



AMERICAN ACADEMY™
OF OPHTHALMOLOGY

Academy
MOC Essentials®
Practicing
Ophthalmologists
Curriculum
2017–2019

Pediatric Ophthalmology/Strabismus

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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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The Academy gratefully acknowledges the contributions of the American Association for Pediatric Ophthalmology and Strabismus.

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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 protect sight and empower lives by serving as an advocate for patients and the public, leading ophthalmic education, and advancing the profession of ophthalmology.

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.

Organization of the POC

The Practicing Ophthalmologists Curriculum comprises 10 practice emphasis areas (PEA), plus Core Ophthalmic Knowledge.

- Core Ophthalmic Knowledge (a required segment for the ABO's MOC examinations.)
- 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. The ABO defines Core Ophthalmic Knowledge as fundamental knowledge every practicing ophthalmologist should have regardless their practice focus.

Each PEA is categorized into topics presented in an outline format for easier reading and understanding. 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 that may be helpful in preparing for MOC examinations.

Creation of the POC

The POC was developed by panels of Academy members who are practicing ophthalmologists in each of the ten practice emphasis areas. The panels reflect a diversity of background, training, practice type and geographic distribution. Additionally, all panel members are time-limited certificate holders actively participating in the MOC process.

The panels have reviewed the [ABO's content outlines](#) for the MOC examinations and developed and clinical review topics that they feel are most likely to appear on MOC examinations. These clinical topics also were reviewed by representatives from each subspecialty society.

Revision Process

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

Prior to a scheduled review the POC may be changed 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

Pediatric Ophthalmology / Strabismus

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Visual acuity testing

Preliterate figures (Allen cards, LEA symbols)

I. List the indications/contraindications

A. Indications

1. Need to quantitate vision in a normal 2 - 3-year-old child or an older child with a delay in development

B. Contraindications

1. Child who is too young to cooperate or a child who can perform optotype vision testing

II. Describe the pre-procedure evaluation

A. Assessment of level of cooperation

B. Review of optotypes with child

III. List the alternatives to this procedure

- A. If young and lacking in cooperation, use the Teller acuity cards (may not correlate with Snellen) or assess fixation preference with base-up or base-down prism

IV. Describe the instrumentation and technique

A. Preliterate figures such as the LEA cards, Allen cards or computerized video device

B. A temporary occluder or an eye patch

C. A 6 meter, well-illuminated, distraction-free testing area

D. Review cards with child binocularly

E. Occlude an eye

F. Present cards, initially at 1 meter and then increase distance quickly to preserve child's attention or interest

G. Repeat for fellow eye

H. Provide positive feedback and rewards for effort

V. Describe the considerations in interpretation of this diagnostic procedure

- A. Provides an estimate, and a comparison of the recognition acuity in children who cannot cooperate for higher levels of testing for recognition acuity

B. Does not equate with Snellen acuity values

C. In amblyopia, the crowding phenomenon makes recognition of isolated figures easier than discriminating a figure in a row

1. Care should be taken not to overestimate visual acuity in amblyopes if single figures are presented
2. Crowding bars surrounding isolated figure provides better estimate of visual acuity

D. LEA/Wright figures correlate to Snellen acuity better than Allen pictures in amblyopes

HOTV/ Sheridan Gardiner

I. List the indications/contraindications

A. Indications

1. Need to quantify vision in a normal 3 - 4-year-old child, an older child with a delay in development or a patient in the hospital, at the bed side.

B. Contraindications

1. Ability to cooperate for Snellen acuity testing if available

II. Describe the pre-procedure evaluation

A. Review cards, assess facility with letter recall

B. If poor recall, teach child to use card to "match" letters with presented letter

III. List the alternatives to this procedure

A. Non-lettered symbols (Allen or LEA)

B. Fixation preference

IV. Describe the instrumentation and technique

A. HOTV letters in graded sizes, and a card with the same letters that are presented by the book of graded letters

B. A 6 meter, well-illuminated, distraction-free testing area

C. Review letters with child binocular

D. Temporarily occlude an eye, measure smallest letter card accurately seen

E. Repeat for fellow eye

F. Provide positive feedback and rewards for effort

V. Describe the considerations in interpretation of this diagnostic procedure

A. Provides an estimate of recognition acuity

B. The crowding phenomenon exists in amblyopes, and care should be taken in interpreting visual acuity measures if single optotypes are presented rather than a line of letters (see above)

Snellen Acuity

I. List the indications/contraindications

A. Indications

1. A need to quantify visual acuity in a cooperative patient

B. Contraindications

1. Inability to cooperate or lack of space or equipment

II. Describe the pre-procedure evaluation

A. Trial with large letters test to observe response

III. List the alternatives to this procedure

- A. If unable, use other ability-appropriate testing method**

IV. Describe the instrumentation and technique

- A. Snellen optotypes or a Snellen optotypes wall chart**
- B. Occluder or eye patch**
- C. Occlude an eye with correction of refractive error, if used, in place**
- D. Have patient read a full line of letters until majority of letters on the line are missed**
- E. Record acuity as the smallest line on which at least half of letters are accurately identified**
- F. Repeat for fellow eye**

V. Describe the considerations in interpretation of this diagnostic procedure

- A. The "gold" standard for recording recognition visual acuity**
- B. The crowding phenomenon exists in amblyopes, and care should be taken in interpreting visual acuity measures if single optotypes are presented rather than a line of letters (see above)**
- C. Fixation preference in free space may indicate a difference in acuity between the two eyes**
- D. Patients with latent nystagmus may test more accurately with non-opaque occluder (e.g., + 8D lens)**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Clarke MP, Sright CM, Hrisos S, et al. Randomised controlled trial of treatment of unilateral visual impairment detected at preschool vision screening. *BMJ* 2003;327:1251.
3. Holmes JM, Beck RW, Repka MX, et al. The amblyopia treatment study visual acuity testing protocol. *Arch Ophthalmol*. 2001;119:1345-53.
4. Harvey EM, Dobson V, Tung B, et al. Interobserver agreement for grating acuity and letter acuity assessment in 1-5.5-year-olds with severe retinopathy of prematurity. *Invest Ophthalmol Vis Sci*. 1999;40:1565-76.
5. Dobson V, Quinn GE, Siatkowski RM, et al. Agreement between grating acuity at age 1 year and Snellen acuity at age 5.5 years in the preterm child. Cryotherapy for Retinopathy of Prematurity Cooperative Group. *Invest Ophthalmol Vis Sci*. 1999;40:496-503.
6. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Pediatric Eye Evaluations Preferred Practice Pattern, 2012.
7. AAPOS Vision Screening Committee Recommended Guidelines.

Abnormalities and delays of fixation behavior

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Abnormal fixation behavior develops as a result of genetics and abnormal visual experiences in a normal visual environment

B. List the pertinent elements of the history

1. Age and quality of eye contact with parent
2. Family history of vision problems
3. History of premature birth
4. Evidence of 'forceful' eye rubbing
5. History of staring at bright lights
6. Prenatal history
7. Perinatal history
8. Delayed developmental milestones

C. Describe pertinent clinical features

1. By 6 weeks of age, (normal) term infants should exhibit good fixation behavior
2. By 2-3 months of age, infant should be interested in bright objects
3. By 3-4 months of age, child should have good horizontal following behavior
4. Premature infants may develop visual attention later, depending on degree of prematurity
5. Disconjugate eye movements should not persist after 4 months
6. Fixation behavior usually determined by examination
7. Normal babies may have delayed maturation

II. Define the risk factors

A. Prematurity

B. Any ocular abnormality

C. Any central nervous system abnormality

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

A. Describe medical therapy options

1. Treat etiology of defect causing poor fixation, if possible

B. Describe surgical therapy options

1. Treat etiology of defect causing poor fixation, if possible

IV. Describe appropriate patient instructions

A. Visual development is a complex process

- B. Consultation with geneticist, neurologist, primary care physician as indicated**
- C. Visual training exercises or devices do not promote normal fixation behavior**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Pediatric Eye Evaluations Preferred Practice Pattern, 2012.

Corneal light reflex

I. List the indications/contraindications

A. Indications

1. Evaluation of binocular alignment
 - a. Infants
 - b. Poor vision in one or both eyes
 - c. Vision screening
2. Evaluation of monocular visual axis
 - a. Eccentric fixation
 - b. Angle kappa
 - i. Angle between line of sight and pupillary axis
 - ii. Positive angle kappa (light displacement nasally) may create a pseudoexotropia; no actual deviation on cover testing
3. Evaluation of fixation preference
 - a. Strabismus

B. Contraindications

1. Photophobia

II. Describe the pre-procedure evaluation

- A. Ability to cooperate with more accurate testing alternatives (Refer to section III List the alternatives to this procedure)

III. List the alternatives to this procedure

- A. Acuity testing
- B. Prism cover/uncover testing

IV. Describe the instrumentation and technique

- A. Muscle light
- B. 14-18 inches from child
- C. Evaluate location of light reflex
- D. May need to flash light and make noise to garner attention
- E. May use accommodative target in combination with above techniques
- F. 1mm deviation = 15 prism diopters
- G. Caution interpreting with anisocoria as pupillary landmarks altered

V. Describe the considerations in interpretation of this diagnostic procedure

- A. Use pupil/iris landmarks
- B. Light displacement opposite eye shift (i.e., temporally for esotropia)

- C. Krinsky testing (light reflex testing with prism alignment)**
- D. Bruchner**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Cover tests

I. List the indications/contraindications

A. Indications

1. Assess binocular alignment in patients with suspected strabismus
2. Determine the eye preferred for fixation
3. Distinguish between and measure both manifest tropic and underlying phoric components of a deviation
4. Distinguish between monocular and binocular diplopia
5. Assess monocular excursions in dissociated vertical deviation (DVD)
6. Diagnose latent nystagmus
7. Determine if secondary deviations are present in paretic or restrictive forms of strabismus

B. Contraindications

1. Profound vision loss in one or both eyes (see below)
2. Insufficient cooperation for fixation on accommodative target

II. Describe the pre-procedure evaluation

A. Determine whether fixation is central or eccentric prior to performing a cover test

B. If sensory testing of stereopsis or binocularity is planned, it should be performed first before interrupting fusion with cover testing

C. Assess visual acuity in each eye before proceeding with full cover testing

1. If visual acuity is too poor in one or both eyes to maintain fixation on a target, corneal light reflex or Krimsky testing may be substituted
2. The presence of subnormal acuity in one eye may indicate amblyopia, and may help determine the eye preferred for fixation in cover testing

III. List the alternatives to this procedure

A. Corneal light reflex tests

1. Hirschberg
2. Krimsky
3. Bruckner

B. Sensory measures of eye alignment

1. Hess screen
2. Red filter test
3. Maddox rod evaluation
4. Lancaster Red-Green Test
5. Amblyoscope (orthoptist)

IV. Describe the instrumentation and technique

A. Instrumentation

1. Accommodative fixation targets at distance and near

- a. For children, several different fixation targets may be required to maintain interest
 2. Opaque occluder
 3. Prisms (loose, bar, or Risley)
 4. Corrected refractive errors (spectacles, contact lenses)
- B. Technique**
1. Cover-uncover test (CUCT)
 - a. The main purpose of this test is to determine whether a manifest deviation (tropia) exists
 - b. With the patient fixating an accommodative target at distance, cover one eye and observe
 - i. The fellow eye
 - i) A re-fixation movement indicates a manifest tropia. However, distinguish from latent nystagmus with fast phase in the direction of the viewing eye
 - ii. The covered eye
 - i) DVD, if latent, becomes manifest with occlusion and have vertical, torsional and/or horizontal components
 - ii) If no manifest tropia, observe the covered eye as cover is removed. If covered eye makes a refixation movement, phoria is present
 - iii) Example, with exophoria, cover eye. Look at uncovered eye, no movement. When removing cover, look at covered eye. A refixation movement nasally will be seen, indicating phoria (i.e., exophoria)
 - c. If patient is diplopic, question whether diplopia resolves with monocular occlusion
 - d. Now occlude fellow eye and repeat same observations
 - e. In the strabismic patient, observe fixation pattern when cover is removed
 - i. If the patient consistently resumes fixation with one eye only, suspect amblyopia in the non-preferred eye
 - ii. If patient is able to alternate fixation, amblyopia is less likely
 - f. Repeat the above with fixation on a near target
 2. Simultaneous prism-cover test (SPCT)
 - a. This test measures the angle of a manifest tropia, separate from any underlying phoria and is most commonly used for patients with the monofixation syndrome
 - i. Especially useful in patients with small manifest deviations with an accompanying phoric component
 - ii. Should be performed after completing the CUCT which will alert the examiner to the eye, if any, that is preferred for fixation
 - b. With patient fixating an accommodative target at distance, simultaneously occlude the fixating eye and present an appropriate prism before the deviating eye
 - i. The apex of the prism is placed in the direction of the deviation
 - ii. The amount of prism is increased or decreased with subsequent simultaneous presentations until no further re-fixation movement is seen with the prism over the deviating eye
 - iii. A small amount of time is given between simultaneous presentations to allow the patient to recover motor fusional control over any underlying phoria that may have decompensated during occlusion
 - c. Repeat the above at near fixation
 - d. Example, with exophoria, SPCT, in theory, no movement should be seen as no tropia present
 3. Prism and alternate cover test (PACT)
 - a. In this test, the total magnitude of the deviation (both phoric and tropic components together) is measured
 - b. Should be performed after the CUCT and the SPCT

- c. With the patient fixating an accommodative target at distance, alternately cover the eyes while placing increasing amounts of prism before one eye until no further re-fixation movement of the fellow eye is seen
 - i. After apparent neutralization, increase the amount of prism further until the direction of the re-fixation movement reverses, then back off until no further re-fixation movement is seen
 - i) This technique allows accurate testing for the angle that slowly builds with alternate cover testing
- d. In the patient with restrictive or parietic strabismus, first perform the PACT with the prism over the affected eye, if this can be determined (primary deviation)
 - i. Then place the prism over the unaffected eye and repeat the measurement (secondary deviation)
 - ii. The secondary deviation will be greater than the primary as a consequence of Hering Law
- e. Repeat the PACT at near fixation
- f. Example, with exophoria, no tropia present. However, if perform PACT, deviation measured with be completely phoria alone

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

- A. **Inaccurate measurements will be obtained if the patient does not maintain sufficient fixation effort**
 - 1. Especially in children, communicate with the patient during the procedure to ensure that they are engaged in the process and attentive to the target
- B. **Variable measurements may be obtained in patients with large amounts of uncorrected refractive error, in certain myopathic conditions, or if large incomitancies are present and head position is not held constant**
 - 1. Have patient return for subsequent visits and repeat testing
 - 2. Repeat measurements after correcting refractive errors

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Pediatric Eye Evaluations Preferred Practice Pattern, 2012.
- 3. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Positions of gaze/evaluation of ductions and versions

I. Anatomy of the extraocular muscles

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. Arise from Annulus of Zinn
 - a. Superior rectus 7.7 mm from limbus
 - i. Superior branch of CN III
 - ii. Primary action elevation, secondary action intorsion
 - b. Lateral rectus 6.9 mm from limbus
 - i. Cranial Nerve (CN) VI
 - ii. Abduction
 - c. Inferior rectus 6.5 mm from limbus
 - i. Inferior branch of CN III
 - ii. Primary action depression, secondary action extorsion
 - d. Medial rectus 5.5 mm from limbus
 - i. Inferior branch of CN III
 - ii. Adduction

B. Obliques muscles

1. Superior oblique
 - a. Longest cranial nerve
 - b. CN IV
 - c. Extraconal
 - d. Primary action intorsion, secondary action depression, tertiary action abduction
2. Inferior oblique
 - a. Runs along lateral orbital floor lateral to lacrimal sac, ventral to inferior rectus, overlies macula
 - b. Inferior branch of CNIII
 - c. Primary action extorsion, secondary action elevation, tertiary action abduction

II. List the indications/contraindications

A. Indications

1. Ocular motility evaluation
2. Ductions are monocular rotations of the eye
3. Versions are conjugate binocular eye movements

B. Contraindications

1. Neck and spine problems might confound examination

2. Suspected or confirmed open globe

III. Describe the pre-procedure evaluation

- A. The following tests can be performed prior to evaluation of ductions and versions, except when patient history indicates these tests would cause significant dissociation**
 1. Visual acuity
 2. Cover testing
 3. Sensory testing

IV. List the alternatives to this procedure

- A. Forced ductions**
- B. Forced generations**
- C. Vestibular ocular reflex (Doll's eyes maneuver)/oculocephalic maneuvers**

V. Describe the instrumentation and technique

- A. Outpatient setting**
 1. Fixation target object that stimulates accommodation is used when assessing binocular alignment
 2. Optical correction in place, if needed
 3. Eye patch, or other occlusion device, such as occluder, as necessary
 4. Prisms
 5. When measuring ductions and versions, the head can remain stable and the eyes move into the separate positions of gaze or the eyes can remain fixed on a target as the head is rotated into the different gaze positions
 6. Measure ductions and versions in all fields of gaze and note graphically or descriptively in chart
 7. Note that when performing version testing, a change of fixating eye may cause a large change in deviation for parietic or restrictive strabismus, or dissociated vertical deviation
 - a. The primary deviation occurs when the non-parietic eye is fixating
 - b. The secondary deviation occurs when the parietic eye is fixating
 - c. Usually, secondary deviations are larger than primary deviations
 8. If ductions or versions are abnormal, quantitative measurement of deviations at 1/3 meter as well as in the cardinal positions (primary), secondary and tertiary position of gaze at 20 ft are performed with prism and cover testing

VI. Describe the considerations in interpretation of this diagnostic procedure

- A. Primary position of gaze**
 1. Observe the position of the eyes when gazing straight ahead at a fixation target at distance
 2. Observe the positions of the eyes when gazing straight ahead at a fixation target at near (1/3-meter)
 3. If ocular positions are abnormal, use prism and cover testing to quantitate the abnormality
- B. Cardinal positions**
 1. Observe the six positions of gaze wherein one muscle of each eye is the prime mover
 2. This test is done with both eyes open (versions) and can be repeated with either eye occluded (ductions) to help identify the muscle with under- or overaction
 3. If ocular positions are abnormal, use prism and cover testing to quantitate the abnormality

C. Diagnostic positions of gaze

1. Observe the eyes in the nine gaze positions that incorporate the six cardinal positions plus straight up and down and primary position
2. Also observe alignment in head tilt right and left
3. If any abnormality, test ductions monocularly
 - a. A and V patterns are identified as are other forms of incomitant strabismus from such movement
4. If ocular positions are abnormal, use prism and cover testing to quantitate the abnormality

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Pediatric Panel: Pediatric Eye Evaluations Preferred Practice Pattern, 2012.
3. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Accommodative convergence/accommodation ratio measurement

I. List the indications/contraindications

A. Indications

1. Esotropia and exotropia
2. Excess crossing at near
3. Reading difficulties
4. Unsatisfactory cosmesis due to esotropia
 - a. Assess accommodative convergence reflex
 - b. Assess any abnormalities of accommodative convergence/ accommodation (AC/A) ratio in patients who have a greater relative esotropia at near than at distance or reduced exotropia at near

II. Describe the pre-procedure evaluation

- ### A. Evaluation of ocular motility including measurements at distance and at near using an accommodative target with the refractive error corrected

III. Describe the instrumentation and technique

A. Heterophoria method

1. AC/A ratio calculated by the distance-near relationship
2. Greater relative esotropia at near indicates a high AC/A ratio
3. Greater relative exotropia at near indicates a low AC/A ratio
4. Inner pupillary distance measurement in centimeters

B. Gradient method

1. AC/A ratio calculated by the change in deviation in prism diopters divided by the change in lens power
2. An accommodative target must be used and accommodation maintained
3. The working distance is held constant
4. Plus, or minus lenses are used to vary the accommodative requirement

IV. Describe the considerations in interpretation for this diagnostic procedure

- ### A. High AC/A ratio type of esotropia. An esodeviation at 1/3 meter that measures greater than 10 prism diopter of esotropia more than the distance deviation with hypermetropia corrected

1. Treat with bifocals
2. Less commonly, treat with phospholine iodide
3. Posterior fixation suture (Faden)

B. High AC/A ratio type of exotropia

1. For accurate diagnosis, measurement at near should be done after a period of monocular patching or with

- +3.00 lenses at near
- 2. Treat with minus lenses +/- bifocals
- 3. Conventional surgery may cause this rare group of patients to overcorrect to a high AC/A ratio accommodative esotropia

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Forced ductions

I. List the indications/contraindications

A. Indications

1. Incomitant strabismus with ductional deficits
2. Need to determine restrictive vs. paretic disease
3. Appropriate planning of diagnostic workup or surgical treatment

B. Contraindications

1. In office setting
 - a. Uncooperative patient
2. Intraoperatively
 - a. Open globe

II. Describe the pre-procedure evaluation

A. Evaluation of ductions/versions/strabismic deviation

B. Discussion of test and reassurance in office setting

III. List the alternatives to this procedure

A. Intraoperative forced ductions if not cooperative for outpatient testing

IV. Describe the instrumentation and technique

A. Outpatient setting

1. Topical anesthetic
2. Cotton-tip applicator or forceps
3. Ask patient to look in direction of ductional deficit
4. Place cotton-tip or forceps at limbus 180 degrees from ductional deficit (e.g., at medial limbus if can't abduct) and attempt to move eye
5. Take care not to damage cornea or conjunctiva

B. Intraoperatively

1. Perform after patient anesthetized for surgery
2. Use forceps instead of cotton-tip applicator in manner described above. Use forceps to proptose globe and attempt to move eye.
3. May also use two forceps to proptose globe and attempt to move (e.g., if ductional deficit is horizontal, place forceps at 12:00 and 6:00 positions to move eye)
4. Exaggerated traction test for oblique muscle evaluation

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

A. Patient discomfort

1. Use adequate anesthesia or defer until intraoperatively

- B. Subconjunctival hemorrhage
- C. Conjunctival laceration
- D. Corneal abrasion
- E. Oculocardiac reflex

VI. Describe the considerations in interpretation of this diagnostic procedure

A. Forced ductions positive

1. Restrictive disease present
2. May need orbital imaging (computed tomography, magnetic resonance imaging) to determine cause (e.g., thyroid eye disease, orbital fracture with entrapment)
3. Surgical correction must consist of relieving restriction
 - a. Therefore, post-procedure, repeat test intraoperatively

B. Forced ductions negative

1. Paretic disease present
2. Must determine cause of muscle weakness
 - a. Consider neurologic conditions like cranial neuropathies or myasthenia gravis
 - b. Consider lost or slipped muscle if in postoperative setting
 - c. Consider atrophy or agenesis of extraocular muscle

C. Forced generation test

1. This test determines the amount of force generated by muscle
2. The preliminary steps same as above for forced duction testing
3. Patient asked to look in direction opposite of field of action
4. Forceps hold globe stable as muscle 'works' (little movement, paretic; strong movement, normal muscle)
5. Helps to evaluate muscle weakness coexisting with restriction
6. Helps confirm paretic disease when no restrictive disease present

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Worth four dot test

I. List the indications/contraindications

- A. Test of gross fusion, suppression and/or anomalous retinal correspondence

II. Describe the pre-procedure evaluation

- A. Visual acuity
- B. Ocular alignment

III. List the alternatives to this procedure

- A. Red lens test
- B. Bagolini lens test

IV. Describe the instrumentation and technique

- A. Glasses worn with red lens over one eye and green lens over the other eye
- B. Target shown consisting of
 - 1. One red light
 - 2. Two green lights
 - 3. One white light
- C. Target shown at near (33 cm) and distance (20 feet)
- D. Patient asked to describe number and color of lights

V. Describe the considerations in interpretation of this diagnostic procedure

- A. All four lights appreciated
 - 1. Normal response (fusion) in the patient with aligned eyes.
- B. Three green lights or 2 red lights
 - 1. One eye suppressed
- C. Three green lights, then two red lights
 - 1. Alternate suppression
- D. Five lights
 - 1. Diplopic response but no suppression
- E. More than five lights
 - 1. Functional Disease
- F. Distance targets tests central fusion/suppression
- G. Near target tests peripheral fusion/suppression
- H. Monofixation syndrome
 - 1. Usual response is suppression at distance and fusion at near

I. Results of Worth 4-dot test can sometimes be combined with results of cover test to

determine retinal correspondence

1. Anomalous retinal correspondence occurs when ocular deviation is present and near response is fused

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Arthur BW, Marshall A, McGillivray D. Worth vs Polarized four-dot test. J Pediatr Ophthalmol Strabismus, 1993;30:53-5.

Stereoacuity testing

I. List the indications/contraindications

A. Indications

1. To determine the status of binocular function
2. To help differentiate congenital from acquired strabismus in older children and adults
3. To determine whether a change in patient management is necessary
4. May be useful as an aid in vision screening

B. Contraindications

1. Patient too young to cooperate
2. Poor vision in one or both eyes

II. List the alternatives to this procedure

- A. Worth 4 dot testing and 20 prism-diopter base-out testing (tests of suppression, which do not measure stereoacuity, but can often be performed at a younger age)

III. Describe the instrumentation and technique

- A. Appropriate lighting
- B. Appropriate refractive correction for distance and near
- C. Polarized glasses with Titmus (a contour stereopsis test) or Randot tests (a random-dot stereopsis test) for near stereo
- D. Near tests calibrated for 14 inches
- E. Polarized vectograph slide or equivalent device for distance stereo
- F. Measure with prisms or anomalous head posture in place to get true stereo potential in patients with strabismus or nystagmus

IV. Describe the considerations in interpretation of this diagnostic procedure

- A. Normal near stereoacuity is 40 arc seconds or better
- B. There are monocular clues on the first several targets of the Titmus test, but not on the Randot test
- C. Normal distance stereoacuity is less than near stereoacuity
- D. If stereoacuity is subnormal or if previously normal stereoacuity is decreasing in a patient with intermittent strabismus, further treatment may be indicated
- E. Patients with anisometropia often have limited stereoacuity
- F. Again interpret responses from younger children cautiously as understanding or cooperation may be limited

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Pediatric Eye Evaluations Preferred Practice Pattern, 2012.

Three-step test for cyclovertical muscle palsies

I. List the indications/contraindications

A. Indications

1. Diagnosis of isolated cyclovertical muscle palsies in patients with hyperdeviations

B. Contraindications

1. Previous strabismus surgery, especially of the cyclovertical extraocular muscles
2. Restrictive forms of strabismus
3. C-spine instability or fixed C-spine deformity

II. Describe the pre-procedure evaluation

A. History

1. Date of onset of symptoms
2. Presence of diplopia
3. Anomalous head posture
4. Facial or orbital trauma
5. Head injury
6. Stroke
7. Other neurological symptoms (headache, vomiting, etc.)
8. Cervical spine instability
9. Prior strabismus or orbital surgery
10. History consistent with restrictive strabismus (i.e., thyroid disease)

B. Review old photographs if available

C. Assess visual acuity in each eye

D. Evaluate ductions and versions

E. Double Maddox rod test for subjective torsion

III. Describe the instrumentation and technique

A. Instrumentation

1. Accommodative fixation target at distance
 - a. For children, several different fixation targets may be required to maintain interest
2. An opaque occluder
3. Prisms (loose, bar, or Risley)

B. Technique

1. Step 1
 - a. Using the cover-uncover test or the alternate cover test, determine which eye is hypertropic
 - b. This eliminates the elevators in the hypertropic eye and the depressors in the contralateral eye from

consideration

- c. Use alternate prism cover test to quantify deviation
- 2. Step 2
 - a. With the alternate prism cover test, using an accommodative target at distance fixation, determine if the hypertropia is greatest in right or left gaze
 - b. This eliminates the cyclovertical muscles with principal action in gaze opposite the direction where the vertical deviation is greater
- 3. Step 3 (Bielschowsky head-tilt test)
 - a. Determine if the hypertropia is greater with forced head-tilt to the right or to the left
 - b. Use alternate prism cover test to quantify deviation

IV. Describe the considerations in interpretation for this diagnostic procedure

- A. This test can easily be misinterpreted if there has been previous cyclovertical muscle surgery, a restrictive strabismus process, or more than one cyclovertical muscle is involved**
 - 1. Test can only be interpreted appropriately in the absence of vestibular or supranuclear disease

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Cycloplegic refraction

I. List the indications/contraindications

A. Indications

1. Determination of objective refractive error by relaxing accommodation
 - a. Young or uncooperative patients
 - b. Strabismus (even in adult patients)
 - c. Suspected malingeringer
 - d. Accommodative spasm/pseudo myopia
 - e. Infantile glaucoma (surrogate for axial length)
 - f. Young adults

B. Contraindications

1. Previous adverse effects
2. All drugs have possibility of anticholinergic side-effects
 - a. Central nervous system changes (mad as a hatter)
 - b. Flushing (red as a beet)
 - c. Dry mouth and eyes (dry as a bone)
 - d. Fever (hot as a pistol)
 - e. May be avoided with pressure over the puncta and canaliculi secondary to absorption through the nasal mucosa
3. As strength is increased, the side effects increase for cyclopentolate

II. Describe the pre-procedure evaluation

- A. Ensure no contraindications exist to cycloplegic agents

III. List the alternatives to this procedure

- A. Manifest refraction
- B. Dry retinoscopy
- C. Autorefraction (with accommodative control)
- D. None of the above is satisfactory in a young child or in a patient with strabismus

IV. Describe the instrumentation and technique

A. Instillation of cycloplegic agents (not simple dilating agents)

1. Tropicamide
 - a. Maximum onset in about 20-30 minutes
 - b. Poor cycloplegic choice in children; not sufficient alone in children
2. Cyclopentolate
 - a. Maximum onset in about 30-40 minutes
 - b. Excellent cycloplegic agent

- c. Should be used with caution in kids with developmental delay, Down's syndrome
 - d. May last anywhere from 2 hours up to several days
 - e. Associated with respiratory depression in premies (use lower dosages)
- 3. Atropine
 - a. Multiple doses over 2-3 days
 - i. "Gold" standard cycloplegic

B. Use of retinoscope

- 1. Knowledge of working distance
- 2. Evaluate dynamic retinoscopy to confirm cycloplegia
- 3. Experience

V. Describe the considerations in interpretation of this diagnostic procedure

A. May require confirmation not under cycloplegia

- 1. Result does not necessarily equal prescription

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Strabismic amblyopia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Constant or intermittent misalignment (tropia)

B. Define the relevant aspects of epidemiology of the disease

1. More common in esotropia

C. List the pertinent elements of the history

1. Constant or intermittent nonalternating strabismus

D. Describe pertinent clinical features

1. Decreased monocular visual acuity in a structurally sound eye

II. Define the risk factors

- A. Constant, nonalternating strabismus or strong fixation preference

III. List the differential diagnosis

A. Deprivation amblyopia

B. Eccentric fixation

C. Anisometropic amblyopia

D. Isoametropic amblyopia

E. Organic amblyopia

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

A. Describe medical therapy options

1. Eyeglasses, if necessary, for refractive error
 - a. Many children have significant improvement with spectacles alone, even when the deviation persists in the spectacles
 - b. Bilateral refractive amblyopia improves significantly with spectacles alone.
2. Amblyopia therapy - full or partial occlusion
 - a. Two and 6 hours daily of prescribed patching are equally effective for moderate amblyopia
 - b. Six and 12 hours of prescribed patching are equally effective for severe amblyopia
 - c. Near activities while patching do not improve visual outcomes
 - d. After a period of treatment with spectacles until vision stops improving, usually by 12 weeks, children with moderate amblyopia treated with 2 hours of daily patching have more improvement than a control group without patching.
3. Cycloplegic agents
 - a. Patching and atropine are equally effective as initial treatments for amblyopia
 - b. Weekend and daily atropine are equally effective for moderate amblyopia
 - c. Visual acuity improvement is similar with weekend atropine with or without use of a plano lens over the sound eye

- d. In older children (age 7 to <13), atropine and patching produce improvements of similar magnitude.
- 4. Bangerter filters
 - a. In children with moderate amblyopia, visual acuity improvement is similar with patching or Bangerter filters.
- 5. Combined treatment
 - a. For children age 7 to <13 with moderate amblyopia, optical correction plus patching plus weekend atropine results in more visual acuity improvement than optical correction alone.
 - b. In children with residual amblyopia who have stopped improving with 6 hours daily patching or daily atropine, visual acuity improvement is similar with combined patching and atropine or with weaning then stopping of treatment.
- B. Describe surgical therapy options**
 - 1. Surgical correction of inciting strabismus (generally after initiation of amblyopia therapy and stability of the surgical deviation)

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

- A. Reverse amblyopia (occlusion amblyopia)**
 - 1. Routine follow-up to monitor visual acuity
 - 2. Reverse patching or "patch holiday" to treat
 - 3. Consider switching to part-time occlusion or atropine when therapy resumes
- B. Unresponsiveness to therapy**
 - 1. Consider non-compliance
 - a. Patient education about importance of compliance
 - b. Change modalities
 - 2. Repeat full examination to determine other causes for amblyopia, a "masquerade syndrome" causing decreased vision, or inaccurate refraction (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe disease-related complications

- A. Suppression**
- B. Anomalous retinal correspondence**
- C. Eccentric fixation**
- D. Loss of binocularity**
 - 1. Loss of vision
 - 2. Loss of stereoacuity
- E. Psychosocial impact/implications**

VII. Describe appropriate patient instructions

- A. Continue therapy as prescribed**
- B. Keep routinely scheduled appointments**
- C. Watch for changes in fixation behavior (reverse amblyopia)**
- D. Don't worry about change to alternate fixation from constant strabismus**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Bhartiya P, Sharma P, Biswas NR, et al. Levodopa-carbidopa with occlusion in older children with amblyopia. *J AAPOS*. 2002;6:368-72.
3. Procianoy E, Fuchs FD, Procianoy L, Procianoy F. The effect of increasing doses of levodopa on children with strabismic amblyopia. *J AAPOS*. 1999;3:337-40.
4. Metzler U, Ham O, Flores V, et al. Blue filter amblyopia treatment protocol for strabismic amblyopia: a prospective comparative study of 50 cases. *Binocul Vis Strabismus Q*. 1998;13:241-8.
5. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Amblyopia Preferred Practice Pattern, 2007.
6. Pediatric Eye Disease Investigator Group. A randomized trial to evaluate 2 hours of daily patching for strabismic and anisometropic amblyopia in children. *Ophthalmology* 2006;113:904-12.
7. Awan M, Proudlock FA, Gottlob I. A randomized controlled trial of unilateral strabismic and mixed amblyopia using occlusion dose monitors to record compliance. *Invest Ophthalmol Vis Sci* 2005; 46:1435-9.
8. Sprunger DT, Hainline BC, Plager DA, Sondhi N, Neely DE. Reverse amblyopia with atropine treatment. *JAAPOS* 2006; 10:81.
9. Pediatric Eye Disease Investigator Group. A randomized trial of atropine vs patching for treatment of moderate amblyopia in children. *Arch Ophthalmol* 2002; 120: 268-278.
10. Pediatric Eye Disease Investigator Group. A randomized trial of prescribed patching regimens for treatment of severe amblyopia in children. *Ophthalmology* 2003;110:2075-87.
11. Pediatric Eye Disease Investigator Group. A randomized trial of patching regimens for treatment of moderate amblyopia in children. *Archives of Ophthalmology* 2003;121:603-611.
12. Pediatric Eye Disease Investigator Group. Randomized trial of treatment of amblyopia in children aged 7 to 17 years. *Archives of Ophthalmology* 2005; 123:437-47.
13. Pediatric Eye Disease Investigator Group. A randomized trial of atropine regimens for treatment of moderate amblyopia in children. *Ophthalmology* 2004;111:2076-85.
14. Pediatric Eye Disease Investigator Group. A Randomized Trial of Near Versus Distance Activities While Patching for Amblyopia in Children Aged 3 to Less Than 7 Years. *Ophthalmology* 2008;115:2071-8.
15. Pediatric Eye Disease Investigator Group. Treatment of Bilateral Refractive Amblyopia in Children 3-<10 Years Old. *Am J Ophthalmol* 2007;144:487-496.
16. Pediatric Eye Disease Investigator Group. Pharmacological Plus Optical Penalization Treatment for Amblyopia: Results of a Randomized Trial. *Arch Ophthalmol* 2009;127:22-30.
17. Pediatric Eye Disease Investigator Group. Patching vs atropine to treat amblyopia in children aged 7 to 12 years: a randomized trial. *Arch Ophthalmol* 2008;12:1634-42.
18. Pediatric Eye Disease Investigator Group. A Randomized Trial Comparing Bangerter Filters and Patching for the Treatment of Moderate Amblyopia in Children. *Ophthalmology* 2010;117:998-1004.
19. Pediatric Eye Disease Investigator Group. Randomized Trial to Evaluate Combined Patching and Atropine for Residual Amblyopia. *Archives of Ophthalmology* 2011;129:960-962.
20. Pediatric Eye Disease Investigator Group. Optical Treatment of Strabismic and Combined Strabismic-Anisometropic Amblyopia. *Ophthalmology* 2012;119:150-8.

Anisometropic amblyopia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Unequal refractive errors cause the image on one retina to be chronically defocused

B. List the pertinent elements of the history

1. Eyes usually look normal
2. May be associated with strabismus or microstrabismus
3. Diagnosis often delayed if no obvious strabismus

C. Describe pertinent clinical features

1. Amblyopia more common with hyperopic or astigmatic anisometropia than with myopic anisometropia
2. In hyperopic anisometropia, the eye with greater hyperopia is usually the amblyopic eye
3. Unilateral high myopia is associated with amblyopia
 - a. Extensive myelination of retinal nerve fibers
 - b. Morning glory syndrome
 - c. Retinal coloboma

II. List the differential diagnosis

- A. Strabismic amblyopia
- B. Eccentric fixation
- C. Bilateral high refractive errors
- D. Deprivation amblyopia
- E. Organic amblyopia

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

A. Describe medical therapy options

1. Optical correction with spectacles alone can result in significant improvement in best-corrected visual acuity
2. Optical correction with monocular contact lens or binocular contact lenses of differing powers
3. Possibly combine with patching or atropine penalization
 - a. Two and 6 hours daily of prescribed patching are equally effective for moderate amblyopia
 - b. Six and 12 hours of prescribed patching are equally effective for severe amblyopia
 - c. Near activities while patching do not improve visual outcomes
 - d. Patching and atropine are equally effective as initial treatments for amblyopia
 - e. Weekend and daily atropine are equally effective for moderate amblyopia

B. Describe surgical therapy options

1. Consider refractive surgery in select cases

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

- A. Usual complications of contact lens wear
- B. Non-compliance of patients with recommended treatment
- C. Reverse(occlusion) amblyopia

V. Describe disease-related complications

- A. Amblyopia not responsive to treatment secondary to delay in diagnosis
- B. Recurrence of amblyopia following cessation of amblyopia treatment
- C. Missed organic cause of poor vision; recheck complete dilated eye exam

VI. Describe appropriate patient instructions

- A. Encourage full-time spectacle wear
- B. Appropriate amblyopia follow-up exams

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Bhartiya P, Sharma P, Biswas NR, et al. Levodopa-carbidopa with occlusion in older children with amblyopia. J AAPOS. 2002;6:368-72.
3. Pediatric Eye Disease Investigator Group. The clinical profile of moderate amblyopia in children younger than 7 years. Arch Ophthalmol. 2002;120:281-7.
4. AAO, Preferred Practice Patterns Committee. Amblyopia Preferred Practice Patterns, 2007.
5. Wallace DK; Edwards AR, Cotter SA and the Pediatric Eye Disease Investigator Group. A randomized trial to evaluate 2 hours of daily patching for strabismic and anisometropic amblyopia in children. Ophthalmology 2006; 113:904-12.
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7. Pediatric Eye Disease Investigator Group. Randomized Trial of Treatment of Amblyopia in Children Aged 7 to 17 Years. Arch Ophthalmol. 2005;123:437-447.

Bilateral amblyopia due to high refractive error

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Uncorrected, bilateral high refractive error of approximately equal magnitude

B. Define the relevant aspects of epidemiology of the disease

1. Relatively uncommon cause of amblyopia

C. List the pertinent elements of the history

1. Often asymptomatic
2. Parent or physician may have noted difficulty with vision

D. Describe the pertinent clinical features

1. Hyperopia typically greater than 4.00-5.00D
2. Myopia typically greater than 5.00-6.00 D
3. Astigmatism typically greater than 2.00-3.00D Reduction in visual acuity usually mild to moderate
4. Strabismus usually not present
5. Child usually has gross stereopsis

II. Define the risk factors

- A. Family history of amblyopia or high refractive errors**

III. List the differential diagnosis

- A. Bilateral retinal or optic nerve disease**
B. Bilateral deprivation amblyopia

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

A. Describe medical therapy options

1. Full time optical correction (eyeglasses or contacts)

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

- A. Difficulty of compliance (effort, psychological impact, cost)**
B. Discuss importance of promoting normal visual development and binocularity with caretakers

VI. Describe disease-related complications

- A. Best corrected vision achieved may be 20/25 to 20/40**
B. Reduced binocularity even after treatment

VII. Describe appropriate patient instructions

- A. Encourage compliance with eyeglasses**
- B. Inform family that vision usually improves slowly with full-time wear of eyeglasses**
- C. Appropriate follow up with ophthalmologist**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Amblyopia Preferred Practice Pattern, 2012.
3. Pediatric Eye Disease Investigator Group. Treatment of bilateral refractive amblyopia in children three to less than 10 years of age. *Am J Ophthalmol* 2007;144:487-96.

Deprivation amblyopia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Amblyopia caused by any abnormality that occludes the visual axis and interferes with the formation of a clear, focused retinal image during the critical period of visual development
2. Reverse amblyopia: occlusion amblyopia caused by patching or atropine penalization

B. Define the relevant aspects of epidemiology of the disease

1. The least common form of amblyopia

C. List the pertinent elements of the history

1. Family history of cataracts or amblyopia. Ocular or other developmental abnormalities, ocular trauma
2. Previous treatment of amblyopia

D. Describe pertinent clinical features

1. Visually significant cataracts (occupying the central 3mm or more of the lens) are the most common cause, have a greater chance of causing severe amblyopia, especially when monocular or occurring in children under 6 years of age.
2. Ptosis or other eyelid disorders such as capillary hemangioma
3. Corneal opacity
4. Vitreous hemorrhage (may have resolved by time of presentation)
5. Posterior segment abnormality

II. Define the risk factors

A. Congenital, developmental, or traumatic abnormalities of the anterior and posterior segment

B. Treatment of amblyopia

III. List the differential diagnosis

A. Strabismic amblyopia

B. Refractive amblyopia

C. Organic causes

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

A. Describe medical therapy options

1. Occlusion patch or atropine penalization of sound eye.
 - a. Bangerter foil
2. Close follow-up to prevent occlusion deprivation amblyopia and to titrate therapy

B. Describe surgical therapy options

1. Treatment of primary cause of deprivation amblyopia

V. Describe appropriate patient instructions

- A. Stress the importance of follow-up and length of treatment required**
- B. Follow up dictated by etiology of amblyopia**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Focal Points: Practical Management of Amblyopia, Module #2, 2000.
3. Holmes JM, Kraker RT, Beck RW, et al. A randomized trial of prescribed patching regimens for treatment of severe amblyopia in children. *Ophthalmology*. 2003;110:2075-87.
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11. Pediatric Eye Disease Investigator Group. A randomized trial of atropine vs. patching for treatment of moderate amblyopia in children. *Arch. Ophthalmol*. 2002;120:268-78.
12. Holmes JM, Beck RW, Repka MX, et al. The amblyopia treatment study visual acuity testing protocol. *Arch Ophthalmol*. 2001;119:1345-53.
13. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Amblyopia Preferred Practice Pattern, 2012.
14. Hatt S; Antonio-Santos A; Powell C, Vedula SS: Interventions for stimulus deprivation amblyopia. *Cochrane Database Syst Rev* 2006; 3: CD005136.

