns**

VI. Describe appropriate patient instructions

- A. Avoid sun exposure when indicated**
- B. Protective eye shield at bedtime while skin graft bolster in place, postoperatively**
- C. Topical and/or oral antibiotics**
- D. Smoking cessation**

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. 1, 1993, p.142-146.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.74-82.
4. AAO, Focal Points: Ectropion and Entropion, Module #10, 1994, p.2.

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
 - a. Diagnosis of exclusion (see below)
 - b. May be infectious etiology (Herpes viruses)
2. Congenital
3. Accidental trauma
4. Inflammatory
5. Tumors
 - a. Brainstem or cerebellopontine angle (CPA) tumors may cause cranial nerve V, VII, and/or VIII involvement
 - b. Parotid tumors
 - c. Squamous cell carcinoma
6. Compressive
7. Vascular (stroke)
8. Infectious (e.g. Ramsey Hunt syndrome)
9. Iatrogenic
 - a. Facial surgery
 - b. Intracranial surgery
 - c. Forceps delivery
10. Lyme disease

B. Define the relevant aspects of epidemiology of the disease

1. Isolated facial nerve palsy with no clear etiology is termed "Bell palsy"
 - a. Occurs in both sexes with equal frequency
 - b. Less common in children
 - c. Usually unilateral
 - d. Thought to be inflammatory or a sequel to herpes zoster infection
2. May occur following surgical excision of brainstem or parotid tumors
3. Hemifacial spasm due to aberrant facial nerve regeneration may be among the presenting complaints

C. List the pertinent elements of the history

1. Patients with stroke or intracranial surgery may have concurrent anesthetic corneas, making them more likely to develop corneal exposure (neurotrophic keratitis)
2. Ipsilateral dry eye, hearing loss, and vestibular dysfunction are associated with cerebellopontine angle (CPA) masses
3. Loss of taste and salivary function with lesions between the geniculate ganglion and the salivary nerve
4. Removal of acoustic neuroma, cerebellar tumor, parotid tumor
5. Skin eruption on pinna of ear, dizziness, hearing disturbance, and moderate to severe facial pain occur in Ramsey Hunt syndrome

D. Describe the pertinent clinical features

1. Reduced or lack of movement of the involved side of the face with eyebrow ptosis, lagophthalmos, mild retraction of the upper eyelid, ectropion and/or retraction of the lower eyelid, poor blink, or mouth droop
2. Loss of vision due to superficial keratitis, and/or distortion in downgaze from "pooled" tears
3. Epiphora due to reflex tearing and/or lacrimal pump insufficiency
4. Chronic facial nerve palsy may include loss of nasolabial fold on affected side, deviation of nasal tip to contralateral side
5. Aberrant facial nerve regeneration may cause variants of abnormal movement including spasm

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Measurement of facial nerve function and tone on each side of the face for the four primary motor branches of VII: frontal, zygomatic, buccal, and marginal mandibular
2. Neuro imaging is usually not recommended for isolated first episode of idiopathic Bell Palsy
 - a. MRI with gadolinium covering course of facial nerve and posterior fossa is indicated if
 - i. The palsy is progressive or recurrent
 - ii. Associated with other cranial neuropathies or neurologic findings
3. Lyme disease
 - a. Positive anti-spirochetal antibody titer by enzyme-linked immunoabsorbent assay (ELISA) method is diagnostic
 - b. Sedimentation rate and aspartate transaminase levels may be elevated

II. Define the risk factors

A. Viral syndrome

B. Forceps delivery

C. CPA tumor

D. Parotid tumors

E. Head trauma

III. List the differential diagnosis

A. Other causes of ectropion and lagophthalmos

B. Ichthyosis, scleroderma

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

A. For new onset of facial palsy, consider ear, nose and throat (ENT) evaluation for possible medical (antiviral and or corticosteroid) or surgical treatment (decompression)

B. Medical prevention and management of corneal exposure

1. Topical lubrication
2. Supportive taping of eyelids
3. Moisture chambers

C. Surgical management of eyelid malpositions and loss of function

1. Lateral (and rarely medial) tarsorrhaphy
 - a. Temporary suture technique
 - b. Eyelid marginal adhesion technique
 - c. Tarsal transposition technique
2. Lower lid retraction and paralytic ectropion repair
 - a. Horizontal tightening
 - b. Silicone (Arion) or fascia lata sling
 - c. Lower eyelid mid/posterior lamella support using ear cartilage, hard palate graft, mucous membrane graft or other spacer materials for eyelid elevation
3. Treatment of lagophthalmos
 - a. Upper eyelid load weights
 - i. Typically gold or platinum
 - ii. Provide passive eyelid closure
 - b. More dynamic lid closure can be provided by palpebral spring to upper eyelid or Arion sling for upper and lower eyelid
 - c. Effect may deteriorate over time

D. Correction of eyebrow ptosis and/or upper eyelid dermatochalasis

1. Eyebrow or forehead lift with possible upper lid skin excision
2. May be primarily considered for patients in whom there is no corneal decompensation, since the sagging brow and redundant upper lid tissue may help the paretic lids to close

E. Improvement in epiphora and/or "pooling of tears"

1. Treatment of superficial keratitis
2. Horizontal lower eyelid tightening
3. Silicone lacrimal intubation

F. General facial reanimation in patients with long standing palsies (often in collaboration with ENT, neurosurgery, plastic surgery)

1. Dynamic reanimation procedures
 - a. Cross face 7th nerve transfers
 - b. XII-VII anastomosis
 - c. Gracilis muscle grafting
 - d. Temporalis muscle transfer
2. Static facial support
 - a. Facelifting with facial or other slings
 - b. Midface lifting techniques

G. Improvement in symptoms related to aberrant regeneration

1. Botulinum toxin
 - a. To the periocular orbicularis for spastic winking, upper eyelid ptosis, and reverse ptosis of the lower eyelid
 - b. For lower facial spasm, including peri-oral and platysma (lower facial and perioral injection carry significant risk of lower facial and mouth droop resulting in difficulties with pursing of lips (ex/drinking, speech, kissing)
 - c. To the lacrimal gland for gustatory tearing ("crocodile tears")

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

A. All tightening procedures eventually relax over time

B. Palpebral load weight and spring/sling procedures

1. Early or late migration, extrusion
2. Prevention- meticulous surgical technique including layered closure and adequate tissue (orbicularis muscle) coverage of eyelid weight
3. Management: remove and replace in separate operative procedures

C. Failure of tarsorrhaphy

1. Prevention
 - a. The adhesion must be made securely, preferably with tarsal transfer

D. Infection

- E. Hematoma, hemorrhage

VI. Describe disease-related complications

- A. Corneal exposure, ulceration, scarring
- B. Decreased visual field
- C. Drooling
- D. Biting cheek
- E. Deformity
- F. Aberrant regeneration

VII. 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 of facial nerve function
- C. Surgical corrections are indicated when minimal recovery of facial nerve function is expected, or if ocular surface compromise is severe

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.
6. Rahman I, Sadiq SA. Ophthalmic management of facial nerve palsy: a review. *Surv Ophthalmol* 2007;52:121-44.

Full-thickness block resection and repair

I. List the indications/contraindications

A. Indications

1. For excision of eyelid lesions or defects
2. For removal of localized regions of trichiasis
3. For horizontal tightening of the eyelid
 - a. Ectropion, entropion
 - b. Floppy eyelid syndrome

B. Contraindications

1. Defect or lesion that is too large for this technique

II. Describe the pre-procedure evaluation

A. Evaluate the potential size of the defect for surgical reconstruction

B. Assess the amount of eyelid laxity for horizontal tightening

C. Standard ocular and periocular exam

1. Skin laxity, eyelid malpositions, etc.
2. Tear lake and lacrimal drainage system, corneal integrity, adequacy of corneal protective mechanisms, etc.

III. List the alternatives to the procedure

- A. Other methods of tumor or defect removal (e.g., Mohs micrographic surgery for skin cancers, electroepilation or radiofrequency ablation for trichiasis)
- B. Other methods of horizontal tightening of the eyelid (e.g. lateral canthoplasty, tarsal strip procedure)
- C. Ocular surface disease management for floppy eyelid syndrome

IV. Describe the instrumentation, anesthesia and technique

- A. Local anesthetic typically used
- B. Make vertical (perpendicular to the eyelid margin) or slightly diverging incisions full thickness through the eyelid
- C. Complete the block with converging incisions (pentagonal)
- D. Repair the eyelid margin with soft (silk, polyglactin or equivalent) sutures with care to approximate the lash line and gray line. Vertical mattress sutures may be used at margin to evert

the wound edges and avoid margin depression/ notching

- E. Keep suture ends away from the cornea
- F. Repair of tarsus and other deep tissues with polyglactin 910 (Vicryl) or equivalent, partial thickness through tarsus to avoid ocular irritation
- G. Close skin defect without tension

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

- A. Premature suture removal may result in wound dehiscence
 - 1. Generally wait 6 to 10 days before removing lid-margin sutures
- B. Poor alignment of wound may result in notching of lid margin
 - 1. Prevented by meticulous surgical technique
 - 2. Treatment may include re-excision and realignment

VI. Describe follow-up care

- A. If excision for malignant tumor, the patient needs arrangements for regular follow-up for possible recurrence

VII. Describe appropriate patient instructions

- A. Don't pull on reconstructed eyelid; protective eye shield may be helpful
- B. Return if patient notes a new lesion, localized recurrence, or wound dehiscence

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Tarsorrhaphy

I. List the indications/contraindications

A. Indications

1. Severe ocular exposure with corneal decompensation
 - a. Paralytic lagophthalmos
 - b. Lid retraction e.g.
 - i. Thyroid eye disease (thyroid orbitopathy)
 - ii. Facial burns or other causes of cicatricial lid retraction
 - c. Neurotrophic ulcer, non-healing corneal defect
2. Moderate ocular exposure in the setting of corneal anesthesia
3. Post-surgical (temporary)

B. Contraindications

1. Active infected corneal ulcer is a relative contraindication for a complete tarsorrhaphy

II. Describe the pre-procedure evaluation

- A. Document persistent ocular/corneal exposure in spite of aggressive conservative measures, or inability to perform supportive measures
- B. Determine whether tarsorrhaphy should be permanent or temporary. Is the underlying condition expected to improve, if so, how soon?
- C. Document lid retraction with lateral scleral show
- D. Determine corneal sensation, active infection, degree of decompensation
- E. Several options are available
 1. Permanent vs. temporary
 2. Complete vs. partial
 3. Lateral vs. medial
- F. Medial or lateral tarsorrhaphy may be used to give coverage and not limit use of topical medications
- G. Consideration of newer procedures for facial reanimation following cranial nerve (CN) VII palsy may offer advantages over the traditional tarsorrhaphy technique

III. List the alternatives to the procedure

- A. Aggressive medical measures for ocular exposure
- B. Botulinum toxin to induce temporary ptosis

1. May be useful adjunct to suture tarsorrhaphy
- C. Lower eyelid tightening and elevation using medial or lateral canthoplasties**
- D. Recession procedures of upper and lower eyelid**
 1. Lower eyelid elevation with posterior lamella spacer grafting
 2. Standard anterior or posterior approach eyelid retractor (Mullers muscle or levator aponeurosis) recession for the upper eyelid
- E. Midface elevation to support lower eyelid in facial palsy or anterior lamellar insufficiency**
- F. Gold or platinum eyelid weight implantation in the upper eyelid**
- G. Implantation of eyelid spring or sling device**

IV. Describe the technique

- A. Temporary**
 1. Local anesthesia
 2. Place horizontal mattress sutures (one suture may be enough) through upper and lower lids and tie over bolsters on skin
- B. Permanent**
 1. Local anesthesia
 2. De-epithelialize portion of eyelid margin for adherence
 3. A variety of incisions can be used to isolate and/or rotate portions of the tarsal plates, which are then sutured together
 - a. Can split lid into anterior and posterior lamella and suture upper and lower lids together by lamella

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

- A. Temporary**
 1. Premature loosening of sutures
 2. Suture infection or inflammation
 3. Corneal abrasion
- B. Permanent**
 1. Tarsorrhaphy dehiscence
 2. Wound infection
 3. Decompensation of the exposed portion of cornea
 4. Madarosis
 5. Trichiasis

6. Temporal visual field impairment
7. Cosmesis

VI. Describe follow-up care

- A. Follow regularly for corneal improvement**
- B. Follow for dehiscence of tarsorrhaphy or relaxation of wound**

VII. Describe appropriate patient instructions

- A. Patient should have continued follow-up and continue use of lubricants and other eye medications unless instructed otherwise; protective eye shield may be helpful until tarsorrhaphy healed**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
2. Seiff, SR, Carter, SR. Reanimation of the paretic eyelid complex. *Facial Plastic Surgery Clinics of North America. Oculoplastic Surgery.* 1998;6:21-30.
3. Seiff SR, Chang J. Management of ophthalmic complications of facial nerve palsy. *Otolaryngol Clin North America.* 1992;25:669-690.
4. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.110,127.
5. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.185-187.
6. Seiff SR, Chang JS. The staged management of ophthalmic complications of facial nerve palsy. *Ophthal Plast Reconstr Sur.* 1993;9:241-9.

Involucional entropion

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Defined as inward rotation of the eyelid margin with direction of the lashes or keratinized lid margin against the ocular surface
2. Three components to etiology of involutional entropion
 - a. Lower lid horizontal laxity
 - b. Disinsertion of lower eyelid retractors
 - c. Preseptal orbicularis overriding pretarsal orbicularis muscle

B. Define the relevant aspects of epidemiology of the disease

1. Older population, with significant tissue laxity
2. Generally only affects the lower eyelid
 - a. Upper eyelid tends to become ptotic, not to rotate inward as a result of involutional changes

C. List the pertinent elements of the history

1. Foreign body sensation, pain, red eye
2. Epiphora

D. Describe pertinent clinical features

1. Lower eyelid margin rotated inward against cornea and conjunctiva
2. Lower eyelid horizontal laxity
3. Corneal and conjunctival irritation, superficial keratitis, occasionally bacterial keratitis

II. Define the risk factors

- A. Elderly
- B. Lower eyelid horizontal laxity

III. List the differential diagnosis

- A. Spastic entropion
- B. Cicatricial entropion
- C. Trichiasis without entropion
 1. Distichiasis
 2. Misdirected lashes

3. Aberrant lashes

D. Epiblepharon

1. The presence of a roll of lower eyelid skin and orbicularis muscle that pushes the lashes against the cornea
2. Most commonly occurs in children of Asian descent
3. May improve with age (and enlargement of the nasal bridge), without surgery

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

A. Describe medical therapy options- useful as temporizing measures for patients waiting for or unable to undergo surgical correction

1. Ocular lubrication to protect the cornea from lashes
2. Bandage contact lens
3. Eyelid taping

B. Describe surgical therapy options

1. Temporizing surgical procedures
 - a. Quickert-Rathbun rotational sutures
 - b. Eyelid skin cautery
2. Definitive surgical correction
 - a. Lower lid retractor advancement/ reattachment
 - b. Lateral lid tightening (e.g. lateral tarsal strip procedure), usually in conjunction with retractor reinsertion
 - c. Quickert modification of Wies (horizontal shortening with margin rotation) (4 snip procedure)
 - d. Weiss procedure (eyelid margin rotation)

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

A. Recurrence

1. Procedures that address more aspects of the etiology of involutional entropion have lower failure rates

B. Wound dehiscence

C. Infection

D. Hematoma or hemorrhage

VI. Describe disease-related complications

A. Constant irritation of lashes against ocular surface can lead to corneal scarring, corneal ulcer,

and vision loss

VII. Describe appropriate patient instructions

A. Follow for recurrence

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Ectropion and Entropion, Module #10, 1994, p.3.

Cicatricial upper and lower eyelid entropion

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Defined as inward rotation of the eyelid margin due to posterior lamellar abnormality with direction of the lashes or keratinized lid margin against the ocular surface
2. Congenital (not truly cicatricial)
 - a. Upper eyelid entropion (Tarsal Kink Syndrome)-relatively unusual-due to deformity of the tarsal plate
 - b. Also seen in incomplete development of the upper lid tarsal plate with an absent eyelid crease and overriding of the orbicularis muscle
3. Acquired - inflammatory or scarring processes cause shortening of the posterior lamella of the eyelid. The posterior shortening causes inward rotation of the eyelid margin with direction of the lashes or keratinized lid margin against the ocular surface
 - a. Conjunctival cicatrizing disorders (Stevens-Johnson Syndrome, ocular cicatricial pemphigoid (OCP), drug induced pseudo ocular cicatricial pemphigoid
 - b. Chemical burns
 - c. Post-traumatic injuries
 - d. Infectious processes (Trachoma)
4. Mechanical entropion due to mass lesions that secondarily invert the lid margin
 - a. May be neoplastic or inflammatory

B. Define the relevant aspects of epidemiology of the disease

1. Congenital - relatively uncommon
2. Acquired - also relatively uncommon in the USA
 - a. Trachoma- travel to endemic areas, exposure to infected individuals
 - b. Stevens Johnson Syndrome- exposure to inciting event or medications
 - c. Ocular cicatricial pemphigoid- bilateral mucosal cicatrizing disease of older patients (>55 yo), with recurrent and relapsing course.
 - d. Drug induced pseudo ocular cicatricial pemphigoid-discontinue inciting agent when possible
3. Postsurgical - lower eyelid transconjunctival orbitotomy or blepharoplasty (posterior and middle lamellar cicatrix formation); upper eyelid conjunctival-Mueller muscle resection ptosis repair
4. Mechanical entropion- dependent upon pathologic process affecting upper eyelid

C. List the pertinent elements of the history

1. Symptoms of ocular irritation, corneal damage, and visual loss are more likely and more severe with upper eyelid involvement than with lower eyelid involvement
2. History of an inflammatory or traumatic process such as
 - a. Autoimmune (ocular mucous membrane pemphigoid)
 - b. Inflammatory (Stevens-Johnson syndrome)
 - c. Infectious (trachoma, herpes zoster)
 - d. Surgical (enucleation, posterior ptosis repair, transconjunctival blepharoplasty, transconjunctival blowout fracture repair)
 - e. Traumatic (thermal or chemical burns, scarring)
 - f. Glaucoma drugs
 - g. History of eyelid neoplasm
3. May involve upper and/or lower lid, unlike involutional entropion which only involves lower lid

D. Describe pertinent clinical features

1. Upper eyelid involvement may have similar signs and symptoms as lower eyelid entropion
 - a. Inability to correct entropion by downward traction (lower lid) or upward traction (upper lid)
 - b. Scarring and shortening of the conjunctiva and/or deformation of the tarsus; symblepharon
 - c. Mechanical corneal abrasion, superficial keratitis, ulcer, corneal scar
 - d. Chronic exposure with lagophthalmos when scarring is significant
2. Additionally, dry eye may be more severe due to the loss of the accessory lacrimal glands

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Testing directed by history
2. Immune studies for mucous membrane pemphigoid
3. Cultures or PCR test for trachoma, herpes in cases that appear active
4. Conjunctival or eyelid biopsy or scraping

II. Define the risk factors

- A. Travel to endemic areas for trachoma**
- B. Chemical exposure**
- C. History of trauma**
- D. Use of topical drugs to treat glaucoma**

III. List the differential diagnosis

- A. Involutional entropion**

B. Spastic entropion

C. Trichiasis without entropion

1. Distichiasis
2. Misdirected lashes from chronic posterior blepharitis, eyelid margin scar, etc.
3. Aberrant lashes
4. Floppy eyelid syndrome - lash ptosis, loss of parallelism

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

A. Describe the natural history, outcome and prognosis-

1. Variable depending upon the etiology and activity of disease. Surgical treatment should only be accomplished when disease process has been controlled

B. Describe medical therapy options

1. Lubrication, bandage contact lens
2. Trachoma- treat infectious etiology
 - a. Azithromycin, doxycycline or tetracycline (avoid latter in children under 8 y/o, pregnant women and nursing mothers)
3. Ocular cicatricial pemphigoid
 - a. Treat blepharitis aggressively
 - b. Topical steroids (caution to follow cornea closely)
 - c. Systemic steroids
 - d. Dapsone (progressive increase to 100-150mg/d)
 - e. Immunosuppressives (e.g. cyclophosphamide, methotrexate, azathioprine, mycophenolate mofetil, rituximab)

C. Describe surgical therapy options

1. Mild cases with minimal to mild tarsal abnormalities
 - a. shortening of anterior lamella (skin muscle resection with epitarsal fixation) or anterior lamella repositioning via eyelid crease incision
 - b. Margin rotation/marginal tarsotomy- useful for marginal cicatricial entropion
 - c. Tarsal fracture- useful for tarsal kink and tarsal deformities
2. Moderate to severe cases of posterior lamella shortening
 - a. More severe cases may require posterior lamellar lengthening procedures with mucous membrane grafting, amniotic membrane grafting, and or tarsal grafting, hard palate or other graft, tarsal modification, or combination
 - b. Tarsal fracture and rotation procedure

- i. This procedure creates a horizontal incision(s) to evert the lid margin, held in place with everting sutures
 - ii. More aggressive eversion of 2 mm of eyelid margin held with everting sutures
 - iii. Posterior lamellar release with/without mucous/amniotic membrane grafting
- c. Marginal rotation procedures (depending upon the severity of marginal entropion) may include super advancement of the inferior tarsal edge

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

- A. Undercorrection
- B. Overcorrection
- C. Recurrence of entropion- make certain etiology of entropion has resolved
- D. Eyelid margin irregularities
- E. Retraction
- F. Potential lid margin necrosis, corneal dellen formation, eyelid contour abnormality, and pyogenic granuloma in tarsal rotation or tarsotomy procedures

VI. Describe disease-related complications

- A. Vision loss
- B. Chronic discharge
- C. Trichiasis
- D. Corneal abrasions
- E. Corneal ulceration
- F. Corneal scarring and decreased vision
- G. Lagophthalmos
- H. Severe dry eye
- I. Recurrence of entropion
- J. Upper lid retraction
- K. Lid margin necrosis due to vascular compromise

VII. Describe appropriate patient instructions

- A. Keep eye well lubricated
- B. Observe for appropriate healing and possible recurrence of entropion as tissues heal
- C. Topical corticosteroids may be useful to limit scar formation and recurrence

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. 1, 1993, p.142-146.
3. AAO, Focal Points: Ectropion and Entropion, Module #10, 1994, p.4.
4. AAO, Focal Points: Management of Cicatricial Entropion, Trichiasis and Distichiasis, Module #12, 1993.
5. Dortzbach RK, Callahan A. Repair of cicatricial entropion of upper eyelids. *Arch Ophthalmol* 85;82, 1971
6. Fox SA. Cicatricial entropion of the upper eyelid. *Am J Ophthalmol* 57;379, 1964.
7. Tenzel RR, Miller GR, Rubenzik R. Cicatricial upper lid entropion. *Arch Ophthalmol* 93; 999, 1975.
8. Beyer CK, Carroll JM. Modreately severe cicatricial entropion. *Arch Ophthalmol* 89; 33, 1973.
9. Wies FA. Surgical Treatment of entropion. *J INT Coll Surg* 21; 758, 1954.
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11. McCord CD, Chen WP. Tarsal polishing and mucous membrane grafting for cicatricial entropion, trichiasis and epidermalization. *Ophthal Surg* 14:1021, 1983.
12. AAAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.26.
13. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.84,101,108,174-176.
14. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.130-131,205,209-217,228-229,232.
15. AAO, Focal Points: Management of Eyelid Trauma, Module #10, 1996, p.3.
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17. Thorne JE, Anhalt GJ, Jabs DA: Mucous membrane pemphigoid and pseudopemphigoid. *Ophthalmology*. 2004; 111:45-52.