Medical treatment of amblyopia: refractive correction

I. List the indications/contraindications

A. Indications

1. Presence of amblyogenic refractive error
2. Presence of amblyopia (reduced visual acuity, in one or both eyes, secondary to an amblyogenic refractive error in a patient young enough to respond to treatment)
3. Poor fixation behavior

B. Contraindications

1. None

II. Describe the pre-therapy evaluation

A. Full examination including

1. Age appropriate visual acuity of each eye
2. Strabismus evaluation
3. Evaluation for structural abnormalities of the eye or visual pathways
4. Determination of cycloplegic refractive error

III. Describe the instrumentation and technique

- A. Dispense the appropriate prescription for eyeglasses (polycarbonate lens and protective frames when patient is functionally monocular)

IV. List the complications of this therapy, their prevention and management

A. Poor compliance

1. Counsel families on the importance of treatment, follow-up care and the risks of non-compliance

B. Unresponsiveness

1. Recheck refraction
2. Look for abnormalities of the macula, optic nerve or pathways
3. Institute additional amblyopia therapy such as occlusion or atropine to the sound eye
4. Can terminate primary therapy after an appropriate length of time if there's no response in a compliant patient

C. Recurrence

1. Reinitiate treatment or continue maintenance regimen

D. Development of amblyopia in the originally better eye

1. Recheck refraction
2. Initiate appropriate amblyopia treatment

V. Describe the follow-up care

- A. **Appropriate follow-up care based on age and severity of amblyopia**

VI. Describe appropriate patient instructions

- A. **Encourage compliance with eyeglasses**
- B. **Stress importance of follow-up as instructed**
- C. **Obtain polycarbonate lenses when the vision in the amblyopic eye is poor**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Clarke MP, Wright CM, Hrisos S, et al. Randomised controlled trial of treatment of unilateral visual impairment detected at preschool vision screening, BMJ. 2003;327:1251.
3. Atkinson J, Braddick O, Robier B, et al. Two infant vision screening programmes: prediction and prevention of strabismus and amblyopia from photo and videorefractive screening. Eye. 1996;10:188-89
4. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Amblyopia Preferred Practice Pattern, 2012.
5. Cotter SA, Edwards AR, Wallace DK and Pediatric Eye Disease Investigator Group. Treatment of anisometropic amblyopia in children with refractive correction. Ophthalmology 2006; 113:895-903.
6. Holmes JM1, Lazar EL, Melia BM and Pediatric Eye Disease Investigator Group. Effect of age on response to amblyopia treatment in children. Arch Ophthalmol. 2011 Nov;129(11):1451-7.
7. Amblyopia - AAPOS. American Association for Pediatric Ophthalmology and Strabismus Web site. <http://www.aapos.org/terms/conditions/21> Updated 05/2015.

Medical treatment of amblyopia: occlusion and optical degradation

I. List the indications/contraindications

A. Indication

1. Reduced visual acuity secondary to amblyopia

B. Contraindication

1. None

II. Describe the pre-procedure/therapy evaluation

A. Evaluation of visual acuity (adequate occlusion of each eye)

B. Obtain comprehensive birth history and family history

C. Comprehensive eye examination to rule out other causes of visual loss other than amblyopia

III. Describe the instrumentation and technique

A. Patching

1. Place adhesive patch on skin or felt patch on eyeglasses over non-amblyopic eye for prescribed time
2. Titrate patching schedule based on patient response

B. Atropine penalization to non-amblyopic eye

C. Optical degradation with Bangerter occlusion foil

D. Occlusion contact lens to non-amblyopic eye

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

A. Patching

1. Occlusion may cause reverse amblyopia in the occluded eye
 - a. Closely follow vision in occluded eye
 - b. Rule of thumb, can occlude one-week full time per year of age before developing amblyopia
2. Contact dermatitis
 - a. May be prevented with skin protectant

B. Atropine

1. Systemic absorption (high fever, redness, hyperactivity)
2. Photophobia
3. Possible retinal/lens phototoxicity
 - a. Use ultraviolet protection, hat etc.
4. Hypersensitivity reactions
5. Reverse amblyopia

C. Induced Strabismus, or greater angle of strabismus can occur after either patching or penalization treatment

- D. Treatment failures generally due to non-compliance (see below) or to delayed institution of treatment

V. Describe the follow-up care

A. Follow-up periods to be determined by

1. Age of patient
2. Treatment prescribed
3. Severity of amblyopia
4. Response to treatment

B. When plateau reached, period of maintenance patching may be helpful to prevent recurrence

C. Treatment generally terminated when no response in compliant patient after appropriate length of time

VI. Describe appropriate patient instructions

A. Patching

1. Encourage patching
2. Emollients for skin irritation

B. Atropine penalization

1. Compliance with drops
2. Watch for side effects

C. Optical degradation

1. Wear eyeglasses as prescribed

D. Non-compliance is a significant issue

1. Parental education on importance of therapy to avoid permanent visual loss
2. Suggestions for improving compliance (e.g., linking to rewards, restraints, materials to cover patch, getting through the toughest first few weeks, etc.)

E. Educate on

1. Possibility of recurrence
2. Need for follow-up exams
3. Possible reinstatement of treatment

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Newsham D. A randomized controlled trial of written information: the effect of parental non-concordance with occlusion therapy. Br J Ophthalmol. 2002;86:787-91.
3. Leguire LE, Rogers GL, Walson PD, et al. Occlusion and levodopa-carbidopa treatment for childhood amblyopia. J AAPOS. 1998;2:257-64.
4. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Amblyopia Preferred Practice Pattern, 2012.
5. Stankovic B, Milenkovic C. Continuous full-time occlusion of the sound eye vs. full-time occlusion of the sound eye periodically alternating with occlusion of the amblyopic eye in treatment of amblyopia; a prospective randomized study. Eur J Ophthalmol 2007; 17:11-9.
6. Repka MX, Holmes JM, Melia BM and the Pediatric Eye Disease Investigator Group. The effect of amblyopia treatment on ocular alignment. J AAPOS 2006; 113:903-12.
7. Holmes JM, Edwards AR, Beck RW and the Pediatric Eye Disease Investigator Group. A randomized pilot study of near activities versus non-near activities during patching therapy for amblyopia. J AAPOS 2005; 9:129-36.

8. Wallace DK, Edwards AR, Cotter SA et al. A randomized trial to evaluate 2 hours of daily patching for strabismic and anisometropic amblyopia in children. *Ophthalmology* 2006; 113:904-12.

Pseudoesotropia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Inherited facial features that cause tissue to obscure the nasal sclera (narrow set eyes)
 - a. This is often associated with a reduced interpupillary distance and wide epicanthal folds

B. List the pertinent elements of the history

1. Eyes apparently crossed in lateral gaze

C. Describe the pertinent clinical features

1. Full ductions and conjugate versions with a cover test at distance and near that does not show a tropia
2. Well centered pupil light reflex
3. Review of photographs with magnification demonstrates normal pupil light reflex, and a cycloplegic refraction within normal range for age
4. Normal dilated fundus examination

II. Define the risk factors

- A. Parent with narrow set eyes
- B. Prominent epicanthal folds
- C. Facial asymmetry
- D. Asian ethnicity

III. List the differential diagnosis

- A. Infantile esotropia
- B. Accommodative esotropia
- C. Duane syndrome
- D. Sixth nerve palsy
- E. Sensory esotropia

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

A. Describe medical therapy options

1. Observe on at least one more occasion to exclude true esotropia if initial examination is not conclusive
 - a. Parents can send "suspicious" photos for evaluation

V. Describe appropriate patient instructions

- A. Demonstrate the pupil reflex to the parent and have them observe for abnormalities
- B. Inform parents that as child grows, bridge of nose displaces epicanthal folds and pseudoesotropia appearance will improve
- C. Consider repeat examination. There is a higher incidence of strabismus or refractive amblyopia in the pseudoesotropes than in the general population

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Infantile esotropia

I. Describe the approach to establishing the diagnosis

A. Define the relevant aspects of epidemiology of the disease

1. Incidence less than 1%
2. No sex predilection
3. Family history of strabismus and amblyopia
4. Children with neurologic and developmental problems have a slightly higher incidence

B. List the pertinent elements of the history

1. Present in first six months of life
2. Cross-fixation common
3. May alternate fixation

C. Describe pertinent clinical features

1. Equal visual acuity common; amblyopia in < 50%
2. Full abduction in each eye (may need doll's head maneuver or vestibulo-ocular reflex to elicit) unless there is secondary medial rectus restriction
3. Angle of deviation often greater than 30 PD
4. Dissociated vertical deviation (DVD) and inferior oblique overaction (IOOA) eventually present in 50%, typically after 1 to 2 years of age
5. Generally low hyperopia
6. Latent nystagmus in up to 50%
7. Horizontal/rotary congenital nystagmus may be present

II. Define the risk factors

- A. Family history of strabismus and decreased binocular function common, including monofixation syndrome
- B. Cerebral palsy, hydrocephalus, prematurity

III. List the differential diagnosis

- A. Ocular instability of infancy
- B. Accommodative esotropia
- C. Partially accommodative esotropia
- D. Cranial nerve (CN) VI palsy
- E. Duane syndrome
- F. Möbius syndrome
- G. Congenital fibrosis syndrome
- H. Sensory esotropia
- I. Nystagmus blockage

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

- A. Verify stability of strabismic angle**
- B. Describe medical therapy options**
 - 1. Eyeglasses for significant refractive errors
 - 2. Amblyopia treatment, if necessary
- C. Describe surgical therapy options**
 - 1. Surgery typically necessary
 - 2. Goal is correction to within 8 -10 PD of orthotropia
 - 3. Recess of both medial rectus muscles most common, but recess-resect procedure on one eye may be appropriate
 - 4. 3-4 muscles considered if deviation > 60 prism diopter (PD)
 - 5. Optimal surgical intervention by 24 months of age, preferably earlier in order to minimize duration of ocular misalignment and achieve maximal benefit to binocular vision
 - 6. Inferior oblique weakening if significant overaction present
 - 7. Early botulinum to both medial recti

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

- A. Undercorrection/persistent esotropia**
 - 1. Ensure no accommodative component by cycloplegic refraction
 - 2. Look for missed nystagmus blockage syndrome
 - 3. Second surgery (e.g., resect lateral rectus OU or re-recess 1 MR) based on measurements
- B. Overcorrection/consecutive exotropia**
 - 1. Ensure no slipped or lost muscle by ductions and versions
 - 2. Stimulate accommodation optically
 - 3. Second surgery (e.g., recess lateral rectus OU or advance medial rectus) based on measurements
- C. Scleral perforation, infection, etc. (See Ocular complications of strabismus surgery, including consecutive deviations)**

VI. Describe disease-related complications

- A. Amblyopia in < 50%**
- B. Decreased stereoacuity**
- C. Development of an accommodative component**
- D. Fusion maldevelopment (latent and manifest latent) nystagmus**
- E. Decompensation of alignment later in life**
- F. Development of DVD or IOOA**

VII. Describe appropriate patient instructions

- A. Amblyopia treatment**
- B. Refractive error correction may necessary**
- C. Discussion of risks and benefits of surgery**
- D. Awareness of possible need for multiple reoperations**
- E. Development of DVD or IOOA**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Birch EE, Stager DR, Everett ME. Random dot stereo acuity following surgical correction of infantile esotropia. *J Pediatr Ophthalmol Strabismus* 1995;32:231-235.
3. Kushner BJ. Botulinum toxin management of essential infantile esotropia [editorial]. *Arch Ophthalmol* 1997;115:1458-1459.
4. McNeer KW, Tucker MG, Spencer RF. Botulinum toxin management of essential esotropia in children. *Arch Ophthalmol* 1997;115:1411-1418.
5. Pediatric Eye Disease Investigator Group. The clinical spectrum of early-onset esotropia: experience of the Congenital Esotropia Observational Study. *Am J Ophthalmol* 2002;133:102-108.
6. AAO, Preferred Practice Pattern Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.
7. Ing MR, Norcia A, Stager D Sr, et al. A prospective study of alternating occlusion prior to surgical alignment for infantile esotropia: one-year postoperative motor results. *J AAPOS* 2006;10:49-53.
8. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.
9. AAO, Focal Points: Botulinum Toxin in Ophthalmology, Module #3, 2007.
10. Tychsen L. Can ophthalmologists repair the brain in infantile esotropia? Early surgery, stereopsis, monofixation syndrome, and the legacy of Marshall Parks. *J AAPOS*. 2005 Dec;9(6):510-21.
11. Louwagie CR, Diehl NN, Greenberg AE, Mohny BG. Is the incidence of infantile esotropia declining?: A population-based study from olmsted county, minnesota, 1965 to 1994. *Arch Ophthalmol*. 2009;127:200-203.
12. MacEwen CJ, Chakrabarti HS. Why is squint surgery in children in decline? *Br J Ophthalmol*. 2004;88:509-511.
13. Carney CV, Lysons DA, Tapley JV. Is the incidence of constant esotropia in childhood reducing? *Eye (Lond)*. 1995;9 (Pt 6 Suppl):40-41.
14. Lam GC, Repka MX, Guyton DL. Timing of amblyopia therapy relative to strabismus surgery, *Ophthalmol*. : 1993; 100: 1751-1756.

Refractive accommodative esotropia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Accommodation due to uncorrected hyperopia causes convergence which overcomes fusional divergence and leads to esotropia
2. Driven by choice of clear vision over alignment
3. Esotropia leads to amblyopia via suppression of non-preferred eye

B. Define the relevant aspects of epidemiology of this disease

1. Begins in early childhood
2. Positive family history
3. May be precipitated by illness or trauma

C. List the pertinent elements of the history

1. Orthophoric initially
2. Esotropia typically begins between 6 months and 7 (average 2 ½) years of age
3. Adults with resolved childhood accommodative esotropia may become symptomatic again near the onset of presbyopia

D. Describe pertinent clinical features

1. Comitant moderate (20 - 30 PD) esotropia
2. Normal accommodative convergence/ accommodation (AC/A) ratio
3. Hyperopia usually greater than 2-3 diopters
4. Although less common than when associated with infantile ET, DVD and inferior oblique overaction may be present

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Must do full cycloplegic refraction/retinoscopy

II. Define the risk factors

- A. High hyperopia
- B. Insufficient fusional divergence mechanism (low hyperopia)
- C. Family history of strabismus or amblyopia

III. List the differential diagnosis

- A. Infantile esotropia
- B. Pseudoesotropia
- C. Decompensated esophoria/intermittent esotropia
- D. Nystagmus blockage syndrome
- E. Non-refractive accommodative esotropia (high AC/A ratio)
- F. Partial accommodative esotropia
- G. Acquired esotropia (neurologic or metabolic disease, trauma)

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

A. Describe medical therapy options

1. Full- time wear of cycloplegic hyperopic correction
 - a. Short term atropine may help initially to relax accommodation and facilitate spectacle compliance
2. Amblyopia therapy, if required
3. Decrease hyperopic refraction maybe attempted in school age children if alignment and binocularity are maintained

B. Describe surgical therapy considerations

1. No strabismus surgery necessary if fully controlled in glasses
2. If ocular alignment is unacceptable with appropriate refractive correction
3. To discontinue low power correction
4. Refractive surgery to eliminate eyeglass use when age appropriate

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

A. Increase in esotropia initially when glasses removed

B. Shattering of eyeglasses

1. Use of polycarbonate

VI. Describe disease-related complications

A. Amblyopia

B. Loss of binocularity

C. Loss of stereopsis

D. Diplopia (uncommon or intermittent)

VII. Describe appropriate patient instructions

A. Must wear eyeglasses full time

B. Increase in esotropia initially when glasses are removed

C. Amblyopia therapy as needed

D. If a non-accommodative component arises, surgery might be needed in the future

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Coats DK, Avilla CW, Paysse EA, et al. Early-onset refractive accommodative esotropia. J AAPOS 1998;2:275-8.
3. AAO, Preferred Practice Pattern Committee, Pediatric Ophthalmology Panel: Esotropia and Exotropia Preferred Practice Pattern, 2012.
4. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Nonrefractive accommodative esotropia (high AC/A ratio)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. High accommodative convergence/ accommodation ratio (AC/A)

B. Define the relevant aspects of epidemiology of the disease

1. Seen more commonly in hyperopic individuals, but can be seen in emmetropes or myopes

C. List the pertinent elements of the history

1. Esotropia at near greater than distance, which is usually noted in early childhood
2. May be associated with other strabismus complexes

D. Describe pertinent clinical features

1. Esotropia greater at near than distance
2. Accommodative convergence is reduced by bifocals

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Cycloplegic refraction
2. Alignment must be checked with accommodative target

II. Define the risk factors

- A. Abnormally high level of convergence per diopter of accommodation

III. List the differential diagnosis

- A. Refractive accommodative esotropia
- B. Partially accommodative esotropia
- C. Non-accommodative acquired esotropia
- D. V-pattern esotropia (near measurements checked in downgaze where esotropia worse)

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

A. Describe medical therapy options

1. Bifocals
 - a. Bifocal should bisect the pupil (executive style) or progressive lens
 - b. Initial bifocal power usually +2.50 up to +3.00
 - c. Weaning from bifocal can be started when appropriate (surgery may be considered in resistant cases)
2. Long-acting cholinesterase inhibitors (rarely used)

B. Describe surgical therapy options

1. Strabismus surgery to correct esotropia
 - a. Can normalize AC/A ratio to allow discontinuation of bifocal

- b. Consider Faden suture
- c. Consider medial rectus recession

C. Observation

- 1. Patients who are orthotropic and fuse at distance may resolve over time

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

A. Suboptimal alignment secondary to inappropriate use of bifocals

B. Long-acting cholinesterase inhibitors (phospholine iodide) side effects

- 1. Lower pseudocholinesterase levels in the blood make patients susceptible to depolarizing muscle relaxants (succinylcholine)
 - a. Instruct parents to inform anesthesiologist if surgery needed, or wear a Medic-alert bracelet
- 2. Iris cysts in some patients
 - a. Concomitant use of phenylephrine drops helps to prevent cysts

C. Over- or under-correction with surgery

- 1. Prism adaptation prior to surgery

VI. Describe disease-related complications

A. Development of non-accommodative esotropia, requiring surgery

B. Amblyopia

C. Diminished binocular function

- 1. Suppression
- 2. Abnormal retinal correspondence
- 3. Loss of stereopsis

VII. Describe appropriate patient instructions

A. Routine follow-up to monitor refraction and alignment

B. If prescribed glasses, patient should wear at all times when awake

C. Review risks of long-acting cholinesterase inhibitors if appropriate.

D. Instruct parents to inform surgeons/anesthesiologists that they are on cholinesterase inhibitors, if surgery necessary

Additional Resources

- 1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Partially accommodative esotropia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Residual esotropia despite full hyperopic correction and amblyopia correction
2. Decompensation of a previously correctable accommodative esotropia
3. A non-accommodative esotropia subsequently develops a new accommodative esotropia component

B. Define the relevant aspects of epidemiology of the disease

1. Common
2. No gender or racial predilection

C. List the pertinent elements of the history

1. Childhood onset (onset after infancy)
2. ET may be intermittent at onset
3. May have family history of esotropia, amblyopia, glasses in childhood
4. May have prior history of infantile esotropia

D. Describe the pertinent clinical features

1. Residual esotropia with full hyperopic correction in place
2. May have amblyopia
3. Hypermetropia

II. Define the risk factors

A. Delay in initiation of treatment following onset of accommodative esotropia

B. High accommodative convergence/ accommodation (AC/A) type accommodative esotropia

1. Esotropia onset before 2 years of age

III. List the differential diagnosis

A. Under corrected accommodative esotropia

B. High AC/A esotropia

C. Sensory esotropia

D. Acquired non-accommodative esotropia

E. Infantile esotropia

F. Cranial Nerve (CN) VI palsy

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

A. Describe medical therapy options

1. Full hyperopic correction
2. Amblyopia management
3. Observe if esotropia small and patient demonstrates binocularity

B. Describe surgical therapy options

1. Medial rectus weakening procedures
2. Recess/resect procedure
3. Lateral rectus resections

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

A. Under corrected accommodative esotropia

1. Repeat cycloplegic refraction to ensure full hyperopic correction is prescribed prior to surgery

B. Surgical undercorrection or overcorrection

1. Reoperation

C. Scleral perforation, infection (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe disease-related complications

A. Amblyopia

B. Decreased binocular vision

VII. Describe appropriate patient instructions

A. Compliance with glasses

B. Amblyopia treatment

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.

Basic (acquired) esotropia in childhood

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Unknown

B. List the pertinent elements of the history

1. Development of esotropia after age 6 months
2. May have diplopia
3. May have a precipitating cause resulting in prolonged disruption of binocular vision

C. Describe pertinent clinical features

1. Comitant esotropia with near deviation same as distance deviation
2. Accommodative component is usually absent
3. Amount of hyperopia is not significant
4. May be associated with central nervous system disorders

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Consider neuroimaging if lateral incomitance, divergence insufficiency, abnormal head position, diplopia or headaches

II. List the differential diagnosis

- A. Cranial nerve (CN) VI palsy
- B. Accommodative esotropia
- C. Undiagnosed Duane syndrome
- D. Cyclic esotropia
- E. Spasm of the near reflex
- F. Acute comitant esotropia

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

A. Describe medical therapy options

1. Correct hyperopia if present
2. Treat amblyopia if present
3. Observe carefully for noncomitant features and other oculomotor abnormalities on follow-up examinations

B. Botulinum toxin injection

C. Describe surgical therapy options

1. Unilateral or bilateral medial rectus recession, unilateral medial rectus recession/ lateral rectus resection
2. Preoperative prism adaptation testing may be considered

IV. List the complications of treatment, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

V. Describe disease-related complications

- A. Amblyopia**
- B. Loss of binocular function**
- C. Psychosocial implications**
- D. Diplopia**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.
3. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Acute esotropia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Decompensation of preexisting esophoria or intermittent esotropia (ET) with or without accommodative component
2. Acquired neurologic disease
3. Idiopathic disease

B. List the pertinent elements of the history

1. Sudden onset of ET
2. Possible recent psychological / emotional trauma
3. Possible recent systemic illness, often severe
4. Recent history of patching (amblyopia therapy, trauma)
5. New onset diplopia usually present
6. Usually normal binocular vision prior to onset

C. Describe the pertinent clinical features

1. New onset diplopia usually present
2. Generally normal stereoacuity returns with prism correction
3. Comitant deviations in non-neurologic disease
4. Incomitant deviation in cranial nerve (CN) VI palsy, low accommodative convergence/ accommodation in divergence insufficiency
5. Oblique dysfunction uncommon
6. Dissociated vertical deviation very rare

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Consider testing for myasthenia gravis and forced ductions in cases with notable abduction deficit
2. Brain magnetic resonance imaging if neurologic disease suspected

II. Define the risk factors

- A. Prior esophoria or intermittent ET
- B. Recent illness
- C. Recent psychological/emotional trauma
- D. Monocular occlusion or monocular visual loss
- E. Aging
- F. Acquired neurologic disease

III. List the differential diagnosis

- A. Accommodative ET
- B. Partially accommodative ET
- C. Myasthenia gravis
- D. Thyroid eye disease (thyroid orbitopathy)

- E. CN VI palsy
- F. Divergence insufficiency
- G. Accommodative spasm

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

A. Describe medical therapy options

1. Prisms
2. Hyperopic eyeglasses if indicated
3. Long-acting cholinesterase inhibitors

B. Describe surgical therapy options

1. Surgery indicated in persistent deviations that do not respond to medical or optical management
2. Chemodenervation (Botulinum toxin) of medial rectus muscle in select cases

V. Describe disease-related complications

- A. Diplopia
- B. Loss of stereoacuity
- C. Amblyopia (if occurs at young age)

VI. Describe appropriate patient instructions

- A. Monocular occlusion in non-amblyogenic patients
- B. Prismatic correction if appropriate
- C. Surgery necessary in majority of cases
- D. Restoration of normal binocular vision often possible

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.
3. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Sensory deprivation esodeviation

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Loss of vision in one or both eyes from amblyopia, trauma or structural lesion

B. Define the relevant aspects of epidemiology of the disease

1. Obstacles preventing clear and focused retinal images
2. Anisometropia and amblyopia common

C. List the pertinent elements of the history

1. Constant esotropia
2. Decreased vision in esotropic eye

D. Describe pertinent clinical features

1. Usually large-angle esotropia
2. Decreased vision in esotropic eye
3. Early in course, comitant
4. Late in course, "tight" medial rectus
5. Often with an afferent pupil defect

II. Define the risk factors

A. Cataract, corneal scar, optic nerve and retinal disorders

B. Uncorrected large refractive error

III. List the differential diagnosis

A. Accommodative or infantile esotropia with amblyopia

B. Long-standing problem with tight medial rectus

1. Cranial Nerve (CN) VI palsy, or paresis
2. Esotropic Duane syndrome

C. Consecutive esotropia

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

A. Correct amblyopia as appropriate

B. Polycarbonate lenses to protect "good" eye

C. Monocular recess-resect procedure preferably on affected eye to correct strabismus

D. After surgical recovery, annual evaluations for health of sound eye

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

A. Unstable alignment and need for reoperation

B. Standard strabismus surgical risks

VI. Describe appropriate patient instructions

- A. Polycarbonate protective lenses should be used**
- B. Sports goggles**
- C. Annual examinations to ensure health of sound eye**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. Lam GC, Repka MX, Guyton DL: Timing of amblyopia therapy relative to strabismus surgery, Ophthalmol.1993; 100: 1751-1756

Nystagmus associated with esotropia

I. Describe the approach to establishing the diagnosis

A. Define the relevant aspects of epidemiology of the disease

1. Latent nystagmus (fusion maldevelopment nystagmus syndrome) often seen in infantile esotropia
2. Sensory-defect nystagmus often accompanies sensory-defect esotropia
3. Esotropia may accompany congenital nystagmus (from any etiology)

B. List the pertinent elements of the history

1. Parents may note manifest nystagmus as early as 6-8 weeks of life
2. Head nodding or anomalous head posture may be seen
3. History of profound visual deficit (e.g., optic nerve hypoplasia, retinal dystrophy, congenital cataract, etc.) with subsequent development of sensory esotropia (ET) and nystagmus
4. History of congenital nystagmus with acquired ET due to convergence dampening (nystagmus blockage syndrome)
5. Crossed eyes

C. Describe pertinent clinical features

1. Latent (fusion maldevelopment nystagmus syndrome) and manifest latent nystagmus (See Latent nystagmus (Fusion maldevelopment nystagmus syndrome))
2. Congenital nystagmus and sensory-defect nystagmus (See Infantile nystagmus syndrome (infantile idiopathic nystagmus, congenital nystagmus))
3. Esotropia with nystagmus
 - a. May be a consequence of
 - i. Convergence dampening (nystagmus blockage syndrome)
 - ii. Sensory defect
 - b. Clinical appearance of the nystagmus is indistinguishable in congenital nystagmus and sensory-defect nystagmus
4. Ciancia syndrome
 - a. Large-angle constant esotropia
 - b. Fixing eye held in adduction
 - c. Minimal nystagmus with fixing eye in adduction
 - i. Amplitude increases as fixing eye is moved into abduction
 - d. Mild symmetric abduction deficits

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. If neurological, genetic, or metabolic etiologies suspected, refer appropriately
2. Consider electroretinogram testing if retinal dystrophy suspected

II. Define the risk factors

- A. May be underlying neurological, genetic, or metabolic disease (rare)

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

- A. Medical therapy options

1. Correct significant refractive errors
2. Treat amblyopia if present
- B. Surgical therapy options (See Latent nystagmus (Fusion maldevelopment nystagmus syndrome)) (See Infantile nystagmus syndrome (infantile idiopathic nystagmus, congenital nystagmus))**
 1. Esotropia surgery
 2. Nystagmus surgery including supra-maximal recessions of all rectus muscles
 3. Kestenbaum if a constant or stable anomalous head position, in order to keep the fixating eye in the null position

IV. List the complications of treatment, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

V. Describe appropriate patient instructions

- A. Emphasize need for periodic follow-up examinations**
- B. Referral to other pediatric sub-specialists as needed**
- C. Explain cause of anomalous head posture**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Ciancia AO. On infantile esotropia with nystagmus in abduction. J Pediatr Ophthalmol Strabismus. 1995;32:280-288.

Incomitant esodeviation

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Cranial nerve (CN) VI palsy
2. Esotropic Duane syndrome
3. Möbius syndrome
4. Strabismus fixus
5. Thyroid eye disease (Graves ophthalmopathy)
6. Ocular trauma
 - a. Orbital fracture with muscle/orbital content entrapment
 - b. Extraocular muscle damage
7. Previous strabismus surgery
8. A and V patterns
9. Myasthenia gravis
10. Orbital myositis, pseudotumor
11. Orbital tumor
12. High myopia

B. List the pertinent elements of the history

1. Diplopia (acquired deviations)
2. Head turn
3. Head trauma
4. Onset (congenital vs. acquired)
5. Other neurologic symptoms

C. Describe pertinent clinical features

1. Esodeviation varies in different fields of gaze

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Neuro-imaging in selected cases
2. Forced ductions when indicated

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

A. Describe medical therapy options

1. Prisms
2. Patching/occlusion for amblyopia and/or diplopia
3. Systemic treatment when indicated

B. Describe surgical therapy options

1. Consider botulinum toxin in the short term in select cases
2. Strabismus surgery to normalize ductions and versions, expand the range of single binocular vision

III. List the complications of treatment, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

IV. Describe disease-related complications

- A. Diplopia
- B. Limited binocular field
- C. Head turn (torticollis)
- D. Decreased abduction
- E. Loss of stereoacuity and binocular vision
- F. Amblyopia

V. Describe appropriate patient instructions

- A. likelihood of restoration of full function is dependent upon the specific etiology

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Esotropic (Type 1) Duane syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Congenital absence or hypoplasia of cranial nerve (CN) VI nerve and nucleus with anomalous innervation by aberrant branches of CN III
2. Simultaneous firing of both horizontal rectus muscles causes globe retraction and palpebral fissure narrowing
3. Most often sporadic, but approximately 10% of cases are autosomal dominant

B. Define the relevant aspects of epidemiology of the disease

1. Most common form of Duane retraction syndrome
2. More common in females
3. Left eye more common in unilateral cases
4. Discordance in monozygotic twins

C. List the pertinent elements of the history

1. Present since birth
2. There may be a family history

D. Describe pertinent clinical features

1. Poor or absent abduction
2. Incomitant esotropia (<30 PD in primary gaze)
3. Face turn to the ipsilateral side
4. Approximately 15% bilateral
5. Globe retraction on adduction with relative enophthalmos and lid fissure narrowing
6. Upshoots and downshoots (probably due to vertical slippage of a tight Lateral Rectus)
7. Small exotropia in gaze away from affected eye
8. Possible amblyopia

II. Define the risk factors

A. Anything that affects fetal development in the early fetal development

B. Associated systemic defects: dermoids, Goldenhar syndrome (oculo-auriculo-vertebral), Wildervanck syndrome (cervico-oculo-acoustic), Marcus Gunn jaw-winking

III. List the differential diagnosis

A. CN VI palsy (>30 PD, no globe retraction, no exotropia in adduction)

B. Orbital disease (medial wall fractures, orbital myositis or metastases)

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

A. Describe medical therapy options

1. Patching or penalization for amblyopia if present
2. Prisms

3. Hyperopic eyeglasses may improve esotropia and abnormal head position

B. Describe surgical therapy options

1. Reserved for abnormal head position, marked globe retraction or large upshoots or downshoots
2. Medial rectus recession (unilateral or bilateral)
3. Vertical rectus muscle transposition
4. Y - splitting of lateral rectus muscle for up and down shoots
5. Posterior fixation sutures

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

A. Consecutive diplopia

1. Undercorrection or overcorrection requiring additional surgery or prisms

VI. Describe disease-related complications

- A. Head turn**
- B. Fissure narrowing, globe retraction**
- C. Diplopia**
- D. Restricted binocular field of single vision**
- E. Amblyopia**

VII. Describe appropriate patient instructions

A. Compliance with amblyopia therapy

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.
3. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Möbius syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Hypoplasia of Cranial Nerves (CN) VI and VII
2. Chromosomal translocation (in some families)

B. Define the relevant aspects of epidemiology of the disease

1. Usually bilateral

C. List the pertinent elements of history

1. Eye misalignment gaze palsies (CN VI)
2. Poor facial muscle function (CN VII)

D. Describe pertinent clinical features

1. CN VI palsy with decreased abduction (persists with Doll's head)
2. CN VII palsy with a mask-like facies
3. Variable strabismus pattern: most are esotropic, but orthotropia (with abduction and adduction defects) or more rarely, large exotropia (with absence of convergence) may also occur
4. Amblyopia and decreased binocular vision
5. Abnormal head turn
6. Exposure keratitis
7. Many patients have limb, chest, and tongue defects
8. Other cranial nerves may be involved

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Diagnosis usually clinically based
2. Consider forced ductions to check for a tight medial rectus

II. Define the risk factors

A. Illness or maternal ingestion of drugs

B. Small percentage of cases hereditary

III. List the differential diagnosis

A. Congenital CN VI palsy

B. Congenital horizontal gaze palsy

C. Ocular motor apraxia

D. Duane syndrome

E. Infantile esotropia with medial rectus contracture

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

A. Describe medical therapy options

1. Treat refractive errors

2. Treat amblyopia
3. Lubricants for corneal exposure

B. Describe surgical therapy options

1. Medial rectus weakening procedure
2. Bilateral transposition of vertical rectus muscles to insertion of lateral rectus
3. Consider adjustable suture surgery
4. Appropriate surgical management of lagophthalmos if necessary

V. List the complications of treatment, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

A. Postoperative healing may be complicated by ocular surface disease

1. Treat keratitis preoperatively
2. Use lubricants postoperatively

VI. Describe disease related complications

- A. Exposure keratitis**
- B. Amblyopia**
- C. Decreased binocularity**

VII. Describe appropriate patient instructions

- A. Follow-up for amblyopia, strabismus, keratitis, other ocular abnormalities**
- B. Follow-up with pediatrician/specialist for associated systemic abnormalities**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Intermittent exotropia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Unknown
2. Can be familial or hereditary

B. Define the relevant aspects of epidemiology of the disease

1. Onset in childhood
2. May decompensate with age
3. Most common type of exodeviation

C. List the pertinent elements of the history

1. Deviation often manifest during visual inattention
2. Worse when tired or sick
3. Reflex closure of one eye with bright lights
4. Usually worse at distance

D. Describe pertinent clinical features

1. Full ductions and versions
2. Divergent form of strabismus controlled by fusion mechanisms under conditions of normal binocular vision
3. Amblyopia uncommon
4. Classified into basic type exotropia, true divergence excess type, simulated divergence excess type, and convergence insufficiency type based upon the difference in deviation at near vs. distance
5. Typically, good stereopsis
6. Diplopia uncommon due to suppression of deviating eye

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Prism and alternate cover testing at distance and near with proper refractive correction in place
2. Uncover full deviation at distance and nearby measuring after a period of monocular occlusion and/or +3.00 lenses at near
3. Qualitative assessment of control of deviation

II. List the differential diagnosis

- A. Exophoria
- B. Infantile exotropia
- C. Pseudoexotropia/positive angle kappa
- D. Chronic progressive external ophthalmoplegia
- E. Partial Cranial Nerve (CN) III palsy
- F. Duane syndrome Type II
- G. Myasthenia Gravis

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

A. Describe medical therapy options

1. Correct significant refractive error
2. Consider over minus lenses to stimulate accommodative convergence
3. Patching
4. Orthoptic treatments for convergence insufficiency
5. Prisms may be indicated in some cases
6. Observation alone with close follow-up may be indicated in cases with low frequency of the deviation and no associated refractive errors

B. Describe surgical therapy options

1. Horizontal muscle strabismus surgery—recession of one or two lateral rectus muscles or recession of 1 lateral rectus muscle combined with resection of the ipsilateral medial rectus muscle
2. Lateral rectus recessions if true divergence excess

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

A. Progression to constant exotropia with medical therapy options

1. Prevented by careful follow-up

B. Loss of stereopsis with medical therapy options

1. Prevented by careful follow-up

C. Development of amblyopia with medical therapy options

1. Prevented by careful follow-up

D. Surgical over/under correction--(See Ocular complications of strabismus surgery, including consecutive deviations)

V. Describe disease-related complications

A. Associated A and V patterns

B. Associated oblique muscle dysfunction

C. Progression to constant exotropia, with loss of stereopsis and development of amblyopia

D. Horizontal diplopia

E. Asthenopic symptoms/headaches

VI. Describe appropriate patient instructions

A. Comply with any treatment of optical error, amblyopia therapy, or orthoptics

B. Caregivers to monitor frequency of exodeviation

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Kushner BJ. Selective surgery for intermittent exotropia based on distance/near differences. *Arch Ophthalmol* 1998;116:324-8.
3. Kushner BJ. The distance angle to target in surgery for intermittent exotropia. *Arch Ophthalmol* 1998;116:189-94.
4. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.
5. Gnanaraj L; Richardson SR. Interventions for intermittent distance exotropia: review. *Eye* 2005;19:617-21.
6. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Infantile exotropia

I. Describe the approach to establishing the diagnosis

- A. Describe the etiology of the disease: unknown
- B. Define relevant aspects of epidemiology of the disease
 - 1. Much less common than infantile esotropia
 - 2. High association with neurological problems
 - 3. May be associated with craniofacial syndromes
- C. List the pertinent elements of the history
 - 1. Large angle exodeviation
 - 2. Present in first year of life
- D. Describe pertinent clinical features
 - 1. Exotropia (XT) usually large deviation
 - 2. Full versions, perhaps mild adduction deficit
 - 3. Alternate fixation common
 - 4. Normal refractive error
 - 5. Inferior oblique overaction, latent nystagmus, and/or dissociated vertical deviation may be present or develop later

II. Define the risk factors

- A. Neurological disease/developmental delay
- B. Craniofacial syndromes

III. List the differential diagnosis

- A. Cranial nerve (CN) III palsy
- B. Intermittent exotropia with early onset
- C. Internuclear ophthalmoplegia
- D. Exotropic Duane syndrome
- E. Sensory exotropia

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

- A. Describe medical therapy options
 - 1. Alternate patching as anti-suppression
 - 2. Correct refractive errors
 - 3. Amblyopia treatment
- B. Describe surgical therapy options
 - 1. List indications/contraindications
 - a. Indications
 - i. Mostly tropic

- ii. 6 months of age or older
- 2. Surgical options
 - a. Monocular recess-resect
 - b. bilateral lateral rectus recessions;
 - c. 3-4 muscles may be required for large deviations

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

- A. Amblyopia
- B. Surgical over/under correction (See Ocular complications of strabismus surgery, including consecutive deviations) surgery, including consecutive deviations

VI. Describe disease-related complications

- A. Dissociated vertical deviation (DVD)
- B. Oblique dysfunction
- C. Amblyopia
- D. Poor stereopsis or loss of stereopsis

VII. Describe appropriate patient instructions

- A. Importance of compliance with amblyopia therapy
- B. Post-operative patient, follow up as instructed

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.

Sensory exotropia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Reduced visual acuity in one or both eyes
2. Common causes include
 - a. Dense amblyopia
 - b. Ocular injury
 - c. Corneal opacities
 - d. Cataract
 - e. Macular or retinal disease
 - f. Optic nerve hypoplasia or atrophy

B. List the pertinent elements of the history.

1. Exo-drift of an eye that may develop weeks to decades after visual loss occurs
2. Diplopia may occur

C. Describe pertinent clinical features

1. Large-angle exotropia
2. Adduction deficits common (due to lateral rectus contracture)
3. Pseudo-overaction and true overaction of the oblique muscles
4. A, V, or X patterns common

II. Define the risk factors

A. Monocular or binocular poor vision

III. List the differential diagnosis

- A. Infantile exotropia
- B. Cranial Nerve (CN) III palsy
- C. Consecutive exotropia
- D. Exotropic Duane syndrome
- E. Internuclear ophthalmoplegia
- F. Ocular myasthenia gravis
- G. Dissociated horizontal deviation
 1. Slipped, detached or stretched scar syndromes

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

A. Describe medical therapy options

1. Occlusive (for diplopia and/or appearance)
 - a. Black patch or adhesive patch

- b. Bangerter filter
 - c. Frosted Scotch tape on lens of glasses
 - d. Occlusive contact lens
- 2. Treat amblyopia as indicated
- 3. Base-in Fresnel or ground-in prism
- 4. Fogging with over- or under-powered contact lens or eyeglass lens
 - a. if globe microphthalmic or pre-phthisical, consider scleral shell (for diplopia and/or appearance of the eye)
- B. Describe surgical therapy options**
 - 1. Recess-resect procedure on non-preferred eye
 - 2. Other options, including oblique weakening procedures, transpositions, or surgery on the opposite eye

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

- A. Standard strabismus surgical risks including risk of sympathetic ophthalmia**

VI. Describe disease-related complications

- A. Psychosocial dysfunction, job discrimination**
- B. Diplopia**

VII. Describe appropriate patient instructions

- A. Because fusion is compromised, exotropia may recur, or consecutive esotropia may develop**
- B. Inform patient preoperatively that neither visual acuity nor binocularity will likely improve with strabismus surgery**
- C. Protection of sound eye in functionally monocular patient (polycarbonate eyeglasses)**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.
- 3. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Exotropic Duane syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Abnormal development of the abducens nerve/nucleus
 - a. Anomalous innervation of the lateral rectus by aberrant branches of oculomotor nerve
 - b. Co-contraction of the horizontal recti resulting in globe retraction and palpebral fissure narrowing

B. Define relevant aspects of epidemiology of this disease

1. Most cases are sporadic
 - a. Occasional families show an autosomal dominant inheritance pattern
2. Left eye is more common
3. Female preponderance
4. Much less common than esotropic Duane syndrome

C. List the pertinent elements of the history

1. Typically present at birth
2. May have positive family history
3. Chronic head turn

D. Describe pertinent clinical features

1. Incomitant strabismus
2. Ipsilateral adduction deficit
3. Deviation often present in primary position
4. Head turn to maintain single binocular vision
5. Globe retraction on attempted adduction
6. May present with over-elevation or over-depression on adduction (upshoot & downshoot)
7. May have limb abnormalities, cardiac abnormalities, neurosensory deafness & other syndromic associations (e.g. Goldenhar syndrome)

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Can confirm with electromyogram but not required

II. Define the risk factors

A. Family history

III. List the differential diagnosis

- A. Cranial nerve (CN) III palsy
- B. Congenital exotropia
- C. Myasthenia gravis
- D. Internuclear ophthalmoplegia
- E. Trauma/restrictive motility
- F. Congenital fibrosis of the extraocular muscles

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

A. Describe medical therapy options

1. Treat amblyopia if present
2. Correct refractive errors
3. Prism eyeglasses for head turn/primary position deviation

B. Describe surgical therapy options

1. Lateral rectus recession (one or both eyes)
2. Avoid medial rectus resection (often worsens retraction)
3. Match "defect" in contralateral eye
4. For up/down shoot:
 - a. Fixation of lateral rectus muscle to orbital wall may be effective.
 - b. The "tether" phenomenon can be treated by splitting the lateral rectus into a "Y" and resuturing sections above and below the original axis along with recession of the lateral rectus

V. List the complications of treatment, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe disease-related complications

- A. Head turn/ neck problems
- B. Amblyopia
- C. Difficulty with social interactions
- D. Restricted binocular field of single vision
- E. Abnormal binocular function

VII. Describe appropriate patient instructions

- A. Surgical intervention is generally not performed when binocular vision is present with the eyes in primary position or if binocularity can be maintained with a slight head turn
- B. Surgery may improve face turn but won't necessarily normalize ocular motility
- C. Reading generally not impaired
- D. Appropriate amblyopia therapy instructions

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. DeRespinis PA, Caputo AR, Wagner RS, et al. Duane's retraction syndrome. *Surv Ophthalmol* 1993;38:257-288.
3. Velez, FG, Thacker N, Britt MT, Alcorn D, Foster RS, Rosenbaum AL. Rectus muscle orbital wall fixation: a reversible profound weakening procedure. *JAAPOS* 2004 Oct. 8(5):473-80.
4. Jampolsky A. Duane syndrome. In: Rosenbaum AL, Santiago AP, eds. *Clinical Strabismus Management: Principles and Surgical Techniques*. Philadelphia: Saunders; 1999:325-346.
5. AAO, Preferred Practice Patterns Committee, Pediatric Panel. Esotropia and Exotropia Preferred Practice Pattern, 2012.
6. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Convergence insufficiency

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Often idiopathic
2. Fatigue or illness
3. Eyeglasses inducing a base-out prism effect
4. Patients with increased reading requirements such as students or computer workers
5. Often occurs in patients with Parkinson's disease
6. May have an association with ADHD

B. List the pertinent elements of the history

1. Usually presents in the older child or teenager
2. Common symptoms: asthenopia, reading problems, blurry near vision and diplopia
3. May have a history of exotropia
4. May complain of headaches after long periods of close work
5. May complain of having to close one eye to read

C. Describe pertinent clinical features

1. Alternate cover testing usually shows an exophoria at near with no significant distance deviation
2. Exophoria at near often intermittently breaks down into a frank exotropia, especially after prolonged near work
3. Remote near point of convergence
4. May also be associated with reduced fusional convergence and/or accommodative amplitudes

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Manifest and cycloplegic refractions
2. Determine near point of convergence (normal value is $\leq 8-10$ cm)
3. Determine convergence amplitudes (average values are 14 PD at distance and 38 PD at near)
4. Determine near points of accommodation
5. Determine accommodative amplitudes
6. Dynamic retinoscopy

II. Define the risk factors

- A. Large amount of time spent on near tasks

III. List the differential diagnosis

- A. Accommodative insufficiency
- B. Uncorrected refractive error
- C. Intermittent exotropia with convergence insufficiency
- D. Uncorrected anisometropia or other causes of asthenopia
- E. Medial rectus weakness due to previous recession

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

A. Describe medical therapy options

1. Appropriate refractive correction
2. Orthoptic exercises
 - a. Convergence exercises
 - b. Base out prisms at near
 - c. Brock string
3. Computer-based vergence exercises
4. Reading eyeglasses with base-in prism

B. Describe surgical therapy options

1. Strabismus surgery
 - a. Surgical options
 - i. Medial rectus muscle resection (unilateral or bilateral)
 - ii. Medial rectus resection + lateral rectus recession

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

A. Complications of surgical therapy (See Ocular complications of strabismus surgery, including consecutive deviations)

1. Diplopia in distance viewing
 - a. Prevent by conservative medial rectus resection surgery

VI. Describe disease-related complications

- A. Diplopia**
- B. Blurred vision**
- C. Headaches**
- D. Poor school performance**

VII. Describe appropriate patient instructions

- A. Compliance in orthoptic exercises**
- B. Goal of treatment is to provide long lasting symptom relief without the need for prisms in eyeglasses or surgery**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Granet DB, Gomi CF, Ventura R, Miller-Scholte A. The Relationship Between Convergence Insufficiency and ADHD. *Strabismus* 2005;13 (4): 163-8.
3. Scheiman M, Mitchell GI, Cotter S, et al and the Convergence Insufficiency Treatment Trial Study Group. A randomized clinical trial of treatments for convergence insufficiency in children. *Arch Ophthalmol* 2005;123:14-24.
4. Scheiman M, Cotter S, Rouse M and the Convergence Insufficiency Treatment Trial Study Group. Randomised clinical trial of the effectiveness of base-in prism reading glasses versus placebo reading glasses for symptomatic convergence insufficiency in children. *Br J Ophthalmol* 2005;89:1318-23.

5. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.
6. Scheiman M, Hertle R, Kraker R, et al and the Pediatric Eye Disease Investigator Group. [Patching vs atropine to treat amblyopia in children aged 7 to 12 years: a randomized trial](#). Arch Ophthalmol 2008;126(12):1634-42.
7. Convergence Insufficiency Treatment Trial Study Group. [Randomized clinical trial of treatments for symptomatic convergence insufficiency in children](#). Arch Ophthalmol 2008;126(10):1336-49.
8. Wallace DK. [Treatment options for symptomatic convergence insufficiency](#). Arch Ophthalmol 2008;126(10):1455-6.

V pattern strabismus

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Etiology is unknown, except in cases of extraocular muscle palsies. Multiple theories suggest primary or secondary muscle dysfunction as noted below
 - a. Inferior oblique muscle overaction
 - b. Superior oblique muscle underaction
 - c. Horizontal rectus muscle dysfunction
 - i. Increased lateral rectus muscle action in upgaze
 - ii. Increased medial rectus muscle action in downgaze
 - d. Vertical rectus muscle dysfunction
 - i. Based on tertiary effect of adduction
 - e. Extorsion of globe and bony orbit as seen in Crouzon or Apert syndrome

B. Define the relevant aspects of epidemiology of this disease

1. A or V pattern occurs in up to one quarter of all strabismus cases
2. V pattern most frequently associated with infantile esotropia

C. List the pertinent elements of the history

1. Anomalous head posture - chin up for V pattern exotropia, chin down for V pattern esotropia
2. Prior strabismus surgery
3. Head trauma

D. Describe pertinent clinical features

1. Horizontal deviation that is more convergent (less divergent) in downgaze compared with upgaze
2. Considered clinically significant when there is a difference of 15 prism diopters between upgaze and downgaze, each measured 25 degrees from primary position while fixing on an accommodative target at distance with the proper refractive correction

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Prism and alternate cover testing including measurements in up- and down-gaze, assessment of ductions and versions
2. Computed tomography (CT) or magnetic resonance imaging (MRI) scan of orbit, with attention to coronal views if orbital anatomic abnormality suspected
3. Consider neuroimaging of brain if bilateral superior oblique palsy

II. Define the risk factors

- A. Infantile esotropia
- B. Prior strabismus surgery
- C. Craniosynostosis (Apert or Crouzon syndrome)
- D. Superior oblique (SO) palsy, especially bilateral
- E. Brown syndrome (divergence seen in attempted elevation from primary position)

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

A. Describe surgical therapy options

1. Weakening of inferior oblique muscles
 - a. Indicated when significant overaction of these muscles is present
 - b. Corrects up to 15-25 prism diopters of V pattern
 - c. Tends to be "self-adjusting"; overcorrection is rare
 - d. Anterior transposition should be considered if DVD present or likely to develop
2. Horizontal rectus muscle displacement
 - a. Indicated when there is no or little oblique dysfunction
 - b. Amount is usually one-half to one full tendon width
 - c. Medial rectus muscles are displaced downward
 - d. Lateral rectus muscles are displaced upward
 - e. Useful acronym is MALE: *m*edial rectus to the *a*pex, *l*ateral rectus to the *e*mpy space
3. SO tuck(s)
 - a. Sometimes indicated when significant SO muscle underaction is present (e.g. bilateral SO palsies)
 - b. Somewhat unpredictable result; can result in Brown syndrome
4. Horizontal displacement of vertical rectus muscles (less commonly performed)
 - a. Sometimes indicated in the absence of a horizontal deviation
 - b. Superior rectus muscles moved nasally; inferior rectus muscles moved temporally (for ex. for V pattern esotropia)

IV. List the complications of treatment, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

V. Describe disease-related complications

- A. Psychosocial impact
- B. Poor binocular vision
- C. Abnormal head position

VI. Describe appropriate patient instructions (See Surgery of the extraocular muscles: weakening procedures)

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

A pattern strabismus

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Superior oblique muscle overaction
2. Inferior oblique muscle underaction
3. Horizontal rectus muscle dysfunction
 - a. Increased medial rectus muscle action in upgaze
 - b. Increased lateral rectus muscle action in downgaze

B. Define the relevant aspects of epidemiology of this disease

1. A or V pattern occurs in up to one-fourth of all strabismus cases

C. List the pertinent elements of the history

1. Anomalous head posture (chin up for A-esotropia, chin down for A-exotropia)
2. Prior strabismus surgery

D. Describe pertinent clinical features

1. Horizontal deviation that is more convergent (less divergent) in upgaze compared with downgaze
2. Considered clinically significant when there is 10 prism diopters (PD) difference between upgaze and downgaze, each measured 25 degrees from primary position while fixing on a distance target with accommodation controlled by the proper refractive correction

II. Define the risk factors

- A. Prior strabismus surgery (especially bilateral inferior rectus or inferior oblique weakening procedures)
- B. Primary superior oblique muscle overaction
- C. Inferior oblique muscle palsy
- D. Craniosynostosis
- E. Trisomy 21

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

A. Describe medical therapy options

1. Management of refractive error and amblyopia as appropriate

B. Describe surgical therapy options

1. Weakening of superior oblique tendons
 - a. Options include tenotomy, recession, spacer
 - b. Indicated when significant overaction of these muscles is present
 - c. Bilateral superior oblique tenotomies produce large esotropic shift in downgaze, little to no esotropic shift in primary gaze; may induce torsional imbalance
2. Horizontal rectus muscle displacement
 - a. Indicated when there is no or very little oblique dysfunction
 - b. Amount is usually one-half to one full tendon width
 - c. Medial rectus muscles are displaced upward

- d. Lateral rectus muscles are displaced downward
- e. Useful acronym is MALE: medial rectus to the apex, lateral rectus to the empty space

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

A. Complications of superior oblique weakening procedures

- 1. Hypertropia in primary gaze
- 2. Excyclodiplopia
- 3. Esotropic shift in primary gaze (See Ocular complications of strabismus surgery, including consecutive deviations)

V. Describe disease-related complications

- A. Diplopia**
- B. Torticollis**
- C. Neck pain**
- D. Headache**
- E. Psychosocial impact**

VI. Describe appropriate patient instructions (See Surgery of the extraocular muscles: strengthening procedures)

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Inferior oblique muscle overaction/overelevation in adduction

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Not well understood in infantile strabismus
2. Primary inferior oblique overaction (IOOA) - not associated with superior oblique palsy
3. Secondary IOOA - associated with paresis or palsy of antagonist superior oblique or yoke superior rectus or restriction of the contralateral inferior rectus

B. Define the relevant aspects of epidemiology of the disease

1. Usually develops in patients with infantile esotropia (ET)

C. List the pertinent elements of history

1. Abnormal head posture/head tilt
2. Abnormal movement of the eyes in extreme positions of gaze
3. Misalignment of the eyes, particularly in side gaze

D. Describe pertinent clinical features

1. Over-elevation of the eye in adduction
2. When the adducting eye is fixating, the abducted eye will be depressed with a hypotropia on alternate cover testing
3. V-pattern strabismus
4. Positive head-tilt test with superior oblique (SO) palsy
5. Fundus excyclotorsion
6. Be alert for IOOA in the child with infantile strabismus, latent nystagmus and/or dissociated vertical deviation or in the child with a head tilt and CN IV palsy

II. Define the risk factors

- A. Infantile strabismus, acquired ET or exotropia
- B. SO palsy
- C. Craniosynostoses
- D. Dysthyroid orbitopathy

III. List the differential diagnosis

- A. Dissociated vertical deviation (DVD)
- B. Duane syndrome with upshoot
- C. Anti-elevation syndrome following anterior transposition of the inferior oblique muscle (ATIO) can produce a restriction to elevation in abduction, which can appear similar to an overelevation in adduction in the fellow eye, mimicking IOOA
- D. Restrictive strabismus/orbitopathy

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

A. Describe medical therapy options

1. Treat significant refractive error
2. Treat amblyopia
3. Observation may be appropriate if there is good alignment in primary gaze, no significant V-pattern and no significant strabismus on side gaze

B. Describe surgical therapy options

1. Weakening of the inferior oblique
 - a. Recession
 - b. Myectomy
 - c. Anteriorization/transposition
 - d. Denervation/extirpation
2. Address above at the same time as any necessary horizontal strabismus surgery

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

A. Consecutive or residual vertical strabismus

1. Persistent inferior oblique overaction (See Ocular complications of strabismus surgery, including consecutive deviations)
2. Possible unmasking of contralateral inferior oblique overaction with unilateral surgery

B. Ocular torsion and occasional torsional diplopia

C. Consecutive A-pattern with overaction of SO muscles

D. Postoperative hypodeviation secondary to limited elevation of the eye,

E. Anti-elevation syndrome associated with anterior transposition of the inferior oblique

VI. Describe disease-related complications

A. Amblyopia

B. Decreased binocularity

C. Abnormal head posture

D. Unacceptable appearance of ocular alignment (psychosocial implications)

VII. Describe appropriate patient instructions

A. Appropriate follow-up for amblyopia and strabismus

B. In a postoperative patient, follow-up as instructed

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Farvardin M, Attarzadeh A. Combined resection and anterior transposition of the inferior oblique muscle for the treatment of moderate to large dissociated vertical deviation associated with inferior oblique muscle overaction. J Pediatr Ophthalmol Strabismus 2002;39:268-72; quiz 293-4.
3. Quinn AG, Kraft SP, Day C, et al. A prospective evaluation of anterior transposition of the inferior oblique muscle, with and without resection, in the treatment of dissociated vertical deviation. J AAPOS 2000;4:348-53.
4. De Angelis D, Makar I, Kraft SP. Anatomic variations of the inferior oblique muscle: a potential cause of failed inferior oblique weakening surgery. Am J Ophthalmol 1999;128:485-488.

5. Stager DR. Costenbader Lecture. Anatomy and surgery of the inferior oblique muscle: recent findings. J AAPOS 2001;5:203-208.
6. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Dissociated strabismus complex (vertical deviation) / dissociated vertical deviation

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Associated with early disruption of binocular development

B. Define the relevant aspects of epidemiology of the disease

1. Common disorder found in patients with infantile horizontal strabismus, but may occur later in life

C. List the pertinent elements of the history

1. Infantile strabismus
2. One or both eyes drifting upward spontaneously

D. Describe pertinent clinical features

1. Either eye may spontaneously and slowly drift upward, outward, and excyclotort
2. Occurs when eye is occluded or during visual inattention
3. When the deviated eye moves downward to fixate, the previously fixating eye makes no downward movement (violating Hering's law)
4. Usually bilateral
5. May be asymmetric
6. Associated with horizontal strabismus and latent nystagmus
7. Patients may have a compensatory head tilt

II. Define the risk factors

A. Infantile horizontal strabismus

III. List the differential diagnosis

A. True hypertropia

B. Overacting inferior oblique muscle

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

A. Treatment indicated if vertical deviation occurs spontaneously, is frequent, and is cosmetically significant

B. Surgical treatment

1. Superior rectus recession +/- Faden
2. Inferior oblique anterior transposition
3. Inferior rectus resection

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

complications of strabismus surgery, including consecutive deviations)

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Unilateral superior oblique palsy

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Congenital
2. Acquired/idiopathic
 - a. Head trauma
 - b. Microvascular disease
 - c. Rarely tumors

B. Define the relevant aspects of epidemiology of this disease

1. Most common vertical strabismus in childhood
2. Most common isolated cyclovertical muscle palsy

C. List pertinent elements of history

1. Binocular vertical diplopia
2. Possible torsional component, especially in acquired cases
3. Head tilt to one side
4. Worsening diplopia with age, fatigue, stress

D. Describe pertinent clinical features

1. Hypertropia worse in contralateral gaze and ipsilateral head tilt --incomitance may diminish over time due to ipsilateral superior rectus contracture
2. Ipsilateral superior oblique underaction and/or inferior oblique overaction
3. Excyclotorsion usually < 10 degrees on double Maddox rod (DMR) and/or dilated fundus examination
4. Torticollis towards contralateral side
5. Increased vertical fusion amplitudes if congenital or long-standing
6. May see facial asymmetry

E. Describe appropriate testing and evaluation for establishing diagnosis

1. Old photograph review can demonstrate longstanding or congenital disease by showing prior torticollis
2. 3-step test
3. Consider neuroimaging if palsy likely to be acquired

II. Define the risk factors

A. Acquired

1. Head trauma
2. Diabetes mellitus
3. Hypertension

III. List the differential diagnosis

A. Contralateral inferior rectus restriction (e.g., thyroid eye disease)

B. Myasthenia gravis

- C. Skew deviation, in which case the hypertropic eye would be incyclotorted
- D. Primary inferior oblique overaction/DVD
- E. Bilateral superior oblique paresis

IV. Describe patient management re treatment and follow-up

- A. Describe medical therapy options
 - 1. Prisms if deviation small, fairly comitant, and with minor torsion
 - 2. Monocular occlusion
 - 3. Neuroimaging if history of trauma or if other neurological signs present
- B. Describe surgical therapy options
 - 1. More than one muscle may be required if deviation in primary > 15 prism diopter
 - a. Inferior oblique weakening
 - b. Superior oblique tuck
 - c. Contralateral inferior rectus recession
 - d. Ipsilateral superior rectus recession if contracture present
 - e. Harada-Ito if excyclotorsion is the primary problem

V. List complications of treatment/prevention/management

- A. Brown syndrome from superior oblique (SO) tuck
- B. Recurrence of deviation
- C. Unmasking contralateral SO paresis
 - 1. Perform careful preoperative three-step test and DMR
 - 2. Consider preoperative Lancaster Red-Green test (See Ocular complications of strabismus surgery, including consecutive deviations)
- D. Overcorrection with vertical diplopia
 - 1. (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe disease-related complications

- A. Headache/neck ache from persistent torticollis
- B. Diplopia

VII. Describe appropriate patient instructions

- A. Follow-up with primary care physician if acquired lesion

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Bilateral superior oblique paralysis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Congenital
2. Acquired
 - a. Trauma
 - b. Vascular (rare)
 - c. Tumor (rare)

B. Define the relevant aspects of epidemiology of the disease

1. Head trauma
2. Craniofacial syndromes

C. List the pertinent elements of the history

1. Head trauma
2. Abnormal head position
 - a. Chin down
 - b. Head tilt if asymmetric
3. Diplopia
4. Unusual eye movements in side gaze
5. Photographs helpful

D. Describe pertinent clinical features

1. V pattern esotropia
2. Chin down posture
3. Right hypertropia in left gaze with left hypertropia in right gaze
4. Right hypertropia in right head tilt and left hypertropia in left head tilt
5. Elevation of each eye in adduction (overaction of inferior oblique muscles)
6. Reduced depression in adduction (underaction of superior oblique muscles)
7. Excyclotorsion of the fundi

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Measurement of alignment of eyes in diagnostic gaze positions
2. Double Maddox rod shows excyclotorsion of more than 10 degrees
 - a. More excyclotorsion in downgaze
3. Excyclotropia on fundus examination
4. "V" Pattern esodeviation in down gaze

II. Define the risk factors

A. Craniosynostosis, Crouzon syndrome and related shallow orbit conditions

B. Significant head trauma

III. List the differential diagnosis

- A. Dissociated vertical deviation**
- B. V-pattern esotropia**

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

A. Describe medical therapy options

- 1. Patch to prevent/treat amblyopia
- 2. Prisms, usually unsuccessful
- 3. Wait 6 months or more from head trauma for recovery

B. Describe surgical therapy options

- 1. Weaken inferior oblique muscles
- 2. Graded tuck of superior oblique muscles
- 3. Harada-Ito for torsion
- 4. Inferior rectus recession
- 5. Correct horizontal deviation if present

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

- A. Brown syndrome, secondary to tuck of superior oblique muscles (See Ocular complications of strabismus surgery, including consecutive deviations)**
- B. Recurrence of inferior oblique overaction**

VI. Describe disease-related complications

- A. Inability to develop fusion and high levels of binocular function.**
- B. Anomalous head posture**
- C. Diplopia**

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Brown syndrome/superior oblique tendon sheath syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Restriction of the superior oblique tendon at the trochlear pulley

B. Define the relevant aspects of epidemiology of the disease

1. 10% are bilateral
2. May be congenital or acquired
3. May be constant or intermittent
4. May spontaneously resolve

C. List the pertinent elements of the history

1. May have abnormal head position, often chin up or face turn to opposite side
2. May have diplopia
3. Patients may note an audible "click" as tendon passes through the trochlea

D. Describe pertinent clinical features

1. Deficient elevation in adduction
2. Elevation improves with abduction
3. Attempted midline elevation may cause divergence (V pattern)
4. Adduction is associated with a widening of the palpebral fissure and a downshoot of the involved eye
5. May have a hypotropia of the involved eye in the primary position

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Alternate cover testing in cardinal gaze positions
2. Assessment of ductions and versions
3. Palpate trochlea
4. Positive forced duction testing
5. May need neuroimaging for acquired disease

II. Define the risk factors

- A. Local trauma in the region of the trochlea
- B. Systemic inflammatory conditions
- C. Sinusitis

III. List the differential diagnosis

- A. Inferior oblique palsy
- B. Double elevator palsy
- C. Orbital floor fracture
- D. Inferior rectus fibrosis

- E. Duane syndrome with downshoot

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

A. Describe medical therapy options

1. Observation
2. Treat any associated amblyopia
3. Consider oral corticosteroids for acquired disease
4. Consider nonsteroidal anti-inflammatory drugs (NSAIDs) for acquired disease

B. Describe surgical therapy options

1. Injection of corticosteroids near the trochlea and for acquired inflammatory disease
2. Ipsilateral superior oblique tenotomy, with or without ipsilateral inferior oblique weakening
3. Superior oblique lengthening, (incisional, with suture, or with silicone spacer)
4. Partial posterior tenectomy of the superior oblique
5. Consider surgical intervention for
 - a. Abnormal head posture
 - b. Symptomatic diplopia
 - c. Vertical strabismus in the primary gaze position

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

- A. Iatrogenic superior oblique muscle palsy
- B. Restrictive strabismus secondary to spacers
- C. Recurrence after corticosteroid injection into trochlea
- D. Systemic complications of corticosteroids

VI. Describe disease-related complications

- A. Abnormal head posture
- B. Amblyopia
- C. Diplopia

VII. Describe appropriate patient instructions

- A. Bring any underlying systemic condition under control
- B. Appropriate amblyopia follow-up and compliance

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Helveston EM. The influence of superior oblique anatomy on function and treatment. The 1998 Bielschowsky Lecture. Binocul Vis Strabismus Q 1999;14:16-26.
3. Wright K. Brown's syndrome: diagnosis and management. Trans Am Ophthalmol Soc 1999;97:1023-1109.
4. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Vertical deviations due to orbital floor fracture (blowout fracture)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Orbital or facial trauma

B. Define the relevant aspects of epidemiology of the disease

1. Objects larger than the orbital opening impact the orbit
2. Motor vehicle accidents account for the majority

C. List the pertinent elements of the history

1. Head and facial trauma
2. Acquired diplopia following injury; may be vertical, oblique, torsional, or horizontal
3. Nausea and or vomiting

D. Describe pertinent clinical features

1. Ecchymosis of the involved eyelid
2. Epistaxis
3. Orbital emphysema
4. Depressed bridge of nose
5. Enophthalmos or proptosis
6. Possible reduced vision from globe or optic nerve injury
7. Possible diplopia in some or all positions of gaze immediately following injury
8. Diplopia may persist in vertical gaze secondary to entrapment of the inferior rectus muscle, inferior oblique muscle, or surrounding tissue
9. Diplopia may persist in horizontal gaze secondary to entrapment of the medial or lateral rectus muscle or surrounding tissue
10. Vomiting, especially in younger individuals with muscle entrapment
11. Bradycardia in children with muscle entrapment
12. Paresthesia or hypoesthesia in the infraorbital area secondary to damage to the infraorbital nerve
13. Nasolacrimal drainage injury with epiphora, lacrimal sac mucocoele, abscess or dacryostenosis
14. Rhinorrhea if orbital roof fracture and cerebrospinal fluid leakage
15. Traumatic ptosis
16. Possible tripod fracture involving zygomatic arch
17. Possible LeFort fracture
18. "White-eyed blowout fracture": marked restriction despite minimal soft tissue findings in the setting of orbital trauma

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan of orbits, direct axial and coronal
2. Magnetic resonance imaging (MRI) scan of orbit with gadolinium contrast, fat saturation technique
3. Forced duction testing can help differentiate restrictive entrapment from cranial nerve paresis

II. Define the risk factors

- A. Lack of proper restraint in automobile**
- B. Not using protective eyewear or headgear in sports-related injury**

III. List the differential diagnosis

A. Motility disturbance due to various types of injury to extraocular muscle

- 1. Entrapment
- 2. Restriction
- 3. Ischemia
- 4. Nerve damage and paralysis

B. Direct injury to muscle from bony fragment

C. Hypoglobus without strabismus

D. Cranial neuropathy leading to motility disturbance

E. Other forms of restrictive or paretic myopathy

- 1. Brown syndrome
- 2. Thyroid eye disease
- 3. Local anesthetic myotoxicity
- 4. Duane syndrome
- 5. Congenital fibrosis syndrome
- 6. Myasthenia gravis

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

A. Describe medical therapy options

- 1. Treat related globe and/or nerve injury
- 2. Rule out closed head or other systemic trauma
- 3. Systemic corticosteroids if significant orbital congestion
- 4. Antibiotic prophylaxis
- 5. Nasal decongestants
- 6. Prism therapy for diplopia

B. Describe surgical therapy options

- 1. Some clinicians advocate immediate exploration and repair of fracture, particularly if fracture is large or clinical/radiological evidence of entrapment
 - a. Prompt surgery if the entrapment is causing bradycardia and/or increased vagal tone, such as in the setting of "white-eyed blowout fracture"
- 2. Some clinicians recommend waiting 5 or more days until orbital edema and hematoma subside, and repair fracture only if persistent diplopia or risk of enophthalmos
- 3. Repair of residual strabismus by standard eye muscle surgical techniques once fracture repaired (if indicated) and/or entrapment relieved and no further spontaneous improvement is likely with stable measurements of motility deficits

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

- A. Persistent diplopia despite repair of fracture, and/or strabismus repair may require occluder or prisms to**

alleviate any residual diplopia

- B. Standard strabismus surgery-related complications (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe disease-related complications

- A. Globe or optic nerve related injury
- B. Orbital cellulitis
- C. Hypoglobus
- D. Enophthalmos

VII. Describe appropriate patient instructions

- A. Acutely, direct patients to not blow nose (thereby preventing orbital emphysema)
- B. Prophylactic systemic antibiotics may be warranted
- C. Counsel patient that residual diplopia may exist following orbital and strabismus repair

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Vertical deviations due to Duane syndrome (Congenital cranial dysinnervation disorders)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Developmental defect of cranial nerve nuclei with subsequent abnormal patterns of innervation of the eye muscles. This may lead to structural changes to the affected muscles, usually stiffening or contractures
2. High muscle tension caused by co-contraction of the medial and lateral rectus muscles
3. Vertical slippage of the tight lateral rectus muscle over or under the globe when the eye is adducted resulting in upshoots and/or downshoots. (See Esotropic (Type 1) Duane syndrome and Exotropic Duane syndrome)

B. List the pertinent elements of the history

1. Onset at birth, manifest with the onset of binocularity
2. Presence of anomalous head posture
3. Appearance of an eye shooting upward or downward or disappearing from view

C. Describe pertinent clinical features

1. Large angle hypertropia or hypotropia with attempted adduction
2. Cornea of affected eye may be covered by upper or lower eyelid
3. Occurs in association with other findings characteristic of Duane syndrome
 - a. Limitation of abduction or adduction or both
 - b. Globe retraction and narrowing of eyelid fissure on attempted adduction
 - c. Relative enophthalmos of affected eye

II. List the differential diagnosis

A. Upshoot

1. Inferior oblique overaction
2. Superior oblique palsy
3. Contralateral superior rectus palsy
4. Other neural misdirection syndromes

B. Downshoot

1. Brown syndrome
2. Superior oblique overaction
3. Inferior oblique palsy
4. Contralateral inferior rectus palsy
5. Other neural misdirection syndromes

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

A. Describe surgical therapy options

1. Recession of lateral rectus muscle
2. "Y-splitting" of lateral rectus muscle +/- recession
3. Posterior fixation suture of lateral rectus muscle +/- recession
4. Recession of medial and lateral rectus muscles

IV. List the complications of treatment, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

- A. Resection of an affected muscle may worsen globe retraction or lead to restrictive strabismus**

V. Describe disease-related complications

- A. Psychosocial stigma**
- B. Decreased binocular vision**
- C. Abnormal head position**

VI. Describe appropriate patient instructions

- A. Surgery is likely to improve, but may not eliminate, severe upshoot or downshoot (See Surgery of the extraocular muscles: weakening procedures)**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Third (oculomotor) cranial nerve palsy

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Congenital
2. Head trauma
3. Inflammatory process involving Cranial Nerve (CN) III
4. Infectious process involving CN III
5. Neoplasm
6. Migraine
7. Aneurysm
8. Microvascular insult (e.g., diabetes mellitus, hypertension)
9. Demyelinating disease

B. Define the relevant aspects of epidemiology of the disease

1. Neoplasm, trauma, and congenital are the most common causes in children
2. Microvascular CN III palsies generally resolve spontaneously

C. List the pertinent elements of the history

1. Onset
2. Head trauma
3. Antecedent infection
4. Headache
5. Other neurological symptoms
6. Diplopia
7. Diabetes mellitus/hypertension
8. History or family history of aneurysm

D. Describe pertinent clinical features

1. Incomitant exotropia
2. Incomitant hypotropia of affected eye
3. Deficient adduction, elevation and depression
4. Ptosis
5. Pupillary involvement possible (dilated/unreactive)
6. Incomplete palsy can occur - motility pattern depends on which muscles affected
7. Aberrant regeneration common (except in microvascular disease), affecting extraocular muscles, eyelid and/or pupil

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Neuroimaging in selected cases
2. Consider work-up for systemic illness
3. Work-up for aneurysm if involving pupil or incomplete palsy and not consistent with known etiology

II. Define the risk factors

- A. Previous infection
- B. Head trauma
- C. Neurological disorder (e.g., aneurysm, neoplasm)
- D. Microvascular disease (e.g., diabetes mellitus)

III. List the differential diagnosis

- A. Myasthenia gravis
- B. Restrictive orbitopathy
- C. Demyelinating disease
- D. Exotropic Duane syndrome
- E. Congenital fibrosis of the extraocular muscles (for congenital cases)

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

A. Describe medical therapy options

- 1. Patching for amblyopia
- 2. Monocular occlusion for diplopia (unnecessary with severe ptosis)
- 3. Fresnel (press on) prism
- 4. Botulinum toxin

B. Describe surgical therapy options

- 1. Large lateral rectus recession and medial rectus resection for exotropia +/- supraplacement of both muscles for hypotropia
- 2. Globe fixation nasally and lateral rectus disinsertion or fixation to periosteum
- 3. Superior oblique tenotomy or transposition of superior oblique to superior-nasal quadrant
- 4. Transposition procedures
- 5. Profound weakening procedure of lateral rectus muscle
- 6. Surgery on contralateral eye
- 7. For incomplete paralysis, surgical plan tailored
- 8. Repair of ptosis to prevent occlusion amblyopia

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

A. Inability to fully correct ductions and versions following strabismus repair.

B. Exposure keratopathy after ptosis repair

- 1. Assess Bell phenomenon preoperatively
- 2. Plan undercorrection of ptosis
- 3. Treat with corneal lubrication postoperatively (See Ocular complications of strabismus surgery, including consecutive deviations)
- 4. Discuss possible intractable diplopia

VI. Describe disease-related complications

- A. Occlusive and/or strabismic amblyopia
- B. Diplopia

VII. Describe appropriate patient instructions

- A. In many cases, adequate alignment in primary gaze and/or in slight downgaze is all that can be expected; diplopia will likely persist in other gaze positions**
- B. Despite treatment incapacitating diplopia may persist (See Surgery of the extraocular muscles: weakening procedures)**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Mudgil AV, Repka MX. Ophthalmologic outcome after third cranial nerve palsy in childhood. J AAPOS 1999;3:2-8.
3. Schumacher-Feero LA, Uyoo KW, Solari FM, et al. Third cranial nerve palsy in children. Am J Ophthalmol 1999;128:216-221.
4. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Sixth nerve (cranial nerve VI) palsy

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Inflammatory process involving CN VI
2. Infectious process involving Cranial Nerve (CN) VI
3. Intracranial mass lesions with compression or infiltration
4. Increased intracranial pressure
5. Head trauma
6. Microvascular insult

B. Define the relevant aspects of epidemiology of the disease

1. More common in childhood than infancy
2. Commonly associated with intracranial lesions
3. Vasculopathic or viral associated lesions resolve over several months

C. List the pertinent elements of the history

1. Diplopia worse at distance than near
2. Head turn toward the paretic CN VI
3. Past head trauma
4. Antecedent infection
5. Associated neurologic symptoms
6. Microvascular risk factors

D. Describe pertinent clinical features

1. Incomitant esodeviation
2. Esotropia increases toward the paretic lateral rectus
3. Slowed saccadic velocities of the affected lateral rectus
4. Versions show limited abduction of the affected eye
5. Anomalous head turn toward the paretic CN VI

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Neuroimaging in selected cases
2. Consider work-up for systemic illness
3. Consider testing for myasthenia gravis
4. Forced duction and forced generation testing to evaluate for restrictive etiology
5. Consider lumbar puncture
6. Coordinate care with primary care provider

II. Define the risk factors

A. Previous infection

B. Head trauma

C. Neurological disorder

- D. Microvascular disease

III. List the differential diagnosis

- A. Congenital esotropia
- B. Duane syndrome
- C. Möbius syndrome
- D. Thyroid eye disease
- E. Medial orbital wall fracture
- F. Cyclic esotropia
- G. Esotropia with medial rectus contracture
- H. Spasm of near reflex
- I. Myasthenia gravis

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

- A. Describe medical therapy options
 - 1. Patching for amblyopia
 - 2. Fresnel press-on prism, or prism ground into the eyeglass lenses
 - 3. Botulinum toxin treatment into the antagonist medial rectus
- B. Describe surgical therapy options
 - 1. Horizontal strabismus surgery when some abduction present
 - 2. Transposition strabismus surgery if poor abduction is present

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

- A. Anterior segment ischemia after transposition procedure (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe disease-related complications

- A. Diplopia
- B. Amblyopia
- C. Tight medial rectus if longstanding
- D. Abnormal head posture

VII. Describe appropriate patient instructions

- A. Follow up with primary care provider
- B. Amblyopia treatment compliance

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Foster RS. Vertical muscle transposition augmented with lateral fixation. J AAPOS 1997;1:20-30.
3. Holmes JM, Beck RW, Kip KE, et al. Botulinum toxin treatment versus conservative management in acute

traumatic sixth nerve palsy or paresis. J AAPOS 2000;4:145-149.

4. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Thyroid eye disease associated with strabismus

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Probably results from an autoimmune myopathy
2. Patients can be hyperthyroid, euthyroid, or hypothyroid at the time of diagnosis
3. Lymphocytic infiltration of extraocular muscles and orbital fat results in edema, inflammation, and fibrosis
4. Smoking aggravates orbital inflammation

B. Define the relevant aspects of epidemiology of this disease

1. Most common cause of acquired vertical deviation in adults
2. Females >> males
3. Bilateral, but may be asymmetric

C. List the pertinent elements of the history

1. History of thyroid dysfunction, including onset and treatment with thyroid ablation and/or medications
2. Diplopia, including type, duration and stability
3. Prior ocular or orbital surgery
4. Tobacco use
5. Associated myasthenia gravis
6. Possible visual loss
7. Possible dry eye symptoms
8. Motility problems can precede known medical diagnosis

D. Describe pertinent clinical features

1. Typically, an active "congestive" phase followed by a quiescent "fibrotic" phase
2. Affected muscles (decreasing order of frequency)
 - a. Inferior rectus
 - b. Medial rectus
 - c. Superior rectus
 - d. Lateral rectus
 - e. Obliques may rarely be affected
3. Combined esotropia and vertical deviation is common presentation
4. Limitation (often severe) of ductions and versions of one or both eyes
5. Forced ductions abnormal in one or more directions
6. Enlarged extraocular muscles and/or orbital engorgement may cause compressive optic neuropathy
7. Proptosis
8. Eyelid retraction
9. Lid lag in downgaze
10. Ophthalmopathy may be markedly asymmetric
11. Exacerbation possible with radioactive iodide treatment

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Clinical history and exam often sufficient
2. Enlargement of muscle bellies with sparing of tendinous insertions demonstrated on orbital computed tomography (CT), magnetic resonance imaging (MRI), or ultrasound
3. Thyroid function tests or referral for evaluation if not previously done or if new diagnosis
4. If myasthenia gravis suspected, edrophonium (Tensilon) test, serologic evaluation (acetylcholine receptor antibodies) or EMG (See Myasthenia gravis (associated with strabismus))

II. Define the risk factors

- A. History of thyroid dysfunction
- B. Female sex
- C. Smoking
- D. Myasthenia gravis

III. List the differential diagnosis

- A. Orbital myositis
- B. Myasthenia gravis
- C. Chronic progressive external ophthalmoplegia
- D. Acquired cranial nerve palsy
- E. Orbital tumor
- F. Carotid cavernous or dural cavernous fistula
- G. Previous orbital fracture with restrictive strabismus

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

A. Describe medical therapy options

1. Fresnel prisms sometimes useful for small deviation and as temporizing measure while waiting for stable alignment
2. Ground-in prism if deviation stable, small, relatively comitant, and no significant torsion
3. Monocular blurring or occlusion

B. Describe surgical therapy options

1. Surgery indicated for diplopia or abnormal head posture
2. Strabismus surgery usually done after orbital decompression (if needed) and before eyelid surgery
3. Stability of alignment before surgery is desirable
4. Recession of restricted muscles
5. Inferior rectus recessions
 - a. Slight initial undercorrection is desirable because of tendency for late overcorrection
6. Resection rarely done
 - a. May worsen restriction

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

- A. A-pattern strabismus after bilateral inferior rectus recession

- B. Late overcorrection especially with inferior rectus recession**
- C. Lower eyelid retraction after inferior rectus recession**
 - 1. Can be minimized by dissection between inferior rectus muscle and lower eyelid retractors
- D. Severe postoperative inflammation**
 - 1. Avoid surgery when significant inflammation present
 - 2. Consider perioperative systemic corticosteroids (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe disease-related complications

- A. Compressive optic neuropathy**
- B. Severe proptosis**
- C. Exposure keratopathy**
- D. Diplopia**
- E. Eyelid retraction**
- F. Ocular hypertension/glaucoma**

VII. Describe appropriate patient instructions

- A. The goal of surgery is to restore single binocular vision in primary and/or downgaze, but diplopia in other positions of gaze usually persists**
- B. More than one strabismus surgery often required**
- C. Lower eyelid retraction often worsens after inferior rectus recession (See Surgery of the extraocular muscles: weakening procedures)**
- D. Assessment for appropriateness of adjustable suture (See Surgery of the extraocular muscles: adjustable sutures)**

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. Coats DK, Paysse EA, Plager DA, et al. Early strabismus surgery for thyroid ophthalmopathy. *Ophthalmol* 1999;106:324-329.
- 3. Meyer DR, Simon JW, Kansora J. Primary infratarsal lower eyelid retractor lysis to prevent eyelid retraction following inferior rectus recession. *Am J Ophthalmol* 1996;122:331-339.
- 4. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Myasthenia gravis (associated with strabismus)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Autoimmune phenomenon
 - a. Acetylcholine receptor sites for neuromuscular transmission are blocked by immune complexes

B. Define the relevant aspects of epidemiology of this disease

1. Can occur at any age
2. Uncommon in children

C. List the pertinent elements of the history

1. Drooping of eyelid
2. Double vision, including type, duration and stability
3. Subacute, insidious onset
4. Variability of symptoms
5. Symptoms increased with fatigue
6. Symptoms improve after sleep/rest

D. Describe pertinent clinical features

1. Can be purely ocular or systemic
2. Ptosis due to weakness of levator muscle
3. Reduced ocular motility due to extraocular muscle weakness
4. Vertical and/or horizontal strabismus
5. Hallmarks of disease are fluctuation and fatigability, resulting in variability of ptosis and/or alignment and motility
6. Ptosis increases after prolonged upgaze
7. Cogan lid twitch
 - a. Overshoot of eyelid when returning to primary gaze from downgaze
8. Associated with thymoma

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Sleep test
 - a. Assess for improvement in ptosis and/or alignment and motility after 20-30 minutes with eyelids closed
2. Ice test
 - a. Assess for improvement in ptosis and/or alignment and motility after external application of ice for a few minutes
 - i. Lack of improvement with ice test does not mean that the patient does not have ocular myasthenia gravis
3. Systemic anticholinesterase inhibitors
 - a. Tensilon (edrophonium) is given intravenously and the patient is observed for change in ptosis and/or alignment/motility
4. Acetylcholine receptor antibodies in serum

- a. Negative titers do not rule out disease, since sensitivity is 90% for systemic disease and only 50% for purely ocular disease
- 5. Chest CT or MRI to rule out thymoma
- 6. EMG - decremental response to repetitive nerve stimulation
- 7. Evaluation of possible concomitant thyroid involvement
 - a. Thyroid function tests
 - b. Antithyroglobulin antibodies

II. Define the risk factors

- A. Association with thyroid eye disease

III. List the differential diagnosis

- A. Chronic progressive external ophthalmoplegia
- B. Acquired cranial nerve palsy
- C. Internuclear ophthalmoplegia
- D. Thyroid eye disease
- E. Other restrictive orbitopathy i.e. fracture, intraorbital tumor

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

- A. Define medical therapy options
 - 1. Systemic treatment in conjunction with neurologist
 - 2. Prisms sometimes useful
 - 3. Monocular occlusion
- B. Define surgical therapy options
 - 1. Strabismus surgery for long-standing, stable deviations unresponsive to medical therapy
- C. Consider thymectomy in conjunction with neuromuscular specialist

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

- A. Side effects of systemic medications
- B. Complications of strabismus surgery if performed

VI. Describe disease-related complications

- A. Ptosis
- B. Diplopia
- C. Conversion of purely ocular disease to systemic form

VII. Describe appropriate patient instructions

- A. Systemic treatment usually necessary
- B. Counseling that purely ocular disease can become systemic

C. Limited success in relieving diplopia with prisms if variability present

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Ellis FD, Hoyt CS, Ellis FJ, et al. Extraocular muscle responses to orbital cooling (ice test) for ocular myasthenia gravis diagnosis. J AAPOS 2000;4:271-281.
3. Golnik KC, Pena R, Lee AG, et al. An ice test for the diagnosis of myasthenia gravis. Ophthalmol 1999;106:1282-1286.
4. Mullaney P, Vajsar J, Smith R, et al. The natural history and ophthalmic involvement in childhood myasthenia gravis at the Hospital for Sick Children. Ophthalmol 2000;107:504-510.
5. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Strabismus following ocular surgery

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Ocular misalignment with or without diplopia
 - a. Scarring of fascial tissues
 - b. Mechanical restriction due to implanted material (e.g., scleral buckle or glaucoma drainage device)
 - c. Direct injury or anesthetic toxicity to extraocular muscle(s)
 - d. Disruption of fusion due to dissimilar sensory input between the two eyes (e.g., monovision, aniseikonia, epiretinal membrane)
 - e. Uncovering or worsening of preexisting strabismus (e.g., decompensated monofixation syndrome)
 - f. Central fusional disruption precipitated by prolonged monocular occlusion (e.g., after cataract surgery)
 - g. Induced prismatic effect in the reading position with eyeglass lenses for anisometropia/anisoastigmatism
 - h. Fixation switch diplopia-preference of previously non-dominant eye
 - i. Changes in accommodation caused by overcorrection during refractive surgery in a phakic patient
2. Rule out monocular diplopia
 - a. Corneal irregularity
 - b. Dislocated intraocular lens
 - c. Tear film disturbance
 - d. Epiretinal membrane

B. List the pertinent elements of the history

1. Characteristics of double vision
 - a. Monocular vs. binocular
 - b. Vertical, horizontal, and/or torsional
 - c. Duration
 - d. Intermittent versus constant
 - e. Onset - sudden or gradual
2. History of double vision, eye misalignment, or amblyopia prior to surgery
3. Previous ocular surgeries, including use of extraocular implants and type of anesthesia (retrobulbar vs. topical)

C. Describe pertinent clinical features

1. Vertical, horizontal, and/or torsional strabismus
2. May be incomitant
3. Frequently limitation of ductions due to mechanical restriction
4. After retrobulbar or peribulbar injection, most common presentation is hypotropia of affected eye with restricted elevation
 - a. Affected muscle often initially paretic with hypertropia, later becomes fibrotic with hypotropia
 - b. History of reversing deviation

II. Define the risk factors

- A. History of strabismus or amblyopia**
- B. History of surgery involving placement of extraocular implant or macular translocation**
- C. Retrobulbar or peribulbar injection**
- D. Prolonged disruption of fusion**
- E. Multiple surgeries**

III. List the differential diagnosis

- A. Preexisting strabismus**
- B. Thyroid eye disease**
- C. Myasthenia gravis**
- D. Acquired cranial nerve palsy**

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

A. Define medical therapy options

1. Prisms sometimes useful for small deviations or as temporizing measure while waiting for stable alignment, but prisms often unsatisfactory due to incomitance
2. Monocular occlusion
3. Observation alone if no diplopia in common positions of gaze

B. Define surgical therapy options

1. Stability of alignment for several months before surgery is desirable
2. Recession of restricted muscles
3. Excision/dissection of scar tissue
4. Adjustable suture technique often useful due to unpredictability of outcome
5. Address torsion if present
6. Removal of scleral buckle or glaucoma drainage device sometimes necessary after consultation with appropriate subspecialist but may not change strabismus pattern

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

- A. Overcorrection/undercorrection after strabismus surgery/persistent diplopia**
- B. Scarring/redness of conjunctiva due to multiple surgeries**
- C. Inadequate control of glaucoma if drainage device removed (See Ocular complications of strabismus surgery, including consecutive deviations)**

VI. Describe appropriate patient instructions

- A. Goal of treatment is to restore single vision in primary and/or downgaze, but double vision in other positions of gaze often persists**
- B. More than one strabismus surgery may be required (See Surgery of the extraocular muscles: weakening procedures)**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Hamed LM. Strabismus presenting after cataract surgery. *Ophthalmol* 1991;98:247-52.
3. Hamilton SM, Elsas FJ, Dawson TL. A cluster of patients with inferior rectus restriction following local anesthesia for cataract surgery. *J of Pediatric Ophthal & Strabismus* 1993;30:288-91.
4. Capo H, Roth E, Johnson T, et al. Vertical strabismus after cataract surgery. *Ophthalmol* 1996;103:918-21.
5. Capo H, Guyton DL. Ipsilateral hypertropia after cataract surgery. *Ophthalmol* 1996;103:721-30.
6. Cadera W. Diplopia after peribulbar anesthesia for cataract surgery. *J of Pediatric Ophthal & Strabismus* 1998;35:240-1.
7. von Noorden GK. *Binocular Vision and Ocular Motility: Theory and Management of Strabismus*. 6th edition St. Louis: Mosby, 2002.
8. AAO, *Ophthalmology Monographs* 17. *Strabismus Surgery: Basic and Advanced Strategies*, 2004.