Trichiasis

I. Describe the approach to establishing the diagnosis

A. Define the disease condition

1. Trichiasis refers to an abnormal condition of eyelashes where the eyelashes are misdirected (often turning inward against the eye) in the absence of entropion, which is defined as inward rotation of the eyelid margin
2. Marginal entropion results from subtle posterior lamellar scarring which inverts the margin slightly, bringing the eyelashes against the eye
3. Entropion - various forms are discussed elsewhere
4. Aberrant lashes: normal position, abnormal direction
5. Distichiasis: second row of lashes from Meibomian gland orifices
 - a. Most commonly a congenital condition
 - b. Acquired forms are also recognized
6. Metaplastic lashes: lashes growing from abnormal position
 - a. Distichiasis can be considered a form of metaplastic lashes but is usually considered a special case
7. For eyelids that have previously undergone surgery or trauma that altered the eyelid margin (e.g. Hughes tarsoconjunctival flap reconstruction), trichiasis may primarily involve Lanugo hairs from the adjacent eyelid skin rather than eyelashes. The patient symptoms and corneal signs are similar to the other forms of trichiasis described above

B. Define the relevant aspects of epidemiology of the disease

1. Trauma or previous eyelid margin surgery can misdirect the eyelash roots, as can a variety of autoimmune (mucous membrane pemphigoid), inflammatory (posterior blepharitis, Stevens - Johnson syndrome), infection (trachoma), and drug induced conditions (glaucoma medications). Frequently the lid margin is also inverted in these conditions
2. Chronic inflammation or infection can result in metaplasia

C. Describe the pertinent clinical features

1. Inward directed lashes
2. Identify the cause of the misdirected eyelashes. Is the lid margin in a normal position? (Upright position)
3. Are there cicatricial posterior lamella changes?
4. Is there evidence of trauma or surgery?
5. Note location and extent of the problem
6. Is there an extra roll of skin pushing the eyelashes against the eye?

7. Corneal changes
8. Any eyelid lesions distorting lash orientation

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Cultures may be useful for infectious processes
2. Basement membrane studies for mucous membrane pemphigoid

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

A. Describe medical therapy options

1. Ocular lubrication
2. Bandage contact lens

B. Describe surgical therapy options

1. Appropriate entropion procedures if present
2. Manual epilation of affected lashes
3. Permanent epilation by electrolysis/ hyfrecation/radiofrequency ablation
4. Cryotherapy
5. Laser ablation
6. Surgical dissection and excision (horizontal lid splitting and lash bulb excision)
7. Block (full thickness block) resection and eyelid margin repair for recalcitrant cases

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

- A. For lubrication, inadequate management
- B. Recurrence is very common with all forms of surgical management
- C. Electrolysis, cryo, and laser may cause additional cicatricial changes including entropion and more rarely, irregularities of the eyelid margin
- D. Surgical management may cause scarring or lid margin irregularity

IV. Describe disease-related complications

- A. Chronic ocular irritation, ulceration, scarring, peripheral corneal neovascularization and blindness can occur

V. Describe appropriate patient instructions

- A. Return if recurrence, ocular irritation

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p.128, 139-140.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.50-52.
4. AAO, Focal Points: Management of Trichiasis, Module #4, 2001.
5. AAO, Focal Points: Management of Cicatricial Entropion, Trichiasis and Distichiasis Module #12, 1993.
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Congenital myogenic ptosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Congenital myogenic ptosis is of unknown cause
2. Characterized by maldevelopment of the levator muscle
 - a. The levator muscle is variably replaced with fibrous or fatty tissue

B. Define the relevant aspects of epidemiology of the disease

1. Usually occurs as an isolated finding
2. Affects males and females equally
3. Usually not inherited in the isolated form
4. Can be seen in various syndromes such as blepharophimosis

C. List the pertinent elements of the history

1. Congenital myogenic
 - a. Constant, nonprogressive ptosis since birth
 - b. May be unilateral or bilateral
 - c. Can be associated with double elevator palsy
2. Blepharophimosis syndrome
 - a. Constant, nonprogressive ptosis since birth
 - b. Family history, usually autosomal dominant
 - c. Other periocular abnormalities present
 - i. Bilateral congenital ptosis (usually poor levator function), epicanthus inversus, telecanthus, horizontal phimosis
 - ii. May also have temporal lower eyelid ectropion, flattened nasal bridge, hypertelorism, lop ears with overhanging helix, and primary amenorrhea

D. Describe pertinent clinical features

1. Variable amounts of upper lid ptosis from mild to severe
2. Frequently bilateral, often asymmetric
3. Triad seen on exam
 - a. Levator function typically reduced
 - b. Eyelid lags on downgaze
 - c. Typically poor lid crease (often reduced proportional to reduction in levator function)

4. May have ipsilateral superior rectus weakness
5. May be associated with amblyopia and strabismus

II. Define the risk factors

- A. Amblyopia (mild to moderate ptosis may induce anisometropia with anisometropic amblyopia; severe ptosis may risk occlusion of the visual axis and occlusion amblyopia; both may be associated with strabismus)**

III. List the differential diagnosis

A. Other forms of congenital ptosis

1. Congenital neurogenic ptosis is due to innervational defects during embryonic development, or interruption of innervation due to perinatal trauma
 - a. Congenital cranial nerve (CN) III palsy
 - i. Birth trauma
 - ii. Ischemic insult to brain
 - iii. Usually unilateral
 - iv. Variable limitation of elevation, depression and adduction of the globe
 - v. Dilated pupil
 - vi. May have aberrant reinnervation
 - vii. Frequent amblyopia
 - b. Congenital Horner syndrome
 - i. Interruption of the sympathetic pathway
 - ii. Birth trauma
 - iii. Neuroblastoma
 - iv. 1 to 2 mm of upper eyelid ptosis
 - v. 1 mm of reverse ptosis of the lower eyelid
 - vi. Miosis with dilation lag
 - vii. Anhydrosis if interruption proximal to carotid bifurcation
 - viii. Iris heterochromia, with ipsilateral iris lighter in color
 - c. Marcus Gunn jaw-winking syndrome
 - i. Aberrant connections between motor division of trigeminal nerve and oculomotor nerve
 - ii. Unilateral upper eyelid ptosis with elevation of the ptotic lid with opening of the jaw, or movement of the jaw to the contralateral side

- iii. Variably decreased levator function
 - iv. May have decreased superior rectus function
- 2. Congenital traumatic ptosis
 - a. May be due to birth trauma, particularly forceps
 - b. Usually aponeurotic type induced by trauma
 - c. Usually aponeurotic with good levator function
 - d. High or indistinct upper eyelid crease
- 3. Congenital mechanical ptosis
 - a. Due to congenital eyelid abnormality such as plexiform neurofibroma or hemangioma
 - b. Congenital eyelid lesion present, mechanically weighing down the upper eyelid
- 4. Pseudoptosis
 - a. Apparent eyelid ptosis due to vertical strabismus, enophthalmos, microphthalmos, anophthalmos, phthisis bulbi, orbital fracture, contralateral eyelid retraction, dermatochalasis
- 5. All forms of congenital ptosis may
 - a. Obstruct the visual axis and cause amblyopia
 - b. Result in a chin-up head posture
- B. In older patients**
 - 1. Acquired myogenic ptosis
 - a. Mitochondrial myopathies typically develop sometime after birth but may be confusing in older children
 - 2. Aponeurotic ptosis

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

A. Describe medical therapy options

- 1. Patching and refraction for amblyopia management

B. Surgical therapy - discuss timing of surgery

- 1. Moderate to good function (4-8 mm)
 - a. Levator resection, anterior or posterior
 - b. Müller's muscle conjunctival resection
- 2. Poor levator function (< 4 mm function)
 - a. Frontalis sling
 - i. Autogenous fascia lata

- ii. Preserved fascia
- iii. Silicone, expanded polytetrafluoroethylene (ePTFE (Gortex)), or other alloplastic materials (ex/Supramid)
- b. Whitnall sling +/- superior tarsectomy

C. Special consideration of unilateral vs. bilateral congenital ptosis

- 1. Unilateral congenital ptosis may be difficult to correct and obtain symmetry in cases of poor levator function
 - a. The patient may not be motivated to use unilaterally suspended lid, leading to need to set lid very high with sling or maximum levator resection
 - i. This is especially common if the affected eye is amblyopic
 - ii. May be complicated by poor lid closure, especially at night, and ocular exposure
 - b. May consider bilateral slings with or without destruction of normal levator (Beard procedure)

D. Timing of congenital ptosis surgery

- 1. All surgeons agree that early ptosis surgery is needed if the ptosis (usually bilateral) is causing developmental delay or amblyopia
- 2. Significant congenital ptosis should be repaired, if possible, prior to the child beginning school
- 3. The exact timing of surgery varies according to surgeon preference and degree of ptosis
 - a. Minor congenital ptosis may be better addressed later in life when procedure can be performed with local anesthesia

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

A. Infection

- 1. Minimize risk with careful, standard, aseptic technique
- 2. Treat with antibiotics

B. Over or under correction

- 1. Revise as needed
- 2. Trial of massage for over-correction, if not effective then recess

C. Wound dehiscence

- 1. Repair wound

D. Poor contour

- 1. Prevent with careful surgical technique
- 2. Surgically revise as needed

E. Lag on downgaze

- 1. Expected side effect with large levator advancement/resection or sling

F. Lagophthalmos

1. Expected side effect with large levator advancement/resection or sling
2. Treat with lubricating drops or ointment

G. Bleeding

1. Prevent with careful surgical technique
2. Open wounds if vision is threatened
3. Observe if vision is not threatened

H. Scar

1. Prevent with careful wound closure
2. Treat with massage, corticosteroids (topical or injected); surgical revision when needed

I. Lid crease asymmetry

1. Prevent with careful planning of incision site
2. Crease fixation with eyelid skin wound closure
3. Revise surgically as needed

J. Conjunctival prolapse

1. Prevent by avoiding damage to the suspensory connections between the levator and the superior conjunctival fornix
2. May resolve with patching
3. Tissue may need to be repositioned and secured with full-thickness sutures passed through fornix

K. Dry Eye

1. Lubrication
2. If not effective, then recession of eyelid may be necessary

L. Granuloma, suture abscess, infected or exposed sling material

1. Antibiotics, topical or oral
2. Removal of sling material
3. Repair of sling exposure or extrusion
4. Medical-surgical management of granuloma or suture abscess

VI. Describe disease-related complications

A. Associated with amblyopia, although this is usually refractive from astigmatism rather than deprivation

1. Occurs in 20% of patients with congenital ptosis, but only a small number are due to the ptosis
2. May result from anisometropia, astigmatism, strabismus, or occlusion of the pupil

3. Visual function and refractive error must be evaluated in all children

VII. Describe appropriate patient instructions

- A. Postoperative ocular lubrication**
- B. Periodic follow-up for assessment of visual function**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.
3. AAO, Focal Points: Congenital Ptosis, Module #2, 2001.

Acquired myogenic ptosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Muscular dystrophy
 - a. Including myotonic dystrophy
2. Chronic progressive external ophthalmoplegia
3. Myasthenia gravis (MG)
4. Oculopharyngeal dystrophy

B. Define the relevant aspects of epidemiology of the disease

1. Chronic progressive external ophthalmoplegia (CPEO) is usually seen in 2 patient groups
 - a. Late teen onset, may be part of Kearns-Sayre syndrome
 - i. Look for heart block. Cardiac pacing may be life saving
 - ii. Look for upgaze limitation
 - iii. Decreased levator function
 - iv. Pigmentary retinopathy - salt and pepper fundus
 - v. Increased cerebrospinal fluid (CSF) protein
 - vi. Deafness, vestibular dysfunction
 - vii. Mitochondrial myopathy- "ragged-red" fibers on muscle biopsy
 - b. Seventh decade
 - i. Look for upgaze limitation
 - ii. Decreased levator function
 - iii. Retinal pigmentary changes (less common)
2. Oculopharyngeal dystrophy
 - a. Originally described with French Canadian ancestry, currently several other lines of inheritance have been identified
 - b. Autosomal dominant
 - c. Genetic mutation has been described
 - d. Decreased levator function
 - e. Difficulty with swallowing (dysphagia)

C. List the pertinent elements of the history

1. Progressive ptosis

2. Frequent diplopia
3. May have generalized weakness with myasthenia
4. May have pharyngeal difficulties with oculopharyngeal dystrophy (OPD)

D. Describe the pertinent clinical features

1. Progressive ptosis
2. Decreasing levator function
3. Frequent diplopia
4. Upgaze limitation
5. Heart block, retinopathy in Kearns-Sayre syndrome
6. Generalized weakness with MG
7. Pharyngeal difficulties with OPD

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Chromosomal testing for OPD
2. Electrocardiogram (EKG) for CPEO
3. Tensilon test or acetylcholine (ACh) receptor antibodies for MG
4. Muscle biopsy for muscular dystrophy

II. Define the risk factors

A. OPD is a familial disorder

III. List the differential diagnosis

- A. Aponeurotic ptosis**
- B. Previously unrecognized congenital form of ptosis**
- C. Traumatic ptosis**
- D. Drug induced ptosis such as botulinum toxin**

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

A. Describe medical therapy options

1. Treat underlying systemic condition
 - a. Pyridostigmine bromide (Mestinon) for generalized myasthenia, although not very effective for ocular component of MG.
 - i. Corticosteroids may also be useful for MG

B. Describe surgical therapy options

1. Can try levator resection if levator function good to fair; however ptosis typically is associated with poor or worsening levator function
2. Adjustable frontalis slings, such as silicone, can be used with poor levator function
3. Eyelid crutch or non-surgical candidates

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

- A. Pyridostigmine (Mestinon) toxicity often occurs prior to therapeutic effect
- B. Frontalis slings may lead to ocular exposure (Adjustable slings are therefore desirable)
- C. If corneal exposure cannot be managed medically, raising the lower eyelid can prevent corneal exposure
- D. Botulinum toxin can be used to lower brows to help manage exposure, although this can decrease the ability to open the eyes
- E. If corneal exposure due to frontalis slings cannot be managed with medical therapy, adjusting the sling, or raising the lower eyelid, the slings should be removed

VI. Describe disease-related complications

- A. Poor vision due to obstruction
- B. Cardiac conduction failure in Kearns-Sayre syndrome
- C. Complications of systemic condition

VII. Describe appropriate patient instructions

- A. Adequate lubrication post-operatively for ocular protection
- B. Notify office if develop signs of exposure

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.
3. AAO, Focal Points: Management of Acquired Ptosis, Module #8, 1999.

Aponeurotic ptosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Defect in levator aponeurosis, redundancy, rarefaction, or complete disinsertion

B. Define the relevant aspects of epidemiology of the disease

1. Involutional changes often associated
2. Trauma
3. Edema or stretching related to eyelid or ocular surgery
4. Long term contact lens use
5. Corticosteroids
6. Longstanding edema as in thyroid-related orbitopathy, blepharochalasis
7. Allergy - eyelid rubbing

C. List the pertinent elements of the history

1. See above

D. Describe pertinent clinical features

1. Good levator function (12-15 mm)
2. Elevated eyelid crease
3. No lagophthalmos on downgaze
4. Thin eyelid
5. Associated dermatochalasis and other involutional changes in affected older adults

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. There is no laboratory test used to confirm the diagnosis of aponeurotic ptosis
2. Assessment of tear film using slit lamp biomicroscopic exam and/or Schirmer's testing should be done prior to correcting blepharoptosis
3. Phenylephrine hydrochloride (HCl) 2.5% to assess Müller muscle stimulation/eyelid elevation for surgical planning

II. List the differential diagnosis

A. Acquired myogenic ptosis

B. Horner syndrome

C. Previously unrecognized congenital ptosis

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

A. Describe surgical therapy options

1. Levator repair (aponeurosis advancement)
 - a. Anterior approach
 - b. Posterior approach
2. Müller muscle conjunctival resection

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

A. Overcorrection

1. Treatment:
 - a. Massage
 - b. Ocular lubrication
 - c. Early suture removal

B. Undercorrection

1. Some surgeons use adjustable sutures
2. Early or late readjustment, revision

C. Asymmetry, including A and B above, plus contour deformities

1. Beware of unmasking ptosis due to subclinical aponeurosis dehiscence in opposite lid (due to Hering's law)

D. Ocular exposure

1. Use lubrication (artificial tears and ointment) in post op period
2. If unresponsive to medical therapy, consider recession of levator aponeurosis

E. Wound infection

V. Describe disease-related complications

A. Visual obstruction (blocking peripheral vision, central vision, or in reading position)

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO Monograph 8, Surgery of the Eyelids, Lacrimal System, & Orbit, 2nd edition, 2011.
3. AAO, Focal Points: Congenital Ptosis, Module #2, 2001.

4. AAO, Focal Points: Management of Acquired Ptosis, Module #8, 1999.

Oculomotor (cranial nerve III) palsy

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Brainstem or subarachnoid lesions such as encephalitis, infarction, tumors, aneurysms
2. Cavernous sinus and superior orbital fissure syndromes usually also involve cranial nerve (CN) IV, V1, V2 and VI. Causes include tumors, aneurysms, inflammatory processes, and vascular lesions such as thrombosis or fistula
3. Orbital lesions usually cause dysfunction of other cranial nerves, visual loss and proptosis
4. Orbital lesions may affect superior or inferior divisions
5. Causes include
 - a. Vasculopathic
 - i. Diabetes mellitus, hypertension, hypercholesteremia, hypertriglyceridemia
 - b. Aneurysm
 - c. Head trauma
 - d. Multiple sclerosis
 - e. Migraine
 - f. Viral or vaccines (children)
 - g. Neoplasm

B. Define the relevant aspects of epidemiology of the disease

1. Can be caused by a broad range of pathologic processes
2. Acquired palsy is far more common than congenital
3. Ischemia is the most frequent cause of pupil-sparing palsies
 - a. Diabetes mellitus
 - b. Ischemia
 - c. Atherosclerosis
 - d. Migraine
4. Severe head trauma is a common cause
 - a. If the trauma is mild, suspect an underlying mass lesion
 - b. Delayed development of pupil involved palsy is suggestive of expanding intracranial mass lesion

C. List the pertinent elements of the history

1. Trauma

2. Pain
3. Progression
4. Systemic disorders such as:
 - a. Diabetes mellitus
 - b. Hypertension
 - c. Multiple sclerosis
 - d. Primary or metastatic carcinomas

D. Describe the pertinent clinical features

1. All patients with congenital CN III palsy have some degree of ptosis and paresis of the muscles innervated by the oculomotor nerve
 - a. Typically the eye is deviated down and out
 - b. Superior division supplies the levator palpebrae superioris and the superior rectus
 - c. Inferior division supplies the medial and inferior rectus and the inferior oblique muscles and supplies parasympathetics via a nerve to the inferior oblique
2. Other neurologic disorders may or may not be present
3. Acquired palsies often are associated with other motor nerve palsies, which may have value in localization of the lesion
4. Determination of pupil involvement
5. Aberrant regeneration may develop with recovery, most commonly producing:
 - a. Lid retraction in downgaze
 - b. Lid elevation or pupil constriction on attempted adduction
 - c. Globe retraction on attempted upgaze
6. If ischemic etiology, aberrant regeneration would not occur

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Magnetic resonance imaging (MRI) scan, magnetic resonance angiography (MRA), or computed tomography angiography (CTA)
2. Angiography
3. Blood glucose
4. Blood pressure (BP)
5. Metastatic work up if history of carcinoma

II. Define the risk factors

A. Systemic disorders such as diabetes, hypertension, hypercholesterolemia, hypertriglyceridemia, multiple sclerosis, primary or metastatic carcinoma

- B. Trauma**

III. List the differential diagnosis

- A. Myasthenia gravis**
- B. Miller Fisher variant of Guillain-Barre'**

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

- A. Neurology or neurosurgical consultation following trauma, especially if progressive**
- B. Expectant observation with pupil-sparing palsy**
 - 1. If worsens within first 2 two weeks after onset or does not resolve in 3-4 months repeat imaging, consider lumbar puncture
- C. Treatment of underlying systemic disease**
- D. Strabismus and eyelid surgery**

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

- A. The incomitant strabismus surgical treatment often produces a compromised result with diplopia in some fields of gaze**
- B. Eyelid ptosis surgery may create exposure keratopathy resulting from lagophthalmos, poor ocular motility and poor postoperative eyelid excursion.**
- C. Ptosis repair usually requires frontalis suspension and is reserved for patients in whom strabismus surgery allows single binocular vision in a useful field of gaze. Silicone rod placement may allow for better eyelid closure than more rigid sling materials**

VI. Describe disease-related complications

- A. Strabismus with diplopia**
- B. Ptosis**
- C. Other neurologic or systemic sequelae**

VII. Describe appropriate patient instructions

- A. Observe for progression of symptoms**
- B. Expectations regarding normalcy of surgical results should be realistic**
- C. Lubrication for an exposed eye**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology; Section 6: Pediatric

Ophthalmology and Strabismus, 2013-2014.