Treatment of strabismus: eyeglasses, including bifocals

I. List the indications/contraindications

A. Indications

1. Refractive accommodative esotropia
2. Nonrefractive accommodative esotropia (high accommodative convergence to accommodation ratio)
3. Partially accommodative esotropia
4. Exotropia (with or without overminusing)
5. Acquired strabismus with symptomatic diplopia requiring prism
6. Sensory strabismus requiring polycarbonate lenses (See Medical treatment of amblyopia: refractive correction)

II. Describe the pre-procedure/therapy evaluation

A. History of eyeglasses wear

B. Visual acuity

C. Cycloplegic Refraction

1. Consider full hyperopic prescription for suspected accommodative esotrope
2. Consider over minus in poorly controlled intermittent exotropes to promote accommodative convergence; prescribe if high hyperopia in intermittent exotropes
3. For adult with strabismus and significant refractive error on cycloplegic, consider post-cycloplegic refraction

D. Ocular alignment at distance and near fixation

E. If diplopia, determination of prism required for single vision at distance and near fixation

III. List the alternatives to this procedure/therapy

A. Alternative to eyeglasses for nonrefractive accommodative esotropia

1. Phospholine iodide
 - a. Consider using phenylephrine 2.5% to prevent iris cysts

B. Alternatives to prism eyeglasses for diplopia

1. Extraocular muscle surgery
2. Botulinum toxin
3. Occlusion or intentional blurring of one eye
 - a. Scotch tape
 - b. Bangerter foil
4. Orthoptics to increase convergence fusional amplitudes
5. No treatment if patient ignores second image

IV. Describe the instrumentation, anesthesia and technique

- A. Single vision eyeglasses for refractive accommodative esotropia**
- B. Bifocal eyeglasses for nonrefractive accommodative esotropia**
- C. Prism eyeglasses**
 - 1. Small amounts of prism can be ground into lenses or prism effect can be created by lens decentration
 - 2. Fresnel prisms useful for
 - a. Large deviations
 - b. Large distance-near disparity
 - c. Variable strabismus for example, in resolving cranial nerve palsy
 - d. Prism adaptation as a trial prior to surgery or prior to grinding prism into lenses
 - e. Patients with diplopia who are poor surgical candidates

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

- A. Poor compliance with prescribed glasses**
- B. Diplopia not relieved**
 - 1. Reevaluate to determine if different strength of prism required
 - 2. Consider distance-near disparity, incomitance, and/or presence of torsion as obstacles to fusion
 - 3. Consider surgery or Botox

VI. Describe the follow-up care

- A. Check eyeglasses on lensometer to insure proper prescription**
- B. Assessment of visual acuity and ocular alignment at each visit**
- C. Reduction of hyperopic power may be possible over time in some cases of accommodative esotropia**

VII. Describe appropriate patient instructions

- A. Eyeglasses with ground-in prism will be heavier and more expensive than standard eyeglasses**
- B. Fresnel prisms blur vision and have noticeable lines**
- C. For poor monocular vision, polycarbonate lenses should be worn at all times**
- D. (See Refractive accommodative esotropia) (See Nonrefractive accommodative esotropia (high AC/A ratio))**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. Ingram RM, Gill LE, Lambert TW. Reduction of astigmatism after infancy in children who did and did not wear glasses and have strabismus. *Strabismus* 2001;9:129-35.
- 3. Ingram RM, Gill LE, Lambert TW. Effect of eyeglasses on changes of spherical hypermetropia in infants who did and did not have strabismus. *Br J Ophthalmol* 2000;84:324-6.
- 4. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Surgery of the extraocular muscles: weakening procedures

I. List the indications/contraindications

A. Indications

1. Restore normal alignment
2. Elimination of diplopia
3. Promotion of stereopsis and binocularity
4. Functional improvement of appearance
5. Relief of asthenopia
6. Improvement of anomalous head posture
7. Improvement of visual acuity (e.g., nystagmus)
8. Expand binocular visual field if esotropic

B. Contraindications

1. Contraindications to anesthesia (e.g. high risk medical status)
2. Recent onset paresis with likelihood of improvement
3. Fully accommodative esotropia
4. Patient with unrealistic expectations

II. Describe the pre-procedure/therapy evaluation

A. History

1. Prior surgery to extraocular muscles, including scleral buckling or glaucoma implant
2. Patient/parental complaints, including presence or absence of diplopia
3. History or family history of complications of anesthesia

B. Complete ocular examination

C. Complete motility evaluation

1. Assessment of ductions and versions
2. Prism and alternate cover test
 - a. In proper refractive correction
 - b. In primary gaze at distance and near
 - c. In gaze directions - at least left, right, up and down gaze
3. For vertical deviations, also include (if attainable)
 - a. Alignment in all 9 cardinal gaze positions (as appropriate and attainable)
 - b. Head tilt test
 - c. Assessment of torsion with Double Maddox Rod and/or by indirect ophthalmoscopy
4. Sensory testing (particularly important in long-standing strabismus cases possibly associated with anomalous retinal correspondence)
 - a. Fusion/suppression/diplopia (e.g., Worth 4-dot)
 - b. Sensory response with prism correcting for objective angle in free space is useful for determining

c. Stereopsis testing (Randot or Titmus for example)

III. List the alternatives to this procedure/therapy

A. Strengthening procedure of an antagonist or yoke muscle

B. Botulinum toxin

C. Prisms

D. Occlusion

E. Observation

IV. Describe the instrumentation, anesthesia and technique

A. Instrumentation

1. Standard strabismus surgery instruments with spatulated needle for sclera

B. Anesthesia

1. General

2. Local

a. Retrobulbar

b. Peribulbar

c. Local infiltrative

3. Topical anesthetic drops

C. Techniques

1. Recession - removal and reattachment of a muscle posteriorly (closer to its origin)

2. Posterior fixation suture (Faden operation)

a. Used to weaken a muscle selectively in its field of action

b. Attachment of a rectus muscle to sclera very posterior to its insertion using a nonabsorbable suture

3. Myotomy/tenotomy - cutting across a muscle/tendon

4. Marginal myotomy - cutting partially across a muscle

5. Myectomy/tenectomy - removing a portion of a muscle/tendon without marginal reattachment

6. Conjunctiva incised for above procedures using fornix or limbal approach

7. Tendon lengthening procedure

8. Recession and resection of the same muscle

a. Weakens muscle in its field of action similar to posterior fixation suture

9. Adjustable suture

10. Fixed suture

11. Hangback suture

V. List the complications of the procedure/therapy, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe the follow-up care

- A. Usually outpatient procedure**
- B. Appropriate follow-up to assess for infection and other surgical complications**

VII. Describe appropriate patient instructions

- A. Compliance with eyedrops if prescribed (1/3 of pediatric ophthalmologists use nothing, 1/3 use antibiotic alone and 1/3 use antibiotic/steroid combo for several days)**
- B. Avoid rubbing eyes**
- C. No swimming in immediate postoperative period**
- D. Mild redness, swelling, and discharge are expected**
- E. Call if signs of infection, such as increasing redness, purulent discharge, severe pain and/or vision loss**
- F. Call if nausea/vomiting persist**
- G. Call if respiratory problem or fever**
- H. Call if sudden shift in eye position**
- I. Result of surgery may not be known for several weeks**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Surgery of the extraocular muscles: strengthening procedures

I. List the indications/contraindications

A. Indications

1. Restore normal alignment
2. Elimination of diplopia
3. Promotion of stereopsis and binocularity (e.g., infantile esotropia)
4. Functional improvement of appearance
5. Relief of asthenopia
6. Improvement of anomalous head posture

B. Contraindications

1. Restrictive strabismus (e.g., thyroid eye disease but this is a relative contraindication)
2. Co-contraction of muscles (Duane syndrome)
3. Contraindications to anesthesia (e.g., malignant hyperthermia, high risk medical status)
4. Recent onset paresis with likelihood of improvement
5. Fully accommodative esotropia
6. Patient with unrealistic expectations

II. Describe the pre-procedure/therapy evaluation

A. History

1. Prior surgery to extraocular muscles, including scleral buckling or glaucoma implant
2. Patient/parental complaints, including presence or absence of diplopia
3. History or family history of complications of anesthesia

B. Comprehensive eye examination

C. Complete motility evaluation

1. Assessment of ductions and versions
2. Alternate prism and cover test
 - a. In proper refractive correction
 - b. In primary gaze at distance and near
 - c. In gaze directions - at least left, right, up and down gaze
3. For vertical deviations, also include
 - a. Alignment in all 9 cardinal gaze positions if attainable
 - b. Head tilt test
 - c. Assessment of torsion with Double Maddox Rod if attainable
4. Sensory testing (particularly important in long-standing strabismus cases possibly associated with anomalous retinal correspondence)
 - a. Fusion/suppression/diplopia (e.g. Worth 4-dot)
 - b. Sensory response with prism correcting for objective angle in free space is useful for determining

c. Stereopsis (Randot or Titmus test for example)

III. List the alternatives to this procedure/therapy

A. Weakening procedure of an antagonist or yoke muscle

B. Botulinum toxin to an antagonist or yoke muscle

C. Prisms

D. Occlusion

E. Observation

IV. Describe the instrumentation, anesthesia and technique

A. Instrumentation

1. Standard strabismus surgery instruments with spatulated needle for sclera

B. Anesthesia

1. General

2. Local

a. Retrobulbar

b. Peribulbar

c. Local infiltrative

3. Topical anesthetic drops

C. Techniques

1. Conjunctiva incised for above procedures using fornix or limbal approach

2. Resection

- a. Absorbable sutures are placed at a predetermined distance posterior to the muscle insertion

b. Muscle segment anterior to the sutures in excised

c. Shortened muscle is reattached to the globe at the insertion

3. Advancement

a. Previously recessed rectus muscle removed and advanced to or closer to its original insertion

4. Resection and advancement

5. Tuck (of superior oblique tendon)

a. Tendon folded on itself

6. Transposition procedures

a. Vertical rectus muscle transposition to lateral rectus muscle

- i. Full tendon transposition

i) With posterior fixation suture (Foster modification)

ii) Without posterior fixation suture

- ii. Partial tendon transposition (Hummelsheim)

b. Horizontal rectus muscle transposition to superior rectus muscle (Knapp procedure)

7. Adjustable vs. fixed suture

8. Plication of rectus muscle

V. List the complications of the procedure/therapy, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe the follow-up care

- A. Usually outpatient procedure
- B. Appropriate follow-up to assess for infection and other surgical complications

VII. Describe appropriate patient instructions

- A. Compliance with drops if prescribed (1/3 of pediatric ophthalmologists use nothing, 1/3 use antibiotic alone and 1/3 use antibiotic/steroid combo for several days)
- B. Avoid rubbing eyes
- C. No swimming in immediate postoperative period
- D. Mild redness, swelling, and discharge are expected
- E. Call if signs of infection, such as increasing redness, purulent discharge, severe pain and/or vision loss
- F. Call if nausea/vomiting persist
- G. Call if respiratory problem or fever
- H. Call if sudden shift in eye position
- I. Result of surgery will not be known for several weeks

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Surgery of the extraocular muscles: adjustable suture techniques

I. List the indications/contraindications

A. Indications

1. Used in selected patients to adjust postoperative alignment (e.g.: patients with diplopia, thyroid ophthalmopathy, restrictive strabismus, incomitant strabismus among others)

B. Contraindications

1. Patient who is unwilling or unable to cooperate for surgeon's chosen method of suture adjustment

II. Describe the pre-procedure/therapy evaluation

- ### A. Assessment of patient's willingness and ability to cooperate for suture adjustment (See Surgery of the extraocular muscles: weakening procedures)

III. List the alternatives to this procedure/therapy

A. Extraocular muscle surgery without suture adjustment

B. Nonsurgical alternatives

1. Botulinum toxin
2. Prisms
3. Occlusion
4. Observation

IV. Describe the instrumentation, anesthesia and technique

A. Instrumentation

1. Standard strabismus surgery instruments

B. Techniques (including anesthesia)

1. Postoperative adjustment
 - a. Surgery completed using standard anesthesia
 - b. Sutures and knots externalized to allow alteration of muscle position during postoperative period
 - c. Allow adequate time for anesthetic effects to subside
2. Operation/reoperation
 - a. Strabismus surgery completed using short-acting or reversible anesthetic agents
 - b. Technique
 - i. Short tag or long tag noose knot
 - ii. Bow-tie
 - c. Patient examined shortly after surgery
 - d. Reanesthetized for suture adjustment
 - i. Topical anesthesia

- i) Immediate or delayed
- ii. Sedated suture adjustment in recovery room or operating room

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

- A. Awake or inadequately anesthetized patient moves during critical part of procedure**
 - 1. Prevent by ensuring adequate anesthesia
- B. Patient unable to cooperate for postoperative suture adjustment**
 - 1. Minimize risk by selecting patients carefully for this technique
 - 2. Management
 - a. Sedation or additional anesthesia may be required
- C. Patient experiences vagal/oculocardiac reflex during postoperative suture adjustment - management**
 - 1. Discontinue suture adjustment
 - 2. Place patient in supine position
 - 3. Ensure adequate airway and circulation
- D. Complications of anesthesia (See Ocular complications of strabismus surgery, including consecutive deviations)**

VI. Describe the follow-up care

- A. Usually outpatient procedure**
- B. Appropriate follow-up to assess for infection and other surgical complications**

VII. Describe appropriate patient instructions

- A. Preoperative**
 - 1. Description of adjustable suture technique
 - 2. Advantages and disadvantages of using an adjustable suture
 - 3. Surgery without adjustable suture is an acceptable alternative
 - 4. If postoperative suture adjustment is planned
 - a. Procedure usually lasts a few minutes
 - b. Topical anesthesia used to minimize discomfort
 - c. Patient and surgeon must be reasonably certain that patient can cooperate for procedure
- B. Postoperative**
 - 1. Foreign body sensation is expected
 - 2. Even if ocular alignment is excellent after suture adjustment, final result of surgery will not be known for several weeks (See Surgery of the extraocular muscles: weakening procedures)

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. Buffenn AN. Adjustable suture use in strabismus surgery. *Curr Opin Ophthalmol* 2005;16:294-7.
- 3. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Anesthesia for strabismus surgery and complications, including malignant hyperthermia

I. List the indications/contraindications

A. Indications

1. General anesthesia
 - a. Incisional surgery
 - b. Chemodenervation such as Botox
 - c. Examination and ancillary testing in children or developmentally challenged adults unable to cooperate in the office
2. Topical anesthesia
 - a. Primary strabismus surgery
 - i. Requires highly cooperative patient
 - ii. Monitored anesthesia care with possible sedation
3. Subconjunctival, sub Tenon, peribulbar or retrobulbar anesthesia
 - a. Cooperative patients
 - b. Patients with risks too high for general anesthesia

B. Contraindications

1. General anesthesia
 - a. Patients with medical conditions that pose undue risk under general anesthesia
2. Topical anesthesia
 - a. Uncooperative or unwilling patients
 - b. Reoperation or multiple muscle strabismus surgery entailing excessive tugging on the extraocular muscle(s)
3. Local infiltration and retrobulbar anesthesia
 - a. Uncooperative or unwilling patients
 - b. Reoperation or multiple muscle surgery entailing excessive tugging on the extraocular muscle(s)
 - c. Desire for immediate postoperative adjustment of muscle position (some surgeons postpone adjustments for at least a half-day because of residual anesthetic effect)

II. Describe the pre-therapy evaluation

- A. Complete physical examination for evaluation of airway, general health, anesthetic risk factors and risk of malignant hyperthermia (usually performed by family physician and/or anesthesia personnel)
- B. Discussion with patient and family of different anesthetic choices and determination of most appropriate form of anesthesia for the situation

III. Describe the instrumentation and technique

- A. Outpatient setting

1. Anesthesia may be administered in an inpatient or day-surgery facility
2. Anesthesia set-up as per anesthesia department or facility
 - a. Age-appropriate equipment
 - b. Malignant hyperthermia cart

B. Intraoperatively

1. Local infiltration, retrobulbar and topical anesthesia are usually administered by the ophthalmologist
2. Anesthesia personnel may use sedative, amnestic and anti-nausea medication to augment
3. General anesthesia is administered by anesthesia personnel

IV. List the complications of this therapy, their prevention and management

A. Postoperative nausea and vomiting (PONV)

1. Intraoperative
 - a. Choose inhalational anesthetic agent to minimize
 - b. Prophylactic anti-emetics
2. Post-op
 - a. Anti-emetics
 - b. Avoid narcotics

B. Oculocardiac reflex

1. Discontinue stimulus
 - a. Release traction on extraocular muscle
2. Intervention
 - a. IV administration of atropine or glycopyrrolate

C. Complications related to underlying medical conditions

1. Risk assessment
 - a. Pre-operative medical clearance

D. Malignant hyperthermia

1. Careful family history for family members with the diagnosis of malignant hyperthermia or unexplained deaths under anesthesia in family members
2. Screen for disorders associated with malignant hyperthermia
 - a. Strabismus
 - b. Ptosis
 - c. Myopathies
3. Avoid triggering agents in susceptible individuals
 - a. Inhalational anesthetic agents
 - b. Succinylcholine
 - c. Local anesthetics of the amide type
4. Recognize early signs of malignant hyperthermia under anesthesia:
 - a. Tachyarrhythmias
 - b. Increased end-tidal carbon dioxide
5. Recognize later signs of malignant hyperthermia under anesthesia
 - a. Temperature rise

- b. Muscular rigidity
 - 6. Once recognized, malignant hyperthermia should be expeditiously treated by standard protocol including
 - a. Discontinuing anesthetic agents
 - b. Hyperventilating with oxygen
 - c. Beginning dantrolene
- E. **Concurrent medications, such as Phospholine Iodide and Succinylcholine, need to be discussed with the anesthesia care team**

V. Describe the follow-up care

- A. **Postoperative nausea and vomiting**
 - 1. Supportive care
 - 2. Antiemetics
- B. **Medical conditions that complicate anesthesia**
 - 1. Refer to primary care provider for management
- C. **Malignant hyperthermia**
 - 1. Consider MedicAlert bracelet
 - 2. Needs to be documented in chart
 - 3. Rarely perform muscle biopsy in family members
 - a. Non triggering protocol is used by anesthesia in all family members at risk for malignant hyperthermia

VI. Describe appropriate patient instructions

- A. **Postoperative nausea and vomiting**
 - 1. Inform patient of anesthetic agents used during their surgery so they can make choices of agents in consultation with anesthesia personnel in future surgeries
- B. **Malignant hyperthermia**
 - 1. Emphasize importance of genetic counseling regarding risks of malignant hyperthermia
 - 2. Caution regarding future anesthetics
 - 3. Medic-alert bracelet

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. Allison CE, DeLange JJ, Koole FD, et al. A comparison of the incidence of the oculocardiac and oculo-respiratory reflexes during sevoflurane or halothane anesthesia for strabismus surgery in children. *Anesth Analg.* 2000;90:306-10.

Ocular complications of strabismus surgery, including consecutive deviations

I. Describe the ocular complications of strabismus surgery

- A. Unsatisfactory alignment (e.g. residual or consecutive deviations)
- B. Refractive changes
- C. Diplopia
- D. Perforation of the sclera
- E. Postoperative infections
- F. Foreign body granuloma
- G. Allergic reaction
- H. Conjunctival inclusion cyst
- I. Conjunctival scarring
- J. Fat adherence syndrome
- K. Dellen
- L. Anterior segment ischemia
- M. Change in eyelid position
- N. Lost muscle
- O. Slipped muscle
- P. Ruptured muscle
- Q. Loss of vision (incidence of 1:5000 or less)

II. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

- 1. Perforation of the sclera
 - a. Inadvertent perforation of the sclera with a suture pass
- 2. Postoperative infection (endophthalmitis, preseptal cellulitis, orbital cellulitis)
 - a. Entrance of bacteria into eye after scleral perforation, or into the orbit and/or preseptal space from the cul-de-sac or periorbita
- 3. Allergic reaction
 - a. Hypersensitivity reaction to suture material or eye drops
- 4. Conjunctival inclusion cyst
 - a. Buried conjunctival epithelium
- 5. Fat adherence syndrome
 - a. Violation of Tenons with prolapse of orbital fat
- 6. Dellen
 - a. Raised conjunctiva prevents adequate resurfacing of the cornea with tears during blinking

7. Anterior segment ischemia
 - a. Interruption of blood supply to anterior segment by surgery on multiple rectus muscles
8. Lost muscle
 - a. Muscle slips out of sutures or surgical instruments
9. Ruptured muscle
 - a. Tight muscle tears while on a muscle hook
10. Slipped muscle
 - a. Muscle inadequately sutured to sclera
 - b. Muscle capsule secured with suture, but muscle slips within capsule post-operatively

B. Define the relevant aspects of epidemiology of the disease

1. Vision loss is possible, but extremely rare
2. Unsatisfactory alignment and need for reoperation are common
3. Risk of diplopia depends on preoperative sensory status

C. List the pertinent elements of the history

1. Diplopia
 - a. Onset
 - b. Frequency
 - c. Type - horizontal, vertical, oblique, torsional
2. Infection
 - a. Redness
 - b. Purulent discharge
 - c. Severe pain
 - d. Vision loss
 - e. Lethargy/fever
3. Granuloma/cyst
 - a. Foreign body sensation
4. Allergic reaction
 - a. Severe itching
 - b. Redness
 - c. Swelling
5. Anterior segment ischemia
 - a. Reduced vision
 - b. Cloudiness of cornea
 - c. Redness
 - d. Change in pupillary size and/or shape
6. Slipped muscle
 - a. Sudden change in eye position
 - b. May be a delay in onset, with consecutive deviation and increasing underaction in field of slipping/slipped muscle

D. Describe pertinent clinical features

1. Unsatisfactory alignment
 - a. Undercorrection

- b. Overcorrection (i.e. consecutive exotropia after surgery for esotropia)
 - c. New deviation (e.g., vertical)
 - d. Early postoperative alignment may not be permanent
- 2. Refractive changes
 - a. Induced astigmatism usually resolves within several months
- 3. Diplopia
 - a. Can occur in patients with or without preoperative diplopia
 - b. Various responses can occur hours to several months later
 - i. Fusion
 - ii. Development of a new suppression scotoma
 - iii. Persistence of diplopia
- 4. Perforation of the sclera
 - a. Usually creates no problem except chorioretinal scar
 - b. Possible complications
 - i. Vitreous hemorrhage
 - ii. Choroidal hemorrhage
 - iii. Retinal detachment
 - iv. Endophthalmitis
- 5. Postoperative infections
 - a. Mild conjunctivitis
 - b. Preseptal/orbital cellulitis rare
 - c. Endophthalmitis
- 6. Foreign body granuloma
 - a. Develops several weeks after surgery, often at suture site
 - b. Localized, elevated, slightly hyperemic mass
- 7. Allergic reaction
 - a. Conjunctival redness and itching
 - b. Secondary to suture material or eye drops
- 8. Conjunctival inclusion cyst
 - a. Translucent subconjunctival mass
 - b. Appears days to years after surgery
 - c. May disappear spontaneously
- 9. Fat adherence syndrome
 - a. May restrict motility
 - b. Incomitant vertical deviation common
 - c. Recognized rent in Tenon should be closed to prevent it
- 10. Dellen
 - a. Corneal thinning just anterior to limbus
- 11. Anterior segment ischemia
 - a. Reduced visual acuity
 - b. Cataract

- c. Corneal epithelial edema
 - d. Folds in Descemet membrane
 - e. Anterior chamber cell and flare
 - f. Change in pupillary size and/or shape
12. Change in eyelid position
 - a. Resecting vertical rectus muscle pulls eyelid forward
 - b. Recessing vertical rectus muscle retracts eyelid
 - c. Minimized by careful dissection of fascial connections as far posteriorly as possible
 13. Ruptured muscle
 - a. Muscle tears intraoperatively
 - b. If edges cannot be reapposed it can lead to duction limitation and worsening of strabismus
 14. Slipped muscle
 - a. Muscle slips within the capsule during the postoperative period
 - b. Weakness of affected muscle with limited rotations and decreased saccades in its field of action
- E. Describe appropriate testing and evaluation for establishing the diagnosis.**
1. Postoperative infections (endophthalmitis)
 - a. Vitreous specimen for gram stain and culture
 2. Lost/slipped muscle
 - a. Imaging study sometimes useful in planning treatment

III. Define the risk factors

- A. Unsatisfactory alignment**
1. Poor fusion
 2. Poor vision
 3. Multiple surgeries/scar tissue
 4. Inaccurate preoperative assessment
 5. Surgical technique
- B. Diplopia**
1. Strabismus acquired in adulthood
 2. Overcorrection in childhood onset strabismus
- C. Perforation of the sclera**
1. Inexperienced surgeon
 2. Patient with very high myopia or thin sclera
- D. Postoperative infections (endophthalmitis)**
1. Perforation of the sclera
- E. Dellen**
1. Resection of muscle
 2. Re-operation
- F. Conjunctival scarring**
1. Reoperation
 2. Large resections

3. Advancement of the plica semilunaris onto the bulbar conjunctiva

G. Anterior segment ischemia

1. Previous surgery on rectus muscles
2. Greatest risk with surgery on all 4 rectus muscles but can occur even with one muscle operation, usually vertical rectus
3. Patient with vascular disease

H. Change in eyelid position

1. Vertical rectus muscle surgery

I. Ruptured muscle

1. Restrictive strabismus (thyroid ophthalmopathy, congenital fibrosis of the extraocular muscles)

IV. List the differential diagnosis

A. Differential diagnosis for severe postoperative vision loss

1. Endophthalmitis
2. Vitreous hemorrhage
3. Retinal detachment
4. Anterior segment ischemia
5. Complications from retrobulbar anesthesia

B. Differential diagnosis for persistent postoperative foreign body sensation

1. Foreign body granuloma
2. Conjunctival inclusion cyst
3. Dellen
4. Loose suture

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

A. Describe medical therapy options

1. Diplopia
 - a. Prism - ground in or Fresnel
 - b. Observation for days to weeks. May resolve spontaneously
2. Postoperative infections
 - a. Mild conjunctivitis - topical antibiotics
 - b. Preseptal/orbital cellulitis - systemic antibiotics
 - c. Endophthalmitis - intravitreal antibiotics
3. Granuloma
 - a. Topical corticosteroids
4. Allergic reaction
 - a. Discontinue or change antibiotic eye drop
 - b. Cool compresses
5. Dellen
 - a. Artificial tears/lubricants
6. Anterior segment ischemia

- a. Topical, subconjunctival, or systemic corticosteroids

B. Describe surgical therapy options

1. Unsatisfactory alignment
 - a. Reoperation
2. Diplopia
 - a. Reoperation
3. Postoperative infections (endophthalmitis)
 - a. Vitrectomy in some cases
4. Postoperative infections (orbital cellulitis)
 - a. Orbitotomy with drainage of abscess if present on imaging studies
5. Foreign body granuloma
 - a. Observation, then excision if symptoms warrant
 - b. Usually resolves spontaneously as suture material dissipates
6. Conjunctival inclusion cyst
 - a. Excision if symptoms warrant
7. Lost muscle
 - a. Attempt to retrieve muscle promptly
 - b. If unable, consider muscle transposition procedure
8. Slipped muscle
 - a. Prompt surgery to retrieve and advance muscle
9. Ruptured muscle
 - a. Attempt to retrieve and reappose muscle edges
10. Scleral perforation
 - a. Intraoperative dilated exam to look for hemorrhage, retinal hole, tear or detachment
 - b. Consider laser retinopexy or cryotherapy for retinal break
 - c. Consider antibiotic prophylaxis

VI. Describe disease-related complications

A. Perforation of the sclera

1. Vitreous hemorrhage
2. Retinal detachment
3. Endophthalmitis

B. Anterior segment ischemia

1. Permanent vision loss
2. Enlarged/abnormally shaped pupil with photophobia
3. Iris atrophy
4. Cataract
5. Phthisis bulbi

VII. Describe appropriate patient instructions (See Surgery of the extraocular muscles: weakening procedures)

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Helveston EM. Surgical management of strabismus. St. Louis, Mosby - Year Book, Inc., 1993, p. 90.
3. Ing M. Infection following strabismus surgery. *Ophthalmic Surgery* 1991;22:41-3.
4. Simon JW, Lininger LL, Scheraga JL. Recognized scleral perforation during eye muscle surgery: Incidence and sequelae. *J Pediatr Ophthalmol Strabismus* 1992;29:273-5.

Infantile nystagmus syndrome (infantile idiopathic nystagmus, congenital nystagmus)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Abnormal calibration of the ocular motor system during its development in the sensitive period

B. List the pertinent elements of the history

1. Family history often positive
2. Presents within first 4 months of life
3. Often associated with strabismus

C. Describe pertinent clinical features

1. Infantile onset
2. Conjugate, horizontal-torsional
3. Accelerating slow phases
4. Increases with fixation attempt
5. Dampened with convergence
 - a. Associated with esotropia (nystagmus blockage syndrome)
6. Null and neutral zones possible (possible anomalous head position)
7. "Paradoxical" inversion of OKN response
8. Waveform of nystagmus may change with age and development
9. Disappearance with sleep
10. Remains horizontal in upgaze and downgaze (uniplanar)

D. Describe appropriate testing and evaluation

1. Appropriate visual acuity testing
 - a. Test distance vision under binocular viewing conditions
 - b. Test distance monocular visual acuity with fogging rather than occlusion
 - c. Test binocular near acuity
2. Complete eye exam
3. Electroretinogram if clinically indicated
4. Neuroimaging if clinically indicated

II. Define the risk factors

- ### A. Family history/genetic

III. List the differential diagnosis

- ### A. Other involuntary oscillations of childhood

1. Manifest latent nystagmus
2. Spasmus nutans
3. Nystagmus with vision loss
4. Acquired brainstem or cerebellar disease
5. Periodic alternating nystagmus

B. Sensory interruptions (some examples follow)

1. Retinal disease
 - a. Albinism
 - b. Aniridia
 - c. Leber congenital amaurosis
 - d. Achromatopsia
 - e. Foveal hypoplasia
2. Structural abnormalities of the eyes
 - a. Cataract (bilateral)
 - b. Optic nerve hypoplasia (bilateral)
 - c. Coloboma

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

A. Describe medical therapy options

1. Correct refractive error
2. Use of prism (induce convergence or correct strabismus)
3. Contact lenses

B. Describe surgical therapy options

1. Correct strabismus
2. Correct head position (i.e., Anderson-Kestenbaum)
 - a. Bilateral yoke recessions or recess/resect procedures
3. 4-muscle large-magnitude recessions (i.e. Helveston)
4. 4-muscle tenotomy-reattach (i.e., Hertle)

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

- A. See Ocular complications of strabismus surgery, including consecutive deviations
- B. Induced strabismus

VI. Describe disease-related complications

- A. Decreased vision
- B. Abnormal head position
- C. Psycho-social consequences

VII. Describe appropriate patient instructions

- A. Parental awareness and education regarding development

- B. Vision rehabilitation services**
- C. Referral for outside support**
- D. School supportive services and intervention when age-appropriate**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Khanna S; Dell 'Osso LF. The diagnosis and treatment of infantile nystagmus syndrome (INS) Scientific World Journal 2006; 6:1385-97.
3. Harris C; Berry D. A developmental model of infantile nystagmus. Semin Ophthalmol 2006; 21:63-9.

Latent nystagmus (Fusion maldevelopment nystagmus syndrome)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Fusion maldevelopment
 - a. Early onset strabismus (congenital esotropia most common)
 - b. Poor vision in one or both eyes

B. Define the relevant aspects of epidemiology of the disease

1. Presents in early childhood, often noticed months or years after strabismus surgery

C. List the pertinent elements of the history

1. Prior history of disruption of fusion, such as infantile strabismus, or vision loss
2. May have anomalous head posture to dampen manifest latent nystagmus (LN)

D. Describe pertinent clinical features

1. Conjugate, horizontal jerk nystagmus
2. Usually occurs when one eye is covered, but can also occur when both eyes are uncovered, but only one eye is being used, such as in amblyopia or poor vision in one eye (manifest LN)
3. Fast phase of the jerk nystagmus is directed toward the uncovered eye
 - a. Slow phase of manifest latent nystagmus shows exponential decrease in velocity
4. This is the only form of nystagmus that reverses direction with changes in fixation
5. Possible reasons for head turn
 - a. Nystagmus dampens with fixing eye in adduction
 - b. Patient may cross fixate, with head turn reversing with change in fixation to place fixing eye in adduction
 - c. Secondary to null point in manifest LN
6. Dampened by fusion and worsened by disruption of fusion
7. Decreased acuity when checked with occlusion; therefore, check with fogging techniques
 - a. High plus lens
 - b. Translucent occluder

II. List the differential diagnosis

A. No differential diagnosis if not manifest

B. If manifest LN present, differential diagnosis

1. Congenital motor nystagmus
2. Acquired nystagmus

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

A. Describe medical therapy options

1. No medical treatment for latent nystagmus, but may treat associated conditions
 - a. Eyeglasses, when appropriate
 - b. Amblyopia therapy when appropriate

B. Describe surgical therapy options

1. Eye muscle surgery for face turn (move eyes in direction of face turn) in manifest LN with torticollis
2. No surgery if LN not manifest

IV. List the complications of treatment, their prevention and management (See Ocular complications of strabismus surgery, including consecutive deviations)

V. Describe disease-related complications

- A. Face turn
- B. Decreased visual acuity

VI. Appropriate Patient Instructions

- A. Advise of importance of binocular viewing conditions and/or using fogging for visual acuity testing

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Acquired nystagmus in children

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Intracranial tumors
2. Increased intracranial pressure with or without hydrocephalus
3. Neurodegenerative disease
 - a. Acute cerebellar ataxia of childhood
4. Epilepsy
5. Meningo-encephalitis
6. Acute febrile illness
7. Trauma
8. Toxins/drugs
 - a. Anticonvulsants
 - b. Tranquilizers
9. Spasmus nutans

B. List the pertinent elements of the history

1. Oscillopsia
2. Family history may be positive for nystagmus
3. Down syndrome
4. Recent febrile illness
5. Weight loss
6. Mental status changes
7. Loss of developmental milestones
8. Seizures
9. Focal neurological symptoms
10. Ataxia
11. Vision loss
12. Headache
13. Vomiting

C. Describe the pertinent clinical features

1. Involuntary, repetitive, sustained eye movement
2. Monocular or bilateral
3. Conjugate or dysconjugate
4. Head nodding
5. Torticollis
6. Variability with gaze position
7. Low, medium, or high frequency
8. Horizontal, vertical, or rotary

9. Periodicity or randomly intermittent

D. Describe appropriate testing and evaluation for establishing diagnosis

1. Visual acuity testing
2. Pupil assessment
3. Clinical assessment of CN III-VII function to assist in localization of inciting process/disorder
4. Visual field testing as possible
5. Neuroimaging
6. Toxin screen/drug levels as indicated by history of exposure/ingestion
7. Lumbar puncture with opening pressure and CSF analysis if indicated
8. Electroencephalogram if indicated

E. Neuroanatomic associations

1. Downbeat with craniocervical junction disease
 - a. Arnold Chiari malformation
2. Spasmus Nutans
 - a. Benign condition but rule out chiasmal glioma
3. "See-saw" nystagmus with parasellar disease
 - a. Rostral midbrain or suprasellar
 - b. Craniopharyngioma
 - c. Optic atrophy
 - d. Bitemporal visual field defect
4. Upbeat may associated with brainstem or cerebellar disease
5. Convergence retraction nystagmus
 - a. Dorsal midbrain syndrome
6. Internuclear ophthalmoplegia (abducting nystagmus) with medial longitudinal fasciculus lesion

II. Define the risk factors

- A. Family history of genetic disease, acquired nystagmus, childhood cancer
- B. Exposure to infectious agents, toxins, or drugs
- C. Factors associated with specific diseases which cause acquired nystagmus
- D. History of hydrocephalus and/or shunt surgery

III. List the differential diagnosis

- A. Late recognition of congenital nystagmus
- B. Manifest latent nystagmus
- C. Voluntary nystagmus (not sustained over long periods)
- D. Nystagmus associated with severe visual loss
- E. Spasmus nutans
- F. Opsoclonus (Associated with occult neuroblastoma)
- G. Ocular flutter

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

A. Correct/treat underlying disease

B. Drug therapy

C. Surgical therapy to move null point, correct head turn, or dampen nystagmus

1. Kestenbaum-Anderson procedure
2. Four muscle recession
3. Large recessions of the medial recti for nystagmus blockage syndrome (with or without posterior fixation sutures)
4. Tenotomy and reattachment
5. Botulinum toxin

D. Prisms

1. Induce convergence
2. Binocular image shifts to correct head position

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

A. Drug Therapy

1. Various systemic side effects
2. Prevention/management
 - a. Screening for pre-existing contraindications for drug use
 - b. Blood count and liver function tests to monitor toxicity
 - c. Prescribing physician should have expertise with anticonvulsant medications

B. Surgical Therapy (See Ocular complications of strabismus surgery, including consecutive deviations)

VI. Describe disease related complications

A. Decreased vision

B. Oscillopsia

C. Abnormal head position

D. Psycho-social consequences

VII. Describe appropriate patient instructions

A. Discuss with parent possible underlying diagnosis and appropriate workup

B. Special consideration for educational needs if vision is reduced

Additional Resources

1. AAO, Basic Science and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Ophthalmia neonatorum caused by Neisseria

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Direct contact with the infecting agent *Neisseria gonorrhea* during passage through the birth canal

B. Define the relevant aspects of epidemiology of the disease

1. Less common in the United States and other industrialized nations

C. List the pertinent elements of the history

1. Usually occurs in the first week of life
2. Maternal history of cervical infection

D. Describe pertinent clinical features

1. Hyperacute conjunctivitis
2. Conjunctival hyperemia
3. Purulent discharge
4. Possible corneal penetration and perforation
5. Lid edema
6. Chemosis

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Gram stain
 - a. Gram negative intracellular diplococci
2. Culture of conjunctival surface

II. Define the risk factors

A. Maternal cervical gonococcal infection

B. Maternal history of poor prenatal care

C. Duration of infant's exposure to *Neisseria* (length of labor; duration of ruptured membranes)

III. List the differential diagnosis

A. Chemical conjunctivitis

B. Chlamydial conjunctivitis

C. Conjunctivitis caused by *Haemophilus species*, *Staphylococcus aureus*, *Streptococcus pneumoniae*, enterococci

D. Viral conjunctivitis

E. Congenital nasolacrimal duct obstruction

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

A. Describe medical therapy options

1. Systemic administration of third generation cephalosporin
2. Saline irrigation of the eyes
3. Consider evaluation and treatment for other sexually transmitted diseases
 - a. Systemic erythromycin until Chlamydia ruled-out
4. Evaluation and treatment of parents and other maternal contacts

B. Describe surgical therapy options

1. For potential corneal complications

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

A. Adverse reaction to medication

VI. Describe disease related complications

A. Rapid development of keratitis, corneal ulceration/perforation

VII. Describe appropriate patient instructions

A. Outpatient follow-up post discharge from the hospital

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Ophthalmia neonatorum from *Chlamydia trachomatis*

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Direct contact with the infecting agent *Chlamydia trachomatis* during passage through birth canal

B. Define the relevant aspects of epidemiology of the disease

1. Most common cause of ophthalmia neonatorum in the United States
2. 30% - 40% risk in untreated mother
3. Obligate intracellular organism, *Chlamydia trachomatis*
4. Exposure through birth canal

C. List the pertinent elements of the history

1. Occurs around 5 to 12 days of age
2. Earlier onset with premature rupture of membranes

D. Describe pertinent clinical features

1. Mild swelling, hyperemia, and papillary reaction
2. Minimal to moderate watery or filmy discharge
3. Rare copious discharge and pseudomembrane formation

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Culture from conjunctival swab using Chlamydial transport medium
2. Intracytoplasmic inclusions by Giemsa stain of conjunctival scrapings
3. Enzyme-linked immunoassays
4. Direct fluorescent antibody test
5. Chest x-ray

II. Define the risk factors

A. *Chlamydia trachomatis* in birth canal or uterus

III. List the differential diagnosis

- A. Chemical conjunctivitis
- B. *Neisseria gonorrhoeae* conjunctivitis
- C. *Staphylococcus* conjunctivitis
- D. Herpes simplex conjunctivitis
- E. Nasolacrimal duct obstruction

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

A. Describe medical therapy options

1. Oral erythromycin

2. Alternative oral trimethoprim-sulfamethoxazole
3. Consider topical erythromycin ointment
 - a. Does not treat possible pneumonitis
4. Consider evaluation for other sexually transmitted diseases
5. Evaluation and treatment of parents and other maternal contacts

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

- A. Adverse reaction to medication**

VI. Describe disease-related complications

- A. Pneumonia**
- B. Gastrointestinal infection**
- C. Potential micropannus or conjunctival scarring in untreated cases**

VII. Describe appropriate patient instructions

- A. Full course of antibiotic therapy**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Isenberg SJ, Apt L, Del Signore M, et al. A double application approach to ophthalmia neonatorum prophylaxis. Br J Ophthalmol. 2003;87:1449-52.