2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.91, 262.
3. AAO, Focal Points: Third, Fourth, and Sixth Nerve Palsies, Module #8, 1996.

Horner syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Disruption of the superior cervical sympathetic chain resulting in ocular sympathetic paresis
 - a. 1st order neuron from hypothalamus to spinal cord
 - b. 2nd order neuron from spinal cord to superior cervical ganglion
 - c. 3rd order neuron from superior cervical ganglion along the internal carotid artery to tarsal muscles and pupillary dilator muscle

B. Define the relevant aspects of epidemiology of the disease

1. 1st order neuron lesions are often caused by vascular lesions or tumors
2. 2nd order neuron lesions are often caused by trauma or tumors affecting the cervicothoracic spinal cord
3. 3rd order neuron lesions are often caused by tumors, inflammatory lesions, trauma or ischemia
4. Can be congenital, usually injury to the brachial plexus head
5. May occur with cluster headaches
6. Often no cause is determined

C. List the pertinent elements of the history

1. Scapula or arm pain classically suggests an apical lung tumor
2. Neck trauma or pain classically suggests a carotid artery dissection

D. Describe pertinent clinical features

1. Mild upper lid ptosis with lower eyelid elevation ("reverse ptosis"), giving appearance of enophthalmos due to narrower vertical palpebral aperture
2. Pupillary miosis
 - a. Accentuated in darkness where dilation is stimulated but there is ipsilateral dilation lag (may appear normal in bright light)
 - b. 10% cocaine drops produce less dilation of involved pupil
 - i. Cocaine blocks reuptake of catecholamines (norepinephrine)
 - ii. Less free norepinephrine available at synapse of involved pupil
 - c. Apraclonidine (0.5% or 1%)
 - i. Practical alternative to cocaine
 - ii. Weak alpha agonist
 - iii. Due to denervation supersensitivity of iris dilator muscle affected pupil dilates

- d. 1% hydroxyamphetamine dilates normal pupils by releasing catecholamines from the motor end plate
 - i. Dilation if 3rd order neuron is intact
 - ii. No dilation if 3rd order neuron is involved
- e. Normal pupillary reactions to light and near (sphincter muscles are intact, no loss of accommodation)
- 3. Ipsilateral facial anhidrosis
- 4. Iris heterochromia
 - a. Associated with congenital Horner syndrome- ipsilateral iris is lighter
 - b. In a child with acquired Horner syndrome without any history of birth trauma, the patient should be screened for neuroblastoma
- 5. Presence of cranial nerve (CN) VI palsy helps to localize to third-order neuron lesions
- E. Describe appropriate testing and evaluation for establishing the diagnosis**
 - 1. Chest x ray
 - 2. Computed tomography (CT) scan of chest, head and neck; resonance angiography (MRA) looking for carotid dissection

II. Define the causes and risk factors

- A. Trauma**
- B. Smoking causing lung tumors**
- C. Forceps delivery**
- D. Neck surgery**

III. List the differential diagnosis

- A. Other causes of ptosis**
- B. Pharmacologic pupil**
- C. CN III palsy**
- D. Contralateral iris sphincter tear**
- E. Physiologic anisocoria**

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

- A. Ptosis repair**
 - 1. Müller muscle resection
 - 2. Levator aponeurosis advancement

V. List the complications of treatment

- A. Ptosis repair under or over correction**

VI. Describe disease-related complications

- A. Sequela associated with trauma, tumors**

VII. Describe appropriate patient instructions

- A. Will require study if new onset**

1. The ophthalmologist will identify cause, direct treatment at specific cause and observe

- B. Return for repair of ptosis, if visually or cosmetically significant**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p.24.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.63, 125-127.
4. AAO, Focal Points: Clinical Importance of Pupillary Inequality, Module #10, 1992, p. 2-6, 9-11.

Myasthenia gravis with ptosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. MG caused by antibodies to acetylcholine receptors of the muscle motor endplate

B. Define the relevant aspects of epidemiology of the disease

1. Women affected in 2nd and 3rd decades
2. Men affected in 6th and 7th decades
3. Symptoms present in 15% of children born to MG mothers
 - a. Usually transient
 - b. True congenital MG is much rarer
4. 50% of MG patients present with ocular involvement
5. 15% of MG involves only the eyes (ocular myasthenia)

C. List the pertinent elements of the history

1. Ptosis
 - a. Worse later in the day
 - b. Worse with fatigue
 - c. Often variable
 - d. Diplopia may be associated

D. Describe the pertinent clinical features

1. Patients may complain of generalized weakness and malaise
 - a. This may indicate generalized myasthenia and patients should be referred for urgent neurologic evaluation
2. Ptosis is the most common clinical feature
 - a. Unilateral or bilateral
3. Variability and fatigability are very suggestive of MG
4. Orbicularis oculi is commonly weak
5. May have diplopia due to involvement of extraocular muscles and this may be variable

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Patients with suggestive findings should undergo testing for myasthenia
2. Edrophonium chloride (Tensilon®) test
 - a. Edrophonium is an anticholinesterase which increases the availability of acetylcholine

- b. Positive test is improvement in muscular deficit
 - c. False positive Edrophonium chloride tests have been described with parasellar meningiomas
 - d. Antidote available for adverse reaction to Edrophonium chloride test
- 3. Ice test
- 4. Acetylcholine receptor antibodies.
 - a. Disease may be present in the absence of detectable antibodies
- 5. Single fiber electromyography often positive in both generalized and ocular MG
- 6. Imaging to rule out thymoma

II. Define the risk factors

- A. Thyroid disease**

III. List the differential diagnosis

- A. Acquired aponeurogenic ptosis**
- B. Horner syndrome**
- C. Chronic progressive external ophthalmoplegia**
- D. Myotonic dystrophy**

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

- A. Neurology consultation**
 - 1. Anticholinesterases (pyridostigmine bromide)
 - 2. Systemic corticosteroids
 - 3. Immunosuppressants
 - 4. Thymectomy in presence of thymoma
- B. Ptosis or strabismus surgery when clinically stable on medical treatment**
 - 1. Poor eye protective mechanisms
 - 2. Use caution to avoid overcorrection
 - 3. Ptosis typically needs frontalis sling
 - 4. Ptosis can also be corrected with lid crutches

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

- A. Anticholinesterase side effects include diarrhea, nausea, abdominal cramping, paradoxical muscle weakness from excess dosage**
- B. Pyridostigmine bromide (Mestinon®) toxicity usually comes before systemic efficacy for ocular myasthenia**
- C. Corticosteroid side effects**
- D. Ptosis over and under correction, lagophthalmos with ocular surface exposure**

VI. Describe disease-related complications

- A. Progressive generalized weakness**
- B. Progressive ptosis and strabismus**
- C. Ocular myasthenia unlikely to progress to systemic disease if stable for 2 years**

VII. Describe appropriate patient instructions

- A. Refer to neurology for further diagnosis and treatment**
- B. Consider ptosis and strabismus surgical treatment when medically treated**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.91.
3. AAO, Focal Points: Management of Acquired Ptosis, Module #8, 1999, p.11-12.
4. Benatar M, Kaminski H. Medical and surgical treatment for ocular myasthenia. Cochrane Database Syst Rev 2006:CD005081.

Pseudoptosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Hypotropia in involved, non-fixating side
2. Eyelid retraction in contralateral fixating eye
3. Inadequate lid support by globe and orbital structures (reduced orbital volume)
 - a. Phthisis bulbi
 - b. Microphthalmos
 - c. Enophthalmos
 - d. Anophthalmos
4. Overhang of skin over the eyelid margin
5. Heavy eyebrow, or fat pads

B. Define the relevant aspects of epidemiology of the disease

1. Dermatochalasis is the most common cause
2. Eyebrow ptosis
3. Strabismus causes (common in the hypotropia seen with congenital fibrosis syndrome)
4. Graves disease is the most common cause of eyelid retraction
 - a. It may be present in contralateral lid

C. List the pertinent elements of the history

1. Vertical diplopia
2. Hyperthyroid symptoms of weight loss, tachycardia, and tremors
3. Longstanding problems with the eye leading to phthisis bulbi
4. Orbital trauma
5. Forehead fatigue due to constant brow elevation in an effort to pull the redundant skin away from the eyelid margin
6. Silent sinus syndrome
7. Strabismus - innervational palsy of the contralateral antagonist if inferior rectus

D. Describe the pertinent clinical features

1. An intact levator mechanism
2. Possibly inadequate support of the ptotic lid from behind
 - a. Exophthalmometry readings reduced

3. Possibly excess upper eyelid skin, brow ptosis
4. Stigmata of Graves disease
5. Collaborative eyelid findings - lid lag on excursion
6. Cover-uncover test to look for vertical strabismus

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Thyroid stimulating hormone assay
2. Orbital computed tomography (CT) scan
3. Orbital echography

II. Define the risk factors

- A. Aging
- B. Anophthalmia
- C. Decreased orbital volume

III. List the differential diagnosis

- A. Blepharoptosis
 1. Aponeurotic ptosis
 2. Congenital ptosis
 3. Myogenic ptosis
 4. Traumatic ptosis
 5. Mechanical ptosis

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

- A. Management of strabismus
- B. Orbital volume augmentation
- C. Orbital fracture repair
- D. Prosthetic volume augmentation
- E. Scleral shell fitting
- F. Functional blepharoplasty
- G. Eyebrow lift

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

- A. Repair of the lid in pseudoptosis without addressing underlying cause will typically result in

VI. Describe disease-related complications

- A. Persistence of pseudoptosis**
- B. In anophthalmic or phthisis cases, failure to retain prosthesis**

VII. Describe appropriate patient instructions

- A. Patient education as to the true etiology of the apparent ptosis**
- B. Discussion of treatment options appropriate for the diagnosis**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.89, 94-95, 262.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p.126-128,146-147.
4. AAO, Focal Points: Management of Acquired Ptosis, Module #8, 1999, p.1.

Floppy eyelid syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Histopathologic studies demonstrate decreased tarsal elastin
2. Tarsus becomes rubbery and lax (See Tarsus)
3. Eyelid laxity allows easy upper eyelid eversion and risk of mechanical irritation
4. Association with obstructive sleep apnea (OSA), and possibly keratoconus with chronic eye rubbing
5. Patients tend to sleep prone or on side resulting in eyelid eversion and mechanical irritation to exposed conjunctiva
6. Constant irritation leads to chronic papillary conjunctivitis

B. Define the relevant aspects of epidemiology of the disease

1. Uncommon, but under diagnosed
2. Slightly more prevalent in men than women
3. Most commonly diagnosed in middle-aged patients (35-55 years)
4. Usually seen in obese patients
5. Strong association with OSA - approximately 25% of OSA patients have FES

C. List the pertinent elements of the history

1. Unilateral or bilateral eye irritation and burning
2. Tearing
3. Ropy, mucoid discharge - usually worse in the morning
4. Decreased vision if there is an associated keratopathy
5. Association with OSA
 - a. Daytime somnolence
 - b. Morning headaches
 - c. Sleep history
 - i. Does patient sleep on side or face down in pillow?
 - ii. Frequent episodes of waking up during the night
 - iii. Does patient already have known OSA or been prescribed nasal continuous positive airway pressure (CPAP) at night?
6. Past ocular history
 - a. Chalazia or hordeola (See Chalazion)

- b. Keratoconus
- c. Contact lens use
- d. Seasonal symptoms
- 7. Past medical history
 - a. Obstructive sleep apnea (OSA)
 - b. Acne rosacea
 - c. Psoriasis
 - d. Hypertension
 - e. Congestive heart failure

D. Describe pertinent clinical features

- 1. Lax upper eyelid that is easily everted when pulling lid superiorly towards eyebrow
- 2. Atrophic, soft and rubbery tarsal plate that can be folded upon itself
- 3. Stringy mucoid conjunctival discharge
- 4. Papillary conjunctival inflammation - especially palpebral conjunctiva of upper and lower eyelids
- 5. Eyelash ptosis with loss of lash parallelism
- 6. Meibomian gland dysfunction
- 7. Periorbital involutional changes
 - a. Brow ptosis (See Brow ptosis)
 - b. Dermatochalasis (See Dermatochalasis)
 - c. Blepharoptosis
 - d. Lower eyelid horizontal laxity involving medial and lateral canthal tendons as well as the tarsal abnormalities noted above
 - e. Lacrimal gland prolapse

E. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Pull upper eyelid towards brow looking for easy eversion of upper eyelid and rubbery consistency of tarsus
- 2. Lower eyelid distraction and snap back test looking for horizontal laxity
- 3. Tear break-up time (TBUT) - may be less than 10 seconds indicating tear instability
- 4. Slit lamp examination
 - a. Lash ptosis and loss of parallelism
 - b. Lash debris (scurf)
 - c. Superior papillary tarsal conjunctival hypertrophy
 - d. Superior bulbar conjunctival injection

- e. Punctate fluorescein staining of superior cornea and conjunctiva
- 5. Conjunctival scrapings
 - a. Papillary conjunctival hypertrophy
 - b. Predominance of polymorphonuclear leukocytes (PMNs) with variable amounts of eosinophils and lymphocytes
- 6. Sleep study - if suspect OSA

II. Define the risk factors

A. Association between floppy eyelid syndrome and OSA

- 1. Experience episodic apnea and hypoxia due to obstruction of airway during sleep
- 2. There is collapse of pharynx during inspiration, resulting in loud snoring and eventual apnea causing patient to awaken
- 3. Difficulty getting into deep restorative phase of sleep leading to daytime somnolence and fatigue
- 4. Patients with OSA are at risk of developing systemic or pulmonary hypertension, congestive heart failure, and cardiac arrhythmias

B. Obesity

C. Keratoconus

D. Chronic eyelid rubbing

E. Meibomian gland dysfunction - possible association with *Demodex brevis* infestation

III. List the differential diagnosis

A. Involutional eyelid laxity

B. Other causes of papillary conjunctivitis

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

A. Describe the natural history, outcome and prognosis

- 1. If unrecognized can develop worsening papillary conjunctival changes, corneal erosions and ultimately ulceration and scarring in severe cases
- 2. Chronic conjunctivitis, punctuate keratopathy , and corneal neovascularization can result in contact lens intolerance
- 3. Associated OSA is a potentially serious problem that can ultimately lead to heart failure and pulmonary hypertension in more severe cases

B. Describe medical therapy options

- 1. Ocular lubrication

2. Trial of a tetracycline if meibomian gland dysfunction is suspected
3. Tape eyelids closed and wear eye shield while asleep
4. Referral to pulmonologist for treatment of any OSA
 - a. May require nasal CPAP at night while sleeping

C. Describe surgical therapy options

1. More conservative medical care often proves inadequate in relieving signs and symptoms
2. Horizontal tightening of the upper eyelid by full-thickness resection/reconstruction or lateral canthoplasty
3. Horizontal tightening of lower eyelid by standard techniques such as lateral tarsal strip procedure if medial canthal tendon laxity is not severe
4. Repair of associated ptosis, dermatochalasis, or lacrimal gland prolapse (some investigators have noted that repair of FES by full-thickness resection/reconstruction often improves blepharoptosis as well; some repair blepharoptosis secondarily if needed while others prefer primary repair at time of horizontal eyelid shortening)

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

A. Poor wound healing

1. Meticulous surgical technique
2. Topical antibiotic/steroid combination
3. Avoidance of eye rubbing post-operatively

B. Unacceptable eyelid height or contour

1. Often improves with time
2. May require revision

C. Recurrence of condition

D. Continued patient symptoms of irritation, burning, and discharge

1. May require concomitant or sequential treatment of coexistent pathology

VI. Describe disease-related complications

- A. Progressive papillary conjunctival changes, mucoid discharge, tear instability, corneal erosions and ultimately ulceration and scarring in severe cases**

VII. Describe appropriate patient instructions

- A. Follow-up as instructed**
- B. Ocular lubrication**

- C. Avoid sleeping with face against pillow or wear eye shield**
- D. Refrain from rubbing eyes**
- E. Possible association with OSA and need for further tests if OSA is suspected**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Ectropion and Entropion. Vol. 12, No. 10. 1994, p. 4-5.
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9. Netland PA, Sugrue SP, Albert DM, Shore JW. Histopathologic features of the floppy eyelid syndrome: involvement of tarsal elastin. *Ophthalmology* 1994;101:174-181.
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Anterior lamellar deficiency

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Describes a deficiency of skin at the upper and/or lower eyelids that results in eyelid retraction, ectropion, cicatricial lagophthalmos, or poor eyelid motility
2. Caused by
 - a. Any entities which inflame and shorten the skin of the eyelids
 - b. Iatrogenic loss of eyelid skin from eyelid or facial surgery (e.g., blepharoplasty, eyelid tumor removal, laser resurfacing, chemical/thermal burns)
 - c. Trauma with subsequent cicatrization
 - d. History of cicatrizing skin disease

B. Define the relevant aspects of epidemiology of the disease

1. Acute dermatologic entities such as allergic dermatitis or infectious dermatitis (often reversible)
2. Chronic dermatologic entities
 - a. Atopy
 - b. Scleroderma, ichthyosis and related disorders
3. Blepharitis and acne rosacea
4. Trauma
5. Chronic actinic damage
6. Postsurgical
 - a. Reconstructive
 - b. Cosmetic

C. List the pertinent elements of the history

1. Speed of onset is important, because acute onset may resolve with acute medical treatment
2. Blepharitis history
 - a. Itching, burning, irritation
 - b. *Staphylococcus aureus*, *Staphylococcus epidermidis*, *Streptococcus*, *Haemophilus*, *Moraxella*, *Neisseria*
3. Atopic history
 - a. Childhood eczema progresses to adult atopic dermatitis
 - b. Triggered by dry skin, foods, wool clothing, anxiety
4. Rosacea history

- a. Chronic acneiform eruption of face
 - b. Erythema, papules, pustules, telangiectasia
 - c. Possibly rhinophyma
 - d. Seborrhea
 - e. Associated blepharitis
- 5. Trauma
 - 6. Chemical or thermal burn
 - 7. Surgery of the eyelids or face
 - a. Cosmetic lower eyelid surgery - blepharoplasty, laser resurfacing, chemical peel
 - b. Upper eyelid surgery with excessive skin removal
 - c. Reconstructive surgery for tumors, trauma or congenital deformity
 - 8. Chronic sun exposure with actinic skin damage
 - 9. Skin neoplasms
 - 10. Rarely congenital, as in blepharophimosis syndrome

D. Describe pertinent clinical features

- 1. Eyelid inadequately covers, protects, and interacts with the eye
- 2. Keratitis, conjunctivitis
- 3. Dry scales
- 4. AM mattering
- 5. Lid margin irritation, keratinization
- 6. Thickening and redness of margins
- 7. Retraction
- 8. Ectropion
- 9. Lagophthalmos and/or weak blink
- 10. Scarring of face or eyelid, traumatic or surgical
- 11. Lateral canthal dystopia, rounding
- 12. Acute inflammation associated with application of medication, cosmetics
- 13. Rubbing of the eyes after handling chemicals, soaps

E. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Intracutaneous testing with protein allergens
- 2. Culture of eyelid margins
- 3. Possible biopsy

II. Define the risk factors

- A. Personal or family history of atopy**
- B. Excessive sun exposure, light skin color**
- C. Eyelid rubbing**
- D. Generalized skin disorders**

III. List the differential diagnosis

- A. Middle lamellar deficiency**
- B. Posterior lamellar deficiency**
- C. Thyroid eyelid retraction**

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

- A. Removal of allergens**
- B. Antibacterial therapies**
- C. Lid hygiene measures**
- D. Anti-inflammatory agents such as topical corticosteroids**
- E. Botulinum toxin relaxation of frontalis muscle may be temporarily helpful for symptomatic upper eyelid anterior lamellar deficiency**
- F. Surgical treatment**
 - 1. Skin grafting
 - 2. Horizontal eyelid tightening- often only temporary
 - 3. Mid face lifting
- G. Treat co-existing middle and posterior lamellar deficiencies concomitantly**
- H. Discourage eyelid rubbing**
- I. Tazarotene 0.1% cream for the treatment of ichthyosis**

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

- A. Atopic reaction to topically applied ointments or eyedrops**
 - 1. Obtain careful allergy history
 - 2. Prompt cessation of medication; consider preservative-containing compounds as possible cause
 - 3. Topical corticosteroid therapy
 - 4. Corticosteroids can cause thinning of skin and vascular changes

- B. Response inadequate to allow eyelid to resume normal position on eye**
- C. Contracture of skin graft**
 - 1. Over-size the graft
 - 2. Massage with corticosteroid ointment
 - 3. Harvest thin skin from eyelid, posterior auricular
- D. Recurrent midfacial ptosis**
 - 1. Repeat midface lift with preauricular facelift

VI. Describe disease-related complications

- A. Corneal exposure sequelae including inflammation with scarring, keratitis sicca, ulcer**
- B. Inferior keratoconjunctivitis**
- A. Cosmetic concerns**

VII. Describe appropriate patient instructions

- A. Ocular lubrication**
- B. Treatment of generalized dermatitis**
- C. Topical corticosteroid therapy when indicated**

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Middle lamellar deficiency

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Shortening of the orbital septum and surrounding tissues, usually produced by trauma or surgery

B. Define the relevant aspects of epidemiology of the disease

1. Eyelid or facial trauma
2. Cosmetic eyelid or facial surgery
3. Reconstructive eyelid or facial surgery

C. List the pertinent elements of the history

1. Eyelid or facial trauma
2. Eyelid or facial surgery
3. Congenital orbitofacial defects
4. Strabismus surgery
5. Thyroid disease

D. Describe the pertinent clinical features

1. Eyelid retraction in the presence of normal amounts of skin and conjunctiva
2. Inferior keratoconjunctivitis
3. Eyelid is usually stiff or "tethered" to the rim
4. Lagophthalmos
5. May be associated with anterior and posterior lamellar eyelid scarring as well

II. Define the risk factors

- A. Surgery
- B. Trauma

III. List the differential diagnosis

- A. Anterior lamellar deficiency
- B. Posterior lamellar deficiency
- C. Thyroid eyelid retraction

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

- A. Acute middle lamellar deficiency (within 6 mos of onset) may be improved with aggressive massage to improve resistance to passive eyelid movement with or without the use of intralesional corticosteroid injection(s)**
- B. Chronic cicatricial middle lamellar deficiency requires surgical treatment**
 - 1. Lysis of cicatrix (including septal scarring) and vertical lengthening procedures
 - 2. Muscle or muscle composite transposition flaps
 - 3. Anterior or posterior lamellar lengthening may be used in conjunction with lysis of middle lamellar cicatrix, if indicated
 - a. Midface lifting, hard palate, ear cartilage, allograft spacer, mucous membrane graft, amniotic membrane
 - 4. Fat pedicle flap or free graft as a middle lamellar spacer to prevent re-formation of the cicatrix
 - 5. Intrasurgical use of antimetabolites
- C. Ocular lubrication**

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

- A. Recurrent vertical eyelid retraction**
 - 1. Massage, corticosteroids
 - 2. Eyelid traction sutures postoperatively
 - 3. Minimization or avoidance of damaging the muscle layer during eyelid procedures
 - 4. Complete release of all cicatrix and septal scarring
- B. Cicatricial entropion or ectropion**
 - 1. Eyelid malposition type is determined by which lamellae are also involved

VI. Describe disease-related complications

- A. Ocular exposure**
 - 1. Keratitis sicca, ocular surface abnormalities
 - 2. Scleral show
- B. Epiphora**
 - 1. Structurally deficient lacrimal outflow system
 - 2. Poor lacrimal outflow pump function
 - 3. Reflex hypersecretion
- C. May be associated with other congenital defects**
- D. Cosmetic deficiencies**

VII. Describe appropriate patient instructions

A. Multiple surgical interventions may be required

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Posterior lamellar deficiency

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Shortening of the conjunctiva or retractor layers of the eyelid resulting in eyelid retraction or entropion
2. See Cicatricial upper and lower eyelid entropion and Thyroid eye disease (eyelid retraction) for more details about these disorders

B. Define the relevant aspects of epidemiology of the disease

1. Most common cause of eyelid retraction is thyroid eye disease (Graves disease)
2. Conjunctival cicatrization may be related to inflammatory disorders, ocular mucous membrane pemphigoid
3. Postsurgical over-correction of upper eyelid ptosis via conjunctival Müller muscle resection
4. Lower eyelid retraction may occur after transconjunctival lower lid blepharoplasty due to scarring of the posterior lamella. Similarly, retraction or tethering of the lower eyelid can follow fracture repairs, although this more commonly involves the middle lamella after an infraciliary approach to orbital fractures.
5. Conjunctival chemical burns
6. Chronic eyelid infections
7. Trachoma
8. Trauma

C. List the pertinent elements of the history

1. Association with thyroid inflammation
2. Acute onset following surgical event, other trauma
3. Slow onset associated with chronic inflammation
4. Conjunctival cicatricial disorder
 - a. Stevens-Johnson syndrome
 - b. Mucous membrane pemphigoid
 - c. For lower eyelid
 - i. Complication of transconjunctival blepharoplasty or transconjunctival approach to blowout fracture repair

D. Describe the pertinent clinical features

1. Cicatricial entropion
2. Eyelid retraction

3. Associated general stigmata of thyroid disease, mucous membrane disease, or trachoma
4. Ocular surface exposure abnormalities

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Immunofluorescent staining techniques for basement membrane immunocomplex reactions

II. Define the risk factors

- A. Endemic area for trachoma**

III. List the differential diagnosis

- A. Anterior lamellar deficiency**
B. Middle lamellar deficiency

IV. Describe patient management in terms of treatment and follow-up (See Cicatricial upper and lower eyelid entropion for additional detail)

A. Withdrawal of offending agent causing Stevens-Johnson syndrome

1. Supportive measures, if necessary, by medical and dermatologic service

B. Irrigation following chemical burn

C. Local and systemic care of cicatrizing conjunctival disease

D. Oculoplastic treatment is primarily surgical

1. Retraction
 - a. Lengthening of the eyelid with spacers such as full thickness buccal mucosa, hard palate, ear cartilage
2. Horizontal eyelid margin tightening
3. Canthal repositioning
4. Cicatricial entropion
 - a. Margin rotation to treat cicatricial entropion
 - i. Tarsotomy
 - ii. Full thickness blepharotomy
 - b. Mucous membrane graft
 - c. Lash excision with partial or full thickness buccal mucous membrane graft, amniotic membrane or conjunctival graft, recession of the anterior lamella (See Cicatricial upper and lower eyelid entropion)

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

A. Recurrent entropion

1. Adequately incise or excise cicatrix

B. Recurrent retraction

1. Place adequate spacer

VI. Describe disease-related complications

A. Ocular surface disease

B. Trichiasis

C. Progressive cicatrix

D. Exacerbation of systemic disease

VII. Describe appropriate patient instructions

A. Lubricate eyes

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Benign essential blepharospasm

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Idiopathic
2. Possible thalamic, basal ganglion, mesencephalic, or brainstem lesions
3. Similar spasms can be associated with Parkinson disease

B. Define the relevant aspects of epidemiology of the disease

1. Mean age is 56 years
2. Women outnumber men 3:1

C. List the pertinent elements of the history

1. Variable periods of bilateral blepharospasm lasting seconds to minutes
2. Progress may be asymmetric
3. Progress to complete eyelid closure with functional blindness in 12% of patients
4. Some patients have inability to open the eyelids (apraxia of eyelid opening)
5. Not present during sleep

D. Describe pertinent clinical features

1. Triggers include sunlight, stress, wind, noise, eyelid, movement, reading, fatigue
2. Some patients learn techniques to diminish the spasm temporarily such as tongue thrusting, humming, mouth opening, extending the neck, closing one eye or rubbing the face. Sleep and rest may improve symptoms
3. Progression is the rule although usually not to complete incapacitation.
4. Social withdrawal with inability to work or drive may result
5. Eventually the lids may clamp shut and must be pried open with fingers
6. Brow protractor spasm associated
7. Eyebrow position in relation to the superior orbital rim differentiates benign essential blepharospasm from apraxia of eyelid opening
8. Associated anatomic problems can result or accompany essential blepharospasm
 - a. Brow ptosis
 - b. Blepharoptosis, associated levator disinsertion
 - c. Dermatochalasis
 - d. Entropion or ectropion
 - e. Eyelid phimosis

9. 70% develop lower facial dystonia (Meige syndrome) orofacial cervical dystonia

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Neurology evaluation may be indicated

II. List the differential diagnosis

A. Hemifacial spasm

B. Aberrant regeneration following facial nerve palsy (Bell's palsy, trauma, surgery, etc)

C. Parkinson disease

D. Apraxia of eyelid opening

E. Orbicularis myokymia

1. Caffeine, stress, physical exertion, fatigue

F. Reflex blepharospasm

1. Corneal/ocular surface irritation
2. Anterior uveitis
3. Dry eye syndrome
4. Medications causing facial dystonia

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

A. Botulinum toxin

1. Average duration of therapeutic effect is 3 months, so patients require repeat injections to control spasms
2. Care should be taken to inject subcutaneously over the target muscle
3. Normal dosage 2.5-5 units per injection site
4. Onset of effect 2-7 days post injection
5. Have patients use ocular lubricants after injection as eyelid closure may be impaired

B. Myectomy

1. Reserved for patients with an inadequate response to botulinum toxin; patients previously minimally responsive to botulinum toxin can get effect after myectomy
2. Most patients still require botulinum neurotoxin even after successful myectomy, though often at decreased dose and frequency.

C. FL-41 Tinted glasses

D. Selective facial neurectomy

1. Historically the first line of treatment, now rarely performed. Success rate is approximately 50%
Selective facial neurectomy targeting the zygomatic branches often results in ectropion and

perioral muscle weakness

E. Muscle relaxants

1. Muscle relaxants such as orphenadrine have been used effectively in mild cases of essential blepharospasm

F. Address any causes of secondary blepharospasm which may accompany essential blepharospasm

G. Some patients may benefit from biofeedback or hypnosis

H. Patients may demonstrate apraxia of eyelid opening after management of blepharospasm

1. Consider frontalis slings (adjustable materials) or eyelid crutches

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

A. Botulinum toxin

1. Primarily involvement of levator, inferior oblique muscles
 - a. Ptosis
 - b. Diplopia
2. Injection should be remote from these muscles
3. The patient should be instructed not to compress the just-injected areas to prevent movement of the botulinum toxin
4. The secondary effects will diminish and resolve as the medication effects subside
5. Superficial keratitis due to reduced orbicularis tone and blinking
6. Ectropion, lagophthalmos

B. Myectomy

1. Recurrence
2. Necrosis of overlying skin
3. Lagophthalmos, retraction, ectropion, ptosis

V. Describe disease-related complications

A. Visual dysfunction with lifestyle limitation

VI. Describe appropriate patient instructions

A. Recommend treatment when functioning becomes limited

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. Bradley EA, Hodge DO, Bartley GB. Benign essential blepharospasm among residents of Olmsted County, Minnesota, 1976 to 1995: an epidemiologic study. *Ophthal Plast Reconstr Surg* 2003;19:177-81.
3. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit and Lacrimal System, Vol. 2, 1994.
4. AAO, Focal Points: Essential Blepharospasm and Related Diseases, Module #5, 2000.
5. Ben Simon GJ, McCann JD. Benign essential blepharospasm. *Int Ophthalmol Clin* 2005;45:49-75.
6. Costa J, Espirito-Santo C, Borges A, et al. Botulinum toxin type A therapy for blepharospasm. *Cochrane Database Syst Rev* 2005:CD004900.
7. AAO, Focal Points: Botulinum Toxin in Ophthalmology, Module #3, 2007.
8. Blackburn MK, Lamb RD, et al. FL-41 tint improves blink frequency, light sensitivity, and functional limitations in patients with benign essential blepharospasm. *Ophthalmology* 2009; 116:997-1001.

Hemifacial spasm

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Vascular compression of the facial nerve at the brainstem
2. Less common: pontine glioma or other mass compressing facial nerve

B. Define the relevant aspects of epidemiology of the disease

1. Usually occurs in older patients, usually as a result of dolichoectatic basilar artery
2. Suspect tumor in young patients with hemifacial spasm

C. List the pertinent elements of the history

1. Uncontrolled spasm on one side of the face, usually present for an variable period of time prior to diagnosis

D. Describe pertinent clinical features

1. Begins with unilateral fasciculations of the orbicularis oculi muscle and later involves the lower facial nerve-served muscles
2. Intermittent synchronous gross contractures of the unilateral face
3. Spasms continue during sleep
4. Often associated with ipsilateral facial nerve-served muscle weakness and aberrant regeneration of the facial nerve
5. Rarely bilateral, but spasms are not synchronous

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Where etiology of hemifacial spasm is unclear, magnetic resonance imaging (MRI)/magnetic resonance angiography (MRA) scan to look for vascular compression of the facial nerve at the brainstem, pontine glioma, or posterior fossa masses is appropriate

II. List the differential diagnosis

A. Benign essential blepharospasm

B. Meige syndrome

C. Aberrant regeneration following facial nerve palsy

D. History of Bell palsy or trauma

E. Parkinson disease

F. Orbicularis myokymia

1. Caffeine
2. Stress

- 3. Physical exertion
- 4. Fatigue
- G. Corneal irritation**
- H. Facial tics**

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

- A. Botulinum toxin**
 - 1. Requires repeated injection often at intervals of 4 to 6 months
 - 2. Less frequently than required for benign essential blepharospasm
- B. Neurosurgical decompression of the facial nerve at the brainstem**
 - 1. Helpful in some patients
 - 2. Trial of orphenadrine may be used prior to botulinum toxin injections
- C. Orbicularis oculi hemi-myectomy sometimes used for patients who do not wish to undergo neurosurgical decompression.**
- D. Correction of any concurrent eyelid ptosis may decrease visual complaints**

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

- A. Botulinum toxin**
 - 1. Involvement of levator, inferior oblique muscles
 - a. Ptosis
 - b. Diplopia
 - 2. Injection should not be directly overlying these muscles
 - 3. The paresis will diminish and resolve as the medication effects subside
 - 4. Ectropion, lagophthalmos may occur
 - 5. If cheek or perioral muscles are injected, transient oral incompetence may occur
- B. Inadequate response to neurosurgical decompression**

V. Describe disease-related complications

- A. Decreased vision of the involved eye**
- B. Social difficulties**

VI. Describe appropriate patient instructions

- A. Return for botulinum toxin injections as needed**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 5: Neuro-Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994.
3. AAO, Focal Points: Essential Blepharospasm, Module #6, 1988.
4. Costa J, Espirito-Santo C, Borges A, et al. Botulinum toxin type A therapy for hemifacial spasm. Cochrane Database Syst Rev 2005:CD004899.
5. AAO, Focal Points: Botulinum Toxin in Ophthalmology, Module #3, 2007.

Dermatochalasis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Laxity (chalasis) of the skin of the eyelids
2. Most commonly involutional

B. Define the relevant aspects of epidemiology of the disease

1. Involutional change genetically related usually
2. Familial facial characteristics
3. Greater in Northern European peoples with less skin pigmentation.
4. Stretching can be exacerbated by conditions causing allergic swelling and in fluid retention conditions

C. List the pertinent elements of the history

1. The patient may complain of aesthetically or functionally displeasing full appearance of the eyelids
2. Forehead straining/ache may be caused by lifting of the excess skin by active frontalis muscle flexion to counteract ptosis or pseudoptosis. Visual functioning and visual fields may be reduced
 - a. A pressure sensation may be present on the upper eyelashes
3. Markedly redundant skin in the lower eyelids occasionally may touch the eye glasses and block bifocal vision
4. Recurrent unilateral or bilateral swelling in a younger patient may suggest a diagnosis of blepharochalasis syndrome.
5. History of eyelid swelling and lid retraction should alert the surgeon to the possibility of thyroid eye disease (Graves disease)
6. Patient complaints of irritation where the skin folds on itself

D. Describe the pertinent clinical features

1. Skin laxity in the upper or lower eyelids
2. Draping of the upper eyelid skin down to or over the eyelashes.
3. Forehead/eyebrow ptosis may be associated
4. Contraction of the frontalis muscle to raise the eyebrows and redundant eyelid skin.
5. Herniation of the eyelid fat (steatoblepharon) may be associated
 - a. Apparent bulging contours in the lower eyelids may be accentuated by descent of midface soft tissues
6. Dermatitis may be present in the excess skin folds
7. Lacrimal gland prolapse may also be seen.

E. Describe appropriate evaluation for establishing the diagnosis and severity of disease

1. Manual or automated perimetry is performed to measure superior visual field obstruction with the eyelids in natural position and then with the excess skin taped up (generally required for insurance coverage)
 - a. There is no established standard for the severity of obstruction that qualifies patients for corrective surgery
2. Photographs, often frontal and side view

II. Define the risk factors

- A. Age
- B. Allergy
- C. Fluid retention states

III. List the differential diagnosis

- A. Eyebrow ptosis
- B. Blepharoptosis
- C. Acute reversible edema
- D. More chronic reversible swelling associated with systemic disease (e.g., thyroid, renal failure, allergy)
- E. Blepharochalasis syndrome

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

- A. Treat the reversible causes of edema
- B. Surgical reduction of the excess eyelid skin (blepharoplasty) and fat, if indicated
- C. Forehead/eyebrow lifting procedures
- D. Eyelid and canthal skin resurfacing procedures, e.g., laser resurfacing, chemical peel, fillers

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

- A. Inadequate or excess skin/fat removal resulting in:
 1. Asymmetry
 2. Under or over correction
 3. Lagophthalmos
 4. Corneal exposure
 5. Inferior scleral show or ectropion

6. Infection
 7. Hematoma or hemorrhage
 8. Wound dehiscence
- B. Prevented by proper skin marking with proper positioning of the eyebrows (See Lower eyelid blepharoplasty and Upper eyelid blepharoplasty)**

VI. Describe disease-related complications

- A. Blepharoptosis of the upper eyelid often is associated**
- B. Horizontal laxity of the lower eyelids may also be present**
- C. Other age-related findings, such as brow ptosis are commonly seen in conjunction**

VII. Describe appropriate patient instructions

- A. Do not rub the eyelids**
- B. Routine postoperative care**
- C. Warnings regarding infection, wound dehiscence, or hemorrhage**

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. 9-10.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.4.
4. AAO, Focal Points: Upper Eyelid Blepharoplasty, Module #8, 1995.

Upper eyelid blepharoplasty

I. List the indications/contraindications

A. Indications

1. Excess skin in the upper eyelids that causes a problem for the patient.
 - a. Vision and superior and lateral visual fields may be compromised
 - b. Skin resting on the eyelashes may annoy the patient and push the eyelashes downward
 - c. Chronic blepharitis may be present in the area beneath the skin fold
 - d. Chronic frontal headache may result from constant elevation of the eyebrows in an effort to pull the redundant skin fold away from the eyelashes
 - e. Often associated with concomitant brow ptosis
2. Cosmetic concerns

B. Contraindications

1. Normal skin volume present, with the apparent excess caused by forehead ptosis
2. Dry eye conditions, as the blink dynamic may be altered postoperatively
3. When the skin will be needed for a grafting source in the future, as in a patient with recurrent periocular neoplasm

II. Describe the pre-procedure evaluation

A. Patient history

1. Effect on driving, reading and other close activities, TV
2. Frontal headache
3. Dry eye problems
4. Ocular irritation
5. Aesthetic concerns

B. Clinical examination

1. Identify the upper lid skin fold inferior extension with the forehead in habitual and relaxed positions
 - a. Resting on the lashes or below indicates a functional indication for the procedure
2. Measure
 - a. Upper eyelid position and levator function
 - i. Rule out or treat coexisting blepharoptosis
 - b. Tear meniscus

- c. Blink action
- d. Lid closure status (lagophthalmos)

C. Preoperative assessment

- 1. Documentation with photographs, visual fields with and without tape elevating the redundant skin away from the lash margin
- 2. Evaluation of lacrimal function

III. List the alternatives to the procedure

A. Forehead lifting, brow plasty

IV. Describe the technique

- A. The upper eyelid crease is marked with attention to symmetry, either at its existing location or its desired location**
- B. The amount of upper eyelid skin to be removed is determined with the eyebrow in the neutral position**
- C. Some surgeons use the "pinch technique" to determine the amount of extra skin that will be excised**
- D. More important than the exact marking technique is leaving adequate and symmetric amounts of eyelid skin for normal closure.**
- E. Skin is excised with or without the underlying orbicularis muscle**
- F. Orbital septum may be opened or tightened to address prolapsing pre-aponeurotic fat pads.**
- G. Address lacrimal gland prolapse if present, with fixation to orbital rim periosteum**
- H. Suture closure**

V. List the complications of the procedure, their prevention, and management-

- A. Inadequate skin removal such that the symptoms are not relieved**
 - 1. Careful measurement with the brow in neutral position prevents this.
 - 2. Additional skin may need to be removed or brow plasty performed
- B. Excess skin removal preventing closure of the eyes**
 - 1. Careful measurement with the brow in neutral position prevents this
 - 2. Lubrication of the eyes and watchful following of the patient will support the patient while orbicularis muscle function returns
 - 3. In extreme cases, skin grafting may be required
- C. Wound separation may occur in the first week postoperatively**
 - 1. A local increase in eyelid inflammation accompanies the separation, and must be differentiated

from infective cellulitis and allergy to the antibiotic ointment

2. Prevention involves subcutaneous suture support, choice of a skin suture which will provide adequate support, and proper skin removal to avoid excess wound tension

D. Asymmetric eyelid creases

1. Seen when crease incision is not symmetric, or undiagnosed aponeurotic ptosis is present
2. Meticulous marking is critical to avoid this complication
3. Crease reformation sutures may be useful in creating symmetric creases

E. Dry eye symptoms

1. Lubrication of the eyes and stressing the importance of blinking help
2. Bandage contact lenses
3. Lubrication of the eyes and watchful following of the patient will support the patient while orbicularis muscle function returns
4. Preoperative discussion of this possibility with the patient should be beneficial
5. Severe dry eye patients should undergo judicious skin removal if at all

F. Allergy to the antibiotic ointment

1. This occurs about 7 days postoperatively or earlier if exposed to the medication before
2. Redness, swelling and itching are present
3. Differentiation from infective cellulitis and wound separation is essential
4. Cessation results in improvement within a few days. May respond more quickly with topical and or oral steroids

G. Infection

1. Rare, as the blood supply to the eyelid is excellent
2. Sterile technique and treatment with antibiotic ointment are standard protocol
3. Broad spectrum oral antibiotics will rapidly improve a cellulitis appearing in the first postoperative week

H. Hematoma

1. Rare, and can be vision threatening
2. If the patient notes a sudden increase in pain they are instructed to call immediately
3. Careful hemostasis is obtained intraoperatively
4. If possible, medications which prolong bleeding time should be stopped preoperatively with the approval of the primary care physician
5. Over-the-counter or herbal medications that could prolong bleeding time should be discontinued in sufficient time before surgery

VI. Describe follow-up care

- A. Follow up is usually done at 1-2 weeks postoperatively
- B. Follow up is complete when the patient is satisfied and when the lid and cornea are stable

VII. Describe appropriate patient instructions

- A. Cold compresses are used for one or two days postoperatively
- B. Follow-up immediately for sudden pain, swelling and decrease in vision,
- C. Antibiotic ointment is used on the incision until it epithelializes by 10 days, or until absorbable sutures are no longer present
- D. After 4-5 days, warm wet compresses are applied
- E. Lubrication of the eyes is stressed

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p.12-20, 29-32.
3. AAO, Focal Points: Upper Eyelid Blepharoplasty, Module #8, 1995.
4. AAO, Ophthalmic Technology Assessment: Functional Indications for Upper and Lower Eyelid Blepharoplasty. Ophthalmology 1995;102:693-695.
5. Bosniak S. Reconstructive upper lid blepharoplasty. Ophthalmol Clin North Am 2005;18:279-89,vi.
6. Purewal BK, Bosniak S. Theories of upper eyelid blepharoplasty. Ophthalmol Clin North Am 2005;18:271-8,vi.