Bacterial conjunctivitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Neonatal
 - a. *Neisseria*
 - i. *N. gonorrhoeae* or *N. meningitidis*
 - ii. gm- intracellular diplococci
 - b. *Chlamydia trachomatis*: Obligate intracellular organism
2. Children
 - a. *Staphylococcus aureus*
 - b. *Streptococcus pneumoniae*
 - c. *Haemophilus* species: Less common w/ immunizations
 - d. *Moraxella N. gonorrhoeae*: Older sexually active or victims of sexual abuse
 - e. *Bartonella henselae*: "Cat-scratch disease," most common cause of Parinaud oculoglandular syndrome (POS)
 - f. *Trachoma*: Uncommon in U.S.

B. Define the relevant aspects of epidemiology of the disease

1. Bacterial conjunctivitis is the most common cause of acute conjunctivitis in children

C. List the pertinent elements of the history

1. Hyperacute to delayed onset of symptoms and signs, depending on organism
2. History of ocular redness, burning, stinging, foreign body sensation, and discharge

D. Describe pertinent clinical features

1. Conjunctival inflammation, edema, and purulent discharge
2. Unilateral or bilateral
3. *Neisseria*: Marked chemosis, discharge, corneal ulceration and perforation, serious systemic infection
4. *Chlamydia*: Pseudomembrane (rare)
5. *N. meningitidis*: High risk of meningitis
6. *Bartonella henselae* (POS): Unilateral granulomatous conjunctivitis w/ preauricular and submandibular adenopathy
7. *Trachoma*: Follicles, papillary hypertrophy, vascularization of cornea, cicatricial changes to cornea and conjunctiva

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. May be treated empirically
2. Gram stain and culture may be indicated for non-responders or more severe cases
3. Chlamydia: Conjunctival scraping to demonstrate intracellular organisms, polymerase chain reaction (PCR), antibody tests, enzyme assays
4. Trachoma: Corneal scrapings for cytoplasmic inclusion bodies

II. Define the risk factors

A. Ophthalmia Neonatorum

1. Sexually transmitted disease in the mother
 2. Prolonged rupture of membranes at time of delivery
- B. POS: Exposure to cats**
- C. Other Bacterial Conjunctivitis: exposure to infected person**

III. List the differential diagnosis

- A. Viral conjunctivitis**
- B. Allergic conjunctivitis**
- C. Atopic keratoconjunctivitis**
- D. Vernal keratoconjunctivitis**
- E. Contact lens-induced giant papillary conjunctivitis**
- F. Phlyctenular keratoconjunctivitis**
- G. Iritis, scleritis, episcleritis**
- H. Nasolacrimal duct obstruction**
- I. Globe trauma**
- J. Foreign body**
- K. Herpetic keratitis**
- L. Corneal ulcer**

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

- A. Describe medical therapy options**
1. Observation, as mild bacterial conjunctivitis and POS may be self-limited
 2. Broad-spectrum topical ophthalmic antibiotic drops or ointment
 3. *Neisseria* species
 - a. Systemic ceftriaxone
 - b. Topical irrigation
 - c. Test for HIV, *Chlamydia*, and syphilis
 4. *Chlamydia*: Oral erythromycin for risk of pneumonia
 5. *Trachoma*: Topical and/or systemic sulfonamides, erythromycin, and tetracyclines

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

- A. Allergic response to topical or systemic antimicrobials**
1. Avoid known allergens
 2. Switch to different class of drugs or discontinue medications
- B. Corneal epithelial toxicity from topical antimicrobials, especially aminoglycosides**
1. Avoid excessively long treatment duration
 2. Discontinue medication or change to alternate drug if further treatment is required
- C. Stevens-Johnson syndrome**

VI. Describe disease-related complications

- A. Corneal scarring
- B. Corneal perforation
- C. Systemic infection in neonate

VII. Describe appropriate patient instructions

- A. Complete full course of antibiotics
- B. Keep follow-up appointments
- C. Limit exposure to prevent contagion
- D. Frequent handwashing and separate hand towels
- E. Public health report for sexually-transmitted disease in Neisserial and chlamydial conjunctivitis

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Høvdig, G. Acute bacterial conjunctivitis. *Acta Ophthalmologica*, 2008; 86: 5-17
3. Michael E. Pichichero, ME. Bacterial conjunctivitis in children: antibacterial treatment options in an era of increasing drug resistance *Clin Pediatr* 2011; 50(1): 7-13.
4. AAO, Focal Points: Neonatal Conjunctivitis: Diagnosis and Treatment, Module #1, 1988, p.2.
5. Isenberg SJ, Apt L, Valenton M, et al. A controlled trial of povidone-iodine to treat infectious conjunctivitis in children. *Am J Ophthalmol*. 2002;134:681-8.
6. AAO, Preferred Practice Patterns Committee, Cornea and External Disease Panel: Conjunctivitis Preferred Practice Pattern, 2013.
7. Epling J, Smucny J. Bacterial conjunctivitis. *Clin Evid* 2005; 14:756-61.

Epidemic keratoconjunctivitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Adenovirus
2. Direct contact
3. Aerosolized
4. Virus survives for weeks
5. Resistant to antiseptics
6. Viral shedding prior to symptoms
7. Highly contagious

B. Define the relevant aspects epidemiology of the disease

1. Closed settings (schools, physician's offices, etc.)
2. Epidemic outbreaks

C. List the pertinent elements of the history

1. Recent exposure within family or school
2. Red eye
3. Ocular and periorbital pain (more severe)
4. Photophobia
5. Foreign body sensation
6. Excessive tearing
7. Eye rubbing
8. Fellow eye tends to be involved
9. Symptoms last for 7-21 days

D. Describe pertinent clinical features

1. Acute follicular conjunctivitis
2. Unilateral at onset, then bilateral
3. Preauricular lymphadenopathy
4. Periorbital ecchymosis
5. Membranous and pseudomembranous conjunctivitis if severe
6. Corneal involvement
 - a. Diffuse superficial keratitis then focal epithelial lesions that stain
 - b. Later (11-15 days) subepithelial (immune) opacities that can last 2 years

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Usually diagnosed clinically
2. Rapid immunochromatography testing in office

II. Define the risk factors

A. Exposure

III. List the differential diagnosis

- A. Allergic conjunctivitis**
- B. Bacterial conjunctivitis**
- C. Other viral conjunctivitis**
- D. Contact lens complications**
- E. Herpes simplex**
- F. Periorbital cellulitis**
- G. Chemical exposure**

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

A. Describe medical/supportive therapy options

- 1. Avoid exposure, contagious for up to 2 weeks
- 2. Cool compress to eye
- 3. Artificial tears
- 4. Topical corticosteroids
 - a. Seldom required in children
 - b. May prolong time to full recovery
 - c. Late subepithelial opacities that reduce vision

B. Describe surgical therapy options

- 1. Extremely rare
 - a. Reserved for severe cases with cicatricial conjunctivitis secondary to symblepharon

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

A. Corticosteroids

- 1. May prolong course of disease
- 2. Glaucoma in corticosteroid responders
- 3. Secondary infections

VI. Describe disease-related complications

- A. Quality of life**
- B. Persistent sub-epithelial opacities**
- C. Conjunctival scarring and symblepharon**

VII. Describe appropriate patient/parent instructions

- A. Frequent handwashing**
- B. Isolation of contaminated objects**
- C. Avoid eye rubbing**
- D. Avoid exposure at school or day care**

VIII. Describe physician instructions

- A. Handwashing**
- B. Clean equipment**
- C. Isolation areas for patients suspected of having adenovirus**
- D. Infected medical personnel should limit exposure to ophthalmic exam areas and neonates**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Ohnsman, CM. Exclusion of students with conjunctivitis from school:policies of state departments of health. *J Pediatr Ophthalmol Strabismus* 2007;44:101-105.

Herpes Zoster / Varicella Zoster

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Varicella-zoster virus (VZV) (chickenpox)

B. Define the relevant aspects of epidemiology of the disease

1. Varicella vaccine prevents severe infection, but vaccinated children still may develop mild disease
2. Severe disease or reactivation rare in children except immunocompromised

C. List the pertinent elements of the history

1. Primary infection
 - a. Fever
 - b. Painful and itchy skin and mucous membrane rash
2. Reactivation of latent VZV
 - a. Past medical history may be negative
 - b. May have history of an immunocompromised state
 - c. May have history of chicken pox infection

D. Describe pertinent clinical features

1. Primary infection (chickenpox)
 - a. Vesicular eruptions of skin and mucous membranes
 - b. Ocular involvement usually mild and similar to HSV primary infection
 - c. Less common ocular manifestations:
 - i. Conjunctival vesicles or ulcerations
 - ii. Corneal epithelial keratitis or dendritic ulcer
 - iii. Interstitial keratitis
 - iv. Mild anterior uveitis
2. Reactivation of VZV from dorsal root and cranial nerve ganglia causes Herpes Zoster
 - a. Unilateral vesicle formation on head or eyelids in a dermatomal pattern
 - b. Ocular involvement most likely if nasociliary branch of CNV₁ involved (Hutchinson sign)
 - i. Epiphora
 - ii. Keratitis
 - iii. Anterior uveitis
 - iv. Episcleritis/scleritis, cranial nerve palsies, keratitis much less common in children than in adults

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Serum titers of varicella zoster virus antibody

II. Define the risk factors

A. Immunocompromised child

III. List the differential diagnosis

A. Herpes simplex (HSV)

1. Herpes Simplex corneal dendrites are thin, branching lesions with club-shaped terminal bulbs that stain well with fluorescein
2. Herpes Zoster corneal lesions are pseudo dendrites of raised mucous plaques. They do not have terminal bulbs and do not stain well with fluorescein
3. Herpes Zoster rash frequently follows a dermatome and does not cross the midline in contrast to HSV rash which does not follow a dermatome or obey the midline

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

A. Describe medical therapy options

1. Primary VZV infection
 - a. Treatment of the conjunctivitis for symptomatic relief
 - b. Topical steroids contraindicated except for late interstitial keratitis
 - c. Topical antibiotics to prevent secondary infections
 - d. Consider systemic antivirals for immunocompromised child
2. Herpes Zoster
 - a. Topical steroids for severe iritis
 - b. Moist compresses, topical antibiotics for cutaneous lesions
 - c. Consider systemic antivirals

B. Describe surgical therapy options

1. Corneal transplant if warranted

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

A. Adverse drug reaction

VI. Describe disease-related complications

- A. Postherpetic neuralgia**
- B. Loss of vision**
- C. Recurrence**
- D. Cranial nerve palsy**

VII. Describe appropriate patient instructions

A. Close follow-up with ophthalmologist and primary care physician

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Herpes simplex viruses

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Herpes simplex virus type 1 (HSV-1): gingivostomatitis, cold sores
2. Herpes simplex virus type 2 (HSV-2): genital
3. Recently more crossover

B. Define the relevant aspects of epidemiology of the disease

1. Most eye infections caused by HSV-1
2. Most neonatal infections caused by HSV-2
3. Many carriers are asymptomatic and shed virus

C. List the pertinent elements of the history

1. Birth history
2. History of exposure to cold sores
3. +/- Skin vesicles

D. Describe pertinent clinical features

1. Primary infection
 - a. May be asymptomatic
 - b. Unilateral blepharoconjunctivitis. Rarely bilateral
 - c. +/- Skin vesicles
 - d. Epithelial keratitis (non-dendritic)
2. Recurrent HSV infection (reactivation)
 - a. Blepharitis (lid disease)
 - b. Conjunctivitis
 - c. Epithelial Keratitis (dendritic or geographic)
 - d. Stromal (immune) keratitis
 - e. Uveitis
 - f. Chorioretinitis
 - g. Decreased vision
3. Systemic involvement
 - a. Up to 2/3 of Neonatal infections have systemic involvement including hepatitis, pneumonia, disseminated intravascular coagulation, encephalitis
 - b. Very high morbidity and mortality in infants
 - c. Eye or cutaneous disease can progress to systemic in infants

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Usually clinical diagnosis
2. Culture, ELISA, or PCR can confirm

II. Define the risk factors

A. Close personal contact with infected individual

- B. History of HSV infection

III. List the differential diagnosis

- A. Viral conjunctivitis
- B. Allergic conjunctivitis
- C. Vernal/atopic conjunctivitis
- D. Trauma
- E. Chemical conjunctivitis

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

- A. Disseminated disease
 - 1. Systemic antivirals
- B. Conjunctivitis
 - 1. Treatment does not alter course of disease
- C. Epithelial keratitis
 - 1. Topical antivirals
 - 2. Oral antivirals
- D. Stromal keratitis and anterior uveitis
 - 1. Oral antivirals
 - 2. Cautious use of topical corticosteroids if indicated

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

- A. Adverse reaction to medication

VI. Describe disease-related complications

- A. Corneal involvement
- B. Uveitis
- C. Chorioretinitis
- D. Vitritis
- E. Cataract (secondary to uveitis)
- F. Optic neuritis and/or optic atrophy
- G. Recurrence
 - 1. Can be associated with acute retinal necrosis (ARN)
- H. Systemic infection in newborns

VII. Describe appropriate patient instructions

- A. Return to clinic for routine follow-up (may be several days to weeks)
- B. Return immediately for increased pain or photophobia, decreased vision, or if lid lesions become infected
- C. If medication is prescribed, do not stop taking the medicine until the full course is finished

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
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3. AAO, Focal Points: Ocular Infections of the External Eye and Cornea in Children, Module #2, 2002, p.4-6.
4. Herpetic Eye Disease Study Group. Predictors of recurrent herpes simplex virus keratitis. *Cornea*. 2001;20:123-8.
5. Arvin AM, Whitley RJ. Herpes simplex virus infections. In: Remington JS, Klein JO, eds. *Infectious Diseases of the Fetus and Newborn Infant*. 5th ed. Philadelphia: Saunders;2001.
6. Overall JC. Herpes simplex virus infection of the fetus and newborn. *Pediatr Ann*. 1994;23:131-136.
7. Schwartz GS, Holland EJ. Oral acyclovir for the management of herpes simplex virus keratitis in children. *Ophthalmology*. 2000;107:278-282.
8. Lambert SR. Ocular manifestations of intrauterine infections. In: Taylor D, Hoyt CS, eds. *Pediatric Ophthalmology and Strabismus*. 3rd ed. Philadelphia: Elseiver Saunders; 2005.

Preseptal cellulitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Infectious or inflammatory process involving the tissues anterior to the orbital septum

B. Define the relevant aspects of epidemiology of the disease

1. Post-traumatic (puncture, laceration, or abrasion of skin): *Staphylococcus* or *Streptococcus* species
2. Severe conjunctivitis (e.g., EKC) or skin infection (e.g., impetigo, herpes zoster, or chalazion)
3. Associated with upper respiratory tract infections or sinusitis: *Haemophilus influenzae* incidence reduced markedly since *H influenzae* vaccine (Hib), but may still be seen. *S. pneumoniae*, other strep species, and *S. aureus* most common

C. List the pertinent elements of the history

1. Acute progressive edema and erythema of eyelid
2. Upper respiratory tract infection symptoms, trauma, local skin infection
3. Immunization status

D. Describe pertinent clinical features

1. Eyelid edema and erythema that may extend to eyebrow and forehead
2. Orbit remains normal with full range of motion, lack of pain on eye movements, no proptosis, normal vision, and no relative afferent pupillary defect
3. Conjunctivitis may be present

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

1. Conjunctival and paranasal sinus cultures
2. Blood cultures if febrile

II. Define the risk factors

A. Upper respiratory tract infection and sinusitis

B. Recent trauma or surgery

C. Severe conjunctivitis or skin infection

D. No Hib vaccine

III. List the differential diagnosis

A. Orbital cellulitis

B. Chalazion

C. Severe conjunctivitis with preseptal edema

D. Dacryoadenitis

E. Tumor

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

A. Describe medical therapy options

1. Non-severe

- a. Oral, broad-spectrum antibiotics (e.g. cephalosporins, ampicillin-clavulanic acid combination)
- 2. Severe, signs of sepsis or meningitis, or infant
 - a. Hospitalization
 - b. Imaging of sinuses and orbit
 - c. IV broad-spectrum antibiotics
 - d. Consider methicillin-resistant *S. aureus*

B. Describe surgical therapy options

- 1. Possible need to drain tissue abscess if develops

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

A. Allergic reaction to antibiotic treatment

VI. Describe disease-related complications

A. Development of orbital cellulitis

B. Intracranial spread via venous drainage system of face

Additional Resources

- 1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, 2008 Focal Points Module, Preseptal and Orbital Cellulitis.

Orbital cellulitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Infectious or inflammatory process involving the tissues posterior to the orbital septum
2. Most commonly associated with ethmoid or frontal sinusitis
3. May follow penetrating injuries of the orbit
4. Neonates: *S aureus* including MRSA and gram-negative bacilli
5. Older children and adults: *S aureus*, *S pyogenes*, *S pneumonia*, anaerobic species
6. Immunocompromised: gram-negative or fungal infections

B. Define the relevant aspects of epidemiology of the disease

1. May occur at any age
2. May have preceding sinusitis
3. Community acquired MRSA becoming more common

C. List the pertinent elements of the history

1. Lethargy, fever
2. Periocular pain and/or headache
3. Swollen, tender eyelid
4. Decreased vision, or diplopia
5. Rhinorrhea
6. May have history of penetrating orbital injury or previous ocular or orbital surgery
7. Sinus disease

D. Describe pertinent clinical features

1. Eyelid edema, erythema, tenderness, warmth
2. Proptosis possible
3. Restricted ocular motility possible
4. Decreased acuity with relative afferent pupillary defect possible

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Orbital imaging to confirm orbital involvement, document sinusitis and/or subperiosteal abscess, rule out foreign body
2. Culture of conjunctival or nasal discharge if present
3. Blood cultures if clinically indicated

II. Define the risk factors

A. Contiguous sinusitis

B. Penetrating orbital trauma or recent sino-orbital surgery

C. Immunocompromised states

III. List the differential diagnosis

- A. Preseptal cellulitis
- B. Idiopathic orbital inflammation (orbital pseudotumor)
- C. Benign orbital tumors: lymphangioma, hemangioma
- D. Malignant tumors: rhabdomyosarcoma, leukemia, metastatic (e.g., neuroblastoma)
- E. Occult orbital trauma
- F. Cavernous sinus fistula or thrombosis
- G. Ruptured dermoid cyst

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

- A. Describe medical therapy options
 1. Hospitalization with broad-spectrum IV antibiotics
 2. Oculoplastics or pediatric otolaryngology consult for sinusitis or subperiosteal abscess
 3. Observe closely for visual compromise
 4. If condition worsens or vision compromised, repeat scan to assess for subperiosteal abscess
 5. If patient immunocompromised consider evaluation for mucormycosis
- B. Describe surgical therapy options
 1. Drainage of subperiosteal abscess if patient not responding to antibiotics and/or vision compromised
 2. Debridement of sinuses if significant disease present
 3. Debridement of necrotic tissue if mucormycosis present; exenteration may be needed

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

- A. Adverse reactions to antibiotics
- B. Complications of orbital surgery (possible visual loss, motility defects)

VI. Describe disease-related complications

- A. Loss of vision from optic neuropathy or corneal exposure
- B. Diplopia
- C. Subperiosteal abscess
- D. Cavernous sinus thrombosis
- E. Intracranial extension which may result in death

VII. Describe appropriate patient instructions

- A. At least 24 hours of antibiotic therapy may be required before clinical stabilization occurs
- B. Complete full prescribed course of oral antibiotic taper
- C. Return at first sign of recrudescence
- D. Follow-up with otolaryngologist as appropriate

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, 2008 Focal Points Module, Preseptal and Orbital Cellulitis.

3. Seltz LB, Smith J, Durairaj VD, Enzenauer R, and Todd J. Microbiology and Antibiotic Management of Orbital Cellulitis. *Pediatrics* 2011; 127:3 e566-e572; doi:10.1542/peds.2010-2117
4. Kobayashi D, Givner LB, Yeatts RP, Anthony EY, Shetty AK. Infantile orbital cellulitis secondary to community-associated methicillin-resistant *Staphylococcus aureus*. *J AAPOS* 2011; 15(2):208-10.

Seasonal allergic conjunctivitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Environmental contact with airborne allergens

B. Define the relevant aspects of epidemiology of the disease

1. Worse in Spring or Fall

C. List the pertinent elements of the history

1. Itching/puffiness, usually bilateral
2. Associated with season, or occurs at a similar time each year
3. Photophobia
4. Watery eyes, tears clear
5. Runny nose

D. Describe pertinent clinical features

1. Mild conjunctival injection and chemosis
2. Allergic shiners i.e., lower-lid ecchymoses
3. Papillary response of the conjunctivae
4. Follicular conjunctival changes if chronic

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Eosinophils in conjunctival scrapings, if obtained
2. Systemic allergy testing to identify allergen

II. Define the risk factors

A. Exposure to allergen

III. List the differential diagnosis

- A. Foreign body
- B. Viral infection
- C. Chemical exposure or other noxious irritant
- D. Glaucoma
- E. Atopic conjunctivitis
- F. Vernal conjunctivitis

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

A. Describe medical therapy options

1. Avoid exposure to allergen
2. Cool compresses to eyes
3. Systemic antihistamines if other symptoms

4. Topical H₁- receptor blocking drops
5. Mast cell stabilizing drops
6. Vasoconstrictors
7. NSAID drops
8. Combination drops
9. Severe cases
 - a. Corticosteroid drops

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

- A. Most treatment is safe**
- B. Vasoconstrictors may cause pupil dilation, blurry vision, and rebound hyperemia.**
- C. If corticosteroids needed**
 1. Glaucoma in corticosteroid responders
 2. Cataracts
 3. Activation of viral keratitis
 4. Secondary infections

VI. Describe disease-related complications

- A. Quality of life**

VII. Describe appropriate patient instructions

- A. Child to avoid allergen or situation causing the symptoms**
- B. Will be a lifelong problem**
- C. Hand and face washing**
- D. Avoid hand to eye transmission of allergen**
- E. Shower if exposed to grasses or pollen (especially at night)**
- F. Close bedroom windows when pollen counts are high**
- G. Compresses to be started as symptoms emerge**
- H. Drops that stabilize mast cells take several doses to become effective**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Bilkhu PS, Wolffsohn JS, Naroo SA. A review of non-pharmacological and pharmacological management of seasonal and perennial allergic conjunctivitis. *Contact Lens and Anterior Eye*. 2012;35:9-16.
3. Yaylali V, Demirlenk I, Tatlipinar S, et al. Comparative study of 0.1% olopatadine hydrochloride and 0.5% ketorolac tromethamine in the treatment of seasonal allergic conjunctivitis. *Acta Ophthalmol Scand*. 2003;81:378-82.
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8. Cirila AM, Cirila PE, Parmiani S, et al. A pre-seasonal birch/hazel sublingual immunotherapy can improve the outcome of grass pollen injective treatment in bisensitized individuals. A case-referent, two-year controlled study. *Allergol Immunopathol (Madr)* 2003;31:31-43. Erratum in: *Allergol Immunopathol (Madr)*. 2003;31:114.
9. Leonardi A, Papa V, Milazzo G, et al. Efficacy and safety of desonide phosphate for the treatment of allergic conjunctivitis. *Cornea.* 2002;21:476-88.

Vernal keratoconjunctivitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Mast cell/lymphocyte-mediated allergic response

B. Define the relevant aspects of epidemiology of the disease

1. Uncommon
2. More prevalent in hot and dry climates
3. Males affected more frequently than females
4. Usually first 2 decades of life

C. List the pertinent elements of the history

1. Bilateral itching, photophobia, lacrimation, and discharge.
2. Usually spring and fall

D. Describe pertinent clinical features

1. Two forms
 - a. Limbal / Bulbar
 - i. Limbal papillae, especially superiorly
 - ii. Limbal Horner-Trantas dots (eosinophil collections)
 - b. Palpebral
 - i. Giant papillae in superior tarsal conjunctiva
 - ii. Pseudomembrane
 - iii. Mechanical ptosis
 2. Corneal findings
 - a. Punctate epithelial keratopathy
 - b. Micropannus
 - c. Shield ulcer may be present
 3. Thick ropy mucus
- ### E. Describe appropriate testing and evaluation for establishing the diagnosis
1. Diagnosis may be made clinically
 2. Hematoxylin and eosin staining of mucus shows eosinophils and eosinophilic granules

II. Define the risk factors

A. Warm climate

B. Atopic disease

C. Under 10 years

III. List the differential diagnosis

A. Allergic conjunctivitis

B. Atopic keratoconjunctivitis

- C. Contact lens-induced giant papillary conjunctivitis
- D. Phlyctenular keratoconjunctivitis
- E. Other etiologies of corneal ulcer

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

- A. Describe medical therapy options
 - 1. Cool compresses and artificial tears
 - 2. Avoidance of triggers
 - 3. Avoidance of rubbing
 - 4. Medications
 - a. Topical and oral antihistamines
 - b. Mast cell stabilizers
 - c. Topical corticosteroids
 - d. Cyclosporin A
 - e. Supratarsal injection of corticosteroid
- B. Describe surgical therapy options
 - 1. Tarsectomy with or without mucous membrane graft to remove cobblestone papillae
 - 2. Corneal transplant if scarring after shield ulcer impedes vision

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

- A. Elevated intraocular pressure (IOP) due to corticosteroid response
 - 1. Monitor IOP while topical corticosteroids are being used
 - 2. Change to alternative corticosteroid with less risk of corticosteroid-induced ocular hypertension
- B. Cataract
 - 1. Patients with atopic disorders are predisposed to cataract
 - a. Topical corticosteroids increase this risk
 - b. Monitor patients for cataract development
- C. Bacterial keratitis
 - 1. Particularly after prolonged corticosteroid use and presence of shield ulcer

VI. Describe disease-related complications

- A. Corneal scarring from pannus formation and shield ulcer

VII. Describe appropriate patient instructions

- A. Discuss chronic nature of the disease
- B. Air-conditioned environment if possible
- C. Educate regarding potential allergens
- D. Stress need for appropriate follow-up to monitor for complications of treatment and/or disease
- E. Referral to allergist if needed

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Kumar, S. Vernal keratoconjunctivitis: a major review. *Acta Ophthalmologica*, 2009;87:133-147.
3. Gupta V, Sahu PK. Topical cyclosporin A in the management of vernal keratoconjunctivitis. *Eye*. 2001;15:39-41.
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5. Akpek EK, Hasiripi H, Christen WG, et al. A randomized trial of low-dose, topical mitomycin-C in the treatment of severe vernal keratoconjunctivitis. *Ophthalmology*. 2000;107:263-9.
6. Tabbara KF, al-Kharashi SA. Efficacy of nedocromil 2% versus fluorometholone 0.1%: a randomized, double masked trial comparing the effects on severe vernal keratoconjunctivitis. *Br J Ophthalmol*. 1999;83:180-4.
7. Holsclaw DS, Whitcher JP, Wong IG, et al. Supratarsal injection of corticosteroid in the treatment of refractory vernal keratoconjunctivitis. *Am J Ophthalmol*. 1996;121: 243-9.

Congenital eyelid anomalies

I. Describe the approach to establishing the diagnosis

A. Describe the etiology

1. Abnormal development of lid and/or orbital structures
2. Heritable, intrauterine insults, or unknown factors

B. Define the relevant aspects of epidemiology of the disease

1. May be isolated or features of a syndrome

C. List the pertinent elements of the history

1. Detailed prenatal and perinatal history focused on environmental insults (teratogens), family history of similar findings, or other dysmorphisms that may indicate a syndrome

D. Describe pertinent clinical features

1. External exam directed at abnormal findings
 - a. Congenital coloboma of eyelid
 - i. Upper most common
 - ii. Association with Goldenhar syndrome
 - iii. Corneal exposure risk
 - b. Ankyloblepharon
 - i. Complete or partial fusion of lid margin
 - ii. Surgical treatment if does not resolve spontaneously
 - c. Congenital ectropion
 - i. Usually associated with short anterior lamellae
 - ii. Surgical treatment often necessary
 - d. Congenital entropion
 - i. Usually requires surgery
 - e. Epiblepharon
 - i. Extra fold of skin in lower eyelid pushes lashes toward cornea
 - ii. Often well-tolerated and usually spontaneously resolves
 - iii. Surgery required only if corneal integrity threatened
 - f. Epicanthus
 - i. Extra fold of skin over medial canthus
 - ii. May give appearance of pseudo-esotropia
 - g. Blepharophimosis syndrome
 - i. Ptosis, telecanthus, epicanthus inversus, and short horizontal palpebral fissures
 - ii. Autosomal dominant inheritance or sporadic
 - iii. Often requires surgery early for ptosis
 - h. Ptosis (See Congenital ptosis)
2. Anterior segment examination for vision threatening corneal issues
3. Cycloplegic refraction for associated amblyogenic anisometropia and/or astigmatism

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. **Diagnosis based on clinical examination**

II. Define the risk factors

- A. **Family history of heritable eyelid disorder**
- B. **Exposure to known teratogenic agent**

III. Describe disease-related complications

- A. **Amblyopia with unilateral conditions, particularly if anisometropia is present**
- B. **Astigmatism**
- C. **Exposure keratitis**
- D. **Social, developmental, or musculoskeletal effects of anomalous head position**

IV. Describe appropriate patient instructions

- A. **Education regarding specific disorder, natural history, likely outcomes, and treatment options and timing**
- B. **Follow-up arrangements**
- C. **Appropriate referral for consultation**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Byrnes GA, Wilson ME. Congenital Distichiasis. Arch Ophthalmol. 1991; 109 (12): 1752-1753.
3. Dollfus H, Verloes A. Dysmorphology and the orbital region: a practical clinical approach. Surv Ophthalmol 2004; 49 (6): 546-561.

Congenital ptosis

I. Define the approach to establishing the diagnosis

A. Define the relevant aspects of epidemiology of the disease

1. No sex predilection

B. List the pertinent elements of the history

1. Family history of ptosis
2. Congenital vs. acquired
3. Chin up head position
4. Associated pupillary abnormalities
5. Associated strabismus
6. Lid changes with sucking/chewing (Marcus-Gunn jaw winking syndrome)
7. Variability during the day
8. Old photograph review may be helpful

C. Describe pertinent clinical features

1. Unilateral or bilateral and often asymmetric
2. Decreased levator function
3. Poorly formed lid crease
4. May use brow to elevate lid
5. May use chin up head position
6. May have superior rectus weakness and poor Bells phenomenon
7. May have lagophthalmos
8. May have deprivation and/or refractive amblyopia
9. Astigmatism
 - a. Most common cause of amblyopia in ptosis patients
 - b. may persist after ptosis surgery
10. May have associated strabismus (CN III palsy, etc)

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Quantification of ptosis, i.e., margin-reflex distance (normal 4-5mm)
2. Measurement of levator function (normal 15mm)
3. Evaluate tear production
4. Evaluate corneal sensitivity
5. Check for Bells phenomenon
6. Cycloplegic refraction to evaluate for astigmatism and/or anisometropia
7. Testing for myasthenia if suspected
8. Pharmacologic pupillary testing if Horner syndrome suspected
9. Neuroimaging if cranial nerve (CN) III palsy or orbital abnormality suspected

II. Define the risk factors

- A. None identified for typical congenital ptosis
- B. Birth trauma risk for perinatal levator dehiscence

III. List the differential diagnosis

- A. Pseudoptosis secondary to microphthalmia or hypotropia
- B. Myasthenia gravis
- C. Chronic progressive external ophthalmoplegia
- D. Horner syndrome
- E. CN III palsy
- F. Marcus-Gunn jaw winking syndrome = congenital trigemino-oculomotor synkinesis of jaw and levator muscles
- G. Blepharophimosis syndrome: blepharophimosis, epicanthus inversus, telecanthus, and ptosis
- H. Trauma (forceps delivery)
- I. Orbit or lid mass (hemangioma)

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

- A. Describe surgical therapy options
 - 1. If no amblyopia, occlusion of visual axis, or significant compensatory head posture, delay surgery until 4-5 years of age
 - 2. If amblyopia present, visual axis occluded, severe chin-up position, prompt surgery recommended
 - a. Levator resection if levator function at least 4 mm
 - b. Frontalis suspension if levator function < 4 mm
 - c. Müller muscle resection if > 12 mm levator function and lid elevates with 2.5% phenylephrine drops
 - d. repeat surgeries are common

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

- A. Undercorrection
 - 1. Additional surgery if visual axis obstructed or significant chin-up position
 - 2. Undesirable lid position or contour
- B. Overcorrection/exposure keratopathy
 - 1. Ensure adequate tear production, corneal sensitivity, and Bells phenomenon preoperatively
 - 2. Lubrication and tears for corneal exposure
 - 3. Downward massage of lid
 - 4. Reoperation if conservative measures fail
- C. Ectropion
- D. Entropion
- E. Abnormal lid folds

VI. Describe disease-related complications

- A. Amblyopia
- B. Headache/neck pain from ocular torticollis

- C. Delays in gross motor development from chin up head position
- D. Astigmatism
- E. Decreased upgaze due to superior rectus dysgenesis

VII. Describe appropriate patient instructions

- A. Discussion of indications for surgery vs. observation
- B. Need for patching/eyeglasses if amblyopia/astigmatism present
- C. Medical treatment of postoperative exposure with artificial tears and ocular lubricants

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
3. Heher KL and Katowitz JA. Chapter 15 Pediatric Ptosis in Katowitz, J. A. (2002) Pediatric Oculoplastics Surgery. New York: Springer.

Nasolacrimal duct obstruction

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Obstruction of the lacrimal drainage system
2. Most commonly caused by a thin mucosal membrane at the lower end of the nasolacrimal duct (NLD) (valve of Hasner)

B. Define the relevant aspects of epidemiology of the disease

1. Incidence of 5 -20% of newborns
2. No sex predilection
3. No family history
4. High rate of spontaneous resolution
5. Beyond age 1 year, spontaneous resolution becomes less likely

C. List the pertinent elements of the history

1. 80-90% of cases manifest by age 1 month
 2. Bilaterality is common in congenital cases
2. Acquired cases usually present after 4 months of age
3. Acquired cases may follow cases of viral conjunctivitis, acute dacryocystitis, or trauma

D. Describe pertinent clinical features

1. Epiphora and/or mucopurulent discharge with or without an associated conjunctivitis
2. Lower eyelid skin irritation/breakdown
3. Reflux of mucoid material from the puncta with digital pressure over the lacrimal sac

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Fluorescein solution instilled and retained after 5-10 minutes with failure of the dye to appear in the nose or pharynx after 10-15 minutes can confirm an obstruction
2. Clinical history consistent with NLD obstruction

II. Define the risk factors

- A. Craniofacial anomalies
- B. Down syndrome
- C. Mid-face trauma

III. List the differential diagnosis

- A. Glaucoma
- B. Conjunctivitis
- C. Blepharitis
- D. Congenital dacryocele (dacryocystocele)
 1. Beware of associated nasal cysts with possible respiratory problems, may need ENT intervention with nasal endoscopy and cyst marsupialization
- E. Epiblepharon in which lashes rub the cornea and cause tearing

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

A. Describe medical therapy options

1. Digital massage
2. Topical antibiotics as adjunctive therapy
3. Observation

B. Describe surgical therapy options

1. Nasolacrimal duct probing
2. Balloon catheter dilation
3. Infraction of the inferior turbinate in conjunction with probing and/or intubation
4. Silicone intubation of the lacrimal system
5. Dacryocystorhinostomy

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

- A. Complications are uncommon
- B. NLD probing is highly successful
- C. Recurrent or persistent symptoms may require additional procedures.
- D. False passage into the nose, usually requiring no treatment
- E. Minor bleeding from the nose or into the tears, usually requiring no treatment
- F. Punctal erosion

VI. Describe disease-related complications

- A. Chronic epiphora
- B. Anisometropia with resulting amblyopia
- C. Scarring of the lacrimal system secondary to chronic infection/inflammation
- D. Preseptal or orbital cellulitis (uncommon)
- E. Dacryocystitis (uncommon)

VII. Describe appropriate patient instructions

- A. NLD massage technique: downward pressure to attempt to open sac and upward pressure to empty sac
- B. Discussion of the risk/benefit of early probing
- C. Discussion of the risk/benefit of in office vs. operating room probing
- D. Postoperative topical antibiotic/corticosteroid preparations
- E. Risk of displaced silicone tubes
- F. Discussion of the technique of silicone tube removal

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Pediatric Eye disease Investigator Group; Repka MX, Chandler DL, Beck RW, et al. Primary treatment of nasolacrimal duct obstruction with probing in children younger than 4 years. *Ophthalmology*. 2008;115(3):577-584.

3. Kushner BJ. The management of nasolacrimal duct obstruction in children between 18 months and 4 years old. *J AAPOS*. 1998;2:57-60.
4. Gunton KB, Chung CW, Schnall BM, et al. Comparison of balloon dacryocystoplasty to probing as the primary treatment of congenital nasolacrimal duct obstruction. *J AAPOS*. 2001;5:139-142.
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6. Pe MRL, Langford JD, Linberg JV, et al. Ritleng intubation system for treatment of congenital nasolacrimal duct obstruction. *Arch Ophthalmol*. 1998;116:387-391.
7. Gardiner JA, Forte V, Pashby RC, et al. The role of nasal endoscopy in repeat pediatric nasolacrimal duct probings. *J AAPOS*. 2001;5:148-152.

Surgery for nasolacrimal duct obstruction

I. List the indications/contraindications

A. Indications

1. Chronic congenital nasolacrimal duct (NLD) obstruction
2. Previously failed probing procedure
3. Dacryoceles (dacryocystoceles), in the newborn, unresponsive to massage

II. Describe the pre-procedure evaluation

A. Assessment of tear lake

B. Lacrimal sac compression

C. Consider a dye-disappearance test

D. Rule out other causes of epiphora (e.g., infantile glaucoma, trichiasis, iritis, etc.)

III. List the alternatives to this procedure

A. Observation for spontaneous resolution

B. Conservative therapy (massage and topical antibiotics)

IV. Describe the instrumentation, anesthesia, and technique

A. Instrumentation

1. Punctal dilator
2. Irrigating probe/cannula
3. Bowman probes
4. +/- Nasolacrimal Balloon catheter
5. +/- Silastic tubes for nasolacrimal intubation

B. Anesthesia

1. General anesthesia or mask sedation
2. Topical anesthesia may be used for in-office probing of the younger infant

C. Technique

1. Punctal dilation
 - a. If puncta imperforate, occult puncta can often be identified and opened with fine probe or sharp punctal dilator
2. NLD probing
 - a. Bowman probe(s) of appropriate size(s) are passed through the upper and/or lower canaliculus, into the lacrimal sac, and down the NLD into the nose
 - b. An obstruction at the level of the valve of Hasner may be appreciated during probing
 - c. Patency of the system is confirmed by metal-on-metal contact in the nose and/or by successful irrigation of fluorescein solution into the nasopharynx
3. Infractionure of the inferior turbinate may occasionally be required if the inferior turbinate obstructs outlet

4. Balloon dilation of the lacrimal drainage system may be used primarily, as a secondary procedure, or in conjunction with other procedures.
5. Intubation of the lacrimal drainage system may be used primarily, as a secondary procedure, or in conjunction with other procedures
6. Consider nasal endoscopy for marsupialization of associated nasal mucocele with dacryocystocele

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

A. False passage

1. Avoid unnecessary force during probing
2. May occur with anatomic variations

B. Bleeding

1. Usually self-limited
2. Topical nasal vasoconstrictors may be helpful.

C. Recurrence

1. May be treated with repeat probing with or without silastic intubation or balloon catheter dilation

D. Cellulitis

1. Consider prophylaxis with topical or systemic antibiotics

VI. Describe the follow-up care

A. Consider topical antibiotics and/or corticosteroids postoperatively

B. Consider topical nasal vasoconstrictors postoperatively

C. If silicone tubes are placed, they are typically removed after 3 to 6 months

VII. Describe appropriate patient instructions

A. Contact physician for severe or persistent nosebleed

B. Contact physician if swelling or redness of periorbital area occurs

C. Contact physician if silicone tubing is dislodged

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Pediatric Eye Disease Investigator Group. Primary Treatment of Nasolacrimal Duct Obstruction with Nasolacrimal Duct Intubation in Children Less than Four Years Old. *Journal of AAPOS* 2008;12:445-450.
3. Pediatric Eye Disease Investigator Group. Primary Treatment of Nasolacrimal Duct Obstruction with Balloon Catheter Dilation in Children Less than Four Years Old. *Journal of AAPOS* 2008;12:451-455.
4. Pediatric Eye Disease Investigator Group. Primary Treatment of Nasolacrimal Duct Obstruction with Probing In Children Younger than 4 Years. *Ophthalmology* 2008;115:577-584.
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6. Pe MRL, Langford JD, Linberg JV, et al. Ritleng intubation system for treatment of congenital nasolacrimal duct obstruction. *Arch Ophthalmol.* 1998;116:387-391.
7. Crawford JS. Intubation of the lacrimal system. *Ophthal Plast Reconstr Surg.* 1989;5(4):261-265.