Lower eyelid blepharoplasty

I. List the indications/contraindications

A. Indications

1. Usually lower eyelid blepharoplasty is cosmetic. Indications include dissatisfaction with
 - a. Wrinkling or sagging of the skin
 - b. Herniation of orbital fat
 - c. "Dark circles below the eyes"
 - d. Deep or prominent tear trough
 - e. "Sad appearance" due to lateral canthal inferior dystopia
2. Rarely, the excess skin and bulging tissues will interfere with looking through the bifocal and are treated for functional reasons

B. Relative contraindications

1. Eyelid retraction
2. Cicatricial ectropion
3. Dry eye
4. Facial neuropathy
5. Unrealistic patient expectations
6. Contraindications should be addressed prior to blepharoplasty

II. Describe the pre-procedure evaluation

A. Patient history

1. Repeated allergic swelling in the periocular region
2. Alcohol usage resulting in lower eyelid swelling
3. Hepatic, renal, hypoproteinemic, hyper and hypothyroid and other endocrine disorders producing lower eyelid swelling
4. Some medications may be associated with eyelid swelling
5. Dry eye history

B. Clinical examination

1. Eyelid position
2. Horizontal eyelid tension
3. Skin type and redundancy

4. Festoons
5. Hypertrophic orbicularis muscle
6. Herniation of fat, if present, for each of the 3 fat pockets
7. Conjunctival and corneal integrity
8. Tear trough and/or visibility of inferior orbital rim contour
9. Midface position
10. Facial nerve function
11. Lacrimal function

C. Preoperative assessment

1. See above
2. Document with photographs

III. List the alternatives to the procedure.

- A. The skin excess and rhytids can, to some degree, be treated with agents such as retinoids, or activated vitamin C
- B. Chemical peels or laser resurfacing can smooth the skin and contract it.
- C. Botulinum toxin can temporarily improve (months) wrinkling, especially at the lateral canthus
- D. Ultrasound and radiofrequency skin tightening procedures may have some benefit for skin laxity
- E. Fat prolapse and tear trough visibility can be temporarily improved (months) with deep submuscular injection of hyaluronic acid gel near the inferior orbital rim
- F. Skin wrinkling can be transiently (hours) improved with moisturizers, or albumin based topical products
- G. Treatment of metabolic or endocrine abnormalities may reduce lower eyelid swelling

IV. Describe the technique

A. Anterior approach

1. An infraciliary incision is created and a skin or a skin/muscle flap is raised
2. Fat herniation may be treated with anterior or posterior approaches. Additionally. Fat transposition/ redraping may be indicated to improve lower lid-midface contour: typically orbital fat is repositioned to the anterior part of the inferior orbital rim
3. The skin-muscle flap may be released inferomedially with lysis of the orbitomalar ligament and draped superiorly and laterally. The orbicularis is secured to the periosteum of the zygoma or lateral orbital rim and the skin undermined so that its contours are natural. Alternative approaches tighten the orbicularis laterally only with minimal central and medial dissection
4. A judicious skin excision is performed, with the removal primarily at the lateral aspect of the incision

5. Horizontal tightening procedures may be performed concomitantly

B. Posterior approach

1. The lid is everted for a posterior approach to the herniating fat pads
2. A conjunctival incision is made and continued through the lower eyelid retractors to the fat pads, which are typically subtotally excised, contoured, or repositioned
3. Following the fat reduction or transposition, an anterior approach to skin excision and horizontal eyelid tightening can be made or anterior lamellar tightening via laser resurfacing or chemical peeling may be performed

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

A. Inadequate skin removal such that the symptoms are not relieved

1. Careful intraoperative measurement with the mouth in the open position minimizes this
2. Additional skin may be removed

B. Ectropion or retraction of the eyelids

1. Often is caused by excess skin removal
2. Careful intraoperative measurement with the mouth in the open position minimizes this
3. The tissue pull is mostly horizontal when draping the skin for removal
4. Recognition and treatment of horizontal lower eyelid laxity reduces the incidence
5. Lubrication of the eyes and watchful following of the patient will support the patient while orbicularis muscle function returns in many patients
6. Secondary lateral canthopexy may help
7. Midface lifting may decrease the pull of the cheek inferiorly
8. In extreme cases, skin grafting may be required

C. Excess fat removal

1. Will result in hollowing of the orbital space immediately or in the years ahead
2. Conservative estimation of excess fat for removal is best

D. Inadequate fat removal

1. Occurs most commonly in the temporal fat pad region
2. Treatment involves further surgical excision

E. Conjunctival chemosis

1. Conjunctival edema is common after lower blepharoplasty from both anterior and posterior approaches
2. Treatment may include topical steroid or non-steroidal anti-inflammatory drops, patching, lubricating ointment, and rarely incision and drainage

F. Wound separation

1. May occur in the 1st week postoperatively
2. A local increase in eyelid inflammation accompanies the separation, and must be differentiated from infective cellulitis and allergy to the antibiotic ointment
3. Prevention involves choice of a skin suture, which will provide adequate support and proper skin removal to avoid excess wound tension

G. Dry eye symptoms

1. Lubrication of the eyes and stressing the importance of blinking help
2. Bandage contact lenses
3. Lubrication of the eyes and watchful following of the patient will support the patient while orbicularis muscle function returns
4. Preoperative discussion of this tendency in the patient with a dry eye condition will be beneficial
5. Severe dry eye patients should undergo minimal reduction of the skin, if at all

H. Allergy to the antibiotic ointment

1. This occurs about 7 days postoperatively or earlier if exposed to the medication before
2. Redness, swelling and itching are present
3. Differentiation from infective cellulitis and wound separation is necessary
4. Cessation results in improvement within 1 day
5. Occasionally topical and or oral steroids are used to speed up recovery

I. Infection is rare, as the blood supply is excellent

1. Sterile technique and treatment with antibiotic ointment are standard protocol
2. Broad spectrum oral antibiotics will rapidly improve a cellulitis appearing in the first postoperative week

J. Hematoma is rare, and can be vision threatening

1. If the patient notes a sudden increase in pain, they are instructed to call immediately
2. Careful hemostasis is obtained intraoperatively
3. All medications, herbal medicines and nutraceuticals that can prolong bleeding time should be stopped preoperatively with the approval of the primary care physician

K. A disturbance of the inferior oblique muscle may cause temporary or permanent diplopia

1. Careful identification of this muscle and avoidance during surgery will prevent this complication
2. Lockwood ligament may be useful as an anatomical "stop sign" in lower eyelid blepharoplasty.
 - a. Superior and inferior divisions of the capsulopalpebral head envelop the inferior oblique muscle and rejoin anterior to the muscle to form the Lockwood ligament

VI. Describe the follow-up care

A. Follow up is usually done at 1-2 weeks postoperatively with additional visits over successive

months

1. Follow up is complete when the patient is satisfied and when the lid and cornea are stable

VII. Describe appropriate patient instructions

A. Cold compresses are used continuously for 36 hours to minimize swelling.

1. Antibiotic ointment is used on the incision until it epithelializes by 10 days, or until absorbable sutures are melted
2. After 4-5 days, warm wet compresses are applied

B. Lubrication of the eyes is stressed

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. 20-32.
3. AAO, Focal Points: Aesthetic Surgery of the Lower Eyelid, Module #7, 1995.
4. AAO, Focal Points: Current Approaches to Cosmetic Blepharoplasty, Module #5, 1987, p. 5-7.
5. AAO, Ophthalmic Technology Assessment: Functional Indications for Upper and Lower Eyelid Blepharoplasty, *Ophthalmology* 1995;102:693-695.
6. Bedrossian EH Jr. Reconstructive lower lid blepharoplasty. *Ophthalmol Clin North Am* 2005;18:291-300,vi.

Brow ptosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. The forehead soft tissue becomes more elastic with age and descends with gravity
2. The underlying attachments of the scalp to the supraorbital ridge weaken and stretch
3. The frontalis muscle does not extend temporally to the bony temporal line, and does not extend to the tail of the brow, often allowing a lateral brow ptosis to develop with age
4. Chronic contraction of the brow depressors (corrugator muscles, orbicularis oculi muscle, depressor supercilii muscle) facilitates brow ptosis

B. Define the relevant aspects of epidemiology of the disease

1. The forehead, with the eyebrows, may congenitally be ptotic
2. Brow descent is a normal aging change
3. Paralytic forehead ptosis is associated with various lesions of the facial nerve, possibly congenital, but usually acquired

C. List the pertinent elements of the history

1. Difficulty in opening the eyes
2. Overactive frontalis muscle
3. Decreased visual field
4. Decreased vision
5. Cosmetic concerns

D. Describe the pertinent clinical features

1. Descent of the eyebrow tissues into the orbital space, especially temporally
2. Accentuated sagging of the upper eyelid anterior lamella due to a lack of support from the tissues above
3. Decreased superior visual field
4. Decreased vision in some severe cases if the tissues block the primary fixation or if the upper eyelashes are pushed below the eyelid margin
5. Deep forehead furrows due to sustained brow elevation
6. Deep glabellar furrows from chronic corrugator and procerus use, sometimes seen with widening of the upper nasal bridge due to central forehead ptosis
7. Forehead fatigue

E. Describe appropriate testing and evaluation to determine level of functional impairment

1. Visual field testing with and without eyebrow elevation

II. Define the risk factors

- A. Age**
- B. Facial nerve palsy**
- C. Trauma**
- D. Botulinum toxin injections**

III. List the differential diagnosis

- A. Redundant upper eyelid skin (dermatochalasis)**
- B. Paralytic vs. non-paralytic etiology**

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

- A. Surgical elevation by supra-eyebrow, mid-forehead, pretrichial, coronal, temporal, or endoscopic approach**
- B. Routine postoperative care is necessary. With more extensive forehead lifting procedures, swelling and numbness are common in the acute postoperative period**
- C. Warning related to infection, wound dehiscence, hemorrhage**
- D. Appropriate follow-up visits**
- E. Limited medical therapy may be useful for involutional brow ptosis**
 - 1. For mild cases, botulinum toxin to the brow depressors (corrugators and temporal orbicularis oculi muscles) may provide some temporary browlifting
 - 2. Fillers placed at the temporal brow facilitate inflation and brow support
 - 3. Must be repeated

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

- A. Under and over correction**
 - 1. Reference to earlier photographs of the patient are used as a guide
- B. Numbness of the forehead secondary to damage to the (supraorbital) or supratrochlear nerves**
 - 1. Awareness of the sensory nerves during surgery helps prevent damage; however some hypesthesia possible after brow ptosis correction procedures
 - 2. Sensory change often improves over weeks to months
- C. Loss of movement of the forehead secondary to damage to the frontal branch of the facial nerve**
 - 1. Careful awareness of the course of the frontal nerve during surgery is essential to prevent damage
- D. Scarring of the skin and hair loss can be avoided by careful incision choice and meticulous wound closure**

- E. Excessive removal of upper eyelid skin during upper blepharoplasty can result if eyebrow ptosis is not recognized**
 - 1. The eyebrow should be placed in the anatomic position when determining the amount of eyelid skin to be removed
- F. Infection**
- G. Hematoma and hemorrhage**
- H. Lagophthalmos if aggressive browlift and upper blepharoplasty are performed together**

VI. Describe disease-related complications

- A. Progressive decrease in vision and visual field**

VII. Describe appropriate patient instructions

- A. Surgical elevation may be indicated to correct functional problems**

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. 4.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p. 4-12, 281.

Involutional and gravitational changes of the face

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease- "deterioration, descent, and deflation"

1. Deterioration in skin quality with age
 - a. Loss of elastin fibers in skin allows creasing to persist
 - b. Collagen breakdown causes atrophy of skin
 - c. Actinic damage and loss of adnexal structures such as oil glands
2. Descent of skin and soft tissues with age
 - a. Gravitational effects on lax skin causes sagging of structures
 - b. SMAS and fat descends as well
3. Deflation of soft tissues and bone with age
 - a. Loss of subcutaneous fat volume
 - b. Remodeling of facial bones with overall loss of volume
 - c. Weight loss or gain may contribute

B. Define the relevant aspects of epidemiology of the disease

1. Changes increase with age
2. Females tend to show changes earlier than males
3. Changes vary among races
4. Heredity may influence patterns of aging changes

C. List the pertinent elements of the history

1. Patients generally complain of aesthetic concerns
 - a. Rhytids
 - i. Dynamic and passive
 - ii. Deep and fine
 - b. Skin quality
 - c. Excess skin
 - d. Sagging subcutaneous soft tissues
 - e. Facial contour changes
2. May express specific concerns with

- a. Skin
 - i. Quality
 - ii. Rhytids
 - iii. Pigmentary dyschromias
 - iv. Scars
 - v. Telangiectasias
- b. Frontal region (forehead and brow)
 - i. Rhytids
 - ii. Brow position or contour
 - iii. Secondary dermatochalasis from brow ptosis
- c. Periorbital region
 - i. Excess skin and anterior herniation of fat of upper and lower lids (steatoblepharon)
 - i) Upper lid skin excess may impair visual field
 - ii. Rhytids of upper, or more commonly, lower lid skin
 - iii. Laugh lines and crow's feet
- d. Midface region
 - i. Tear trough deformity
 - ii. Festoons
 - iii. Sagging cheeks/loss of high cheekbone appearance
 - iv. Midcheek creases
 - v. Skin quality and rhytids
 - vi. Nasal contour changes, including sagging nasal tip
- e. Lower face
 - i. Increased depth of nasolabial folds and creases
 - ii. Jowling/Marionette lines
 - iii. Blurring of jawline
 - iv. Weak or sagging chin
 - v. Skin quality and rhytids
- f. Perioral area
 - i. Perioral rhytids
 - ii. Collapse of upper and lower lip with loss of cupid's bow
- g. Neck

- i. Shape, including fat distribution, and loss of youthful acute angle
- ii. Sagging submandibular salivary glands
- iii. Skin quality and laxity
- iv. Platysmal bands

D. Describe the pertinent clinical features

- 1. Skin
 - a. Loss of youthful elasticity and luster including hyperkeratosis and atrophy
 - b. Rhytids
 - c. Pigmentary dyschromias and seborrheic keratosis
 - d. Actinic damage and actinic keratosis
- 2. Frontal region (forehead and brow)
 - a. Frontal and glabellar rhytids
 - b. Low or asymmetric brow position or unacceptable contour
 - c. Widening and ptosis of the upper nasal dorsum
 - d. Secondary upper eyelid dermatochalasis from brow ptosis
- 3. Periorbital region
 - a. Excess skin and fat of upper or lower lids
 - i. Upper lid skin excess may impair visual field
 - b. Rhytids of upper, or more commonly, lower lid skin
 - c. Prominent laugh lines and crow's feet
 - d. Eyelid laxity with malpositions
 - i. Retraction
 - ii. Ectropion
 - iii. Entropion
 - iv. Lid imbrication
 - v. Floppy eyelid syndrome
- 4. Midface region
 - a. Tear trough deformity
 - b. Cheek ptosis, mid-cheek crease
 - c. Nasal tip ptosis, widening of the lateral alae, rounding of the nasal dorsum
 - d. Festoons
 - e. Poor skin quality and prominent rhytids

5. Lower face
 - a. Increased depth of nasolabial folds and creases
 - b. Jowling/Marionette lines
 - c. Blurring of jawline
 - d. Weak or sagging chin
 - e. Poor skin quality and prominent rhytids
6. Perioral area
 - a. Perioral rhytids
 - b. Collapse of upper and lower lip with loss of cupid's bow
7. Neck
 - a. Obtuse mental angle
 - b. Lipodystrophy
 - c. Ptosis of the submandibular salivary glands
 - d. Anterior position of the hyoid
 - e. Poor skin quality with laxity
 - f. Platysma banding

II. Define the risk factors

- A. Age**
- B. Sun exposure**
 1. Lighter complexion (racial influence)
- C. Smoking**
- D. Females greater than males**

III. List the differential diagnosis

- A. Some features, which are aesthetically less desirable, are inherited characteristics, rather than involutional changes**
 1. Many of these are also correctable by aesthetic techniques
- B. Chronic skin disorders such as allergies, contact dermatitis, psoriasis, eczema, ichthyosis, blepharochalasis, and others**
- C. Facial palsy will lead to facial sagging**
- D. Thyroid related orbitopathy may lead to periorbital swelling, tissue redundancy of lids, eyelid fat prominence, and eyelid malpositions**

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

A. Poor skin quality

1. Regular use of sunscreen
 - a. Helps prevent skin aging and actinic (benign and neoplastic) changes
 - b. Lifetime use
 - c. Complications
 - i. Rare allergic reactions
2. Retinoic acids
 - a. Regular application provides increased skin quality and improves fine wrinkles
 - b. Enhances collagenesis with improved skin quality
 - c. Ongoing use required
 - d. Complications
 - i. Skin sensitivity
 - ii. Photosensitivity
3. Alpha and beta hydroxy/glycolic acids
 - a. Enhance skin quality through turnover
 - b. Lessens fine rhytids
 - c. Ongoing use required
 - d. Complications
 - i. Skin sensitivity
 - e. Chemical peels- Trichloroacetic acid (TCA)/phenol peel
 - f. Significant ablation of superficial skin layers
 - g. Collagenesis improves rhytids and skin quality
 - h. Effects should last for years with maintenance as above
 - i. Complications
 - i. Fairly prolonged recovery
 - ii. Scarring
 - iii. Pigmentation abnormalities
 - iv. Photosensitivity
 - v. Infection
4. Laser skin ablation/ resurfacing
 - a. Vaporizes superficial layers faster than thermal relaxation time of tissue so minimal damage

to skin left behind

- b. Improves skin quality and rhytids
 - c. Depth of ablation correlates with effectiveness, but deeper ablation requires longer recovery and increased complication rates
 - d. Erbium lower energy; CO2 higher energy
 - e. Modern lasers for skin resurfacing are pulsed and/or fractionated
 - f. Complications
 - i. Potentially prolonged recovery
 - ii. Scarring
 - iii. Pigmentation abnormalities
 - iv. Photosensitivity
 - v. Infection
5. Microdermabrasion
- a. Superficial dermabrasion leads to improved skin quality, variable effect
 - b. Ongoing treatments required
 - c. Complications
 - i. Mild skin irritation
 - ii. May worsen telangiectasias
6. Nonablative techniques
- a. Intense pulsed light (especially useful for pigmentary dyschromias), radiofrequency, ultrasound
 - b. Variably improve skin quality through collagen changes
 - c. Some effect on rhytids
 - d. Complications
 - i. Generally minimal
 - ii. Rarely reported burns

B. Facial rhytids

- 1. Sunscreens
 - a. Help prevent skin quality changes and rhytids
 - b. Lifetime of use required
 - c. Complications
 - i. Skin sensitivities
- 2. All techniques described under skin quality have some effect on, at least, fine rhytids

3. Dermabrasion
 - a. Deeper ablation of skin possible
 - b. Can be used for deep rhytids and acne scarring
 - c. Complications
 - i. Prolonged recovery for deep abrasion
 - ii. Scarring
 - iii. Pigment changes
 - iv. Infection
 - v. Photosensitivity
4. Botulinum toxin
 - a. Good for dynamic rhytids resulting from contraction of facial animation muscles
 - b. May take 6 months (2-3 injection sessions) to see optimal results
 - c. Deep rhytids may not respond to botulinum toxin alone
 - d. Need to repeat every 3-6 months
 - e. Complications
 - i. Rare
 - i) Hematoma
 - ii) Facial palsy (time limited)
 - iii) Headache
5. Fillers
 - a. Good for mild, moderate to deep rhytids
 - b. Need to be repeated, depending on agent used
 - c. Hyaluronic acid-based fillers are most popular
 - i. May take multiple sessions to fill defect
 - ii. Usually need to repeat in 6-12 months
 - iii. Low incidence of sensitivities
 - d. Other fillers available
 - i. Bovine and porcine collagen
 - ii. Human dermal allograft
 - iii. Calcium hydroxyapatite
 - iv. Poly-L-lactic Acid (also stimulates collagenesis)
 - e. Complications

- i. Sensitivity (skin test for bovine collagen)
 - ii. For longer lasting fillers, concerns about placing in wrong area
 - i) Each filler should be placed in appropriate skin layer
- 6. Surgical techniques
 - a. Surgical techniques discussed below will also help smooth rhytids as skin is supported and stretched

C. Frontal region (forehead and brow)

- 1. Brow and forehead lifts will rejuvenate the frontal region
- 2. Various techniques may be appropriate for various patients
 - a. Browpexy will give temporary support to brow at time of blepharoplasty
 - b. Direct or adjacent lift
 - i. Primarily lifts central brow
 - ii. Little effect on rhytids
 - iii. May leave visible brow scar
 - c. Midforehead lift
 - i. Will lift medial and central brow
 - ii. Little effect on temporal brow
 - iii. Can improve glabellar rhytids
 - iv. May leave prominent forehead scar
 - d. Temporal lift
 - i. Will lift temporal brow
 - ii. Can be used with facelift, midface lift, or endoscopic brow lift to lift temporal brow and remove redundancies generated from inferiorly
 - iii. Care should be taken to not change dissection planes inferior to the hairline to minimize risk to facial nerve
 - e. Coronal lift
 - i. Will lift entire brow and flatten forehead
 - ii. Release depressors of the brow to minimize glabellar furrows
 - iii. Excise scalp to support brow elevation
 - iv. Complications: coronal scar (generally best in women), hematoma, infection, anesthesia of scalp
 - f. Endoscopic brow lift
 - i. Generally same advantages of coronal lift without large incision
 - ii. Based on release of depressors of brow and repositioning of scalp in elevated

position via subperiosteal undermining

- iii. Can combine with temporal excision of scalp for better temporal brow lift
- iv. Complications: similar to coronal but no large scar
- g. Trichophytic (pretrichial) brow lift
 - i. Whereas coronal, temporal, and endoscopic lifts elevate the hairline, an incision can be placed at hairline and tissue resected from forehead to lower hairline
 - ii. Procedure otherwise similar to coronal
 - iii. Complications: similar to coronal but possible hairline scar

D. Periorbital region

- 1. Excess skin and fat of upper and lower lids corrected by blepharoplasty techniques (See Lower eyelid blepharoplasty and Upper eyelid blepharoplasty)
- 2. Eyelid malpositions including lid retraction, ectropion, entropion

E. Midface region

- 1. Midface descent can be addressed with midface lift via various approaches
- 2. Tear trough deformity can be addressed with fat grafts, implants, fat repositioning, fillers, and resuspension of the orbicularis oculi at lower blepharoplasty
- 3. Festoons can be addressed with direct excision or, when milder, with resuspension of the orbicularis at lower blepharoplasty
- 4. Midcheek creases can be addressed with fillers and fat grafts
- 5. Nasal aging changes can be addressed with rhinoplasty

F. Lower face

- 1. Nasolabial folds/creases can be improved with temporary or permanent fillers or fat grafts
- 2. Nasolabial folds can be improved by midface lifting
- 3. Jowls and marionette lines can be improved with temporary or permanent fillers, fat grafts, chin-jowl implants, and sometimes with botulinum toxin to the depressor anguli oris muscles.
- 4. Jowls, jawline blur, marionette lines, and neck laxity can be improved with lower face lift
 - a. Many approaches including cutaneous, superficial musculoaponeurotic system (SMAS), deep plane, composite

G. Perioral area

- 1. Perioral rhytids can be improved with above techniques including resurfacing, botulinum toxin, and fillers
- 2. Collapse of the upper and lower lip can be improved with injection of temporary or permanent fillers, or with fat grafting

H. Neck

- 1. Neck lipodystrophy can be treated with liposuction

2. Platysmal plication may be necessary to improve neck angle and reduce banding
3. Neck lift
 - a. Usually part of face lift and includes perithelial suspension of lateral platysma as part of SMAS tightening, as well as tightening of lax neck skin with excision of excess
 - b. Can be performed alone with perithelial incision or with midline incision
4. Limited improvement may be gained with non-invasive treatments including radiofrequency and ultrasound skin tightening

V. Describe the disease related complications

- A. Progression of the involutional and gravitational changes**
- B. If tissues in the frontal and periorbital area become sufficiently redundant, visual field may be compromised**

VI. Describe appropriate patient instructions

- A. Patients must understand that skin care is a lifelong process**
- B. Detailed peri-procedural instructions for each of the interventions above are required, but are outside the scope of this monograph**
- C. Care must be taken to assure that the various cosmetic units of the face are treated in an aesthetically homogeneous manner to maintain facial harmony**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Botulinum toxin use in oculoplastics

I. List the indications/contraindications

A. Indications

1. Functional uses
 - a. Benign essential blepharospasm (BEB)
 - b. Hemifacial spasm
 - c. Meige's syndrome (orofacial cervical dystonia)
 - d. Headache
 - e. Hypersecretion of tears
 - f. Strabismus (not further discussed in this section)
 - g. Aberrant facial nerve regeneration
2. Cosmetic uses
 - a. Glabellar furrows (Food and Drug Administration (FDA) approved indication)
 - b. Forehead creases
 - c. Lateral rhytids - "crow's feet"
 - d. Platysma bands
 - e. Hyperhidrosis
 - f. Browlifting
 - g. Lower eyelid orbicularis muscle roll
 - h. Optimizing results of cutaneous resurfacing
 - i. Perioral rejuvenation
 - i. Vertical lip lines
 - ii. Marionette lines
 - iii. Chin dimples and rhytids
3. Other
 - a. Torticollis
 - b. Spastic dysphonia
 - c. Spasticity of cerebral palsy

B. Contraindications

1. Pregnancy

2. Known sensitivity to botulinum toxin
3. Neuromuscular disorders (Myasthenia Gravis, Lambert-Eaton syndrome)
4. Albumin allergy

II. Describe the pre-procedure evaluation

A. Patient history

1. Functional uses
 - a. Blepharospasm (q.v.)
 - b. Hemifacial spasm (q.v.)
 - c. Meige syndrome (orofacial dystonia)
 - i. Blepharospasms associated with lower facial spasm
 - ii. Patients may complain of speech and swallowing difficulty
 - iii. Patients may have neck spasms
 - d. Aberrant facial nerve regeneration
 - i. Eyelid spasm with blinking or hypertonicity causes upper eyelid ptosis and/or lower eyelid reverse ptosis
 - ii. Various patterns of perioral, cheek, and platysma spasm or hypertonicity
 - iii. Gustatory lacrimation or "crocodile tears"
 - i) Found in setting of aberrant regeneration of facial nerve after facial palsy
 - ii) Can be seen with hemifacial spasm without history of palsy
 - iii) Can be seen after herpes zoster infection
 - e. Headache
 - i. Patients complain of headache, tension or migraine
 - f. Hypersecretion of tears
 - i. Patients complain of increased tearing in the setting of patent lacrimal outflow system and functioning lacrimal pump mechanism, often caused by ocular irritation
 - ii. Patients complaining of increased tearing when eating have "crocodile tears" (see above)
 - g. Torticollis
 - i. Patients complain of sternocleidomastoid and trapezius contractions
 - h. Other conditions
 - i. Spastic dysphonia
 - i) Difficulty with phonation

- ii. Spasticity associated with cerebral palsy
- 2. Cosmetic uses
 - a. Patients complain of glabellar furrows and medial forehead ptosis
 - i. This is a common presentation for injection, and the only cosmetic use approved by the FDA
 - b. Forehead wrinkles
 - c. Lateral canthal rhytids
 - d. Perioral rhytids
 - e. Neck bands

B. Clinical examination

- 1. Benign essential blepharospasm
 - a. Brow spasm and ptosis
 - b. Associated levator disinsertion
 - c. 70% develop lower facial dystonias
 - d. Typically have normal facial strength
- 2. Hemifacial spasm
 - a. Intermittent synchronous gross contractures of the unilateral face, including dimpling of chin
 - b. Often associated with ipsilateral facial nerve-served muscle weakness
 - c. Rarely bilateral, but spasms are not synchronous
 - d. If present with other cranial neuropathies (e.g., cranial nerve (CN) VIII) or significant progression, this may indicate an intracranial mass
 - e. Etiology - may be due to facial nerve compression by aberrant vascular loop, rarely neoplasm
 - f. Diagnostic studies - neurology/neuro-ophthalmology consult, magnetic resonance imaging (MRI)/magnetic resonance angiography (MRA)
- 3. Meige syndrome
 - a. Findings similar to BEB but spasms extend into face, perioral area, and neck
 - b. Puckering of lips
 - c. Difficulty with speech
 - d. May have torticollis-like features
- 4. Headaches
 - a. Little on examination except identification of possible trigger points/inciting muscle groups
- 5. Hyperlacrimation
 - a. Increased tear lake

- b. Tearing with salivation (crocodile tears)
 - c. Patent lacrimal outflow system
 - d. Normal lid positions with functioning lacrimal pump
 - e. Normal Jones 1 and 2 tests
- 6. Cosmetic
 - a. Ask the patient to demonstrate the areas of concern
 - b. Rhytids appropriate for treatment are related to contraction of treatable muscles

C. Preoperative assessment

- 1. Spasms
 - a. Location of spasms will determine the site of injections given inciting muscles
 - b. Severity and time to recurrence of spasms may affect dosage
- 2. Headaches
 - a. Assess nature of headaches and neurologic exam/imaging to rule out structural cause
- 3. Hypersecretion
 - a. Lacrimal outflow patency
 - b. Eyelid position and function of lacrimal pump
 - c. Jones testing to demonstrate hyperhidrosis as possible cause of increased tearing
- 4. Cosmetic treatment of rhytids
 - a. Determination of which muscle groups are causing the rhytids
 - b. The number of treatment areas and the depth of the wrinkles
 - c. Some rhytids are too fine to respond and some are too deep to show an initial effect after muscle weakening or paralysis
 - i. These patients may be treated by other modalities
- 5. In functional periocular cases, patients must be assessed for dry eye syndrome and prophylactically given artificial tears, as increased lagophthalmos may develop post injection

III. List the alternatives to this procedure

A. BEB

- 1. Trial of artificial tears and ocular surface evaluation, typically prior to botulinum injections
- 2. Oral neurotropic agents: some insurance companies require a trial of these agents prior to authorizing ongoing use of botulinum
- 3. Protractor myectomy
 - a. Reserved for patients with an inadequate response to botulinum toxin

- b. Many patients find increased effect of botulinum after myectomy
 - c. Initial surgery may be limited to upper eyelid protractor myectomy, sparing the orbicularis of the lower lids
- 4. Selective facial neurectomy
 - a. Rarely used except in severe cases unresponsive to other treatments
- 5. Oral neurotropic agents

B. Hemifacial spasm

- 1. Neurotropic agents, as in blepharospasm
- 2. If there is MRI evidence of neuro-vascular compression in posterior fossa, patient may choose surgical decompression (Jannetta procedure)

C. Meige syndrome

- 1. Treatment approaches are similar to BEB above but myectomy is not used for lower facial spasms
- 2. Heavier reliance on neurotropic agents in these patients

D. Headaches

- 1. Analgesics
- 2. For migraine, ergots and avoidance of triggers

E. Hypersecretion of tears

- 1. In severe cases, surgical removal of lacrimal gland, punctoplasty or dacryocystorhinostomy (DCR) to increase outflow

F. Cosmetic

- 1. Glabellar and frontal furrows
 - a. Brow and forehead lift
 - b. Corrugator resection
 - c. Tissue fillers
 - d. Skin resurfacing, chemical peel dermabrasion, or non-ablative techniques
- 2. Lateral rhytids (Crows feet)
 - a. Temporal browlift
 - b. Tissue fillers and fat grafting
 - c. Skin resurfacing, chemical peel dermabrasion, or non-ablative techniques
- 3. Perioral
 - a. Fillers and fat grafting
 - b. Platysmal bands
 - c. Platysma plication
 - d. Facial and cervical rhytidectomy (face and neck lift)

4. Hyperhidrosis
 - a. Oral pharmacologic management of sweat glands

IV. Describe the technique

- A. Botulinum toxin is available in several distinct subtypes that are prepared by mixing with concentration-dependent amount of saline, following manufacturer's guidelines
- B. For onabotulinum toxin A, and incobotulinum toxin A, injection aliquots of 1.25 to 5.0 units per injection site are typical
- C. For abobotulinum toxin A, injection aliquots of 2 to 15 units per injection site are typical
- D. Injection should be placed so as to maximize effect on target muscles and minimize effect on other structures

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

A. Periocular injections

1. Primarily involvement of levator (ptosis), inferior oblique muscles (diplopia)
 - a. Injection should be remote from these muscles
 - b. The unwanted paresis will diminish and resolve as the medication effects subside
 - c. Apraclonidine 0.5% eye drops may be helpful
2. Ectropion, lagophthalmos
 - a. Artificial lubrication of the eyes is recommended to prevent sicca syndrome
 - b. Resolves over time

B. Facial injections

1. Can create facial weakness or paralysis with drooping of corner of mouth, drooling, difficulty with speech
2. Ecchymosis

VI. Describe the follow-up care

- A. Patient may benefit from artificial tears after periocular injections

VII. Describe appropriate patient instructions

- A. The patient should be instructed not to compress the just-injected areas to prevent movement of the botulinum
- B. Call for reinjection when spasms or movement (cosmetic) just begins to return.
- C. Patients should be aware of the temporary nature of the treatment and retreatment will be required in 3 to 4 months typically

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993.
3. AAO, Ophthalmology Monographs 8. Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994.
4. AAO, Focal Points: Essential Blepharospasm and Related Diseases, Module #5, 2000.
5. AAO, Focal Points: Lasers in Periorbital Surgery, Module #7, 2000.
6. Physician's Desk Reference®, 58th Ed. 2004, p. 552.
7. Sadick N, Sorhaindo L. The cosmetic use of botulinum toxin type B. *Int Ophthalmol Clin* 2005;45:153-61.
8. Carruthers J, Carruthers A. Adjunctive botulinum toxin type A: fillers and light-based therapies. *Int Ophthalmol Clin* 2005;45:143-51.
9. Foster JA, Proffer PL, Proffer LH, et al. Modifying brow position with botulinum toxin. *Int Ophthalmol Clin* 2005;45:123-31.
10. Khan JA. Cosmetic botulinum neurotoxin application. *Int Ophthalmol Clin* 2005;45:107-22.
11. Mannor GE. Practical aspects of cosmetic botulinum toxin. *Int Ophthalmol Clin* 2005;45:99-106.
12. Kaltreider SA, Kennedy RH, Woog JJ, and the Ophthalmic Technology Assessment Committee Oculoplastics Panel. Cosmetic oculofacial applications of botulinum toxin: a report by the American Academy of Ophthalmology. *Ophthalmol* 2005;112:1159-67.
13. Costa J, Espirito-Santo C, Borges A, et al. Botulinum toxin type A therapy for blepharospasm. *Cochrane Database Syst Rev* 2005:CD004900.
14. Costa J, Espirito-Santo C, Borges A, et al. Botulinum toxin type A therapy for hemifacial spasm. *Cochrane Database Syst Rev* 2005:CD004899.
15. AAO, Focal Points: Botulinum Toxin in Ophthalmology, Module #3, 2007.

Main lacrimal gland and lacrimal ductules

I. Describe relevant aspects of anatomy

A. Main lacrimal gland

1. Exocrine gland
2. Located in the superolateral quadrant of the orbit in the lacrimal gland fossa of the frontal bone
3. Gland lined by outer connective tissue pseudocapsule
4. Indentation of the lacrimal gland by the lateral horn of the levator muscle divides the gland into orbital and palpebral lobes
5. Whitnall ligament inserts into the indentation of the lacrimal gland and projects onto the lateral orbital tubercle
6. Septa divide lobules within lacrimal gland
7. Contraction around acinar unit- (lined with columnar epithelial cells) drains aqueous fluid into ductules
8. Innervation
 - a. Afferent innervation, lacrimal nerve from the trigeminal ophthalmic branch(V1) of the trigeminal nerve(CNV)
 - b. Efferent
 - i. Parasympathetic fibers from the cranial nerve (CN) VII in the greater superficial petrosal nerve send fibers to the sphenopalatine ganglion and then by way of the zygomatic nerve
 - ii. Sympathetic pathways are not clearly defined
9. Accessory lacrimal glands
 - a. Krauss- located in superior conjunctival fornix
 - b. Wolfring- located in subconjunctival tissue near superior border of the upper lid tarsus and inferior edge of lower lid tarsus
10. Arterial blood supply
 - a. Served by a single artery - lacrimal artery
 - i. This unique anatomical feature permits selective cannulation of the lacrimal artery for intra-arterial delivery of chemotherapy in treating adenoid cystic carcinoma of the lacrimal gland
 - ii. The lacrimal artery, a branch of the ophthalmic artery and therefore, of the internal carotid system, anastomose in the lids with the anterior deep temporal artery, a branch of the external carotid system.
11. Fascial support

- a. Fibrous septae connecting lacrimal gland to lacrimal gland fossa
 - i. Edge of the levator aponeurosis and its lateral horn
 - ii. Superior transverse ligament = Whitnall's ligament which is condensed fascia of levator muscle which extends nasally from fascia in trochlear area and laterally to capsule of lacrimal gland
 - iii. Fascial band inferior to gland

B. Lacrimal ductules

- 1. Two to six ductules extend from the orbital lobe of the lacrimal gland, into the palpebral lobe, and join the palpebral lobe ducts
- 2. Eight to twelve major palpebral lobe ductules
 - a. Ducts - pseudostratified squamous epithelium
- 3. Ductules located 4-5 mm above the superior margin of the upper tarsus
- 4. Ductules drain into the lateral superior fornix cul-de-sac

II. Describe clinical correlations

- A. Removal or damage to the palpebral portion of the gland can severely reduce secretion from the lacrimal gland**
- B. Ductal cysts can simulate a lacrimal gland mass**
 - 1. Can be treated by marsupialization

III. Function of the lacrimal gland - secretion of aqueous layer of tear film

- A. Tear film comprised of**
 - 1. Mucin layer- secreted by the goblet cells of the conjunctiva
 - 2. Aqueous middle layer - secreted by the lacrimal gland and accessory lacrimal glands
 - 3. Oily outer layer- secreted by the Meibomian glands

IV. Tumors of the lacrimal gland

- A. Epithelial**
 - 1. 50% are benign mixed epithelia tumors (pleomorphic adenomas)
 - 2. 50% are carcinomas (of the carcinomas, 50% are adenoid cystic carcinoma, the rest are malignant mixed tumor, primary adenocarcinoma, mucoepidermoid carcinoma, or squamous carcinoma)
- B. Non-epithelial**
 - 1. Lymphoid proliferations

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 4: Ophthalmic Pathology and Intraocular Tumors; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Superior and inferior puncta and canaliculi

I. Describe relevant aspects of anatomy

A. Puncta

1. Opening of nasolacrimal excretory system-approximately 1.5 mm diameter
2. Located at medial upper and lower eyelid margin
3. Puncta are slightly inverted against the globe
4. Puncta are located in the tear lake
5. Puncta opens into an ampulla which is 2 mm long and perpendicular to the margin of the eyelid

B. Canaliculi

1. 8 to 12 mm in length
2. 90% of canaliculi combine to form a common canaliculus at the lacrimal sac
3. Traditionally, the valve of Rosenmüller was thought to prevent reflux from the sac to the canaliculi (but the acute angle of entrance of the canaliculi into the sac may lead to a valve-like effect and prevent reflux)
4. Lacrimal sac expansion may also kink canaliculi, leading to functional blockage
5. Epithelium- layers of non keratinous stratified squamous epithelium

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

Medial canthal tendon

I. Describe relevant aspects of anatomy

- A. Medial Canthal Tendon (MCT) is comprised of an elastic lateral portion which supports the lacrimal canaliculi, and a medial portion, which splits into anterior, superior, and posterior limbs**
 - 1. Anterior limb MCT
 - a. Inserts on the maxillary bone
 - b. Pretarsal and preseptal orbicularis oculi muscle fibers insert onto anterior limb of MCT
 - 2. Superior limb MCT
 - a. Extends to the lacrimal sac apex and covers the anterosuperior portion of the lacrimal sac
 - 3. Posterior limb MCT
 - a. Inserts on the posterior lacrimal crest of the lacrimal bone
 - b. Posterior attachments of the MCT and orbicularis muscle result in the normal posterior pull on each medial eyelid which maintains the eyelids against the globe and the puncta in the lacrimal
 - 4. The lateral portion of the MCT supports the lacrimal canaliculi
- B. The MCT is closely associated with a complicated, interdigitating network of orbicularis oculi insertions**
- C. An anatomic plane is present in the multiple fascial extensions from the medial rectus muscle sheath to the MCT, orbicularis oculi muscle, plica semilunaris, and caruncle**
 - 1. Surgical dissection in this plane avoids the medial rectus muscle posteriorly and laterally and the lacrimal drainage anteriorly and medially
 - 2. This plane is utilized in the transcaruncular approach to the medial orbit
- D. The angular vessels(which anastomose the vascular system of the face and orbit) are located 7 to 8 mm anterior to the medial canthal tendon**
- E. Anatomy of medical canthal tendon may play a crucial role in lacrimal pumping**

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. 11-15.
- 3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p. 85-86, 182-185.
- 4. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p. 61-63, 236-238.
- 5. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p. 127-128.

Nasolacrimal duct (NLD) and Valve of Hasner

I. Describe relevant aspects of anatomy

A. Nasolacrimal duct

1. Interosseous portion NLD - 12 mm in length
2. The NLD traverses in an inferior lateral direction from the lacrimal sac into the inferior meatus
3. NLD travels the bony nasolacrimal canal and terminates below the inferior turbinate 2.5 cm posterior to the nares
4. The valve of Hasner is located at the distal edge of the nasolacrimal duct
5. Tear drainage
 - a. 10% evaporation in children
 - b. 20% evaporation in adults
 - c. Remainder through nasolacrimal pump mechanism

B. Valve of Hasner

1. Mucosal flap over the distal ostium of the nasolacrimal duct below the inferior turbinate of the nose
 - a. During normal blink, the lacrimal sac is compressed, emptying its contents
 - b. Persistent membrane at valve of Hasner is a common site of congenital nasolacrimal duct obstruction (up to 50% at birth)

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Fundamentals and Principles of Ophthalmology; Section 5: Neuro-Ophthalmology; Section 6: Pediatric Ophthalmology and Strabismus; Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgical Anatomy of the Ocular Adnexa, 1996, p. 99-100.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p. 98-99.
4. AAO, Focal Points: Evaluation of the Tearing Adult Patient, Module #8, 2002.
5. AAO, Focal Points: Management of Eyelid Trauma, Module #10, 1996.
6. AAO, Focal Points: Evaluation and Surgery of the Lacrimal Drainage System in Adults, Module #12, 1995.