Aniridia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Defect in PAX6 gene

B. Define the relevant aspects of epidemiology of the disease

1. Rare
2. Bilateral
3. 2/3 Familial
 - a. Familial form is autosomal dominant
4. 1/3 Sporadic
 - a. Associated with Wilms tumor (nephroblastoma) in 1/3 of cases

C. List the pertinent elements of the history

1. Infant with nystagmus
2. Photophobia
3. Family history of aniridia, nystagmus, and/or poor vision

D. Describe pertinent clinical features

1. Near total absence to mild hypoplasia of iris
 - a. Appears to have dilated pupils
 - b. Has a rudimentary iris stump, so not true aniridia
2. Foveal hypoplasia
3. Sensory nystagmus
4. Poor visual acuity
5. Optic nerve hypoplasia common
6. Glaucoma common
7. Cataracts common
8. Corneal pannus
 - a. May progress to opacification
 - b. Secondary to stem cell deficiency
9. Persistent pupillary membrane fibers

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Chromosome analysis for all patients with aniridia
 - a. To determine if deletion in adjacent Wilms tumor gene
 - b. Positive results
 - i. Consult oncologist
 - ii. Repeated abdominal ultrasounds and clinical exam

II. Define the risk factors

A. Family history of aniridia

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

A. Describe medical therapy options

1. Monitor for glaucoma
2. Genetic testing and counseling

IV. Describe disease-related complications

- A. Glaucoma**
- B. Wilms tumor**
- C. WAGR complex (Wilms, aniridia, genitourinary malformations, and mental retardation)**
- D. Cataracts**
- E. Progressive corneal opacification**

V. Describe appropriate patient instructions

- A. Close follow up with ophthalmologist and with primary care provider**
- B. Genetic counseling**
- C. Provide information for low vision services**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Lee H, Khan R, O'Keefe M. Aniridia: current pathology and management. [Acta Ophthalmol](#). 2008 Nov;86(7):708-15.
3. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.

Primary congenital (infantile) glaucoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Basic pathologic defect controversial
2. Developmental anomaly of tissue arising from neural crest cells (anterior segment and angle)

B. Define the relevant aspects of epidemiology of the disease

1. 2/3 of cases bilateral
2. More common in males
3. No racial or geographic prevalence
4. Most cases are sporadic but several gene loci have been identified, which are usually autosomal recessive

C. List the pertinent elements of the history

1. May be a family history of primary congenital glaucoma (PCG)
2. Classic clinical triad: Epiphora, photophobia, and blepharospasm

D. Describe pertinent clinical features

1. Corneal edema
2. Haab striae
3. Corneal enlargement: depends on age, but >13 mm in any child is abnormal
4. Buphthalmos
5. Reduced visual behaviors
6. Elevated intraocular pressure (IOP)
7. Epiphora
8. Optic nerve cupping
9. Myopia
10. Indistinct landmarks on gonioscopy

II. Define the risk factors

A. Family history of congenital glaucoma

III. List the differential diagnosis

A. Nasolacrimal duct obstruction

B. Congenital hereditary endothelial dystrophy (CHED)

1. Autosomal dominant and recessive inheritance
2. Bilateral
3. Diffuse stromal edema with increased corneal thickness

C. Birth trauma (Forceps injury causing breaks in Descemet membrane)

1. Watch for amblyopia secondary to haze and or anisometropia

D. Megalocornea

E. Sclerocornea

1. Sporadic, autosomal dominant and autosomal recessive
2. Usually bilateral
3. Scleralization of cornea with ill-defined limbus, and vascularization which can affect partial or complete cornea
4. Central cornea clearer than periphery
5. Commonly associated with other ocular or systemic abnormalities

F. Corneal ulcer

G. Mucopolysaccharidoses

H. Peters anomaly

1. Most sporadically, also autosomal dominant and recessive
2. Bilateral in 60% of cases
3. Congenital central cornea opacity with iridocorneal adhesions
4. Other anomalies include keratolenticular touch, cataract, congenital glaucoma, microcornea, aniridia, persistent fetal vasculature, and skeletal anomalies

I. Dermoid (limbal)

1. Usually unilateral, inferotemporal
2. Elevated fibrofatty tissue covered by keratinized epithelium

J. Ocular inflammation

1. Intrauterine keratitis
 - a. Sporadic
 - b. Unilateral or bilateral
 - c. Posterior corneal defect, with corneal infiltrates, vascularization, keratic precipitates, iris adhesions and uveitis
 - d. May be associated with teratogenic effects with ocular malformation

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

A. Describe medical therapy options

1. Usually not successful unless accompanied by surgical therapy
2. Oral carbonic anhydrase inhibitors
3. Topical glaucoma medications

B. Describe surgical therapy options

1. Angle surgery is the preferred initial surgery for PCG
 - a. Goniotomy
 - b. Trabeculotomy
2. Trabeculectomy, with or without mitomycin-C
3. Glaucoma implant surgery
4. Cycloablation

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

A. Systemic complications of medical glaucoma treatment

- B. Postoperative infection, cataract, failure of IOP control, strabismus, hypotony, retinal detachment, phthisis, etc. may follow IOP lowering procedure

VI. Describe disease-related complications

- A. Blindness
- B. Amblyopia
- C. Strabismus
- D. Dislocated lens
- E. Phthisis/loss of eye

VII. Describe appropriate patient instructions

- A. Close and frequent follow-up exams are required
- B. Examinations under anesthesia may be required to monitor glaucoma
- C. Correction of any refractive error is mandatory
- D. Treatment of amblyopia is critical

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
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Congenital and acquired cataracts in children

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Bilateral
 - a. Idiopathic
 - b. Hereditary
 - i. Autosomal dominant (most common)
 - ii. Also autosomal recessive, X-linked
 - c. Chromosomal abnormality
 - d. Metabolic (e.g., galactosemia)
 - e. Syndromes (e.g., Lowe, myotonic dystrophy, Down syndrome)
 - f. Maternal infection
 - i. TORCH
 - ii. Varicella
 - iii. Syphilis
 - g. Ocular anomalies
 - i. Aniridia
 - ii. Anterior segment dysgenesis syndrome
 - h. Iatrogenic
 - i. Corticosteroids
 - ii. Radiation (may be unilateral)
2. Unilateral
 - a. Idiopathic
 - b. Ocular anomalies
 - i. Persistent fetal vasculature (PFV)
 - ii. Anterior segment dysgenesis
 - iii. Posterior segment tumors
 - c. Trauma
 - i. Accidental or Non-accidental

B. List the pertinent elements of the history

1. Red reflex anomalies
2. Family history
3. Strabismus
4. Nystagmus
5. Patient's health history
6. Previous trauma
7. Pregnancy history

C. Describe pertinent clinical features

1. Morphology of cataract
 - a. Anterior polar
 - i. Usually small (1mm) and stable
 - ii. Not visually significant
 - iii. Look for anisometropia
 - b. Nuclear
 - i. Size variable
 - ii. Density is variable
 - iii. Unilateral or bilateral
 - iv. Often associated with microcornea and increased risk of glaucoma
 - c. Lamellar
 - i. Usually 5 mm or more
 - ii. Unilateral, bilateral, or asymmetric
 - iii. Usually acquired
 - d. Posterior lenticonus
 - i. Thinning of posterior capsule
 - ii. Often progressive
 - iii. Usually unilateral
 - iv. Look for refractive errors
 - e. Persistent fetal vasculature (PFV)
 - i. Failure of the fetal hyaloid vascular complex to regress with retrolental membrane of varying size and density
 - ii. Congenital
 - iii. Unilateral almost always
 - iv. Microphthalmia, microcornea
 - v. High incidence of glaucoma
 - f. Posterior subcapsular cataracts
 - i. Not common in children
 - ii. Usually acquired, bilateral, progressive
 - iii. Often secondary
 - i) Steroids, uveitis, retinal degenerations, radiation, neurofibromatosis II
2. Density of cataract
 - a. retinoscopy and indirect ophthalmoscopy can help determine if dense enough to be visually significant
3. Location of cataract
 - a. central or posterior opacities are usually more visually significant than anterior opacities
4. Size of cataract
 - a. Less than 3 mm may not be visually significant
5. Other ocular pathology
6. Presence of nystagmus, may indicate visually significant
7. Presence of strabismus, may indicate visually significant

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Slit lamp examination, intraocular pressure, corneal diameter
2. Fundus exam or B-Scan ultrasound to assess posterior pole
3. Bilateral congenital cataracts without family history
 - a. Examine family members for possible hereditary etiology
 - b. Consider TORCH titers and VDRL
 - c. Consider metabolic workup
 - d. Consider further systemic evaluation by geneticist or primary care physician

II. Define the risk factors

- A. Family history
- B. Intrauterine infection
- C. Trauma
- D. Medications
- E. Genetic or systemic disease
- F. Ocular anomalies
- G. Uveitis

III. List the differential diagnosis

- A. Congenital versus acquired
- B. Other causes of leukocoria

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

- A. Determine if cataract is visually significant
 1. Central dense cataracts 3 mm or greater
 2. Reduced visual acuity
 3. Effect on activities of daily living
- B. Nonsurgical management
 1. Observation
 2. Pharmacologic dilation
- C. Surgical management (See Pediatric cataract extraction) (See Intraocular lens implantation)
 1. Lensectomy
 2. Possible intraocular lens implantation
- D. Refractive correction
- E. Amblyopia therapy as required

V. List the complications of treatment (See Pediatric cataract extraction) (See Intraocular lens implantation)

- A. Endophthalmitis
- B. Glaucoma

- C. Corneal decompensation
- D. Secondary membrane formation
- E. Retinal detachment
- F. Cystoid macular edema
- G. Uveitis
- H. IOL dislocation
- I. Refractive surprises
- J. Poor compliance with amblyopia treatment

VI. Describe disease-related complications

- A. Amblyopia
- B. Strabismus
- C. Glaucoma
- D. Retinal detachment
- E. Nystagmus
- F. Surgical complications as described above
- G. Subnormal vision

VII. Describe appropriate patient instructions

- A. Early intervention
- B. Prescribed medications
- C. Postoperative instructions
 - 1. Watch for signs of infection
 - 2. Begin optical correction as soon as possible
 - 3. Instruct in proper use of drops (and oral medications if used)
 - 4. Keep all postoperative appointments
- D. Constant refractive correction is mandatory
- E. Patching compliance is mandatory
- F. Examinations under anesthesia are frequently needed
 - 1. Refraction
 - 2. Glaucoma
 - 3. Retinal disease
 - 4. Secondary opacification
- G. Appropriate ocular protection
- H. Lifelong monitoring required

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Sections 6 (Pediatric Ophthalmology and Strabismus) and 11 (Cataract), 2015-2016.
- 2. AAO, Focal Points: Pediatric Cataracts, Module #2, 2011.
- 3. Wright KW, Speigel PH. Pediatric Ophthalmology and Strabismus. New York. Springer, 2003.

4. Wilson EA, Trivedi RH and Pandey SK (Eds), Pediatric Cataract Surgery. Philadelphia, JB Lippincott Co, 2005.
5. Long V, Chen S, Hatt S. Surgical interventions for bilateral congenital cataract. Cochrane Database Syst Rev 2006;19:3:CD003171.
6. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.

Pediatric cataract extraction

I. List the indications/ contraindications

A. Indications

1. Visually significant lens opacity (See Congenital and acquired cataracts in children)
2. Lens-induced inflammation or glaucoma
3. Capsular rupture with lens swelling
4. Lens dislocation if visually significant or causing glaucoma
5. Timing is critical in pediatric patients

B. Contraindications

1. Lens opacities not visually significant
2. Medically unstable child
3. Active inflammation

II. Describe the pre-procedure evaluation

A. History

1. Child's visual behavior
2. Family history
3. Trauma
4. Age of onset of visual signs and symptoms
5. Pictures with abnormal red reflex

B. Clinical examination: note some elements may be performed under anesthesia prior to surgery

1. Visual/sensory functions
 - a. Acuity
 - b. Nystagmus
 - c. Strabismus
2. Quality of red reflex
3. Size and morphology of cataract before and after pharmacologic dilation
4. Pupillary evaluation
5. Corneal diameter
6. Intraocular pressure
7. Cycloplegic refraction
8. Fundus examination
9. B-Scan if no view to the posterior pole
10. Potential acuity meter if age appropriate
11. A-scan and keratometry if implanting an intraocular lens (IOL)
12. Systemic work up if warranted (See Congenital and acquired cataracts in children)

III. List the alternatives to this procedure

- A. **Chronic dilation (to see around opacity or around dislocated lens)**
- B. **Amblyopia therapy**

IV. Describe the instrumentation, anesthesia and technique

- A. **General anesthesia**
- B. **Scleral tunnel or clear cornea incision**
- C. **Anterior capsulorhexis**
 - 1. Continuous-tear capsulorhexis
 - 2. Vitrectorhexis
- D. **Lens aspiration**
- E. **Posterior capsulotomy and anterior vitrectomy**
 - 1. May omit if anticipate child's cooperation for Nd:Yag capsulotomy
 - 2. Expect posterior capsular opacification in young children
- F. **IOL implantation (See Intraocular lens implantation)**
 - 1. Depends upon age, laterality, eye size, comorbidities, family preference
- G. **Incision closed with sutures**
- H. **Subconjunctival antibiotic and/or steroid injections may be used**
- I. **Hard shield at the end of the procedure**

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

- A. **Inflammation**
 - 1. Increased in younger children and/or with IOL implantation
 - 2. Treat with topical and sometimes oral steroids
- B. **Secondary opacification of the visual axis**
 - 1. Common in infants when an IOL placed
 - 2. May require surgery
- C. **Glaucoma (aphakic or pseudophakic)**
 - 1. Common, especially with microphthalmos or persistent fetal vasculature (PFV)
 - 2. Onset may be rapid or years after lensectomy
- D. **Endophthalmitis: incidence similar to adults**
- E. **Retinal detachment**
- F. **Wound dehiscence**
- G. **Astigmatism**
- H. **Lens dislocation**

VI. Describe the follow-up care

- A. **Immediately postoperative period**
 - 1. Protection with a hard shield or eyeglasses
 - 2. Topical antibiotics
 - 3. Anti-inflammatory medications

4. Dilation to prevent synechiae
5. Optical rehabilitation
 - a. Aphakic spectacles for bilateral aphakia
 - b. Aphakic contact lenses for unilateral or bilateral aphakia
 - c. Spectacles for residual refractive error for unilateral or bilateral pseudophakia

B. Long term

1. Lifelong screening for glaucoma
2. Amblyopia therapy
3. Protective eyewear if indicated
4. Some aphakic children may be candidates for secondary IOL implantation

VII. Describe appropriate patient instructions

- A. Postoperative medications, patching/optical rehabilitation, compliance is essential for good outcome**
- B. Protect eye in immediate postoperative period**
- C. Report visual changes, pain, redness, discharge, clouding of cornea, changes in pupil shape, trauma**
- D. Contact lens education if used**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Focal Points: Pediatric Cataracts, Module #2, 2011.
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Intraocular lens implantation

I. List the indications/contraindications

A. Indications

1. Optical correction of aphakia
2. Intraocular lens (IOL) implantation may be considered in children of all ages under appropriate circumstances
 - a. Pediatric IOL implantation is considered investigational by the Food and Drug Administration
 - b. Role of IOL is controversial in infants

B. Contraindications

1. Relatively contraindicated in eyes with associated ocular abnormalities
 - a. Microcornea/sclerocornea
 - b. Microphthalmos/nanophthalmos
 - c. Uncontrolled glaucoma
 - d. Active inflammation/uveitis
2. Poor zonular or capsular support
 - a. Consider a sutured posterior chamber IOL or anterior chamber IOL
 - b. Long-term safety of these options in children is unknown
3. Higher complication rate with IOL implantation in infants less than 7 months of age

II. Describe the pre-procedure evaluation

A. A-scan and keratometry to determine IOL power calculation

1. Myopic shift is to be expected with eye growth
2. Formula and refractive target for IOL calculation is controversial

B. Assess capsular support for secondary IOL implantation

C. B scan if unable to view fundus

D. IOP measurement and pachymetry

III. List the alternatives to this procedure

A. Eyeglasses

B. Contact lenses

IV. Describe the instrumentation, anesthesia and technique

A. General anesthesia

B. Following cataract extraction, the IOL is implanted.

1. Implantation in capsular bag or in sulcus
2. Acrylic IOL's are commonly used
 - a. Foldable so can be inserted through smaller wound
 - b. Decreased incidence of posterior capsular opacification

- C. Primary posterior capsulectomy and limited anterior vitrectomy in young children
- D. Consider pharmacologic pupil constriction
- E. Incisions are usually sutured in children
- F. Anti-inflammatory/antibiotic measures

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

- A. Amblyopia
- B. Iris pigment loss
- C. Corectopia
- D. Vitreous loss
- E. Choroidal or retinal detachment
- F. IOL dislocation
- G. Glaucoma
- H. Uveitis
- I. Endophthalmitis
- J. Incorrect IOL power
- K. Unanticipated refractive error
- L. Opacification of visual axis
- M. Dislocation of sutured IOL

VI. Describe the follow-up care

- A. Immediate postoperative period
 - 1. Patch/shield
 - 2. Topical antibiotic
 - 3. Topical and/or oral anti-inflammatory medications
 - 4. Consider pupil dilation to prevent synechiae
- B. Long term
 - 1. Monitor for the development of glaucoma
 - 2. Treat amblyopia
 - 3. Optical correction with bifocal for refractive error
 - 4. Protective eye wear if unilateral amblyopia

VII. Describe appropriate patient instructions

- A. Postoperative care important
- B. Visual rehabilitation important
- C. IOL exchange or refractive surgery may be helpful later in life to reduce refractive error

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Focal Points: Pediatric Cataracts, Module #2, 2011.

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Common hereditary vitreoretinal disorders

I. Broad group of disorders

- A. Juvenile Retinoschisis
- B. Stickler Syndrome
- C. Norrie disease
- D. Familial exudative vitreoretinopathy

II. Juvenile retinoschisis: Describe the approach to establishing the diagnosis

- A. Describe the etiology and genetic basis for the disease
 - 1. X-linked disease
- B. Define the relevant aspects of epidemiology of the disease
 - 1. Occurs in approximately between 1 in 5000 to 1 in 25000 males
- C. List the pertinent elements of the history
 - 1. Male
 - 2. Decreased vision in early childhood
 - 3. Family history of vision loss
- D. Describe pertinent clinical features
 - 1. Foveal retinoschisis
 - 2. Peripheral retinoschisis in 50%
 - a. Retinoschisis occurs in nerve fiber layer
 - 3. Vitreous findings
 - a. Veils and strands
 - b. Vitreal syneresis or liquefaction
 - c. Hemorrhage
 - 4. Retinal detachment
- E. Describe appropriate testing and evaluation for establishing the diagnosis
 - 1. ERG findings
 - a. Reduction of scotopic b-wave with preservation of the a-wave
 - 2. Optical coherence tomography (OCT)
 - a. Schisis spaces in the middle layers of the macula
- F. Define the risk factors
 - 1. Family history
- G. List the differential diagnosis
 - 1. Stargardt disease
 - 2. Goldman-Favre syndrome
- H. Describe patient management in terms of treatment and follow-up

1. Medical therapy options
 - a. Gene replacement some success in mouse model
 2. Surgical therapy options
 - a. Photocoagulation
 - b. Retinal detachment surgery
 - c. Vitrectomy
- I. List the complications of treatment, their prevention and management**
1. Cataract
- J. Describe disease-related complications**
1. Vitreous hemorrhage
 2. Retinal detachment
- K. Describe appropriate patient instructions**
1. Contact sports should be avoided due to retina susceptible to trauma

III. Stickler Syndrome: Describe the approach to establishing the diagnosis

- A. Describe the etiology and genetic heterogeneity of this disease**
1. Several types with gene defects on chromosomes 1, 6, and 12
- B. Define the relevant aspects of epidemiology of the disease**
1. Autosomal dominant types in majority of types
- C. List the pertinent elements of the history**
1. Progressive arthropathy and degenerative joint changes
 2. Progressive myopia with blurred vision
 3. Progressive hearing loss
 4. Respiratory and feeding difficulties due to association with Pierre Robin sequence
- D. Describe pertinent clinical features**
1. Orthopedic and skeletal manifestations
 - a. Marfanoid skeletal habitus, subgroups with short stature
 2. Flat midface, cleft palate
 3. Progressive hearing loss
 4. Heart defects, mitral valve prolapse
 5. Pierre Robin sequence
 - a. Cleft palate, small mandible, and backward displacement of tongue
 - b. Finding high myopia in an infant with the Pierre Robin sequence is highly suggestive of Stickler syndrome
 6. Ocular manifestations
 - a. Severe myopia
 - b. Vitreoretinal manifestations
 - i. Vitreous liquefaction, fibrillar collagen condensation
 - ii. Lattice retinal degeneration
 - iii. Retinal folds, breaks, giant retinal tears and high risk of retinal detachment
 - iv. Pigment clumping around the retinal vessels

- v. High incidence of proliferative vitreoretinopathy
 - c. Lenticular manifestations
 - i. Cataracts
 - ii. Ectopia lentis
 - d. Strabismus and Amblyopia
 - e. Glaucoma
 - f. Ptosis
- E. Describe appropriate testing and evaluation for establishing the diagnosis**
 - 1. Family history and clinical examination
 - 2. Genetic testing
- F. Define the risk factors**
 - 1. Family history
- G. List the differential diagnosis**
 - 1. Degenerative myopia
 - 2. X-linked retinoschisis
 - 3. Marfan syndrome
 - 4. Other vitreoretinopathies
- H. Describe patient management in terms of treatment and follow-up**
 - 1. Describe medical therapy options
 - a. Conservative treatment
 - b. Eyeglasses or contact lenses for significant myopia
 - c. Patching for amblyopia
 - 2. Describe surgical therapy options
 - a. Laser photocoagulation for retinal breaks and holes
 - b. High incidence of vitreous loss during cataract surgery
 - i. Vitrectomy may be needed
 - c. High rate of subsequent retinal detachment following cataract surgery
 - d. Frequent follow-up is needed
 - e. Retinal detachment surgery may be needed
- I. List the complications of treatment, their prevention and management**
 - 1. Recurrent retinal detachment with proliferative vitreoretinopathy following retinal repair
 - 2. Potential perforation of the globe during strabismus or retinal surgery due to thin sclera
- J. Describe disease-related complications**
 - 1. Amblyopia due to severe myopia
 - 2. Strabismus
 - 3. Deafness
 - 4. Serious respiratory and feeding problems
- K. Describe appropriate patient instructions**
 - 1. Be cautious with full contact sports
 - a. Risk of retinal detachment
 - b. Risk of orthopedic injury

2. Low vision rehabilitation

IV. Norrie disease: Describe the approach to establishing the diagnosis

- A. Describe the etiology and genetic basis for the disease**
 1. X-linked recessive
- B. Define the relevant aspects of epidemiology of the disease**
 1. Rare, exact incidence unknown, no known racial or ethnic associations
- C. List the pertinent elements of the history**
 1. Male
 2. Blindness at birth
 3. Hearing impairment
 4. Intellectual disability
- D. Describe pertinent clinical features**
 1. Ophthalmic manifestations
 - a. Distinctive retinal appearance
 - b. Dystrophic retina
 - c. Pigmentary changes in the avascular periphery
 - d. Retinal detachment within a few days of birth
 - e. Progresses to white mass behind the clear lens
 - f. Progressive opacification of lens and cornea
 - g. Phthisis bulbi may ensue
 - h. Female carriers show peripheral retinal abnormalities
 2. Systemic manifestations
 - a. Hearing loss
 - b. Developmental delay
- E. Describe appropriate testing and evaluation for establishing the diagnosis**
 1. Complete ophthalmic exam
 2. Genetic testing
- F. Define the risk factors**
 1. None known
- G. List the differential diagnosis**
 1. Retinopathy of prematurity
 2. FEVR
- H. Describe patient management in terms of treatment and follow-up**
 1. No known treatment
- I. List the complications of treatment, their prevention and management**
 1. No known treatment
- J. Describe disease-related complications**
 1. Retinal detachment
- K. Describe appropriate patient instructions**
 1. Referral of parents for genetic counseling

V. Familial exudative vitreoretinopathy (FEVR): Describe the approach to establishing the diagnosis

A. Describe the etiology and genetic basis for the disease

1. Abnormal retinal vascularization similar to Retinopathy of Prematurity
2. Autosomal dominant or X-linked recessive

B. Define the relevant aspects of epidemiology of the disease

1. Rare, but prevalence unknown as people with normal vision may not come to medical attention

C. List the pertinent elements of the history

1. Full term infant

D. Describe pertinent clinical features

1. Ophthalmic manifestations are bilateral
 - a. Posterior pole folds
 - b. Retinal traction
 - i. May be associated with positive angle Kappa
 - c. Retinal breaks
 - d. Retinal detachment
 - e. Avascular peripheral retina
 - f. Peripheral intraretinal and subretinal exudates

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Fluorescein angiography shows areas of retinal nonperfusion

F. Define the risk factors

1. Family history

G. List the differential diagnosis

1. Retinopathy of Prematurity
2. Coats disease

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

1. Photocoagulation
2. Cryopexy
3. Retinal detachment surgery
4. Vitrectomy
5. Anti VEG-F

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

1. Bleeding
2. Cataract-may require surgery

J. Describe disease-related complications

1. Retinal detachment

K. Describe appropriate patient instructions

1. Examination of family members is important in the diagnosis

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Treatment of uveitis: corticosteroids

I. List the indications/contraindications

A. Indications

1. Findings consistent with anterior, intermediate, or posterior uveitis
2. To prevent complications of uveitis such as cataract, cystoid macular edema, band keratopathy, etc.
3. Assumes adequate pretreatment or concurrent coverage with antibiotics or antivirals as appropriate for any coexistent infection

B. Contraindications

1. Presence of concurrent infection, especially fungal, which could be worsened by corticosteroids
2. Corticosteroid-induced glaucoma may be a relative contraindication in some cases

II. Describe the pre-therapy evaluation

A. Medical history for causes of uveitis

B. Visual acuity

C. Evaluation of anterior chamber and anterior vitreous with slit-lamp biomicroscope when possible

D. Measurement of intraocular pressure (IOP) when possible

E. Dilated fundus examination

III. List the alternatives or adjuncts to this therapy

A. Topical or systemic nonsteroidal anti-inflammatory drugs (NSAIDs)

B. Systemic immunosuppressive agents

C. Cycloplegics

IV. Describe the technique

A. Topical agents

1. Shake all suspensions prior to use
2. Punctal occlusion
3. Used primarily for anterior uveitis

B. Oral agents

1. Prescribe appropriate corticosteroid taper
2. Consider H2 blocker or similar agent for gastric ulcer prophylaxis
3. Used for more severe uveitis or intermediate and/or posterior uveitis

C. Sub-Tenons injection

1. If done in office, appropriate explanation and topical anesthesia prior to injection
2. If cooperation is limited, perform under conscious sedation or mask anesthesia in operating room or minor procedure room
3. Most commonly used for intermediate uveitis

V. List complications of therapy, prevention, and management

A. Glaucoma

1. Prevention
 - a. Use less potent agents (e.g. fluorometholone or loteprednol) and limit dose and length of therapy of higher potency steroids (e.g. dexamethasone, prednisolone)
2. Treatment
 - a. Switch to corticosteroid sparing treatment, such as methotrexate or a biologic, with close involvement with rheumatologist (See Primary congenital (infantile) glaucoma)

B. Cataract

1. Prevention
 - a. Limit dose and length of therapy
2. Consider surgery if visually significant

C. Infection

1. Prevention
 - a. Limit dose and length of therapy
2. Treat or give prophylaxis with antibiotics or antivirals as appropriate

D. Perforation of globe

1. Possible complication of injection
2. Prevent by appropriate patient selection for office procedure and careful knowledge of ocular and orbital anatomy

E. Complications of systemic corticosteroid therapy

1. Gastritis
 - a. H2 blocker or similar agent
2. Growth retardation
3. Osteoporosis
4. Hyperglycemia
5. Irritability/mood changes
6. Fluid retention
7. Pseudotumor cerebri
8. Pituitary suppression
9. All complications lessened by minimizing dose and length of therapy

VI. Describe the follow-up care

- A. As clinically indicated for uveitis and/or complications of therapy
- B. Measure IOP when possible and clinically indicated
- C. Comanagement with primary care physician to monitor for complications when systemic agents used

VII. Describe appropriate patient instructions

- A. Return for worsening vision, pain, photophobia, etc.
- B. Take all medicines as prescribed with attention to tapering instructions
- C. Some physicians avoid vaccinations while child is on corticosteroids

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Focal Points: Steroid Therapy for Ocular Inflammatory Disease, Module #7, 2006.

Medical treatment of uveitis: mydriatic/cycloplegic agents

I. List the indications/contraindications

A. Indications

1. Anterior, intermediate or posterior uveitis
2. Ciliary spasm
3. Posterior synechiae

B. Contra-Indications

1. Use with caution in children with cardiac issues
2. Previous adverse effects of cycloplegic agents

II. Describe the pre-therapy evaluation

A. Work up for causes for inflammation

B. Degree of anterior chamber reaction or vitritis

C. Extent of synechiae

D. Anterior chamber depth, with gonioscopy if shallow

III. List the adjuncts to this therapy

A. Topical nonsteroidal anti-inflammatory drugs (NSAIDs)

B. Topical corticosteroid preparations

C. Systemic immunosuppression with corticosteroids and/or antimetabolite preparations

IV. Describe the instrumentation and technique

A. Application of short acting (tropicamide or cyclopentolate), intermediate (homatropine) or long term (atropine) cycloplegic drops to the affected eye

B. Application of sympathomimetic in office to break synechiae

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

A. Blurred vision

1. Prescribe correction of refractive error and add bifocal if therapy is protracted

B. Amblyopia

1. Correct refractive error and monitor vision

C. Systemic anticholinergic complications

1. Dry mouth; dry eyes
2. Psychologic symptoms especially in children like hyperactivity or hallucinations
3. Urinary retention
4. Constipation

5. Facial flushing
6. Dizziness

VI. Describe the follow-up care

- A. Weekly to monthly visits depending on degree and response of inflammation to treatment

VII. Describe appropriate patient instructions

- A. Wash hands before and after application
- B. Do not get medication in eyes not being treated
- C. Instruct patient/family on expected duration of cycloplegic effect of medication
- D. Keep drops out of the reach of children
- E. Separate eye drops by 2-5 minutes

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Juvenile idiopathic arthritis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Auto-immune

B. Define the relevant elements of epidemiology of the disease

1. Uveitis diagnosed before onset of arthritis (formerly known as juvenile rheumatoid arthritis), uncommon
2. Uveitis diagnosed after onset of arthritis usually within 7 years of onset, usual presentation
3. More common in girls than boys

C. List the pertinent elements of the history

1. Joint pain
2. Blurred vision
3. Irregular pupil or anisocoria
4. Change in iris color

D. Describe pertinent clinical features

1. Cell and flare in anterior chamber
2. Band keratopathy
3. Synechiae
4. Cataract
5. Heterochromia
6. Joint pain

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Antinuclear antibodies (ANA)
2. Rheumatoid factor
3. Rheumatology consultation

II. Define the risk factors (for development of uveitis)

- A. ANA+, rheumatoid factor negative, early age of onset
- B. Girls at increased risk
- C. Pauciarticular disease

III. List the differential diagnosis, uveitis

- A. Trauma
- B. Sarcoid
- C. Lyme
- D. Spondyloarthropathies
- E. Psoriatic arthritis
- F. Inflammatory bowel disease
- G. Multiple sclerosis

- H. **Bartonella henselae (cat scratch)**
- I. **Idiopathic**

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

A. Describe medical therapy options

1. Topical and/or systemic corticosteroids
2. Systemic immunosuppressives/immunomodulators (Disease modifying anti-rheumatic drug - DMARD or biologics)
3. Nonsteroidal anti-inflammatory drug (NSAID) drops
4. Cycloplegia if indicated
5. Glaucoma medication if high intraocular pressure

B. Describe surgical therapy options

1. Superficial keratectomy or chelation if visually significant band keratopathy
2. Treat glaucoma with appropriate glaucoma surgery
3. Remove cataract when visually significant, intraocular lens (IOL) can be cautiously considered when uveitis under best control

C. Serial screening exams in asymptomatic patients per guidelines

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

A. Cataract

1. Control inflammation
2. Use corticosteroids judiciously
3. Remove if vision decreased

B. Glaucoma

1. Control inflammation
2. Use corticosteroids sparingly
3. Use topical intraocular pressure (IOP) reducing medications and surgery if unable to control

C. Systemic immunosuppression

1. Close communication with pediatrician and rheumatologist

VI. Describe disease-related complications

- A. **Band keratopathy**
- B. **Amblyopia**
- C. **Medication side effects**
- D. **Cataract**
- E. **Synechiae**
- F. **Glaucoma**
- G. **Cystoid macular edema**
- H. **Phthisis bulbi**

VII. Describe appropriate patient instructions

- A. Follow-up is important**
- B. Disease may be present and causing damage even when asymptomatic**
- C. Education about eye irritation, redness or light sensitivity should prompt office visit**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
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Traumatic uveitis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Superficial or blunt trauma to the globe

B. List the pertinent elements of the history

1. History of globe trauma
2. May be unwitnessed and children may deny history; therefore, a high degree of suspicion warranted, especially if other corroborating evidence exists

C. Describe pertinent clinical features

1. Associated symptoms and signs of ocular trauma
 - a. Uveitis
 - i. Pain
 - ii. Photophobia
 - iii. Ciliary flush
 - iv. Cell and flare in the anterior chamber
 - v. Anterior and posterior synechiae
 - b. Hyphema may coexist
 - c. Anterior vitreous cell
 - i. Exclude retinal tear if suspect pigmented cells
 - d. Cataract/lens opacities (lens-related uveitis may be present if capsule open)
 - e. Elevated intraocular pressure (IOP) or hypotony
 - f. Hypopyon (suspect endophthalmitis)
 - g. Abnormalities of pupil size, shape, or reactivity
 - h. Commotio retinae
 - i. Retinal hemorrhage or tear
 - j. Choroidal rupture
 - k. Macular hole

II. Define the risk factors

A. Globe trauma

III. List the differential diagnosis

- A. idiopathic iritis
- B. Postoperative iritis following intraocular surgery
- C. Infectious endophthalmitis
- D. Exclude open globe injury

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

- A. Topical corticosteroids**
- B. Cycloplegics**
- C. Treat coexisting ocular, adnexal injuries**
- D. If hyphema present**
 - 1. Carefully observe for increased IOP or rebleed
 - 2. Sickle prep, especially if African-American
 - 3. Consider hospitalization of children if indicated for compliance
- E. If IOP is elevated, treat with topical and/or systemic agents and address any treatable causes of high IOP (i.e., lens material in eye, hyphema, angle closure)**

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

- A. Corticosteroid-related glaucoma**
 - 1. Treat with topical ocular antihypertensives
 - 2. Switch to alternate topical corticosteroid or taper and discontinue corticosteroid if iritis resolving
 - 3. Suspect other causes of glaucoma
- B. Blurred vision**
- C. Amblyopia if prolonged use of cycloplegic**

VI. Describe disease-related complications

- A. Glaucoma**
 - 1. Gonioscopy to assess risk if angle recession present
- B. Cataract**

VII. Describe appropriate patient instructions

- A. Discuss signs of retinal detachment or increased IOP**
- B. Avoid re-injury**
- C. Long term follow-up as recommended to detect glaucoma, cataract, or retinal complications**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Retinopathy of prematurity

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Normal retinal vascular development begins in utero, with blood vessels reaching nasal ora serrate by about 32 weeks gestation and blood vessels reaching temporal ora serrate shortly after a full-term birth
2. Normal development is altered by premature exposure to the extrauterine environment

B. Define the relevant aspects of epidemiology of the disease

1. Infants born weighing less than 1500 grams are at risk for serious visual sequelae
2. Risk of visual loss increases as gestational age and birth weight decrease

C. List the pertinent elements of the history

1. Birth weight
2. Gestational age at birth
3. Current and postmenstrual age
4. Significant systemic illnesses
5. Selection of patients for screening
 - a. All premature infants with
 - i. A birth weight less than 1500 grams
 - ii. A gestational age less than or equal to 30 weeks
 - iii. With an unstable clinical course felt to be at high risk for retinopathy of prematurity (ROP)
6. Timing of exams
 - a. Infants are first examined at 31 weeks postmenstrual age or 4 weeks chronological age, whichever is later
 - b. Then at least every 3-14 days thereafter, until vascularization proceeds to zone III or the risk of requiring treatment is passed (until postmenstrual age of 45 weeks and no prethreshold disease)

D. Describe pertinent clinical features

1. Zone - location of disease
 - a. Zone I
 - i. Posterior pole; circle centered on optic nerve with radius twice the nerve-macula distance
 - b. Zone II
 - i. Edge of Zone I to a circle centered on optic nerve with radius equal to the distance from the optic nerve to the nasal ora serrata
 - c. Zone III
 - i. Residual crescent anterior to Zone II
2. Stage - severity of disease
 - a. Stage 1 = demarcation line
 - b. Stage 2 = ridge
 - c. Stage 3 = ridge with extraretinal fibrovascular proliferation
 - d. Stage 4 = subtotal retinal detachment
 - e. Stage 5 = total retinal detachment
3. Plus disease - marked vascular dilation and tortuosity of posterior pole vessels; at least 2 quadrants meet or exceed the amount seen in a standard photograph

4. Types of ROP (ET-ROP)
 - a. Type I ROP
 - i. Degree of severity at which laser treatment is generally performed
 - ii. Defined as zone I, any stage with plus disease, or
 - iii. Zone I, stage 3 without plus disease, or
 - iv. Zone II, stage 2 or 3 with plus disease
 - b. Type II ROP
 - i. Zone I, stage 1 or 2 without plus disease, or
 - ii. Zone II, stage 3 without plus disease
 - c. Aggressive, posterior ROP (AP-ROP) - uncommon, rapidly progressing, severe form of ROP characterized by posterior location, prominence of plus disease, and the ill-defined nature of the retinopathy

II. Define the risk factors

- A. Low birth weight
- B. Young gestational age at birth
- C. Supplemental oxygen administration
- D. Multiple birth
- E. Birth outside a hospital with a neonatal intensive care unit
- F. Caucasian race
- G. Respiratory distress syndrome
- H. Intraventricular hemorrhage
- I. Sepsis
- J. Poor weight gain

III. List the differential diagnosis

- A. Early stage
 1. Familial exudative vitreoretinopathy
- B. Late stages
 1. Other causes of leukocoria
 - a. Persistent fetal vasculature (formerly known as persistent hyperplastic primary vitreous)
 - b. Retinoblastoma
 - c. Norrie disease
 - d. Incontinentia pigmenti
 - e. Coats disease
 - f. Coloboma
 - g. Toxoplasmosis
 - h. Toxocara

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

- A. Describe surgical therapy options

1. Laser photocoagulation of peripheral avascular retina
 - a. Early Treatment for ROP Study
 - i. Early treatment of high-risk Type 1 ROP significantly reduced risk of unfavorable visual outcome
 - b. Performed within 48 hours of diagnosis of Type I ROP
 - c. Re-treatment sometimes necessary
2. Anti-VEGF (Vascular Endothelial Growth Factor) drugs:
 - a. Compared with laser, (off-label) bevacizumab reduced the rate of recurrent neovascularization requiring retreatment for zone I eyes;
 - b. Optimal dosage and long-term effects are unknown;
 - c. Need for prolonged surveillance because of risk of late recurrence
3. Treatment options for retinal detachment
 - a. Observation only (some partial detachments)
 - b. Scleral buckle
 - c. Vitrectomy

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

- A. Inadvertent laser burns**
- B. Intense postoperative inflammation**
 1. Prevented by use of topical corticosteroids
- C. Cataract**
- D. Glaucoma**
- E. Posterior synechiae**
 1. Prevented by use of topical cycloplegic
- F. Hyphema**
- G. Vitreous hemorrhage**
- H. Phthisis bulbi**

VI. Describe disease-related complications

- A. Retinal detachment - partial or total**
- B. Macular heterotopia (macular dragging)**
- C. High myopia**
- D. Amblyopia**
- E. Strabismus**
- F. Anisometropia**
- G. Glaucoma**
- H. Phthisis bulbi**
- I. Pseudostrabismus secondary to positive angle kappa**

VII. Describe appropriate patient instructions

- A. Introduction to basic concepts of ROP**

- B. Discussion of risks and benefits of laser treatment**
- C. Discussion of possibility of progression of disease despite treatment and need for re-treatment**
- D. Awareness of importance of timely and appropriate outpatient follow-up**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
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Stargardt disease

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Stargardt disease is thought to be caused by impaired transport of vitamin A between the photoreceptor cells and the retinal pigment epithelium (RPE) resulting in photoreceptor death and the accumulation of lipofuscins in the retinal pigment epithelium
 - a. This is a result of a buildup of A2E, a toxic by product of Vitamin A that is normally released after light exposure during the visual cycle
2. Rhodopsin is found in the photoreceptor cells
 - a. They contain a pigment called retinal (made from vitamin A), which is converted to retinol when a rhodopsin structure is exposed to light
 - b. This process causes the rhodopsin to break down, and the resulting waste material is transported to the RPE for recycling
3. The gene that codes the protein responsible for the transport of retinol to the RPE is the ABCA4 (formerly ABCR) gene
 - a. If this gene is defective, transport cannot take place, and the waste remains in the photoreceptor tissue, where the toxic A2E poisons the healthy cells
 - i. ABCA4 gene encodes ATP binding (ABCR)
 - ii. ABCA4 expressed in cones and rods
 - iii. ABCA4 spectrum retinal diseases mild late onset STGD1 to severe early onset RP
 - iv. Early onset highly predictive of ABCA4 mutations
4. Stargardt disease can be caused by mutations in the ABCA4 gene
5. Autosomal recessive (STGD1)
6. Autosomal dominant
 - a. STGD3 autosomal dominant macular atrophy with flecks. Mutation ELOVL4 6q14
 - b. STGD4 autosomal dominant mutation PROM1 4p15.32

B. Define the relevant aspects of epidemiology of the disease

1. Stargardt disease is one of the most frequent causes of macular degeneration in childhood
2. Most prevalent inherited juvenile-onset retinal dystrophy
3. It has an estimated incidence of 1 in 10,000
4. It is usually autosomal recessive but may be dominant

C. List the pertinent elements of the history

1. The child will describe difficulty in central vision especially reading with onset between 8 and 15 years of age or just a subjective loss of sharpness of vision
2. The child may also describe a prolonged dark adaptation after exposure to sunlight
3. Color vision can deteriorate and the child can be light sensitive

D. Describe pertinent clinical features

1. This is a bilateral symmetrical condition
2. The fundus may appear normal in the early stages even when some vision has been lost. At this stage, the child may be misdiagnosed as having functional visual loss
3. The first fundus signs are loss of foveal reflex, followed by development of a typical bull's eye lesion
 - a. Yellow flecks (lipid rich deposits accumulated in the RPE layer) in the posterior pole are

characteristic but not essential for the diagnosis

- b. Eventually the macula may acquire an atrophic appearance with a 'beaten bronze' appearance
- c. This macula lesion enlarges and deepens and eventually may show choroidal atrophy with prominent choroidal vessels

E. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Fundoscopy for the features described above
- 2. Fluorescein angiography
 - a. May show the characteristic "bull's eye" with macular hyperfluorescence due to a "window effect"
 - b. 'Choroidal silence' or 'dark fundus' may be seen in 50-80% of cases due to a blocking effect in the retinal periphery caused by lipofuscin deposits
 - c. Autofluorescence of lipofuscin
- 3. Electroretinography
 - a. The condition displays variable electroretinogram (ERG) abnormalities depending on disease severity and may be normal in the early stages
 - b. Prolonged periods of dark adaptation can also be found
- 4. Optical coherence tomography
 - a. Thickening of the external limiting membrane
 - b. Photoreceptor abnormalities
 - c. Retinal pigment epithelial abnormalities
- 5. Genetic testing for ABCA4 gene mutation
- 6. Fundus Autofluorescence; there are areas hypofluorescence-dark macula with or without areas of hyperfluorescence