Congenital nasolacrimal obstruction

I. Embryology/development

A. Lacrimal excretory system

1. Core of ectodermal tissue between the medial canthal tendon and nasal cavity
2. Cavitation occurs with the duct canalizing last
 - a. Usually at the time of birth
3. Obstruction at the valve of Hasner is found in 50% of neonates at birth
4. Patency develops within the first few months of life

II. Physiology

- A. 50% of children at birth have an obstructed system but only 2% to 6% of full-term infants present with epiphora at age three to four weeks
- B. One-third are bilateral
- C. 90% resolve by the first year of life

III. Pathophysiology

- A. Obstruction at the valve of Hasner is most common cause
- B. Membranous occlusion of the puncta
- C. Absence/stenosis or duplication of the puncta
- D. Lacrimal - cutaneous fistula from canaliculus or sac drain tears to the skin
- E. Facial clefting abnormalities may involve the nasolacrimal system
- F. Dilated sac at birth may indicate dacryocystocele
 1. Nasolacrimal duct obstruction (NLD) with valve of Rosenmüller preventing egress of fluid and obstruction of the distal system
 2. Resultant trapping of amniotic fluid or mucus in the system
 3. May have nasal mass resulting in respiratory distress for neonates

IV. Evaluation

A. History

1. Tearing since birth or shortly thereafter
2. Constant tearing with minimal mucopurulence suggests punctal or canalicular agenesis

3. Constant tearing with frequent mucous discharge suggests nasolacrimal duct obstruction
4. Intermittent tearing with mucous discharge suggests intermittent obstruction of the nasolacrimal duct, most likely due to swollen inferior turbinate mucosa associated with upper respiratory tract infection (URI) or allergy

B. Are the puncta present and patent?

C. Ensure that there is no reason for reflex tearing secondary to ocular surface irritation

1. Infectious conjunctivitis
2. Epiblepharon and trichiasis
3. Congenital glaucoma - cloudy cornea, increased corneal diameter

D. Examine the medial canthal tendon for distended sac or congenital defect such as encephalocele

E. Digital compression of the sac may elicit mucoid reflux, indicating nasolacrimal duct obstruction

F. Presentation as acute dacryocystitis

V. Differential diagnosis of medial canthal mass

A. Myelomeningocele or encephalocele if there is swelling above the medial canthal tendon

B. Hemangioma

C. Nasal dermoid cyst

D. Malignancy

VI. Treatment

A. Probing

B. General anesthesia

C. Oxymetazoline, not cocaine due to potential cardiac toxicity in children

D. Lateral traction of the eyelid to straighten canaliculi

E. Avoid kinking the system with passage of the probe

F. Hard stop

1. Once hard stop is encountered, then rotate the probe vertically and direct posterolaterally

G. Direct probe to the floor of the nose

H. Don't force

I. Distance

1. 20 mm from the punctum to the floor of the nose

J. Confirm patency with fluorescein irrigation, metal on metal contact, or direct observation

VII. Management

A. Management of congenital nasolacrimal obstruction

1. Most resolve by age four to six months
2. Treatment during this time should include lacrimal sac compression massage and topical antibiotic as necessary during the first six months
3. Systemic antibiotics are indicated if dacryocystitis with orbital cellulitis/preseptal cellulitis develop
4. Drainage of abscess may be necessary
5. For uncomplicated cases of nasolacrimal obstruction, most surgeons probe before one year of age (generally between 9-12 months)
6. General anesthesia is preferred in most cases. In young infants, probing may be performed under restraint
7. Failure of probing should be followed by either silicone intubation or balloon dilation. Some surgeons will repeat probing
8. Failure after silicone intubation may necessitate dacryocystorhinostomy (DCR)

B. Management of dacryocystocele

1. Topical antibiotics and massage for two weeks
2. If persists beyond two weeks, probe and accomplish intranasal examination to look for cysts
3. Urgent intervention if breathing difficulties (from intranasal cyst) arise or infection develops

C. Prognosis for congenital nasolacrimal duct obstruction

1. Treatment success rate is 90%
2. Consider inferior turbinate infraction where impaction is present
3. Silicone intubation
 - a. Recurrent epiphora following initial probing
 - b. Older children with stenosis/scarring at the time of initial probe
4. Balloon dacryoplasty
 - a. Possibly of benefit in difficult cases
 - b. Possibly of benefit in failed cases
5. Dacryocystorhinostomy (DCR)
 - a. Consider DCR for persistent epiphora following silicone intubation and evidence of NLD obstruction
 - b. Children with developmental abnormalities
 - c. CDCR with Jones tube generally deferred in young children

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus;

Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 1, 1993, p. 102-103.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p. 265-268, 286.

Acquired nasolacrimal duct obstruction in adults

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Involutional stenosis (idiopathic)
 - a. Most common
 - b. Women are two times more frequently affected than men
2. Trauma
 - a. Particularly midface and nasal orbital ethmoid fractures
3. Drugs
4. Inflammatory disease
5. Granulomatous disease
 - a. Sarcoidosis
 - b. Wegener granulomatosis
6. Tumors
 - a. Lymphoma
 - b. Squamous cell carcinoma (CA)
 - c. Inverting papilloma
 - d. Sinonasal tumors
7. Radiation therapy for malignancies
8. Nasal pathology
 - a. Cocaine inhalation abuse
 - b. Tumor
 - c. Prior nasal/sinus surgery, including endoscopic
 - d. Chronic sinusitis/rhinitis

B. Describe appropriate testing and evaluation for confirmation of the diagnosis

1. Ophthalmic exam
 - a. Increased tear lake
 - b. Evidence of dacryocystitis with conjunctivitis or purulence
 - i. Red eye/chronic conjunctivitis

- ii. May palpate enlarged lacrimal sac or abscess cavity
 - iii. Expression of pus with medial canthal pressure
- 2. Physiologic tests
 - a. Dye disappearance test
 - i. Procedure
 - i) Instill fluorescein
 - ii) Observe under cobalt blue light for five minutes
 - iii) Look for asymmetry of dye clearance
 - ii. Test does not determine site of lacrimal drainage abnormality
 - iii. Interpretation
 - i) Significant delay in dye clearance time suggests lacrimal obstruction
 - ii) Asymmetric dye disappearance time suggests unilateral obstruction to outflow
 - b. Jones I
 - c. Jones II
 - d. Irrigation
- 3. Intranasal examination
- 4. Imaging studies
- 5. Results of these tests may suggest a functional partial and/or anatomic (complete) block

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

A. Define surgical therapy options

- 1. Probing/tube placement and balloon dacryoplasty sometimes work
- 2. Most commonly, dacryocystorhinostomy (DCR) is necessary
- 3. Consider malignancy if blood tinged tears are present
- 4. Biopsy of lacrimal sac and intranasal lesion if malignancy is suspected
 - a. DCR may be contraindicated in presence of tumor

Additional Resources

- 1. AAO, Basic and Clinical Sciences 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.

Diagnostic probing (palpation) of the upper system

I. List the indications/contraindications

A. Indications

1. Used in the evaluation of epiphora
 - a. Possible canalicular stenosis/obstruction
 - b. Possible common canalicular stenosis/obstruction
2. Used in conjunction with irrigation to evaluate the lacrimal system

B. Possible contraindication

1. Acute infection

II. Describe the pre-procedure evaluation

A. History

1. Epiphora, dacryocystitis
2. History of
 - a. Viral keratoconjunctivitis
 - b. Glaucoma medication
 - c. Anti-viral topical medication
 - d. Trauma
 - e. Ocular surface disease
 - f. Previous punctal/intracanalicular plug insertion
 - g. Chemotherapy - Docetaxel (Taxotere®), 5-Fluorouracil
 - h. Mega dose I131 therapy (>150 mCi)

B. Exam

1. Punctal stenosis
2. Punctal dilation or discharge
3. Canalicular erythema
4. Elevated tear meniscus (increased tear lake)
5. Delayed fluorescein dye clearance

III. List the alternatives to this procedure

- A. Lacrimal irrigation without probing, although probing of upper lacrimal system may be done concurrently with a cannula used for irrigation**
- B. Physiologic patency of the lacrimal excretory system should include dye disappearance test, Jones I and II tests, and a nasal speculum exam**
- C. Dacryocystography**
- D. Dacryoscintigraphy**

IV. Describe the instrumentation and technique

A. Anesthesia

- 1. Adults
 - a. Topical anesthesia
- 2. Infants
 - a. Usually general anesthesia

B. Technique

- 1. Punctal dilation
- 2. Lateral traction on eyelid
- 3. Gentle probe passage medially
- 4. Demarcation of length of probe which may be inserted prior to resistance ("soft stop") or successful passage of probe into lacrimal sac with contact with bone ('hard stop")

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

- A. False passage creation prevented by gentle probing technique**
 - 1. Generally, no treatment required

VI. Describe the considerations in interpretation of this diagnostic procedure

- A. Soft stop identifies the presence and position of obstruction**
- B. Hard stop demonstrates a patent canaliculus into the sac**
- C. A stenotic portion of the canaliculus can be identified**

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. 3, 1995, p. 266.

Nasal endoscopy

I. List the indications

- A. Used to rule out intranasal causes of lacrimal obstruction
- B. Used to rule out nasal septum or turbinate abnormalities which might require correction at time of lacrimal surgery
- C. Used to monitor status of dacryocystorhinostomy (DCR) ostium or Jones tube after lacrimal surgery
- D. Facilitate removal of silicone tubing
- E. Other potential nasal pathology

II. Describe the pre-procedure evaluation

- A. History suggestive of lacrimal outflow obstruction and/or intranasal pathology
- B. Clinical examination of lacrimal outflow system including pertinent, external/slit-lamp biomicroscope examination, fluorescein dye disappearance testing, lacrimal irrigation +/- upper system probing

III. List the alternatives to this procedure

- A. Nasal speculum examination
- B. Ear, nose, and throat (ENT) consultation
- C. Sinus computed tomography (CT) studies (complementary)

IV. Describe the instrumentation and technique

- A. +/- topical vasoconstrictor/anesthetic spray
- B. Use of zero or "thirty" degree, 2.7-4.0 mm endoscope to evaluate nasal mucosal health, and nasal anatomy including inferior/middle turbinates/meatus, and the septum

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

- A. Inability to visualize above structures due to nasal septal deviation, mucosal edema, etc.
- B. Infrequent abrasion of the nasal mucosa
- C. Some discomfort

VI. Describe the considerations in interpretation of this diagnostic procedure

- A. Interpretation of nasal endoscopy requires knowledge of the intranasal anatomy

1. Inferior meatus
 - a. The nasolacrimal duct drains here via the Valve of Hasner
2. Inferior turbinate
3. Middle meatus
 - a. Receives drainage from the frontal sinuses, anterior and middle ethmoids, and maxillary sinus (via the infundibulum/osteomeatal complex)
4. Middle turbinate
 - a. The anterior root of the middle turbinate is medial to the lacrimal sac
5. Uncinate process
 - a. Ridge like elevation of lateral wall in middle meatus
6. Superior meatus
 - a. Receives drainage from the posterior ethmoid air cells
7. Superior turbinate
8. Sphenoethmoid recess
 - a. Receives drainage from the sphenoid sinus
9. Nasal septum

B. Visualization of intranasal abnormalities (e.g., significant septal deviation, mucosal hyperemia/discharge/ulceration, masses, synechiae) may prompt ENT consultation for further evaluation and medical/surgical treatment of sinonasal abnormalities as indicated (e.g., possible septoplasty at time of DCR, sinus carcinoma, Wegener granulomatosis)

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p. 263-265.
3. AAO, Focal Points: Evaluation and Surgery of the Lacrimal Drainage System in Adults, Module #12, 1995.

Evaluation of reflex tearing

I. List the indications/contraindications

- A. Reflex tearing is considered when tearing is present but the outflow system appears patent
 - 1. Possible decreased basal tear production
 - 2. Chronic ocular irritation
 - 3. Possible reflexive secondary hypersecretion

II. Describe the pre-procedure evaluation

- A. History with attention to circumstances associated with tearing
- B. External and slit-lamp biomicroscopic examination

III. List the alternatives to these procedures

- A. Procedure performed in conjunction with history, external and slit-lamp biomicroscopic examinations
- B. Dye disappearance test, lacrimal irrigation, canalicular probing, intranasal examination often performed concurrently

IV. Describe the instrumentation and technique

- A. Basic secretion test
 - 1. Evaluates tear production with minimal ocular surface stimulation
 - 2. Topical anesthetic instilled in eye
 - 3. Conjunctival cul-de-sac dried with tissue paper
 - 4. Filter paper strip placed in inferotemporal fornix for 5 minutes
 - 5. Normal tear production > 15 mm
 - 6. Less than 10 mm: Patient's symptoms likely to respond to artificial tear drops if applied on a regular basis (e.g. QID)
- B. Schirmer I
 - 1. Evaluates tear production but variable response due to irritation by filter paper
 - 2. No topical anesthetic
 - 3. Dry conjunctival cul-de-sac with tissue paper
 - 4. Filter paper strip placed in inferotemporal fornix for 5 minutes

5. Normal tear production greater than 15 mm
6. Patients with less than 15 mm may have positive response to regular use of artificial tear drops

C. Schirmer II

1. Evaluates potential for any tear production in patients with severe dry eyes (e.g. Sjögren syndrome)
2. This test is only done in patients with a significantly decreased Schirmer 1 test
3. No topical anesthetic
4. Dry conjunctival cul-de-sac with tissue paper
5. Filter paper strip placed in inferotemporal fornix for 5 minutes
6. A cotton applicator is used to irritate the inside of the nose and stimulate tearing
7. A moistened filter paper represents some ability of the main lacrimal gland to respond to a noxious stimulus

D. Tear breakup time

1. This test is used to qualitatively assess the quantity and quality of tears produced
2. Fluorescein is instilled in the affected eye
3. At the slit lamp, the tear film is assessed
4. Patient is asked to blink and then stare
5. Initial lack of corneal tear coating with blink may indicate a general deficiency of tear production
6. If initially smooth coating of cornea occurs, but breaks up after 15 seconds, this is likely normal
7. If tear film breaks up in less than 10 seconds, this may represent a deficiency of mucin or lipid components of tears

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

A. Ocular irritation

VI. Describe the considerations in interpretation of this diagnostic procedure

- A. The interpretation of results suggested above is generalized**
- B. A trial of artificial tear drops is clearly indicated in these symptomatic patients**
- C. Patients with basic secretion test results less than 5 mm should be considered for systemic workup for conditions such as Sjögren syndrome**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.

2. AAO, Focal Points: Evaluation of the Tearing Adult Patient, Module #8, 2002, p.3.
3. AAO, Focal Points: Evaluation and Surgery of the Lacrimal Drainage System, Module #12, 1995, p. 2.

Punctal ectropion

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Commonly associated with lower lid ectropion
2. Laxity of medial canthal tendon, horizontal lower eyelid laxity, segmental lower eyelid retractor disinsertion
3. Keratinization of eyelid margin
 - a. Actinic skin change
 - b. Chronic blepharitis
4. Anterior lamellar shortening
 - a. Chronic inflammation
 - b. Cicatrizing disease or post trauma
 - c. Post tumor excision

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

1. Elderly population with generalized involutional skin processes
2. Actinic changes of the face

C. Pertinent elements of the history

1. Epiphora
2. Ocular irritation

D. Describe the clinical features

1. Punctal eversion with possible stenosis
2. Lower lid laxity
3. Lower lid cicatricial changes

II. Define the risk factors

A. Age

B. Sun exposure

III. List the differential diagnosis

A. Generalized ectropion of the lid

B. Lid retraction

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

A. Describe the natural history, outcome and prognosis

1. Epiphora
2. Progressive cicatrization of the conjunctiva
3. Progressive ocular irritation

B. Describe the medical therapy options

1. Massage steroid (antibiotic) ointment into the affected lid daily for 6 weeks
2. General management of skin conditions such as rosacea and actinic changes
3. Avoid sun exposure
4. Avoid potential causes of contact irritation

C. Describe the surgical options

1. Resect conjunctiva and lower lid retractors below the punctum with re-approximation of the margins of the wound
2. Probe may be placed in the canalicular system to protect it
3. May be combined with horizontal lid tightening
4. May need skin grafting to lower lid if anterior lamella shortened
5. Z-plasty for scar revision
6. May benefit from bicanalicular intubation to help reapproximate the puncta to the tear lake

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

A. Entropion from posterior lamella resection

1. Caution to not shorten inferior fornix

B. Anesthetic complications

C. Aesthetic compromise with skin graft to lower eyelid

1. Careful technique for grafting

VI. Describe disease related complications

A. Persistent epiphora

B. Ocular exposure

VII. Describe appropriate patient instructions

A. Post operative followup

B. Adequate management of ongoing skin pathology that contributed to the problem originally

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. 99.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 2, 1994, p. 59.

Punctal stenosis and atresia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disorder

1. Punctal stenosis
 - a. Chronic conjunctival exposure and keratinization (e.g. ectropion)
 - b. Dry eyes with low tear outflow
 - c. Chronic blepharitis
 - d. Recurrent or severe conjunctivitis
 - e. Tumor
2. Congenital punctal atresia or agenesis
 - a. May be inherited
 - b. Congenital lack of one punctum may be very common due to low incidence of symptoms

B. Describe relevant aspects of the epidemiology

1. Punctal stenosis common in elderly and female population with dry eyes and ongoing lid/conjunctival irritations such as blepharitis

C. List the pertinent elements of the history

1. Patients may not complain of epiphora due to underlying dry eyes or compensation to congenital condition
 - a. Patients with congenital absence of a punctum may be extremely common but not recognized due to absence of symptoms
2. Patients with acquired stenosis may complain of ocular irritation and epiphora

D. Describe pertinent clinical features

1. Punctal stenosis-very small punctal opening
2. Punctal atresia
 - a. May be acquired or congenital
 - b. There may be a fine membrane over top of punctal mound
3. Punctal agenesis - congenital
 - a. There may be no evidence of punctal mound or opening

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Slit lamp evaluation of punctal region
2. Assessment of tear production

II. Define the risk factors

- A. Patients with dry eyes and ocular surface disease**
- B. Family history of punctal agenesis**

III. List the differential diagnosis

- A. Prior punctal closure**
- B. Chronic marginal inflammation causing punctal scarring**

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

- A. Describe the natural history, outcome and prognosis**
 - 1. Many patients are asymptomatic with congenital forms and in patients with dry eyes
- B. Describe medical therapy options**
 - 1. Patients with dry eyes may benefit from use of artificial tear drops and ointments
 - 2. Management of underlying lid and ocular irritant conditions may improve stenosis and symptoms
- C. Describe surgical options**
 - 1. Membrane may be punctured with a needle, sharp probe or dilator
 - 2. For absent punctum
 - a. Vertical cut down through lid margin, looking for canalicular lumen
 - b. Silicone intubation after above
 - c. If no system found, conjunctivodacryocystorhinostomy (CDCR) when child or adult is able to undergo this
 - d. Some will defer CDCR until preteen or teenage years

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

- A. Damage to opposite patent punctacanalicular system**
 - 1. Minimize manipulation of patent part of the system
- B. Epiphora by opening system in patients with very dry eyes**
 - 1. Evaluate for dry eyes preoperatively

VI. Describe disease related complications

- A. Epiphora**
- B. Conjunctivitis**

VII. Describe appropriate patient instructions

A. Follow post operative instructions

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Evaluation of the Tearing Adult Patient, Module #8, 2002.
3. AAO, Focal Points: Evaluation and Surgery of the Lacrimal Drainage System in Adults, Module #12, 1995.

Canalicular obstruction

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Infection
 - a. Viral
 - i. Herpes virus
 - ii. Adenovirus
 - b. Bacterial
2. Medications
 - a. 5- fluorouracil and other chemotherapy drugs
 - b. Idoxuridine (Herplex®)
 - c. Physostigmine (Eserine®)
 - d. Docetaxel (Taxotere®)
 - i. Effect appears to be dose dependent
3. Trauma
 - a. Accidental
 - b. Iatrogenic
 - i. Punctal or canalicular plugs
 - ii. Surgery
 - iii. Cautery
4. Congenital
5. Inflammatory
 - a. Pemphigoid
 - b. Stevens-Johnson syndrome
6. Tumors

B. Describe appropriate testing and evaluation for confirmation of the diagnosis

1. Probe palpates a soft-stop in the canaliculus
2. Common canalicular obstruction
 - a. 8 mm soft-stop
 - b. Irrigant refluxes through opposite punctum but not into the nasolacrimal sac

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

A. Constriction - silicone intubation after dilation

B. Limited obstruction

1. Silicone intubation after dilation may be useful in some cases
2. 2. Resect obstruction and repair ends of anastomosis over a stent

C. Total obstruction - conjunctivodacryocystorhinostomy (CDCR) or suture of canaliculus to the lacrimal sac

1. CDCR typically involves creation of a fistula between the "medial conjunctival fornix" in the region of the caruncle and the lateral wall of the nose in the region of the anterior tip of the middle turbinate
2. Tear passage through the fistula is facilitated with the positioning of a Pyrex glass (Jones) or equivalent tube
3. Care should be taken to be certain that the bypass tube is well positioned in the medial fornix and that the end of the tube is free in the nasal vault
 - a. Beware obstruction by the middle turbinate or septum

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Evaluation of the Tearing Adult Patient, Module #8, 2002.
3. AAO, Focal Points: Evaluation and Surgery of the Lacrimal Drainage System in Adults, Module #12, 1995.

Canaliculitis

I. Describe the approach to establishing diagnosis

A. Describe the etiology of this disease

1. Bacterial, viral, chlamydial, mycotic organisms
 - a. *Actinomyces israelii* very common
2. Can occur after punctal plugs
 - a. Intracanalicular plugs pose most risk
3. Can occur after trauma

B. Define the relevant epidemiology of the disease

1. Patients at risk for dacryocystitis
2. Females more common

C. List the pertinent elements of the history

1. Tearing
2. Tenderness in medial canthal and medial lid
3. Chronic discharge and ocular redness
4. Recurrent swelling of lids

D. Describe pertinent clinical features

1. Erythema and swelling of medial lids
2. Dilated and pouting punctum
3. Follicular conjunctivitis
4. Pyogenic granuloma may present out of punctum

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Ophthalmic exam demonstrates above features
2. Milking canaliculus produces purulent discharge and/or stones
3. Grating sensation on probing of canaliculus (due to stones)
4. Culture of discharge

II. Define the risk factors

A. Females

III. List the differential diagnosis

- A. **Dacryocystitis**
- B. **Preseptal cellulitis**
- C. **Acquired lacrimal outflow obstruction**
- D. **Chronic conjunctivitis**

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

A. Describe the natural history, outcome, and progress

1. Ongoing inflammation leading to canalicular obstruction
2. Possible preseptal cellulitis of lids
3. Ongoing ocular inflammation
4. Epiphora and discharge
5. Recurrent episodes

B. Describe medical therapy

1. Antibiotic drops
 - a. Steroid drops may help quiet inflammation
 - b. Penicillin formulated drops may be particularly helpful with Actinomyces
2. Systemic antibiotics may be indicated for associated cellulitis or dacryocystitis
3. Warm compresses
4. Milking of canaliculus

C. Describe surgical options

1. Curettage/milking of canaliculus to remove debris and stones.
2. Incision of canaliculus (marsupialization) may be helpful
3. Lacrimal intubation in severe or recurrent cases

V. Complications of treatment, their prevention and management

A. Recurrent canaliculitis

1. Appropriate antibiotics for post op management
2. Lacrimal intubation may help minimize recurrences

B. Epiphora

1. Minimize canalicular incision
2. Keep incision on conjunctival side of canaliculus

VI. Describe disease related complications

- A. Ongoing and potentially permanent lacrimal outflow obstruction**
- B. Epiphora**
- C. Preseptal cellulitis**

VII. Describe appropriate patient instructions

- A. Finish appropriate antibiotic course**
- B. Early followup if symptoms return**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014
2. AAO, Focal Points: Eyelid Trauma, Module #10, 1991, p. 2-3.
3. AAO, Focal Points: Management of Malignant Eyelid Tumors, Module #6, 1989, p. 6.

Lacrimal outflow system trauma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Traumatic traction on the eyelid may result in medial canthal tendon and canalicular avulsion
 - a. Such traction may result in bicanalicular avulsion from the lacrimal sac and direct injury to the sac
2. Sharp injury to the lids may result in direct canaliculus laceration
3. Injury to the nasolacrimal duct usually occurs as a result of midface trauma
 - a. Medial zygomatic complex fractures
 - b. Nasal orbital ethmoid complex fractures
 - c. Nasal orbital ethmoid fractures
 - d. LeFort II and III fractures
 - i. Be aware of potential airway obstruction with LeFort III fractures

B. Define the relevant aspects of epidemiology of the disease

1. Dog bites frequently result in canalicular lacerations
2. Those at risk for midface trauma may frequently suffer injury to the nasolacrimal duct
3. Nasolacrimal duct obstruction may result after orbital decompression

C. Pertinent elements of the history

1. History of eyelid or midface trauma
2. If old injury, patient may complain of epiphora or symptoms of dacryocystitis (may result in cellulitis)

D. Describe pertinent clinical features

1. Evidence of new or old medial eyelid laceration
2. Post traumatic changes of the midface including nasal deviation or traumatic telecanthus
3. Evidence of tearing or dacryocystitis.

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. High index of suspicion in setting of trauma history
2. Canalicular irrigation and probe
3. Examination under anesthesia for a child with a laceration medial to the punctum
4. Irrigation of the opposite canaliculus in a wet field with air or viscoelastic material as a method of identifying the proximal lacerated canaliculus
5. Rule out injury to the globe

6. Rule out intracranial injury
7. CT scan to evaluate midface pathology

II. Define the risk factors

- A. Children playing with dogs close to their face**
- B. Boxers and others at risk for midface trauma**

III. List the differential diagnosis

- A. In acute setting**
 1. Eyelid laceration without canalicular or sac involvement
 2. Midface injury without involvement or displacement of nasolacrimal duct
- B. In chronic setting**
 1. Dacryocystitis or epiphora unrelated to traumatic injury but with suspicious history

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

- A. Describe the natural history, outcome and prognosis**
 1. Untreated isolated laceration of the upper canaliculus may not result in long term symptoms
 - a. Repair of an upper canaliculus should not place the lower canaliculus in jeopardy
 2. Untreated isolated laceration of the lower canaliculus will typically lead to canalicular obstruction and epiphora
 - a. Some patients may do well with upper canaliculus only, especially if dry eyes
 3. Untreated bicanalicular laceration or sac injury will frequently lead to epiphora
 4. Untreated injury to the nasolacrimal duct, especially in the setting of midface fractures, will lead to symptoms in 25% of cases
 - a. Epiphora
 - b. Dacryocystitis
 - c. May not manifest for about 1 year
- B. Define the medical therapy options**
 1. This is a surgical problem
 2. Repair of potential injury to nasolacrimal duct can be deferred until symptomatic
- C. Define surgical therapy options**
 1. For canalicular repair
 - a. Generally, to the operating room for lighting, instrumentation, and general anesthesia or

local anesthesia with intravenous sedation

- b. Occasionally, can repair upper canalicular system in treatment room under local anesthesia
 - c. Approximate ends with suture over a silicone stent
 - i. Bicanalicular nasolacrimal intubation is preferred for bicanalicular and lower canalicular lacerations
 - ii. Monocanalicular intubation is appropriate for upper canalicular lacerations; may also be appropriate in other situations
 - d. Difficulty in determining medial cut canalicular ends may be addressed by use of air, KY jelly, or viscoelastic substance injected from patent canaliculus and look for material at cut end.
 - i. A smooth-tipped pigtail probe may be used with caution to help find the cut end
 - e. Surgery preferred over first 24-48 hours but may be delayed
 - f. Suture canaliculus with fine sutures over silicone stent
 - g. Reattachment of medial canthal tendon (MCT) essential if avulsed
 - h. Repair lid margin in standard fashion
 - i. Stent in place for minimum six weeks (Remove if stent erodes punctum or canaliculitis, or stent-induced pyogenic granuloma formation)
2. For injury to the nasolacrimal duct
- a. Suspect in the presence of midface and nasal fractures and open medial canthal wounds, especially "compound" fractures
 - b. Early intubation/DCR probably indicated for open defects and disruption of the upper NLD
 - c. Some advocate prophylactic lacrimal intubation in setting of closed midface fractures
 - i. May be preferred to treat only those that become symptomatic
 - ii. These can generally be managed with simple DCR if late obstruction occurs
 - d. Manage fractures in a timely fashion

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

A. Risks of general anesthesia

- 1. Delaying repair until elective anesthesia protocol can be followed will reduce these risks

B. Intubation may cause damage to uninjured canaliculus or late erosion

- 1. Monocanalicular intubation is acceptable for upper canalicular laceration

VI. Describe disease related complications

A. Epiphora

- B. Dacryocystitis**
- C. Cellulitis**

VII. Describe appropriate patient instructions

- A. Post repair followup for up to 2 years is needed to detect late obstruction**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Focal Points: Periorbital Animal Bites, Module #3, 1992.
3. Reifler DM: Management of canalicular laceration. *Surv Ophthalmol.* 1991; 36:113-32.

Dacryocystitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Nasolacrimal duct obstruction
 - a. More common in females
 - b. Trauma
 - c. Congenital
 - d. Iatrogenic
 - e. Tumors
2. Chronic tear stasis and retention
3. Secondary bacterial infection
4. Mucopyocele (dacryocystocele)
5. Gram-positive most common organism (including MRSA) but consider gram-negatives in diabetic patients, immunocompromised patients, or nursing home residents
6. Candida occasional pathogen

B. Describe pertinent clinical features

1. Acutely, erythema, edema inferior to the medial canthal tendon
 - a. May present with cellulitis
2. Pain
3. Mass (the normal lacrimal sac usually is not palpable)
4. Chronic conjunctivitis mucopurulent discharge
5. Epiphora
6. Increased tear lake
7. Nasal speculum exam to exclude intranasal pathology

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

- A. Do not irrigate or probe during acute infection
- B. Warm compresses
- C. Oral antibiotics
- D. IV antibiotics for severe or persistent cellulitis
- E. Incision and drainage of localized abscess with packing of the abscess

F. Dacryocystorhinostomy

1. Delay intraocular surgery until infection subsides

G. Biopsy of the lacrimal sac or nasolacrimal duct may be helpful to diagnose the underlying cause of the lacrimal drainage obstruction

III. Describe disease-related complications

A. Recurrent/persistent dacryocystitis

1. Consider methicillin resistant *Staphylococcal aureus* (MRSA) and atypical mycobacteria

B. Mucocele

C. Chronic conjunctivitis

D. Progression to orbital cellulitis/abscess

E. Cutaneous fistula may form if abscess ruptures externally

F. Chronic epiphora

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System; Section 8: External Disease and Cornea, 2013-2014.
2. AAO, Focal Points: Evaluation of the Tearing Adult Patient, Module #8, 2002.
3. AAO, Focal Points: Evaluation and Surgery of the Lacrimal Drainage System in Adults, Module #12, 1995.

Endonasal dacryocystorhinostomy for acquired nasolacrimal duct obstruction

I. List the indications/contraindications

A. Indications

1. Complete/partial nasolacrimal duct (NLD) obstruction with tearing
2. NLD obstruction, acute dacryocystitis following antibiotic treatment
3. NLD obstruction with chronic dacryocystitis
4. Useful in younger patients requiring dacryocystorhinostomy (DCR) when any visible scar is undesirable
5. May be preferable to external DCR in setting of previous medial canthal skin tumor resection/radiation therapy

B. Contraindications

1. Complete canalicular obstruction
2. Suspected/diagnosed lacrimal sac tumor
3. Management of lacrimal system stone or foreign body may be more difficult via endonasal route
4. May be more difficult in setting of trauma/previous trauma repair
5. May be more difficult in setting of nasal septal deviation, unless concurrent septoplasty performed

II. Describe the pre-procedure/therapy evaluation

A. Complete ophthalmic/medical history with attention to history of:

1. Topical glaucoma/anti-viral therapy
2. Ocular surface disease
3. Herpes simplex virus (HSV) infection
4. Paranasal sinus disease/surgery
5. Facial trauma/surgery/radiation therapy
6. Acetylsalicylic acid (ASA)/anti-coagulant therapy
7. Lacrimal tumor

B. Complete lacrimal evaluation

C. Potential imaging options (on case-by-case basis)

1. Orbit/sinus computed tomography (CT)/magnetic resonance imaging (MRI) scan
2. Dacryocystography

3. Radionuclide dacryoscintigraphy

III. List the alternatives to this procedure/therapy

- A. Observation**
- B. Medical therapy of dacryocystitis**
- C. Silicone intubation +/- balloon dacryoplasty (partial NLD obstruction)**
- D. Dacryocystectomy**
- E. External DCR**
- F. Conjunctivodacryocystorhinostomy (CDCR)**
 1. Consider where there is severe canalicular obstruction
 2. Can be done in conjunction with endoscopic DCR

IV. Describe the instrumentation, anesthesia and technique

- A. Anesthesia options**
 1. Monitored anesthesia care (MAC) anesthesia
 - a. Nasal mucosal anesthesia/decongestion: 4% cocaine vs. oxymetazoline or phenylephrine with topical Xylocaine
 - b. Infraorbital +/- anterior ethmoidal nerve blocks
 - c. Local infiltration around canaliculi/lacrimal sac
 2. General anesthesia
- B. Technique**
 1. Nasal mucosal decongestion/anesthesia with topical agents
 2. Nasal mucosal decongestion/anesthesia with injectable agents
 3. Punctal dilation
 4. Introduction of endoilluminator through canaliculus into lacrimal sac (optional)
 5. Nasal mucosal incision anterior to base of anterior tip of middle turbinate which is typically medial to lacrimal sac/duct
 6. Nasal mucosal removal/flap creation; submission of tissue for pathologic examination
 7. Removal of bone overlying lacrimal sac/duct with drill/rongeurs/osteotome
 8. Lacrimal sac mucosal incision/flap creation; submission of tissue for pathologic examination
 9. Silicone intubation
 10. Mitomycin C application (optional)

V. List the complications of the procedure/therapy, their prevention and management

A. Complications and preventive measures

1. Recurrent/progressive dacryostenosis
 - a. Adequate ostium size and flap formation help prevent failure
2. Cheese-wiring through puncta/canaliculi
 - a. Appropriate tension on silicone tubing loop in medial commissure for prevention
3. Infection
 - a. Possible perioperative antibiotic treatment; not necessarily standard of care
4. Intraoperative/postoperative hemorrhage
 - a. ASA avoidance for prevention
5. Sinusitis
6. Intranasal synechiae
7. Cerebrospinal fluid (CSF) rhinorrhea

B. Complications and management

1. Recurrent/progressive dacryostenosis
 - a. Appropriate surgery
2. Cheese-wiring through puncta/canaliculi
 - a. Stent removal, canalicular repair, possible stent replacement under less tension
3. Infection
 - a. Stent removal +/- antibiotic treatment
4. Perhaps slightly lower success rate than external DCR, surgeon dependent

VI. Describe the follow-up care

A. Office exam regarding presence of above complications

B. Nasal endoscopic exam in office with crust removal

C. Lacrimal irrigation (optional)

D. Stent removal (6-8 weeks to 6 months after surgery)

1. Office
2. Transnasal vs. transcanalicular

VII. Describe appropriate patient instructions

A. Perioperative antibiotics (antibiotic vs. antibiotic-corticosteroid drops, possible oral antibiotics)

B. No eye-rubbing; possible use of eye shield at bedtime

1. Avoid inadvertent silicone tube displacement

C. No/limited nose-blowing in perioperative period

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Ophthalmic Technology Assessment: Endonasal Dacryocystorhinostomy, *Ophthalmology* 2001;108:2369-77.
3. Watkins LM, Janfaza P, Rubin PA. The evolution of endonasal dacryocystorhinostomy. *Surv Ophthalmol* 2003;48:73-84. Comment in: *Surv Ophthalmol* 2004;49:543; author reply 544.

External dacryocystorhinostomy for acquired nasolacrimal duct obstruction

I. List the indications/contraindications

A. Indications

1. Complete/partial NLD obstruction with tearing
2. NLD obstruction with acute dacryocystitis
3. NLD obstruction with chronic dacryocystitis
4. Canalicular/common canalicular obstruction (in association with canalicular/common canalicular surgery/silicone intubation)
5. Lacrimal system stone or foreign body
6. Selected cases lacrimal sac trauma

B. Contraindications

1. Complete canalicular obstruction
2. Suspected/diagnosed lacrimal sac epithelial tumor
3. Some surgeons prefer to avoid external DCR in setting of acute lacrimal infection (e.g., dacryocystitis, diverticulitis)
 - a. Endoscopic approach in such cases may be preferred
4. Caution in setting of previous medial canthal resection/reconstruction/radiation therapy

II. Describe the pre-procedure/therapy evaluation

- A. Complete ophthalmic/medical history with attention to history of topical anti-glaucomatous/anti-viral therapy, ocular surface disease, herpes simplex virus (HSV) infection, paranasal sinus disease/surgery, facial trauma/surgery/ radiation therapy, acetylsalicylic acid (ASA)/anti-platelet/anti-coagulant/anti-inflammatory therapy
- B. Complete lacrimal evaluation
- C. Nasal speculum or endoscopic examination
- D. Potential imaging options (on case-by-case basis)
 1. Orbit/sinus computed tomography (CT)/magnetic resonance imaging
 2. Dacryocystography
 3. Radionuclide dacryoscintigraphy

III. List the alternatives to this procedure/therapy

- A. Observation**
- B. Medical therapy of dacryocystitis**
- C. Dacryocystostomy**
- D. Silicone intubation +/- balloon dacryoplasty (partial NLD obstruction)**
- E. Dacryocystectomy**
- F. Endonasal DCR**
- G. Conjunctivodacryocystorhinostomy (CDCR)**
 - 1. CDCR can be used in cases of severe canalicular obstruction
 - 2. External DCR can provide fistulized mucosal tract for passage of Jones tube

IV. Describe the instrumentation, anesthesia and technique

- A. Anesthesia options**
 - 1. Monitored anesthesia care (MAC) anesthesia
 - a. Nasal mucosal anesthesia/decongestion: 4% cocaine vs. oxymetazoline or phenylephrine with topical Xylocaine
 - b. Infraorbital +/- anterior ethmoidal nerve blocks
 - c. Local infiltration around canaliculi/lacrimal sac
 - 2. General anesthesia
- B. Technique**
 - a. Nasal mucosal decongestion
 - b. Skin marking, incision
 - c. Dissection down to periosteum of anterior lacrimal crest
 - d. Elevation of lacrimal sac from lacrimal sac fossa
 - e. Osteotomy
 - f. Lacrimal sac incision and flap creation
 - g. Nasal mucosal incision and flap creation
 - h. Lacrimal and nasal mucosal biopsy
 - i. Suturing of posterior lacrimal and nasal mucosal flaps (optional)
 - j. Silicone tubing placement (optional)
 - k. Suturing of anterior lacrimal and nasal mucosal flaps
 - l. Incision closure

V. List the complications of the procedure/therapy, their prevention and management

- A. Complications and preventive measures**

1. Recurrent/progressive dacryostenosis
2. Cheese-wiring through puncta/canaliculi
 - a. Appropriate tension on silicone tubing loop in medial commissure for prevention
3. Infection
 - a. Possible perioperative antibiotic treatment; not necessarily standard of care
4. Intraoperative/postoperative hemorrhage
 - a. ASA avoidance for prevention
 - b. Stop anticoagulants pre op
 - i. May require heparinization and reversal in patients at high risk for thromboembolic disease
 - c. Post operative hemorrhage can be managed with
 - i. Elevation of head, ice compresses
 - ii. Nasal decongestant spray (e.g. oxymetazoline)
 - iii. Nasal packing in severe cases
5. Unsightly cutaneous scar, canthal webbing
 - a. Massage
 - b. Topical or injectable corticosteroids
6. Suture granuloma
 - a. Warm compresses
 - b. Antibiotics
 - c. Possible removal
7. Sinusitis
8. Intranasal synechiae
9. Cerebrospinal fluid (CSF) rhinorrhea
10. Sump syndrome
11. Stent prolapse

B. Complications and management

1. Recurrent/progressive dacryostenosis
 - a. Appropriate surgery
2. Cheese-wiring through puncta/canaliculi
 - a. Stent removal
 - b. Canalicular repair
 - c. Possible stent replacement under less tension

3. Infection
 - a. Stent removal +/- antibiotic treatment

VI. Describe the follow-up care

- A. Office exam regarding presence of above complications**
- B. Nasal endoscopic exam/crust removal (optional)**
- C. Lacrimal irrigation (optional)**
- D. Suture removal (if non-absorbable skin sutures used) 1 week postoperatively**
- E. Stent removal (6 weeks to 6 months after surgery)**
 1. Office
 2. Transnasal vs. transcanalicular

VII. Describe appropriate patient instructions

- A. Perioperative antibiotics (ointment on incision, possible oral antibiotics, possible steroid antibiotic drops); postoperative iced compresses**
- B. No eye-rubbing; possible use of eye shield at bedtime**
- C. No/limited nose-blowing in perioperative period**
- D. Restart anticoagulants day after surgery**

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. 240.
3. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p. 199-200, 268, 271-276.

Silicone intubation of the nasolacrimal drainage system

I. List the indications/contraindications

A. Indications

1. Congenital dacryostenosis
 - a. Failed previous probing
 - b. In conjunction with balloon dacryoplasty
 - c. As primary procedure in selected patients (e.g., multifocal obstruction, canalicular stenosis noted on probing)
 - d. In conjunction with punctal reconstruction
2. Acquired dacryostenosis
 - a. Partial nasolacrimal duct obstruction
 - b. Canalicular stenosis
 - c. In conjunction with balloon dacryoplasty
 - d. In conjunction with dacryocystorhinostomy (DCR) or canalicular stenosis repair. In conjunction with canaliculotomy for canaliculitis
3. In conjunction with trauma repair, eyelid reconstruction following tumor resection
4. Prophylactically prior to systemic chemotherapy/local radiation therapy. Major agents of concern would be docetaxel (Taxotere®) and 5-fluorouracil

B. Contraindications

1. Complete canalicular/common canalicular obstruction (without ability to restore patency)
2. Complete nasolacrimal duct obstruction
3. Acute lacrimal infection (e.g., dacryocystitis/diverticulitis; procedure may be useful in management of canaliculitis)

II. Describe the pre-procedure/therapy evaluation

A. Complete lacrimal evaluation

III. List the alternatives to this procedure/therapy

- A. Above surgical procedures alone (without intubation)
- B. Conjunctivodacryocystorhinostomy (CDCR) with Jones tube placement in the setting of high-grade canalicular obstruction, although salvaging of the native canalicular system should

be attempted

IV. Describe the instrumentation, anesthesia and technique

A. Intubation options

1. Monocanalicular stents
 - a. Into lacrimal sac
 - b. Down nasolacrimal duct
2. Bicanalicular stents
 - a. Down nasolacrimal duct
 - b. Into lacrimal sac (ring intubation, or "pigtail probe" of canaliculi)

B. Anesthesia

1. General anesthesia
2. Local anesthesia with/without sedation
3. Nasal decongestion/vasoconstriction for procedures involving entry into nasal cavity

C. Technique

1. Monocanalicular intubation
 - a. Punctal dilation
 - b. Placement of probe attached to tubing through punctum into canaliculus, lacrimal sac +/- nasal cavity
 - c. Seating of punctal plug portion of monocanalicular stent in punctum, or suturing of free end of tube anterior to punctum
2. Bicanalicular intubation (ring intubation)
 - a. Punctal dilation
 - b. Introduction of pig-tail probe
 - c. Careful passage of pig-tail probe through both canaliculi
 - d. Placement of silicone tubing loop containing internal suture in lumen
 - e. Tying of internal suture with rotation of tied ends of tubing into canaliculus
3. Bicanalicular intubation (nasolacrimal duct)
 - a. Nasal decongestion/anesthesia
 - b. Punctal dilation
 - c. Passage of probe attached to tubing through canaliculus into lacrimal sac, and down nasolacrimal duct with retrieval of probe from inferior meatus (with/without endoscopy)
 - d. Passage of second probe through other canaliculus in similar fashion

- e. Tying/suturing of tubing ends in nasal vestibule
- f. +/- suturing of tubing knot to mucosa of lateral nasal wall
- 4. Bicanalicular intubation performed with DCR, etc.
 - a. Passage of probes through canaliculi into DCR ostium in middle meatus
 - b. Intranasal probe retrieval from ostium
 - c. Tying/suturing/securing of tubes as above

V. List the complications of the procedure/therapy, their prevention and management

A. Complications and preventive measures

- 1. Recurrent/progressive dacryostenosis
- 2. Cheese-wiring through puncta/canaliculi
 - a. Appropriate tension on tubing loop in medial commissure
- 3. Infection
- 4. False passage creation
- 5. Epistaxis

B. Management of complications

- 1. Recurrent/progressive dacryostenosis
 - a. Appropriate surgery
- 2. Cheese-wiring through puncta/canaliculi
 - a. Stent removal
 - b. Canalicular repair
 - c. Possible stent replacement under less tension
- 3. Infection
 - a. Stent removal
 - b. +/- antibiotic treatment
- 4. False passage creation
 - a. Stent removal
 - b. Possible antibiotic treatment/additional surgery

VI. Describe the follow-up care

A. Office exam regarding presence of above complications

B. Stent removal (4 weeks to 1 year, or more, after surgery)

1. Office/ operating room (OR)
2. Transnasal vs. transcanalicular removal

VII. Describe appropriate patient instructions

- A. Perioperative antibiotics**
- B. No eye-rubbing; possible use of eye shield at bedtime**
- C. No/limited nose-blowing**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 7: Orbit, Eyelids, and Lacrimal System, 2013-2014.
2. AAO, Surgery of the Eyelid, Orbit & Lacrimal system, Vol. 3, 1995, p. 275.
3. AAO, Focal Points: Evaluation and Surgery of the Lacrimal Drainage System in Adults, Module #12, 1995, p. 12-13.