II. Define the risk factors

- 1. Family history

III. List the differential diagnosis

- A. Early-onset progressive cone-rod dystrophies can be difficult to differentiate from Stargardt disease: the former usually progresses to below 20/200 vision whereas the latter usually stabilizes at 20/200**
- B. Neuronal ceroid lipofuscinosis (Batten disease) may have a bull's eye maculopathy sign but there is systemic neurological involvement and this must be excluded**
 - 1. It is worth following a child for 6-12 months after the initial diagnosis before giving a definitive diagnosis
 - 2. Children with progressive neurological deficits should be worked up for neurodegenerative diseases such as Batten disease
- C. Fundus flavimaculatus (FFM) is an allelic condition very similar to Stargardt disease but tends to spare the macula with peripheral fleck-like lesions**
 - 1. FFM has a later age of onset
 - 2. If loss of visual acuity begins in the first 2 decades, the designation Stargardt disease is preferred
 - 3. If loss of visual acuity begins later in life and has a more progressive course, the term FFM is preferred

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

A. Describe the natural history, outcome and prognosis

- 1. A progressive condition with central vision leveling out at 20/200 with normal peripheral vision

B. Describe medical therapy options

1. No known treatment
2. There has been a suggestion that reduction of exposure to ultraviolet (UV) light in animal model decreases accumulation of lipofuscin
3. Supplemental Vitamin A may accelerate visual loss and should be avoided

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

A. No established treatment

VI. Describe disease-related complications

- A. Loss of central vision but maintenance of peripheral vision**
- B. Rarely, choroidal neovascularization may form**

VII. Describe appropriate patient instructions

- A. Low visual aid guidance, with registration as legally blind to help with educational needs**
- B. Support groups may be helpful**

Additional Resources

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Albinism

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Oculocutaneous albinism (OCA)
 - a. Autosomal recessive
 - b. Eyes, skin, hair lack pigment
2. Ocular Albinism (OA)
 - a. X-linked recessive
 - b. Hypopigmentation mainly affects eyes
3. Error in melanin metabolism
 - a. Misrouting of optic nerve fibers
 - b. Hypopigmentation of retina, iris, skin and hair

B. Define the relevant aspects of epidemiology of the disease

1. Hermansky-Pudlak
 - a. Autosomal recessive
 - b. OCA plus bleeding diathesis, pulmonary fibrosis and granulomatous colitis
 - c. Puerto Rican heritage
2. Chediak-Higashi
 - a. Autosomal recessive
 - b. OCA 2
 - c. Recurrent infections
 - d. Neurologic deficits

C. List the pertinent elements of the history

1. Present at birth
2. Nystagmus in first few months of life
3. Blue or light colored eyes common
4. Another family member affected

D. Describe pertinent clinical features

1. Skin, hair, and ocular hypopigmentation
2. Photophobia
3. Decreased acuity due to foveal hypoplasia
4. Strabismus
5. Positive angle kappa
6. Nystagmus
7. History of easy bruising (Hermansky-Pudlak syndrome)
8. Recurrent infections (Chediak-Higashi syndrome)
9. Iris transillumination defects

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Visual evoked potential (VEP) may help confirm diagnosis
 - a. Patients show an asymmetry of VEP
 - b. Secondary to misrouting of optic pathways
2. Hematologic workup in selected cases if Hermansky-Pudlak or Chediak-Higashi syndromes are suspected

II. Define the risk factors

- A. Positive family history

III. List the differential diagnosis

- A. Aniridia
- B. Nystagmus (congenital), idiopathic
- C. Vitiligo
- D. Cone dystrophy
- E. X-linked juvenile retinoschisis

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

- A. Describe medical therapy options
 1. Eyeglasses/contact lenses for significant refractive errors (possible bifocals)
 2. Tinted lenses
 3. Vision rehabilitation referral
 4. Patching for amblyopia
- B. Describe surgical therapy options
 1. Strabismus surgery
 2. Nystagmus surgery

V. Describe disease-related complications

- A. Amblyopia
- B. Photophobia
- C. Strabismus
- D. Low vision
- E. Sun-related skin damage

VI. Describe appropriate patient instructions

- A. UV protection
- B. Need for eyeglasses/contact lenses
- C. Need for vision rehabilitation aids
- D. Consider genetic counseling

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Kerr R, Stevens G, Managa P, et al. Identification of P gene mutations in individuals with oculocutaneous albinism in sub-Saharan Africa. *Hum Mutat* 2000;15:166-172.
3. Oetting WS, Summers CG, King RA. Albinism and the associated ocular defects. *Metab Pediatr Syst Ophthalmol* 1994;17:5-9.
4. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
5. Up To Date 2015 Genodermatoses. <http://www.uptodate.com/contents/the-genodermatoses>

Optic nerve hypoplasia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Multifactorial
2. First trimester intrauterine insult
3. Idiopathic

B. Define the relevant aspects of epidemiology of the disease

1. Unilateral or bilateral
2. Often asymmetric if bilateral
3. No sex predilection

C. List the pertinent elements of the history

1. Congenital visual impairment
2. Abnormal eye movements (nystagmus)
3. Mother with insulin-dependent diabetes mellitus
4. Maternal drug/alcohol use in pregnancy
5. Associated neurological symptoms
 - a. Neonatal seizures
6. Strabismus

D. Describe pertinent clinical features

1. Decreased number of optic nerve axons
2. Small and pale optic nerve
3. Yellow/white ring around optic disc ("double ring sign")
4. Frequent association with retinal vascular tortuosity
5. Wide range of visual acuity levels (normal to no light perception)
6. Visual field defects
7. Associated strabismus may be present
8. May be associated with nystagmus
9. Relative afferent pupillary defect if asymmetric
10. Risk for hypothalamic/pituitary dysfunction and associated midline central nervous system anomalies (Septo-optic dysplasia)

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Neuroimaging to assess for ectopic pituitary bright spot and structural midline defects
2. Consider visual field testing if age appropriate
3. Endocrine evaluation even if MRI normal

II. Define the risk factors

A. Maternal diabetes mellitus

1. Associated with superior segmental optic nerve hypoplasia

B. Fetal alcohol syndrome

- C. Maternal drug ingestion**
 - 1. Phenytoin
 - 2. Quinine
 - 3. LSD
- D. Periventricular leukomalacia (PVL)**
- E. Primiparity**
- F. Young maternal age**

III. List the differential diagnosis

- A. Optic atrophy**
- B. Tilted optic discs**
- C. High hyperopia**

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

- A. Describe medical therapy options**
 - 1. Safety eyeglasses for protection and/or refractive error correction
 - 2. Appropriate endocrine treatment
 - 3. Amblyopia treatment
 - 4. Strabismus surgery if ocular misalignment present

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

- A. Standard endocrine management**

VI. Describe disease-related complications

- A. Poor vision**
- B. Nystagmus**
- C. Visual field loss**
- D. Central nervous system anomalies, e.g., septo-optic dysplasia**
- E. Endocrine dysfunction**

VII. Describe appropriate patient instructions

- A. Amblyopia treatment compliance**
- B. Follow up with primary care provider (PCP)**
- C. Discuss with family and PCP the importance of following growth chart and establishing care with Endocrinology**
- D. Provide patient and family with low vision assessment and vision rehabilitation services as required**

Additional Resources

- 1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. Hellstrom A, Wiklund LM, Svennson E. The clinical and morphologic spectrum of optic nerve hypoplasia. J

AAPOS. 1999;3:212-220.

Optic atrophy

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Tumor compression or infiltration
2. Hereditary
3. Anoxic encephalopathy
4. Intraventricular hemorrhage (IVH)
5. Periventricular leukomalacia (PVL)
6. Hydrocephalus
7. Glaucoma
8. Retinal degenerative condition (secondary)
9. Demyelination
10. Inflammatory optic neuropathy
11. Trauma
12. Toxic
13. Previous meningitis
14. Nutritional (i.e., vitamin B deficiency)

B. Define the relevant aspects of epidemiology of the disease

1. Dependent on underlying causative diagnosis
 - a. Hereditary
 - b. Acquired
2. Can be unilateral or bilateral

C. List the pertinent elements of the history

1. Age of onset
2. Duration
3. Perinatal difficulties
4. Family history of decreased vision
5. Drug use
6. Results of vision testing in school
7. Associated neurologic diagnosis
8. Associated systemic diagnosis
9. Hydrocephalus
10. Headache
11. Toxic exposures
12. Diet history

D. Describe pertinent clinical features

1. Decreased or decreasing vision
2. Impaired color vision

3. Nystagmus
4. Sluggish or paradoxical pupil response
5. Pale, but normal sized optic nerve head
6. Attenuated retinal arterioles and / or pigmentary abnormalities if associated with retinal degenerative syndromes

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Acuity
2. Color vision
3. Pupils
4. Intraocular pressure
5. Consider visual field
6. Examine other family members
7. Neurological evaluation
8. Neuroimaging
9. When familial, genetic evaluation with DNA evaluation if Lebers hereditary optic neuropathy or dominant optic atrophy are suspected
10. Laboratory testing for nutritional and toxic etiologies if clinically indicated
11. Electroretinogram if associated with retinal pathology

II. Define the risk factors

- A. Optic nerve or chiasmal compressive lesion
- B. Infiltrative tumor i.e. optic nerve glioma in neurofibromatosis
- C. Cerebral palsy
- D. Prematurity (IVH, PVL)
- E. Optic neuritis - demyelinating
- F. Autoimmune disorder
- G. Infectious disorder
- H. Chronic hydrocephalus
- I. End stage glaucoma

III. List the differential diagnosis

- A. Optic nerve hypoplasia
- B. Optic nerve coloboma

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

- A. Describe the medical therapy options
 1. Treat underlying etiology if possible
 2. Provide low vision assessment

V. Describe disease-related complications

- A. Progressive loss of all vision

- B. Loss of vision with stabilization**
- C. Visual field defects**

VI. Describe appropriate patient instructions

- A. Eye examinations at appropriate intervals**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
3. AAO, Focal Points: Optic Atrophy, Module #2, 2006.

Optic neuritis in children

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Inflammatory disorder of the optic nerve
2. Post infectious or post immunization
3. Multiple sclerosis
4. Acute disseminated encephalomyelitis (ADEM)
5. Neuromyelitis optica (NMO)

B. Define the relevant aspects of epidemiology of the disease

1. Presentation either unilateral or bilateral
 - a. < 10 years old most often bilateral
 - b. > 10 years old most often unilateral
2. Presence or absence of MRI white matter abnormalities outside the visual system
 - a. Higher risk of the development of MS with MRI white matter abnormalities outside the visual system
3. Risk of development of MS
 - a. Unilateral vs. bilateral is not a risk factor, although greater age and MRI white matter abnormalities at a higher risk

C. List the pertinent elements of the history

1. Age of child
2. Time of onset visual loss
3. Area of visual loss, central or peripheral field
4. Pain on eye movements
5. Other neurologic or systemic symptoms
6. Recent infections or immunizations, i.e. mumps, rubella, hepatitis B vaccine
7. Family history of autoimmune disorders

D. Describe pertinent clinical features

1. Decreased vision, unilateral or bilateral
2. Possible optic disc swelling
3. Possible relative afferent papillary defect
4. Impaired color vision
5. Possible pain on eye movements
6. Probable visual field defect
7. Absence of retinal or cortical lesions
8. Presence or absence of MRI abnormalities outside the visual system
9. Possible other neurologic findings
 - a. Isolated neurologic findings
 - b. Transverse myelitis (Neuromyelitis Optica) (NMO)
 - c. Acute disseminated encephalomyelitis (ADEM)

E. Describe appropriate laboratory testing for establishing the diagnosis

1. Neuro-imaging
2. Visual evoked potentials
3. Serologic studies to rule out infectious or autoimmune optic neuropathy
4. Cerebrospinal fluid analysis to include cell count, protein and oligoclonal bands
5. NMO-IgG/Aquaporin-4 autoimmunity
6. Consider Leber and OPA testing

II. Define the risk factors

- A. Post infectious
- B. Family history

III. List the differential diagnosis

- A. Compressive optic neuropathy
- B. Infectious optic neuropathy e.g. Lyme, Bartonella
- C. Toxic / metabolic optic neuropathy
- D. Leber's optic neuropathy
- E. Autosomal dominant optic neuropathy
- F. Autoimmune optic neuropathy
- G. Functional visual loss
- H. Papilledema/Increased intracranial pressure (when macular edema present)
- I. Neuroretinitis

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

- A. Describe the medical therapy options
 1. Intravenous Solu-Medrol followed by oral prednisone taper
 2. Other immune modulating agents

V. Describe disease-related complications

- A. Loss of vision
- B. Visual field defects
- C. Recurrence
- D. Development of other neurologic defects

VI. Describe appropriate patient instructions

- A. Comply with treatment regimen
- B. Keep follow up visits with subspecialists
- C. Report development of other neurologic symptoms

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Brady KM, Brar AS, Lee AG, Coats DK, Paysse EA, Steinkuller PA: Optic Neuritis in Children: Clinical features and Visual Outcome. JAAPOS 1999;3;98-103.

Papilledema

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Increased intracranial pressure

B. List the pertinent elements of the history

1. Headache
2. Nausea
3. Vomiting
4. Transient visual obscurations
5. Diplopia
6. Pulsatile tinnitus

C. Describe pertinent clinical features

1. Optic disc elevation
2. Opacification of peripapillary nerve fiber layer
3. Flame-shaped hemorrhages at the disc margin
4. Disc hyperemia
5. Obscuration of retinal vessels at disc margin
6. Loss of venous pulsations
7. Venous dilation and tortuosity
8. Exudates
9. Nerve fiber layer infarcts
10. Esotropia due to a sixth nerve palsy
11. Enlarged blind spot on visual field
12. Visual acuity usually normal except in chronic disc swelling or atrophy, or lipid/fluid extension into macula (macula star)

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) or magnetic resonance imaging (MRI) of the head
2. Consider MRI/MRV
3. Lumbar puncture if above is normal with opening pressure and CSF analysis

II. Define the risk factors

- A. Presence of intracranial mass causing obstructive hydrocephalus
- B. Hydrocephalus
- C. Venous sinus thrombosis
- D. Malignant hypertension
- E. Obesity and associated IIH
- F. Meningitis
- G. Increased intracranial pressure
- H. Craniosynostosis

III. List the differential diagnosis

- A. Pseudopapilledema**
- B. Optic nerve sheath tumor**
- C. Optic neuritis**
- D. Other infectious or inflammatory optic neuropathies**
- E. Infiltrative optic neuropathy**
- F. Intrinsic optic nerve tumor**

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

- A. Describe medical therapy options**
 - 1. Specific treatment dependent on underlying cause of papilledema
 - 2. Monitor visual acuity
 - 3. Follow visual field
 - 4. Follow appearance of optic nerve heads - photos
 - 5. Follow nerve fiber layer thickness - OCT
- B. Describe surgical therapy options**
 - 1. Dependent on underlying cause of papilledema

V. Describe disease-related complications

- A. Loss of visual acuity**
- B. Loss of visual field**

VI. Describe appropriate patient instructions

- A. Follow up with ophthalmologist to monitor visual function and response to any specific treatment**
- B. Follow up with primary care provider or appropriate specialist for any systemic condition**

Additional Resources

- 1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. AAO, Focal Points: Optic Atrophy, Module #2, 2006.

Idiopathic intracranial hypertension (pseudotumor cerebri)

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. Equal incidence male: female prepubertal
2. Prepubertal not necessarily obese
3. Weight (obesity) and female gender post pubertal
4. Predominance increase during adolescence

C. List the pertinent elements of the history

1. Headaches
2. Transient visual obscurations
3. Diplopia
4. Nausea/vomiting
5. Tinnitus
6. Recent weight gain
7. Snoring
8. Use of medications that have been associated with pseudotumor cerebri (section II A)

D. Describe pertinent clinical features

1. Papilledema
2. Preserved central vision unless chronic disease
3. Diplopia secondary to cranial nerve (CN) VI palsy
4. Macular star possible

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Neuroimaging
 - a. To rule out hydrocephalus
 - b. Assess for slit-like ventricles
 - c. Assess for empty sella
 - d. Consider magnetic resonance venogram to rule out venous sinus thrombosis
2. Elevated opening pressure with normal cerebrospinal fluid chemistry on lumbar puncture exam
3. Defects may or may not be present on visual field testing
 - a. Enlarged blind spot
 - b. Arcuate defects

II. Define the risk factors

A. Associated with medication use

1. Tetracyclines
2. Corticosteroids, especially withdrawal
3. Vitamin A - oral or topical; or derivatives (isotretinoin)
4. Oral contraceptives
5. Growth hormone

- B. Associated with cranial venous sinus thrombosis**
- C. Associated with obesity**
- D. Associated with sleep apnea**
- E. Associated with anemia**

III. List the differential diagnosis

- A. Secondary causes of papilledema with elevated intracranial pressure**
 1. Intracranial mass causing obstructive hydrocephalus
 2. Meningitis
- B. Pseudopapilledema**

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

- A. Describe medical therapy options**
 1. Stop offending agent
 2. Acetazolamide (Diamox®)
 3. Weight loss as appropriate
- B. Describe surgical therapy options**
 1. Neurosurgical shunting procedures
 2. Optic nerve sheath fenestration
 3. Venous sinus stenting

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

- A. Acetazolamide**
 1. Metabolic acidosis
 2. Tingling around mouth, hands and feet
 3. Nausea/dyspepsia
 4. Diuresis
- B. Neurosurgical shunt**
 1. Shunt malfunction
 2. Shunt infection
 3. Other surgical complications
- C. Optic nerve sheath fenestration**
 1. Visual loss and permanent optic neuropathy or central retinal artery occlusion
 2. Infection
 3. Diplopia if EOM disturbed or displaced during procedure

VI. Describe disease-related complications

- A. Progressive and permanent visual loss with associated optic atrophy
- B. Chronic headache

VII. Describe appropriate patient instructions

- A. Compliance with medical therapy
- B. Weight loss as appropriate
- C. Notify appropriate physicians if adverse drug reaction or progressive symptoms despite treatment

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Optic nerve drusen

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Congenital anomaly
2. Usually bilateral

B. List the pertinent elements of the history

1. Suspicion for disc edema in past
2. Deep circumlinear disc hemorrhage in the past
3. Loss of visual acuity or visual field
4. Family history
5. Absence of diplopia or other neurologic dysfunction to suggest increased intracranial pressure

C. Describe pertinent clinical features

1. "Lumpy-bumpy" appearance of the optic nerve
2. Disc elevation
3. Spontaneous venous pulsations helpful if present to indicate within normal intracranial pressure
4. May have a deep circumlinear disc margin hemorrhage
5. Visual field defect can occur
6. Can be calcified
7. Become exposed in teen age years
8. Do not leak fluorescein

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Imaging of optic nerve (ultrasound or computed tomography)
2. Visual field
3. Autofluorescent photography
4. Fluorescein angiography

II. Define the risk factors

A. Parent with drusen of the optic nerve

III. List the differential diagnosis

A. Papilledema

B. Other congenital disc abnormalities

1. Pseudopapilledema
 - a. Anomalous retinal vessel branching pattern
 - b. Extra vessels emanating from the disc
 - c. May see glial tissue overlying disc
 - d. Absent cup

C. Other optic neuropathies - infectious/inflammatory/ischemic/infiltrative

- D. Optic neuritis
- E. Crowded discs in high hyperopia
- F. Astrocytoma of the optic disc (tuberous sclerosis)

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

- A. No proven medical or surgical treatment for progressive visual field loss
- B. Observation

V. Describe disease related complications

- A. Visual field defects may be progressive
- B. Deep circumlinear disc hemorrhages at disc margin
- C. Peripapillary choroidal neovascularization

VI. Describe appropriate patient instructions

- A. Discuss problem, and if permitted, document disc appearance (photo or sketch) for future reference
- B. Discuss possibility of progressive visual field loss

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Davis PL, Jay WM. Optic nerve head drusen. Semin Ophthalmol. 2003 Dec;18(4):222-42.

Orbital rhabdomyosarcoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Malignant neoplasia of extraocular muscle precursor cells found in orbital soft tissue

B. Define the relevant aspects of epidemiology of the disease

1. Most common primary orbital malignancy in childhood
2. Typical age at diagnosis is 5-7 years
3. Slightly higher male predominance

C. List the pertinent elements of the history

1. Sudden onset and rapid evolution of proptosis
2. Periorbital ecchymosis
3. Subacute presentation less common
4. History of trauma may be elicited - a red herring
5. Vision loss

D. Describe pertinent clinical features

1. Unilateral proptosis
2. Globe displacement
3. Ptosis, lid edema
4. Mass may be palpable, most commonly in superior nasal orbit
5. Pain may be present but less commonly

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Neuroimaging
2. Prompt orbital biopsy
 - a. Prognosis varies by histopathologic type
 - b. Cross striations diagnostic if present

II. List the differential diagnosis

- A. Orbital hemangioma
- B. Orbital cellulitis
- C. Inflammatory pseudotumor
- D. Orbital lymphangioma
- E. Orbital dermoid
- F. Ocular/orbital trauma
- G. Neuroblastoma
- H. Ewing sarcoma
- I. Optic nerve glioma
- J. Chalazion (rarely)

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

A. Describe medical therapy option

1. Combination chemotherapy
2. External beam irradiation
3. Corneal exposure
 - a. Artificial tears
 - b. Lubricating ointments

B. Describe surgical therapy options

1. Surgical resection for smaller well-circumscribed tumors
2. Debulking for less well-defined tumors/often contraindicated by possible damage to surrounding tissue

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

A. Chemotherapy

1. Bone-marrow suppression
2. Anemia
3. Immunocompromise

B. Radiation therapy

1. Mid-face bony hypoplasia
2. Radiation retinopathy
3. Optic atrophy
4. Dry eye syndrome
5. Corneal scarring
6. Enophthalmos
7. Cataract

V. Describe disease-related complications

A. Strabismus, diplopia

1. Usually resolves with treatment

B. Corneal exposure

1. Topical lubricants

C. Hyperopia, astigmatism due to tumor compression

1. Usually resolves with treatment

D. Vision loss

1. Lid effects
2. Amblyopia
3. Corneal scarring
4. Direct optic nerve compression/damage

VI. Describe appropriate patient instructions

A. Orbital biopsy is needed urgently

- B. Multidisciplinary approach is critical**
- C. Overall prognosis of rhabdomyosarcoma isolated to the orbit is good**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Breitfeld PP, Lynden E, Raney RB, et al. Ifosfamide and etoposide are superior to vincristine and melphalan for pediatric metastatic rhabdomyosarcoma when administered with irradiation and combination chemotherapy: a report from the Intergroup. Rhabdomyosarcoma Study Group. J Pediatr Hematol Oncol. 2001;23:225-33.
3. Raney RB, Anderson JR, Barr FG, et al. Rhabdomyosarcoma and undifferentiated sarcoma in the first two decades of life: a selective review of intergroup rhabdomyosarcoma study group experience and rationale for Intergroup Rhabdomyosarcoma Study V. J Pediatr Hematol Oncol. 2001;23:215-20.
4. Raney RB, Meza J, Anderson JR, et al. Treatment of children and adolescents with localized parameningeal sarcoma experience of the Intergroup Rhabdomyosarcoma Study Group protocols IRS II through-IV, 1978-1997. Med Pediatr Oncol. 2002;38:22-32.
5. Donaldson SS, Meza J, Breneman JC, et al. Results from the IRS-IV randomized trial of hyperfractionated radiotherapy children with rhabdomyosarcoma -a report from the IRSG. Int J Radiat Oncol Biol Phys. 2001;51:718-28.
6. Mackall CL, Helman LJ. High-dose chemotherapy for rhabdomyosarcoma: where do we go from here. J Pediatr Hematol Oncol. 2001;23:266-7.
7. Crist WM, Anderson JR, Meza JL, et al. Intergroup rhabdomyosarcoma study-IV: results for patients with nonmetastatic disease. J Clin Oncol. 2001;19:3091-102.
8. Baker KS, Anderson JR, Link MP, et al. Benefit of intensified therapy for patients with local or regional embryonal rhabdomyosarcoma: results from the Intergroup Rhabdomyosarcoma Study IV. J Clin Oncol. 2000;18:2427-34.
9. Flamant F, Rodary C, Rey A, et al. Treatment of non-metastatic rhabdomyosarcomas in childhood and adolescence. Results of the second study of the International Society of Paediatric Oncology: MMT84. Eur J Cancer. 1998;34:1050-62.

Neuroblastoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Embryonal malignancy arising from neuroblasts
2. Usually originates in adrenal gland or sympathetic chain

B. Describe the relevant aspects of epidemiology of this disease

1. One of the most common childhood cancers
2. Orbital metastases can occur
3. Usually presents in infants and toddlers
4. Orbital involvement may be the initial manifestation of the tumor

C. List the pertinent elements of the history and clinical features

1. Ocular signs
 - a. Orbital metastases
 - i. Proptosis
 - ii. Periorbital/lid ecchymosis
 - b. Opsoclonus (rapid, multidirectional saccadic eye movements)Paraneoplastic effect from tumor antibodies
 - i. Associated with a more favorable prognosis
 - c. Horner syndrome
 - i. Secondary to tumor compression of the cervical or thoracic sympathetic chain
2. Systemic signs
 - a. Increased abdominal girth
 - b. Neurologic changes
 - c. Emesis
 - d. Weight loss
 - e. Anorexia
 - f. Diarrhea
 - g. Bone pain
 - h. Edema
 - i. Hypertension

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Imaging to identify primary tumor site
2. Urine catecholamine testing
3. Oncology consultation
4. Tumor biopsy may be needed

II. List the differential diagnosis

A. Rhabdomyosarcoma

B. Orbital hemangioma

- C. Lymphangioma
- D. Optic glioma
- E. Orbital cellulitis
- F. Other metastatic tumors
- G. Nonaccidental trauma
- H. Opsoclonus-myoclonus
- I. Post-viral opsoclonus

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

- A. Referral to a pediatric oncology center familiar with treatment
- B. Treatment dependent upon staging
- C. Describe medical therapy options
 - 1. Combination chemotherapy
 - 2. Radiation
- D. Describe surgical therapy options
 - 1. Surgical excision

IV. Describe disease-related complications

- A. Death
- B. Opsoclonus
- C. Side effects or complications of treatment for primary disease
- D. Morbidity (Refer to section IC2 Systemic Signs)

V. Describe appropriate patient instructions

- A. Stage of the tumor at the time of diagnosis and age of the patient are important prognostic factors

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
3. Ahmed A, Goel S, Khandwala M, Agrawal A, Chang B, Simmons IG. Neuroblastoma with orbital metastasis: ophthalmic presentation and role of ophthalmologists. *Eye* 2006; 20: 466-470.
4. Mahoney NR, Liu GT, Menacker SJ, et al. Pediatric horner syndrome: etiologies and roles of imaging and urine studies to detect neuroblastoma and other responsible mass lesions. *Am J Ophthalmol.* 2006; 142: 651-9.
5. Smith SJ, Diehl NN, Smith BD, Mohny BG. Incidence, ocular manifestations, and survival in children with neuroblastoma: a population-based study. *Am J Ophthalmol.* 2010;149:677-682.

Hemangioma (Capillary hemangioma)

I. Describe the approach to establishing the diagnosis

A. Define the relevant aspects of epidemiology of the disease

1. Relatively common
2. Females predominate
3. Rarely associated with family history of capillary hemangioma

B. List the pertinent elements of the history

1. Present at birth or in first six months of life
2. Initially small but grows rapidly in the first six months of life
3. Spontaneous involution begins after first year of life
4. Majority are completely regressed by age 8 years

C. Describe pertinent clinical features

1. Skin may have a raised, red dimpled appearance
2. Subcutaneous lesions are smooth, dark red to blue
3. Most commonly involve the upper eyelid with extension into the orbit and surrounding tissues +/- axial proptosis
4. Mass may swell when the baby cries or is held in Trendelenburg position
5. Similar lesions can be found on other parts of the body
 - a. Visceral lesions can be widespread
 - b. Wheezing indicates airway involvement
6. Association of systemic disease and capillary hemangioma
 - a. PHACE(S) syndrome: *posterior fossa malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, eye abnormalities, sternal clefting and supraumbilical raphe*
 - b. Clinician should have raised level of suspicion when child has larger segmental facial hemangiomas

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Imaging modalities
 - a. Ultrasound
 - b. Neuroimaging

II. List the differential diagnosis

- A. Nevus flammeus
- B. Rhabdomyosarcoma
- C. Lymphangioma (with hemorrhage into lesion)
- D. Cavernous hemangioma

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

- A. Describe medical therapy options

1. Conservative treatment (watchful waiting) if not significant
2. Eyeglasses for significant refractive errors
3. Patching for amblyopia
4. Topical or systemic beta-blocker
5. Oral corticosteroids

B. Describe surgical therapy options

1. Intralesional corticosteroids
2. Complete or partial excision in selected circumscribed lesions
3. Consider laser surface ablation to treat those hemangiomas with prominent skin component

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

A. Corticosteroid related complications

1. Pituitary-adrenal suppression
2. Skin depigmentation, subcutaneous fat atrophy, eyelid necrosis
3. Central retinal artery occlusion by particles of the drug
 - a. Dilated fundus examination at time of injection

B. Beta blocker related complications

1. Hypoglycemia, hypotension, bradycardia

V. Describe disease-related complications

- A. Anisometropic astigmatism
- B. Ptosis
- C. Strabismus
- D. Amblyopia
- E. Thrombocytopenia

VI. Describe appropriate patient instructions

- A. Regular follow-up during the growth phase of the lesion
- B. Need for patching, eyeglasses if significant growth
- C. Need for treatment if sight threatened
- D. Reconstructive treatments deferred until child older as the lesion may spontaneously regress by the second decade
- E. Encourage follow-up with primary care provider

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Haik BG, Karcioğlu ZA, Gordon RA, Pechous BP. Capillary hemangioma (infantile periorbital hemangioma). *Surv Ophthalmol.* 1994;38:399-426.
3. Kushner BJ. Hemangiomas. *Arch Ophthalmol.* 2000;118:835-836.
4. Propranolol for severe hemangiomas of infancy. Leaute-Labreze C, Dumas de la Roque E, Hubiche T, Boralevi F, Thambo JB, Taieb A. *N. Engl. J. Med.* 2008;358:2649-51.

Lymphangioma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease--unknown

B. Define the relevant aspects of epidemiology of the disease

1. No sex predilection
2. Not hereditary

C. List the pertinent elements of the history

1. Presents in infancy or childhood
2. May involve the conjunctiva, lids, or orbit

D. Describe pertinent clinical features

1. Subconjunctival hemorrhage
2. Diffuse hematoma of the eyelid
3. Proptosis
 - a. Result of hyperplasia of lymphoid tissue which may worsen during an upper respiratory infection
 - b. Results from rapid expansion from an intralesional bleed- "chocolate cyst"

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Magnetic resonance imaging(MRI)
 - a. Infiltrative lesion with variable enhancement
2. Computed tomography (CT)
 - a. Low density cyst-like mass
3. Ultrasound
 - a. Cystic spaces
4. Biopsy if available imaging is uncharacteristic of lymphangioma because of risk of hemorrhage
 - a. Histopathology shows thin, endothelium lined vascular channels with lymphatic fluid, poorly encapsulated

II. List the differential diagnosis

A. Capillary hemangioma

B. Rhabdomyosarcoma

C. Neuroblastoma

D. Ewing sarcoma

E. Leukemia

F. Histiocytosis X

G. Optic nerve glioma

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

A. Describe medical therapy options

1. Conservative management

- a. Cold compresses
 - b. Lubrication for corneal exposure
- 2. Systemic corticosteroids
- B. Describe surgical therapy options**
 - 1. Surgical decompression if optic nerve is compromised
 - 2. Carbon dioxide laser to debulk tumor
 - 3. Intralesional injection of a sclerosing agent

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

- A. Complications**
 - 1. Orbital hemorrhage
 - 2. Damage to orbital structures, optic nerve
- B. Prevention**
 - 1. Avoid biopsy unless diagnosis is unclear
 - 2. Avoid aggressive surgery

V. Describe disease related complications

- A. Strabismus**
- B. Ptosis**
- C. Optic nerve compression**
- D. Exposure keratopathy**
- E. Proptosis**
- F. Amblyopia**

VI. Describe appropriate patient instructions

- A. Follow-up based on severity of clinical situation**
- B. Call immediately with any new bleeding, proptosis, change in vision**

Additional Resources

- 1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. Giguere CM, Bauman NM, Sato Y, et al. Treatment of lymphangiomas with OK-432 (Picibanil) sclerotherapy: a prospective multi-institutional trial. Arch Otolaryngol Head Neck Surg. 2002;128:1137-44.
- 3. Muallem MS, Garzosi HJ. Conservative management of orbital lymphangioma. J Pediatr Ophthalmol Strabismus. 2000;37:41-43.
- 4. Wright JE, Sullivan TJ, Garner A, et al. Orbital venous anomalies. Ophthalmology. 1997;104: 905-913.

Idiopathic orbital inflammatory disease (orbital pseudotumor)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Idiopathic

B. Define the relevant aspects of epidemiology of the disease

1. Female predominance

C. List the pertinent elements of the history

1. Acute
2. Painful
3. Diplopia
4. Associated systemic complaints
 - a. Headache
 - b. Fever
 - c. Nausea and vomiting
 - d. Lethargy

D. Describe pertinent clinical features

1. Bilaterality and episodic recurrence are common
2. Proptosis, motility disturbance, injection, lid swelling, tenderness
3. Uveitis may be present

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Imaging studies (computed tomography (CT), magnetic resonance imaging (MRI) or ultrasound)
 - a. Increased density of orbital fat
 - b. Thickening of extraocular muscle and tendon
 - c. Thickening of posterior sclera

II. Define the risk factors

- ### A. Rarely associated with a systemic disorder

III. List the differential diagnosis

- A. Orbital cellulitis
- B. Orbital mass lesion
- C. Asymmetric thyroid eye disease (thyroid orbitopathy)
- D. Cavernous sinus lesion
- E. Scleritis
- F. Infiltrative disease (i.e. lymphoma)

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

A. Describe medical therapy options

1. Systemic corticosteroids
2. Systemic immunosuppressant
3. Topical lubricants
4. Radiation for refractory cases

B. Describe surgical therapy options

1. Diagnostic biopsy

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

A. Complications of corticosteroid and immunosuppressant therapy

VI. Describe disease-related complications

A. Spread of inflammation into ocular structures and posterior structures

VII. Describe appropriate patient instructions

A. Compliance with appropriate therapy

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Retinoblastoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Neuroblastic tumor arising from retinal cells
2. Inactivation of the retinoblastoma (*RB1*) tumor suppressor gene, or its deletion, will permit development of tumor

B. Define the relevant aspects of epidemiology of the disease

1. In children, most common malignant ocular tumor
2. Sporadic (somatic mutation) , about 60% of patients
3. Hereditary (germ-line mutation), in the remaining 40% of patients
4. Less than 25% of affected children will have a positive family history
5. Hereditary cases will frequently have bilateral disease, earlier presentation during the first year of life, and multifocal disease
6. Parents with a bilateral presentation have a higher chance of having a child with this tumor

C. List the pertinent elements of the history

1. Abnormal red reflex
2. Positive family history for tumor or enucleation early in life
3. Unilateral disease detected 18 - 24 months
4. Bilateral disease detected within the first year of life
5. Onset rare after 5 years of age, may present as diffuse infiltrating type retinoblastoma
6. Child not appearing to see well
7. Strabismus, usually esotropia
8. Parent with treated or regressed tumor

D. Describe pertinent clinical features

1. Solitary or multifocal white to cream colored solid masses in the retina or in the vitreous
 - a. Endophytic that breaks through internal limiting membrane with associated vitreous seeding
 - b. Exophytic may occur in the subretinal space
2. Retinal detachment
3. Calcification in mass
4. Anterior chamber seeding, pseudohypopyon, rare
5. Periocular inflammation, proptosis and glaucoma are other ways the tumor presents
6. Enlargement of the optic nerve due to extension posterior through the lamina cribrosa
7. Pineal gland enlarged due to "trilateral retinoblastoma" pineoblastoma

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. B scan useful to detect intraocular mass
2. Neuroimaging
 - a. MRI imaging
 - i. Preferred to decrease radiation exposure
 - ii. Required to rule out pineoblastoma

- b. Intraocular calcification present on computed tomography (CT) scan
- 3. Oncologic consultation
- 4. Genetics consultation

II. Define the risk factors

- A. Parent with tumor in one eye, 15% chance that offspring will be affected
- B. Parent with bilateral tumor, 45% chance that offspring will be affected

III. List the differential diagnosis

- A. Persistent fetal vasculature (previously known as persistent hyperplastic primary vitreous)
- B. Coats disease
 - 1. Yellowish to greenish subretinal exudates with or without hemorrhage
 - 2. Telangiectatic vessels always present, but may be difficult to see on fundoscopy
 - 3. Usually unilateral but can present bilaterally, and in adults
 - 4. Male predilection
 - 5. Average age of 5 years at presentation
 - 6. Exam under anesthesia with fluorescein angiography and angiography showing leakage from telangiectatic vessels, along with ultrasound and MRI may be needed to differentiate from retinoblastoma
- C. Norrie disease
- D. Retinopathy of prematurity
- E. Toxoplasmosis
- F. Toxocara canis
- G. Familial exudative vitreoretinopathy
- H. Cataract

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

- A. Describe medical therapy options
 - 1. Chemo-reduction with systemic chemotherapy followed by local therapy (described below)
 - 2. Intra-arterial chemotherapy
- B. Describe local therapy options
 - 1. Cryotherapy
 - 2. Laser ablation
 - 3. Radiotherapy
 - a. Plaque
 - b. External beam (in cases resistant to other treatments or with local extension)
 - 4. Enucleation
 - 5. Excise as long a segment of optic nerve as possible to ensure clean margins

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

- A. Cytotoxic drugs can alter reproductive genetic codes

B. Radiation can cause

1. Cataract
2. Hypoplasia of bone
3. Optic nerve atrophy
4. Radiation retinitis
5. Secondary tumors (sarcomas)

VI. Describe disease-related complications

- A. Metastasis (via blood, CSF or local invasion)
- B. Secondary tumors (usually osteosarcoma)- higher risk with external beam radiation
- C. Chance of dying with retinoblastoma if not treated approaches 100%, but if treated, survival is 90% or better
- D. Chance of dying because of secondary tumor is greater than death from retinoblastoma

VII. Describe appropriate patient instructions

- A. Follow-up visits will be arranged depending on the size and location and stage of treatment
- B. Appointments must be kept
- C. Periodic examinations under anesthesia will be necessary
- D. All family members must be examined to determine the risk for transmission
- E. Siblings must receive examinations, the frequency will depend on the age, and risk
- F. Protective eyewear must be used in children with loss of vision in an eye

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Laser Photocoagulation of the Retina and Choroid, 1997.
3. Smith BJ, O'Brien JM. The genetics of retinoblastoma and current diagnostic testing. J Pediatr Ophthalmol Strabismus 1996;33:120-123.
4. Shields CL, Shields JA. Recent developments in the management of retinoblastoma. J Pediatr Ophthalmol Strabismus 1999;36:8-18.
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Ophthalmologic manifestations of neurofibromatosis

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Two forms of the disease with distinct genetics, ocular manifestations, and management
 - a. NF-1 autosomal dominant inheritance on chromosome 17
 - b. NF-2 autosomal dominant inheritance on chromosome 22

B. Define the relevant aspects of epidemiology of the disease

1. NF-1 is the most common of the phakomatoses
2. NF-2 rare

C. List the pertinent elements of the history

1. Positive family history
2. Tumors of the skin or nervous system
3. Learning disabilities/developmental delay
4. Pigmented skin lesions

D. Describe pertinent clinical features

1. NF-1
 - a. Diagnostic clinical criteria - two or more of the following:
 - i. Café-au-lait spots
 - ii. Neurofibroma or plexiform neurofibroma
 - iii. Axillary, inguinal, or intertriginous freckling
 - iv. Optic nerve or pathway glioma
 - v. Lisch nodules (2 or more) develop over time with increasing age (2 or more)
 - vi. Distinctive bony lesions including sphenoid dysplasia
 - vii. A first-degree relative with NF-1
 - b. Other ocular manifestations
 - i. Choroidal nevi common
 - ii. Prominent corneal nerves
 - iii. Retinal hamartomas
 - iv. Glaucoma may occur in the setting of an ipsilateral plexiform neurofibroma of the eyelid
 - c. Associated presenting signs
 - i. Amblyopia
 - ii. Proptosis
 - iii. Strabismus
 - iv. Optic nerve atrophy
2. NF-2
 - a. Bilateral acoustic neuromas
 - b. Posterior subcapsular cataracts in young adults

- c. Combined hamartoma of retina and retinal pigment epithelium
- d. Benign tumors (neurofibroma, meningioma, schwannoma)
- e. Epiretinal membrane

E. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Genetic evaluation
- 2. Ophthalmic evaluation for diagnostic features of disease

II. Define the risk factors

A. Genetic predisposition

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

A. In genetically predisposed children with NF-1

- 1. Ophthalmologic examinations with special attention to detection of Lisch nodules and optic disc changes
 - a. Vision
 - b. Visual fields
 - c. Color vision
 - d. Intraocular pressure (IOP)
 - e. Pupillary examination for relative afferent pupillary defect in the setting of an optic nerve glioma
 - f. Consider magnetic resonance imaging (MRI) of brain and orbits

B. In children with established diagnosis

- 1. More frequent follow-up required for
 - a. Optic nerve or chiasmal glioma
 - b. Plexiform neurofibroma involving orbit or eyelid (amblyopia, glaucoma)

C. Optic nerve or chiasmal glioma requires multi-disciplinary care

D. Surgical therapy for plexiform neurofibroma

- 1. Debulk plexiform neurofibroma if amblyogenic or occludes visual axis
- 2. Surgical treatment of secondary glaucoma if topical ocular anti-hypertensives inadequate

E. Standard eye muscle surgery if strabismus present

IV. List the complications of treatment, their prevention and management in NF-1

A. Standard surgical risks for strabismus, glaucoma, lid surgery

V. Describe disease-related complications in NF-1

- A. Open-angle glaucoma**
- B. Anisometropia**
- C. Amblyopia**
- D. Strabismus**
- E. Cataract**
- F. Optic atrophy**
- G. Obstructive hydrocephalus**

- H. Proptosis, exposure keratopathy
- I. Blindness
- J. Increased incidence of malignancies

VI. Describe appropriate patient instructions

- A. Emphasize need for periodic follow-up examinations
- B. Referral to other pediatric subspecialists as needed
- C. Genetic counseling

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Kaye LD, Rothner AD, Beauchamp GR, Meyers SM, Estes ML. Ocular findings associated with neurofibromatosis type II. *Ophthalmol* 1992;99:1424-1429.
3. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
4. Avery RA, Fisher MJ, Liu GT. Optic pathway gliomas. *J Neuroophthalmol* 2011; 31:269-78. Review.

Sturge Weber syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Unknown; sporadic inheritance

B. List the pertinent elements of the history

1. Lesions present at birth
2. Typically, unilateral facial redness/asymmetry
3. Ocular injection
4. Possible ipsilateral enlarged cornea
5. Possible signs of glaucoma (epiphora, photophobia, blepharospasm)
6. Seizures frequent

C. Describe the pertinent clinical features

1. Facial nevus flammeus
2. Increased vascularity of conjunctival/episcleral vessels
3. Possible enlarged cornea with or without Descemet breaks
4. Possible elevated intraocular pressure (IOP) especially if upper eyelid involved with nevus flammeus secondary to presumed elevated episcleral venous pressure
5. Possible choroidal hemangioma - "tomato catsup fundus"—may be progressive with overlying retinal degeneration
6. Possible optic disc cupping
7. Meninges/cortex/white matter may be involved with vascular abnormalities
8. Seizures and mental retardation common
9. Hemiplegia/hemianopia may occur
10. Magnetic resonance imaging (MRI)/computed tomography (CT) scan shows cerebral calcifications
11. All findings typically unilateral but bilateral involvement can occur

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. MRI/CT to rule out intracranial vascular abnormalities
2. Examination under anesthesia often needed for IOP, if cannot be performed during clinic exam

II. List the differential diagnosis

A. Capillary hemangioma vs. nevus flammeus

B. Isolated megalocornea vs. true glaucoma

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

A. Describe medical therapy options

1. Topical beta-adrenergic antagonists or carbonic anhydrase inhibitors
2. Regular monitoring of IOP and refractive status
3. Monitor for amblyopia

B. Describe surgical therapy options

1. Goniotomy/trabeculotomy/trabeculectomy/tube shunt for glaucoma
2. Cycloablative procedures in severe cases
3. Laser therapy for facial hemangioma
4. Choroidal hemangioma
 - a. No treatment available

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

- A. Choroidal hemorrhage a risk of glaucoma surgery: avoid excessive hypotony (See Primary congenital (infantile) glaucoma)**

V. Describe disease-related complications

- A. Glaucoma due to increased episcleral pressure and/or angle malformation**
- B. Anisometropia**
- C. Amblyopia**
- D. Choroidal exudation/retinal degeneration in some cases of choroidal hemangioma**

VI. Describe appropriate patient instructions

- A. Counsel parents regarding absence of genetic role**
- B. Frequent exams to monitor or check for development of glaucoma**
- C. If glaucoma present, multiple procedures often necessary, and blindness can occur**
- D. Amblyopia therapy, eyeglasses often necessary**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.

Shaken baby syndrome/non-accidental trauma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Abrupt acceleration/ deceleration injury ruptures bridging intracranial vessels and compresses brain against skull
2. Poor neck stabilization in youngest children

B. Define the relevant aspects of epidemiology of this disease

1. Usually under 3 years of age

C. List the pertinent elements of the history

1. Often unreliable history
2. Trauma history inconsistent with clinical picture
3. Trauma inconsistent with developmental stage or abilities of child
4. History usually involves inconsolable crying child or obtunded/comatose child

D. Describe pertinent clinical features

1. May have lack of external evidence of trauma
2. Central nervous system findings
3. Subdural or subarachnoid hemorrhage
4. May have fractures of different ages
5. Ocular
 - a. Hemorrhages in multiple retinal layers
 - b. Hemorrhages often concentrated near macula
 - c. Vitreous hemorrhages
 - d. Full thickness perimacular retinal folds
 - e. Traumatic retinoschisis
 - f. Usually bilateral, but may be unilateral

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Alert child protective services
2. Full clinical and ophthalmic examination
3. Head magnetic resonance imaging (MRI)/computed tomography (CT) scan
4. X-ray for spiral fractures in long bones
5. Hematologic evaluation (rule out bleeding disorders, leukemia, etc.)

II. Define the risk factors

A. May occur in children of all socioeconomic levels

B. Trauma under suspicious circumstances or multiple injuries of characteristic patterns

C. Cardiopulmonary resuscitation (CPR) is not a risk factor for retinal hemorrhages in multiple layers

III. List the differential diagnosis

- A. Accidental trauma**
- B. Birth trauma**
- C. Bleeding disorder**
- D. Terson syndrome**

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

- A. Social service involvement**
- B. Describe medical therapy options**
 - 1. Treat amblyopia
 - 2. Systemic treatment as needed
- C. Describe surgical therapy options**
 - 1. Vitrectomy if appropriate

V. Describe disease-related complications

- A. Death**
- B. Neurologic complications**
- C. Vision loss**
 - 1. Traumatic retinoschisis
 - 2. Optic nerve damage
 - 3. Cortical damage
 - 4. Amblyopia

VI. Describe appropriate patient instructions

- A. Child must be removed from at-risk environment**
- B. Child's interests come first**
- C. Recovery is injury dependent**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Superficial ocular injury

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Accidental injury
2. Consider nonaccidental trauma

B. List the pertinent elements of the history

1. Circumstances of injury should be consistent with level of injury
 - a. If not, consider child abuse
2. Sudden onset of pain
3. Foreign body sensation
4. Redness
5. Tearing
6. History of contact lens use

C. Describe pertinent clinical features

1. Corneal abrasion
 - a. Fluorescein staining
 - b. Intact corneal sensation
2. Thermal burns of the cornea
 - a. White corneal epithelium
 - b. Otherwise, appears similar to corneal abrasion
3. Chemical burns of the cornea
 - a. Can be similar to corneal abrasion and thermal burns of the cornea, depending on pH of solution
 - b. pH of cul-de-sac determines alkalinity or acidity
4. Corneal foreign bodies
 - a. Possible corneal abrasion with particulate material on cornea or lodged under the lids

II. Define the risk factors

A. Lack of adult supervision

B. Male sex

C. Lack of appropriate eye protection during injuring activity

III. List the differential diagnosis

A. Corneal abrasion

1. Herpes keratitis

B. Thermal burns

1. Chemical burns
2. Corneal abrasion
3. Corneal ulcer

C. Chemical burns

1. Thermal burns
2. Corneal abrasion
3. Corneal ulcer

D. Corneal foreign body

1. Herpes keratitis

E. Severe dry eye

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

A. Describe medical therapy options

1. In all superficial ocular injuries, consider topical cycloplegic and antibiotic drops or ointment
2. Never patch eye if organic material is the cause of the abrasion or if the cause is unclear
3. In chemical burns, initial treatment includes copious irrigation with a balanced salt or saline solution and removal of particulate matter
 - a. Test the pH of the fornix until pH is neutral
 - b. Patching is contraindicated
4. Shield the eye

B. Describe surgical therapy options

1. Foreign bodies
 - a. Remove the foreign body from the palpebral conjunctiva or the cornea (deeply embedded inert foreign bodies may be left in place)

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

A. Amblyopia

1. Carefully monitor vision during treatment
2. Patching or penalization for amblyopia

VI. Describe disease-related complications

A. Amblyopia

B. Corneal scarring

C. Secondary infection

VII. Describe appropriate patient instructions

A. Administer medications as prescribed

B. Return for scheduled follow-up appointments

C. Watch for signs of infection and return to clinic immediately if they occur

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Penetrating injury in children

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. High velocity projectile
2. Sharp object
3. Metal on metal strike

B. Define the relevant aspects of epidemiology of the disease

1. More common in males than females

C. List the pertinent elements of the history

1. History of trauma
2. May have systemic injuries
3. Decreased vision
4. Photophobia
5. Pain

D. Describe the pertinent clinical features

1. Exposed uvea, vitreous, retina
2. Positive Seidel test
3. Visualization of intraocular foreign body
4. Eyelid laceration
5. Orbital chemosis
6. Conjunctival hemorrhage/laceration
7. Focal iris-corneal adhesion
8. Shallow anterior chamber
9. Iris defect
10. Peaked pupil
11. Hypotony
12. Lens capsule defect
13. Lens opacity
14. Retinal tear or hemorrhage
15. Decreased visual acuity
16. Possible abnormal ocular rotation

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan with thin cuts through the orbit to exclude intraocular foreign body

II. Define the risk factors

A. Lack of adequate eye protection

B. Unsupervised child

C. High risk activities: paintball, bb gun

- D. Child in close proximity to adults working on high risk tasks (Refer to section I.A. Describe the etiology of the disease)
- E. Unrestrained passenger

III. List the differential diagnosis

- A. Perforating injury
- B. Injury due to blunt trauma

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

A. Describe medical therapy options

1. Protective shield over the injured eye
2. Nothing by mouth
3. Pain control
4. Antiemesis medication
5. Intravenous antibiotics
6. Tetanus prophylaxis

B. Describe surgical therapy options

1. Restore the integrity of the globe
2. Explore for muscle and extraocular muscle insertion tears
3. Remove any intraocular foreign bodies
4. Iris repair
5. Cataract extraction+/- intraocular lens insertion
6. Retina repair when indicated

V. Describe disease-related complications

- A. Sympathetic ophthalmia
- B. Retinal detachment
- C. Cyclitic membrane formation
- D. Phthisis bulbi
- E. Endophthalmitis
- F. Retained intraocular foreign body
- G. Amblyopia
- H. Loss of vision
- I. Glaucoma

VI. Describe appropriate patient instructions

- A. Complete course of prescribed antibiotics
- B. Follow-up for refraction and correction with eyeglasses/contact lens
- C. Follow-up for amblyopia treatment
- D. Follow-up as needed for any other ocular problems

E. Appropriate eye protection

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Hyphema

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Superficial, penetrating, or blunt trauma to the globe
2. Child abuse
3. Nontraumatic spontaneous causes
 - a. Retinoblastoma
 - b. Juvenile xanthogranuloma of the iris
 - c. Bleeding disorders

B. Define the relevant aspects of epidemiology of the disease

1. Trauma more common in males
2. Family history for nontraumatic causes of hyphema

C. List the pertinent elements of the history

1. History of trauma
1. Skin lesions
2. Iris heterochromia
3. Tendency toward easy bruising
4. History of eye surgery
5. Sickle trait/anemia

D. Describe pertinent clinical features

1. Other signs of ocular trauma
2. Blood in the anterior chamber
3. Pain
4. Photophobia
5. Ciliary flush
6. Red blood cell and flare in the anterior chamber
7. Anterior vitreous cells
8. Synechiae
9. Increased intraocular pressure (IOP)
10. Other signs of non-accidental trauma

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Ultrasound to evaluate posterior segment if unable to be visualized on clinical exam
2. Pediatric referral if child abuse suspected
3. Complete blood count (CBC) and coagulation studies if bleeding disorder suspected
4. Sickle cell prep in selected patients
5. Measurement of intraocular pressure

II. Define the risk factors

- A. Globe trauma
- B. Child abuse
- C. Family history of nontraumatic causes of hyphema
- D. Juvenile xanthogranuloma

III. List the differential diagnosis

- A. Iritis
- B. Infectious endophthalmitis
- C. Leukemia

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

A. Describe medical therapy options

1. Reasonable limitation of activity
2. Consider topical corticosteroids
3. Consider cycloplegic agents
4. Pressure lowering medications as necessary with special attention to increased risk of elevated IOP in setting of sickle cell trait/anemia to avoid optic neuropathy
5. Cessation of non-steroidal anti-inflammatory drugs or aspirin as these medications may increase the risk of further bleeding
6. Consider antifibrinolytic agents

B. Describe surgical therapy options

1. Appropriate surgical evacuation of hyphema
2. Examination under anesthesia, IOP monitoring if necessary
3. Glaucoma surgery when appropriate

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

A. Corticosteroid-induced glaucoma if corticosteroids used

1. Treat with topical ocular antihypertensives

B. Nausea, vomiting and systemic hypotension if oral antifibrinolytic agent is used or if IOP is high

1. Treat by discontinuing oral agent

VI. Describe disease-related complications

- A. Rebleed, with greatest risk in the first 5 days post hyphema
- B. Synechiae
- C. Angle-recession glaucoma
- D. Corneal blood staining
- E. Deprivation amblyopia
- F. Sensory strabismus
- G. Associated ocular injuries

VII. Describe appropriate patient instructions

- A. Comply with treatment recommendations until hyphema resolves**
- B. Gonioscopy performed as patient cooperation allows in traumatic causes of hyphema**
- C. Lifetime annual follow-up in patients with angle recession**
- D. Follow-up as indicated for etiology of nontraumatic cases of hyphema**

Additional Resources

- 1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
- 2. Pieramici DJ, Goldberg MF, Melia M, et al. A phase III, multicenter, randomized, placebo-controlled clinical trial of topical aminocaproic acid (Caprogel) in the management of traumatic hyphema. *Ophthalmology*. 2003;110:2106-12.
- 3. Teboul BK, Jacob JL, Barsoun-Homsey M, et al. Clinical evaluation of aminocaproic acid for managing traumatic hyphema in children. *Ophthalmology*. 1995;102:1646-563.
- 4. Rahmani B, Jahadi HR. Comparison of tranexamic acid and prednisolone in the treatment of traumatic hyphema: a randomized clinical trial. *Ophthalmology*. 1999;106:375-379.
- 5. Shiuey Y, Lucarelli MJ. Traumatic hyphema. *Ophthalmology*. 1998;105:851-855.

Orbital fractures

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Blunt facial trauma

B. Define the relevant aspects of epidemiology of the disease

1. Objects larger than the orbital opening impact the orbit rim

C. List the pertinent elements of the history

1. Head and facial trauma

D. Describe pertinent clinical features

1. Ecchymosis of the involved eye
2. Epistaxis
3. Orbital emphysema
4. Enophthalmos
5. Possible reduced vision from injury to globe or optic nerve.
6. Possible diplopia in some or all positions of gaze
7. Limited vertical gaze secondary to entrapment of the vertical rectus muscles, inferior oblique muscle, or surrounding tissue
8. Limited horizontal gaze secondary to entrapment of the horizontal rectus muscles or surrounding tissue
9. Paresthesia or hypoesthesia in the infraorbital area secondary to damage to the infraorbital nerve
10. Nasolacrimal system injury with epiphora, lacrimal sac mucocoele, or dacryostenosis
11. Rhinorrhea if orbital roof fracture and cerebrospinal fluid leakage
12. Traumatic ptosis
13. Possible tripod fracture involving zygomatic arch
14. Possible LeFort fracture

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Computed tomography (CT) scan of orbits, direct axial and coronal
2. Magnetic resonance imaging (MRI) scan of orbit with gadolinium contrast, fat saturation technique
3. Forced duction testing can help differentiate restrictive entrapment from cranial nerve paresis

II. Define the risk factors

A. Lack of proper restraint in automobile

B. Not using protective eyewear or headgear in sports related injury

III. List the differential diagnosis

A. Cranial neuropathy leading to motility disturbance

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

A. Describe medical therapy options

1. Treat related globe or optic nerve injury appropriately
2. Antibiotic prophylaxis
3. Nasal decongestants
4. Consider steroids to decrease edema if indicated
5. Prism therapy

B. Describe surgical therapy options

1. Urgent surgical intervention is required in children who have a blowout fracture and a finding of oculocardiac reflex (bradycardia, nausea/vomiting)
2. Urgent surgical intervention is recommended when there is clinical and/or radiographic evidence of muscle entrapment
3. Some clinicians advocate immediate exploration and repair of fracture
4. Some clinicians recommend waiting 5 to 10 days until orbital edema and hematoma subside, and repair fracture only if persistent diplopia or risk of enophthalmos
5. Repair of residual strabismus by standard eye muscle surgical techniques
6. Posterior fixation suture of contralateral inferior rectus (IR) where diplopia occurs in downgaze but not primary position

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

- A. Persistent diplopia despite repair of fracture, and/or strabismus repair may require occluder or prisms to alleviate any residual diplopia**

VI. Describe disease-related complications

- A. Globe or optic nerve related injury**
- B. Orbital cellulitis**

VII. Describe appropriate patient instructions

- A. If medial wall fracture, ask patients to not blow nose, thereby preventing orbital emphysema with possible infection or secretions**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Bonsagi ZC, Meyer DR. Internal orbital fractures in the pediatric age group -characterization and management. *Ophthalmol* 2000;107:829-836.
3. Manson PN, Iliff N. Early repair for selected injuries. In: Dutton JJ, Slamovits T, eds. Viewpoints. Management of blowout fractures of the orbital floor. *Surv Ophthalmol* 1991;35:280-292.
4. Putterman AM. The conservative approach. In: Dutton JJ, Slamovits T, eds. Viewpoints. Management of blowout fractures of the orbital floor. *Surv Ophthalmol* 1991;35:292-298.
5. AAO, Ophthalmology Monographs 17. Strabismus Surgery: Basic and Advanced Strategies, 2004.

Persistent fetal vasculature

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Incomplete regression of hyaloid vasculature system
2. Formerly known as persistent hyperplastic primary vitreous

B. Define the relevant aspects of epidemiology of the disease

1. No sex predilection
2. Not hereditary

C. List the pertinent elements of the history

1. Normal gestation
2. Microphthalmia often noted at birth or in infancy
3. Leukocoria may be present

D. Describe the pertinent clinical features

1. Majority unilateral
2. Microphthalmos
3. Cataract may be present with vessels or vessel remnants on lens
4. Retrolental fibrovascular membrane
5. May have stalk extending from posterior lens to optic disc
6. Central displacement of ciliary processes
7. Prominent radial iris vessels
8. Strabismus common
9. Posterior pole anomalies common
 - a. Macular dysplasia
 - b. Pigmentary disturbances
 - c. Optic nerve dysplasia
 - d. Traction which may progress to peripheral or posterior retinal detachment
10. Milder cases with only Bergmeister papilla or Mittendorf dot
11. Glaucoma due to angle anomaly
12. Glaucoma due to progressive anterior chamber shallowing, rapid onset possible

E. Describe appropriate testing and evaluation for establishing diagnosis

1. Ultrasound to measure axial length and assess posterior segment if cataract present

II. List the differential diagnosis

A. Globe asymmetry

1. Nanophthalmos
2. Isolated microphthalmos
3. Contralateral glaucoma

B. Leukocoria

1. Isolated cataract
2. Retinal detachment
3. Retinopathy of prematurity
4. Coats disease
5. Retinoblastoma (not usually associated with microphthalmos)

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

A. Describe medical therapy options

1. Refractive error correction
2. Amblyopia treatment
3. Polycarbonate eyeglasses
4. Medical therapy alone suffices for only very mild forms of disease

B. Describe surgical therapy options

1. Lensectomy/anterior vitrectomy for cataract
 - a. Tough fibrovascular plaque may require additional instrumentation besides vitrector including intraocular cautery
 - b. Intraocular lens (IOL) controversial and most appropriate for less severely affected eyes
 - c. Most optical rehabilitation is via contact lens
2. Core or posterior vitrectomy for vitreous hemorrhage, retinal traction or detachment

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

- A. Vitreous hemorrhage**
- B. Retinal detachment**

V. Describe disease-related complications

- A. Angle closure glaucoma**
- B. Retinal detachment**
- C. Vitreous hemorrhage**
- D. Amblyopia**
- E. Strabismus**

VI. Describe appropriate patient instructions

- A. Counsel parents regarding potentially guarded visual outcome depending on condition of posterior pole and compliance with/response to amblyopia management**
- B. Note that strabismus is common and often requires surgery**
- C. Reinforce need for protective eyewear at all times**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Ocular coloboma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Defective embryogenesis
2. Ocular coloboma-failure of the fetal or choroidal fissure to close.
 - a. Fifth week of gestation

B. Define the relevant aspects of epidemiology of the disease

1. Eyelid coloboma-Distinct entity embryologically unrelated to ocular coloboma
 - a. Goldenhar Syndrome
 - b. Mandibulofacial dysostosis (Treacher-Collins syndrome)
 - c. Amniotic band syndrome
 - d. Isolated
2. Ocular- iris, ciliary body, choroid, retina, optic nerve
 - a. Numerous chromosomal abnormalities
 - b. CHARGE syndrome
 - i. Ocular coloboma
 - ii. Heart defects
 - iii. Choanal atresia
 - iv. Mental retardation
 - v. Genitourinary anomalies
 - vi. Ear anomalies
 - c. Isolated

C. List the pertinent elements of the history

1. Present at birth
2. Vision variable- normal to poor, depending on involvement of optic nerves or maculae
3. Nystagmus may be present
4. Possible family history
5. Strabismus may be present

D. Describe pertinent clinical features

1. Defect in eyelid(s)
2. Keyhole shaped pupil
3. Typical colobomas occur in the inferonasal quadrant
4. Iris transillumination of radial wedge
5. Microphthalmia
 - a. (microphthalmos with cyst/colobomatous cyst)
6. Abnormal red reflex/leukocoria
7. Flattening or notching of lens periphery with zonules absent in colobomatous area
8. Segmental absence of uveal tissue in posterior segment- may have skip areas

9. Segmental absence of optic nerve tissue, may resemble disc cupping
10. Macula may or may not be involved
11. Refractive Error
12. Possible nystagmus
13. Findings may be unilateral or bilateral

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Consider genetics evaluation

II. Define the risk factors

A. Various syndromes and chromosomal anomalies

III. List of differential diagnosis

A. Eyelid coloboma

1. Amniotic band syndrome
2. Goldenhar syndrome
3. Mandibulofacial dysostosis (Treacher Collins)

B. Ocular

1. Uveal
 - a. Retinal detachment
 - b. Retinopathy of prematurity (ROP)
 - c. Coats disease
 - d. Retinoblastoma
 - e. Myelinated nerve fibers
2. Iris/lens
 - a. Anterior segment dysgenesis
 - b. Trauma
3. Optic nerve
 - a. Glaucomatous cupping
 - b. Optic pit
 - c. Optic nerve staphyloma
 - d. Morning glory disc anomaly

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

A. Eyelid

1. Observe for drying of the cornea
 - a. Treat with lubricants
2. May require surgical closure

B. Ocular

1. Treat refractive errors
2. Treat superimposed amblyopia

3. Monitor for retinal detachments
4. Treat severe microphthalmia
 - a. Scleral shells to expand fornices and increase orbital volume to encourage orbital growth
5. Prescribe safety glasses/goggles if poor vision monocularly
6. Strabismus surgery
7. Cosmesis- custom prosthetic contact lens

V. Describe disease-related complications

A. Eyelid coloboma

1. Exposure keratitis

B. Ocular

1. Amblyopia
2. Refractive Error
3. Retinal detachment
4. Cataract
5. Nystagmus in bilateral cases

VI. Describe appropriate patient instructions

- A. Consider genetic counseling**
- B. Regular follow-up with ophthalmologist**
- C. Describe risk and symptoms of retinal detachment**
- D. Need for eyeglasses/patching if amblyopia/refractive errors/poor vision present**
- E. Refer child to appropriate rehabilitative services if bilaterally poor vision**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Onwochei BC, Simon JW, Bateman JB, Couture KC, Mir E. Ocular colobomata. *Surv Ophthalmol* 2000;45:175-94.
3. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.

Basic concepts in genetics for pediatric ophthalmologists

I. Genetic code

- A. DNA is the molecule that constitutes the genetic code, formed of nucleotides**
 - 1. Nucleotides arranged in single series and form strands
 - 2. Two nucleotide strands form a double helix
- B. Gene is a DNA segment that codes for a specific protein with a defined function**
- C. Human genome contains 23 chromosomes**
- D. Each cell contains 44 autosomal chromosomes (22 from each parent) and a set of sex chromosomes (XX or XY)**
- E. Alleles are different forms of one type of gene which can occupy the same place on homologous chromosomes and are responsible for different traits**
- F. Mutations cause changes in genes which alter the structure and affect the function of the protein produced, thus producing a new allele**
- G. Trait is an identifiable feature of an organism that is determined by genetic and/or environmental factors**
- H. Phenotype is the observable traits of an organism**
- I. Genotype is the alleles producing a certain trait or set of traits under consideration**
- J. Variable expressivity occurs when a phenotype is expressed to a different degree among individuals with the same genotype**

II. Mendelian inheritance

- A. Autosomal dominant (AD) inheritance**
 - 1. Mutation affects a gene located on an autosome
 - 2. Phenotype is expressed when only one allele is mutated
 - 3. Occurs in consecutive generations
 - 4. Males and females equally affected
 - 5. Offspring of an affected parent have a 50% risk of having the disease
 - 6. Male-to-male transmission indicates AD mutation
 - 7. Examples of ophthalmic disease with AD inheritance
 - a. Nonsyndromic bilateral congenital cataracts
 - b. Stickler syndrome
 - i. Cataracts
 - ii. High myopia in a child with Pierre Robin Sequence
- B. Autosomal recessive (AR) inheritance**
 - 1. Mutation affects both alleles of a gene
 - 2. Phenotype is expressed only when both alleles are mutated
 - 3. Occurs in siblings approximately 25% of the time
 - 4. Males and females are equally affected
 - 5. Parents and offspring of the proband are usually unaffected

6. Parents are more likely to be closely related
7. Typically skips generations

C. X-linked inheritance

1. May be dominant or recessive (recessive is more common)
2. X-linked recessive inheritance includes female-to-male transmission
3. Males are usually affected
4. Affected fathers transmit the mutations to their daughters, who become carriers
 - a. A genetic carrier has inherited a genetic trait or mutations but does not display that trait or show symptoms of the disease. They are able to pass the gene on to their offspring, who may then express the gene
 - b. Lowe syndrome (oculocerebral renal syndrome) is an example of X-linked inheritance
5. X-linked dominant inheritance affects males and females equally, although most diseases are uniformly fatal to males, leaving only affected female offspring
 - a. Aicardi syndrome
 - b. Incontinentia pigmenti

III. Non mendelian inheritance

A. Mitochondria contain DNA, and mitochondrial DNA is inherited entirely from the mother (never from the father to the offspring)

1. Diseases resulting from mitochondrial mutations
 - a. Chronic progressive ophthalmoplegia
 - b. Leber hereditary optic neuropathy

B. Multifactorial genetic disorders

1. Result from the interactions of multiple gene products with each other and the environment
2. Show a hereditary component but do not follow a pattern of Mendelian inheritance
3. Diseases resulting from multifactorial inheritance
 - a. Juvenile nonsyndromic myopia
 - b. Nonsyndromic comitant childhood strabismus

IV. Genetic counseling

A. Important in diseases such as retinoblastoma and albinism, where the disease is heritable and the transmission patterns of the gene mutations fairly well established and able to predict risk of disease in siblings or offspring

V. Gene therapy

- A. May involve viral vector to replace an existing mutant gene**
- B. May involve eliminating the protein product of the mutant gene**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 2: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Bollinger K, Traboulsi EI. Molecular genetics for the pediatric ophthalmologist. J Pediatr Ophthalmol Strabismus. 2007 Jul-Aug 44(4):209-15.

Normal milestones of ocular and visual development

I. Describe approach to establishing the diagnosis

A. List the pertinent elements of the history

1. Quality of visual behavior
2. Ocular alignment
3. Presence of nystagmus
4. Normal and symmetric appearance of the globes/adnexae
5. Family history of early-onset eye disease

B. Describe pertinent clinical features

1. Normal anatomic development
 - a. Lids fused in utero
 - b. Lens vascularization disappears during fourth or fifth month of gestation
 - c. Corneas may be cloudy up to 30 weeks gestation
 - d. Above findings may occur in otherwise normal extremely premature babies
 - e. Retina vascularized nasally at 36 weeks, temporally at 40 weeks
 - f. Iris pigmentation continues through first year of life, therefore eye color may continue to darken
 - g. Normal infant corneal diameter 9.5-10.5 mm
 - h. 90% of globe growth occurs by age 2 years
 - i. Fovea matures during first 4 months of life
2. Emmetropization
 - a. Most children are moderately hyperopic
 - b. Axial length increases in the growing eye
 - c. Cornea and lens flatten
 - d. Refractive power of anterior segment and axial length adjust to reach emmetropia
3. Normal functional development
 - a. Visual acuity
 - i. Response to light in early infancy
 - ii. Good foveal fixation/fix and follow behavior by 8-12 weeks
 - iii. Visual acuity improves rapidly in the first six months of life
 - b. Alignment
 - i. Neonates often exotropic
 - ii. Variable alignment in infancy
 - iii. Ocular alignment stable by 4-6 months
 - c. Motility
 - i. Horizontal gaze full at birth
 - ii. Vertical gaze full by 6 months

II. Describe appropriate patient instructions

- A. Educate parents on timeline and visual development**
- B. Premature babies should reach milestones based on adjusted age**

Additional Resources

1. AAO, Basic and Clinical Science Course, Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Congenital infection syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. TORCH Syndrome: Toxoplasmosis, Other agents (Hepatitis B, HIV, Syphilis, Varicella-Zoster, lymphocytic choriomeningitis virus), Rubella, Cytomegalovirus and Herpes Simplex
2. Congenital protozoan, viral, or bacterial infection

B. Define the relevant aspects of epidemiology of the disease

1. Relatively uncommon

C. List the pertinent elements of the history

1. Prenatal maternal infection
2. Prematurity
3. Low Birth Weight

D. Describe pertinent clinical features

1. Toxoplasmosis
 - a. Systemic
 - i. Hepatosplenomegaly
 - ii. Microcephaly
 - iii. Intracranial calcifications
 - iv. Developmental delay
 - b. Ocular
 - i. Retinitis/choroiditis, usually bilateral
 - i) Thickened, cream-colored area of retinal infection with overlying vitritis
 - ii) Satellite lesion at edge of old, atrophic scar
 - ii. Panuveitis
 - iii. Cataract
 - iv. Optic atrophy
 - c. Differential diagnosis
 - i. Scarring from other causes of chorioretinitis or vitritis such as CMV
2. Rubella
 - a. Systemic
 - i. Sensorineural hearing loss
 - ii. Growth and developmental delay
 - iii. Congenital heart defects
 - iv. Microcephaly
 - v. Hepatosplenomegaly
 - b. Ocular
 - i. Congenital cataract
 - ii. Microphthalmos
 - iii. Pigmentary retinopathy

- iv. Glaucoma
 - v. Keratitis
- 3. Cytomegalovirus
 - a. Systemic
 - i. Fever
 - ii. Jaundice
 - iii. Thrombocytopenia
 - iv. Deafness
 - v. Microcephaly
 - vi. Periventricular calcification
 - b. Ocular
 - i. Retinochoroiditis
 - ii. Optic atrophy
 - iii. Microphthalmos
 - iv. Cataract
 - v. Uveitis
- 4. Herpes simplex
 - a. Systemic
 - i. Encephalitis
 - ii. Disseminated disease
 - b. Ocular
 - i. Keratoconjunctivitis/interstitial keratitis
 - ii. Retinochoroiditis
 - iii. Cataract
- 5. Syphilis
 - a. Systemic
 - i. Generalized lymphadenopathy
 - ii. Hepatosplenomegaly
 - iii. Pneumonia
 - iv. Anemia
 - v. Skeletal anomalies
 - vi. Dental anomalies (widely-spaced, peg-shaped teeth)
 - vii. Sensorineural hearing loss
 - b. Ocular
 - i. Bilateral interstitial keratitis (older children and adults)
 - i) Part of Hutchinson's triad
 - (i) Interstitial Keratitis
 - (ii) Dental anomalies
 - (iii) Eighth nerve deafness
 - ii. Pigmentary retinopathy
 - iii. Anterior uveitis

- iv. Glaucoma
 - v. Optic atrophy
- 6. Lymphocytic Choriomeningitis (LCMV)
 - a. Occurs due to exposure to rodents, both domestic and wild
 - b. Systemic
 - i. CNS abnormalities
 - i) Hydrocephalus
 - ii) Microcephalus
 - iii) Intracranial calcifications
 - iv) Cognitive impairment
 - c. Ocular
 - i. Chorioretinal scars similar to those seen in CMV, and toxoplasmosis
 - ii. Consider LCMV when testing for the more common CMV and toxoplasmosis is negative

E. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Serological testing when diagnosis is suspected
- 2. Live virus/organism may be recovered from body secretions

II. Define the risk factors

- A. Maternal infection, especially when acquired early in pregnancy**

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

A. Describe medical therapy options

- 1. Appropriate anti-viral (topical and systemic), anti-protozoan, and anti-bacterial drugs
- 2. Corticosteroids sometimes appropriate
- 3. Multidisciplinary care for immediate post-natal and developmental issues

B. Describe surgical therapy options

- 1. Various surgeries may be required depending on ocular involvement

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

- A. Side effects of drug therapy**
- B. Inflammation**
- C. Immunosuppression, glaucoma, and cataract from steroids**

V. Describe appropriate patient instructions

- A. Emphasize need for periodic follow-up examinations**
- B. Referral to other pediatric subspecialists as needed**

Additional Resources

- 1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.

Cerebral/cortical visual impairment in children (retrogeniculate vision loss)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease

1. Posterior visual pathway/cortex damage
 - a. Optic radiations (subcortical)
 - b. Occipital cortex (cortical)
2. Congenital, prenatal and perinatal causes
 - a. May be idiopathic
 - b. Developmental defects
 - c. Intrauterine infection
 - d. Intraventricular hemorrhage/periventricular leukomalacia in premature infants
3. Acquired causes
 - a. Anoxic damage
 - b. Ischemia
 - c. Hydrocephalus
 - d. Seizure disorder
 - e. Meningitis, encephalitis
 - f. Trauma
 - g. Nonaccidental trauma

B. Define the relevant aspects of epidemiology of the disease

1. High association with prematurity
2. Associated with other neurologic diseases
3. Associated with traumatic brain injury
4. Associated with anoxic brain injury

C. List the pertinent elements of the history

1. Infant misses normal visual milestones
2. Searching eye movements

D. Describe pertinent clinical features

1. Searching eye movements
2. Normal pupils
3. Variable levels of visual attentiveness
4. Peripheral vision better than central
5. May navigate better than vision suggests
6. Vision often improves
7. Co-exists with other neurologic deficits
8. Optic atrophy may be present

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Complete history
2. Magnetic resonance imaging (MRI)
3. Possible electroencephalogram for seizures
4. Visual evoked potential

II. Define the risk factors

- A. Prematurity
- B. Prolonged labor
- C. Congenital infections
- D. Birth trauma
- E. Nonaccidental trauma
- F. Nuchal cord
- G. Neurologic abnormalities
- H. Traumatic or anoxic brain injury

III. List the differential diagnosis

- A. Uncorrected high refractive error
- B. Delayed visual maturation
- C. Organic ocular disease

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

- A. Describe medical therapy options
 1. Treat seizures
 2. Treat meningitis
 3. Refer for early intervention
 4. Refer for parental support/education
 5. Watch and wait
 6. Continued follow up
 7. Correct refractive errors

V. Describe disease-related complications

- A. Decreased vision
- B. Other neurological delay

VI. Describe appropriate patient instructions

- A. Continue optimistic view to parents
- B. Parents to get early intervention/education

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Edmond JC, Foroozan R. Cortical visual impairment in children. Curr Opin Ophthalmol 2006 17:509-12.

The role of the ophthalmologist in learning disabilities

I. Describe the approach to establishing the diagnosis

A. Definition

1. Heterogeneous group of disorders
2. Difficulty understanding and using spoken or written language

B. Describe the etiology of this disease

1. Deficit in phonologic component of language that makes it difficult to use the alphabetic code to decode the written word
2. Multifactorial
3. Genetic influences as well as differences in how the brain functions

C. Define the relevant aspects of epidemiology of the disease

1. Estimates of prevalence vary widely
2. Family history of speech and learning problems

D. List the pertinent elements of the history

1. Failure to achieve average reading level skills or problems in other academic areas, despite average IQ and adequate instruction
2. Poor handwriting and slow writing speed
3. Short attention span and memory problems
4. Low self-esteem or frustration with school work
5. Delays in speech and language development
6. Trouble with coordination
7. Maternal drug or ethanol abuse during pregnancy
8. History of child abuse, shaken baby syndrome
9. Neonatal brain injury in very low birth weight delivery
10. Any medical condition that could interfere with the child's ability to learn
11. Chronic illness that could cause school absences or difficulties concentrating or learning

E. Describe pertinent clinical features

1. No significant ophthalmic abnormality on complete dilated exam with cycloplegic refraction
2. There is no correlation between reading performance, refractive error or strabismus

F. Describe appropriate testing and evaluation for establishing the diagnosis

1. Complete age-appropriate ophthalmologic exam including dilated fundus exam and cycloplegic refraction
2. Referral as early as possible for educational, neuropsychological, psychological, and/or medical diagnostic assessments

II. Define the risk factors

A. No known eye or visual cause for learning disabilities

B. May be associated with other psychological disorders such as depression, anxiety, behavioral disorder, attention deficit hyperactivity disorder

- C. Family history

III. List the differential diagnosis

- A. Developmental delay
- B. Convergence insufficiency
- C. Amblyopia
- D. Uncorrected refractive error
- E. Visual field loss

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

- A. Describe medical therapy options
 - 1. Multidisciplinary approach
 - 2. Treat strabismus, amblyopia, and refractive error
 - 3. Involve the educational system
 - 4. Remedial educational programs
 - 5. Lack of scientific evidence for
 - a. Vision therapy
 - b. Vision training
 - c. Tinted lenses or filters
 - d. Eye exercises
 - e. Behavioral vision therapy
 - f. Diet or herbal preparations
 - 6. Early educational intervention

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

- A. Inappropriate treatment can delay proper educational assistance
- B. If untreated, learning disabilities may lead to low self-esteem

VI. Describe disease-related complications

- A. Failure to achieve age appropriate learning milestones

VII. Describe appropriate patient instructions

- A. Parents should read aloud to their children to help develop language skills
- B. Evaluation and Intervention as early as possible
- C. Complete ophthalmic examination
- D. Treat correctable ocular disorders
- E. Team approach to the specific disorder
- F. Proper assistance can be delayed if these complex problems are attempted to be corrected with simple solutions/vision therapy
- G. Communication with the educational system

- H. Educate parents regarding learning-disabled student's rights to school accommodations and individualized support**
- I. Instruction using a multisensory, structured language approach**

Additional Resources

1. AAO, Joint Technical Report on Learning Disabilities 2011 <http://www.aao.org/guidelines-browse?filter=complementarytherapyassessment>
2. AAO, Focal Points: The Ophthalmologist and Learning Disabilities, Module #2, 2005.
3. Learning Disabilities, Dyslexia, and Vision. Pediatrics 2011;127:e818-e856

Informed consent in strabismus surgery

I. Describe professional responsibilities pertaining to the topic

A. Describe elements of informed consent process

1. Respect for patient autonomy
2. Patient comprehension
 - a. Special role of parent/ guardian for pediatric patients
 - b. Write description in plain English, easily understood to patient (avoid medical jargon); use appropriate translation services as necessary
3. Discusses benefits, risks, alternatives, and complications including, but not limited to, over-and undercorrection, need for additional surgery, infection, bleeding, complications that could affect vision, risk of general anesthesia, diplopia, loss of eye
4. Counsel the parents regarding the possible need for ongoing amblyopia treatment
5. In academic centers, presence of resident/fellow participation
6. Re-emphasize on day of surgery actual procedure being performed!

B. Describe applicable rule(s) of the American Academy of Ophthalmology Code of Ethics - Informed Consent: "The performance of medical or surgical procedures shall be preceded by appropriate informed consent"

II. Discuss legal implications of noncompliance

- A. Procedures performed without adequate informed consent may be legally interpreted as "battery"
- B. Medical liability risk
- C. Write down everything performed in surgery, "If not written down, it did not happen"

III. Describe the potential consequences of failure to obtain informed consent

- A. Breach of patient trust
- B. Loss of patient satisfaction
- C. Unrealistic expectations of outcomes
- D. Report(s) to local, state and National Practitioner Data Bank

IV. Describe potential proactive actions

- A. Education regarding ethical requirements
- B. Education regarding legal requirements
- C. Informed Consent is a process, requiring signatures, witnesses and written documentation

Additional Resources

1. American Academy of Ophthalmology: Code of Ethics. <http://www.aao.org/ethics-detail/code-of-ethics>
2. American Academy of Ophthalmology. Advisory Opinions of the Code of Ethics. Informed Consent. <http://www.aao.org/ethics-detail/advisory-opinion--informed-consent>

Functional vision loss in childhood

I. Describe the approach to establishing the diagnosis

A. List the pertinent elements of the history

1. Girls > boys
2. School age typical
3. May have some sort of secondary gain
 - a. Desire for eyeglasses
4. Need for attention
5. Vague visual complaints
 - a. "Just not seeing" with little inconvenience
 - b. Blurred vision
 - c. Distorted images
 - d. Tunnel vision
 - e. Occasional bilateral blindness
6. Symptoms often come on gradually
 - a. Exception is when a significant emotional event (death) occurs
7. May be family history of eye disease (i.e., retinitis pigmentosa)
8. Psychiatric disorders

B. Describe the etiology of the disease

1. Functional vision loss (nonorganic visual loss) is any visual deficit that cannot be explained by physical findings
 - a. Repeated normal objective ocular examination
 - b. Must rule out amblyopia
 - c. Must rule out structural abnormality
 - d. Diagnosis of exclusion

C. Describe appropriate testing and evaluation for establishing the diagnosis

1. Observation: bilateral blindness testing
 - a. Blink response: throwing ball on a string
 - b. Mirror test (rock or rotate about vertical axis)
 - i. Blind: no movement of eyes when mirror rotated
 - ii. Sighted: eyes will move as mirror rotates
 - c. Optokinetic drum or tape response
2. Unilateral blindness testing
 - a. Prism test to evaluate whether the affected eye is being suppressed
 - b. Worth four dot testing
 - c. Stereoacuity/vectograph testing
 - d. Relative afferent pupillary defect (RAPD) associated with profound monocular visual loss
3. Direct measurement of visual acuity
 - a. Confusion refraction testing

- i. Start acuity testing with 20/10 or 20/15 line
 - ii. Recheck vision with multiple 20/20 lines
 - iii. Different type target with same visual angle
 - iv. Different testing distances
 - b. Check vision using plano lens
 - c. Fogging techniques
 - i. High plus lens fogs good eye
 - ii. Two high power cylinders lens (equal and opposite sign)
 - i) Axes parallel: net power is plano
 - ii) Axes nonparallel: fogging good eye
 - iii. Cycloplegic agent in one eye only
 - i) Check near acuity with both eyes open
 - d. Visual field defects
 - i. Tunnel or tubular vision
 - ii. Spiraling or crossing of isopters on Goldmann kinetic perimetry
 - iii. Tangent screen field testing 1 and 2 meters may be useful in eliciting a non-organic visual field response
 - iv. Binasal and bitemporal defects are rare in functional patients
 - v. Central scotomas are also rare; must rule out organic disease
 - e. Repeat visual acuity without parents
- 4. Confirmatory studies normal (if required)
 - a. Fluorescein angiogram (FA) - rule out Stargardt disease
 - b. Electrophysiology testing (ERG)
 - c. Visual evoked potentials (VEP)
 - d. MRI scan may be needed
 - e. Optical coherence tomography (OCT) - rule out Juvenile X-linked retinoschisis

II. List the differential diagnosis

- A. Amblyopia**
- B. Organic ocular disease**

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

- A. Reassurance**
- B. Elicit parents assistance in finding underlying stresses**
 - 1. Home and family (abuse, sibling rivalry, marital problems)
 - 2. School (bullying, poor academic performance)
- C. Refer back to primary care for psychiatric issues**
- D. Pediatric neurology consultation when appropriate**

IV. Describe disease-related complications

A. Associated psychiatric disorders

1. Depression
2. Conversion disorder

B. Neurological disorders

C. Child abuse

1. Munchausen syndrome: self-inflicted injuries
2. Munchausen by proxy: injuries inflicted by caregivers

V. Describe appropriate patient instructions

A. Continued reassurance to patient and parents

B. Repeated examination if needed

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 6: Pediatric Ophthalmology and Strabismus, 2015-2016.
2. Taylor D, Hoyt CS. Pediatric Ophthalmology and Strabismus. Elsevier and Saunders; 2005.
3. Wright KW and Spiegel PH. Pediatric Ophthalmology and Strabismus. Springer; 2003.
4. O'Hara MA. Cracking the crock: How to deal with factitious eye disease. *Journal of Ophthalmic Nursing and Technology* 1998;17(5):1-5.

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