Practicing Ophthalmologists Curriculum
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Practicing Ophthalmologists Curriculum
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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
Glaucoma

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Aqueous humor dynamics and intraocular pressure

I. Route:
   
   A. Trabecular Outflow (predominant route)
      1. Ciliary Body (CB) ➔
      2. Posterior Chamber ➔
      3. Pupil ➔
      4. Anterior Chamber ➔
      5. Trabecular Meshwork ➔
      6. Schlemm Canal ➔
      7. Collector Channels ➔
      8. Episceral venous system

   B. Uveoscleral Pathway
      1. Ciliary Body ➔
      2. Posterior Chamber ➔
      3. Pupil ➔
      4. Anterior Chamber ➔
      5. Ciliary muscle ➔
      6. Supraciliary and Suprachoroidal space ➔
      7. Intact sclera

II. Aqueous humor formation - site of production
   
   A. Ciliary processes
      1. Double layer of epithelium connected together by intercellular tight junctions (blood aqueous barrier)
         a. Inner non-pigmented (site of aqueous production)
         b. Outer pigmented

III. Aqueous humor formation
   
   A. Active secretion/transport
   B. Ultrafiltration
   C. Diffusion

IV. Active transport
   
   A. Independent of IOP
   B. Majority of aqueous produced this way
V. Ultrafiltration
   A. Pressure dependent
   B. Capillary pressure in CB > IOP
   C. Oncotic gradient forces fluid back in to CB

VI. Diffusion
   A. Passive movement of ions across a gradient

VII. Functions of aqueous
   A. Maintain intraocular pressure
   B. Provide substrates for cornea and lens
   C. Remove metabolic products

VIII. Aqueous characteristics
   A. Very low protein level compared with plasma

IX. Rate of aqueous formation
   A. Fluorophotometry (method of quantifying rate of formation)
   B. 2 - 2.5 µl/min
   C. 1% turnover / minute
   D. Varies diurnally and decreases during sleep
   E. Decreases with:
      1. Age
      2. Trauma
      3. Inflammation
      4. Certain systemic drugs
         a. Anesthetics
         b. Systemic hypotensive agents
         c. Beta Blockers
         d. Carbonic Anhydrase Inhibitors
         e. Alpha 2 Agonists
      5. Carotid occlusive disease

X. Aqueous humor outflow
   A. Facility of outflow varies greatly in normals
   B. Decreases with age
   C. Decreases with surgery, trauma and endocrine factors
   D. Decreased in glaucoma
XI. Calculating IOP - Goldmann equation

A. \( P_0 = \frac{F}{C} + P_v \)
   1. \( P_0 = \text{IOP (mm Hg)} \)
   2. \( F = \text{Aqueous formation (ul /min)} \)
   3. \( C = \text{Facility of outflow (ul / min /mm Hg)} \)
   4. \( P_v = \text{Episcleral venous pressure (mm Hg)} \)
   5. \( R (\text{resistance to outflow}) = \frac{1}{C} \)
   a. Not part of equation

XII. Trabecular outflow

A. Majority of outflow is through trabecular meshwork
   1. Uveal
   2. Corneoscleral
   3. Juxtacanalicular
      a. Major site of outflow resistance

B. Pressure dependent outflow

C. Increased by
   1. Miotics
   2. Trabeculoplasty

XIII. Uveoscleral outflow

A. Non trabecular outflow
   1. Predominantly via CB to supraciliary and suprachoroidal space

B. Pressure independent outflow

C. Increased by
   1. Cycloplegics
   2. Adrenergic
   3. Prostaglandins analogues

D. Decreased
   1. Miotics

XIV. Episcleral venous pressure

A. Relatively stable
   1. Same as central venous pressure
   2. Increases with
      a. Head down position
      b. Disease of orbit
      c. Neck diseases
      d. AV shunts
B. Normal 8 - 10 mm Hg

XV. IOP distribution

A. Mean IOP 15.5mmHg
B. Non gaussian distribution
   1. Skewed toward higher IOPs

XVI. Factors influencing IOP

A. Time of day
B. Heartbeat
C. Breath holding/Valsalva
D. Exercise
E. Fluid intake
F. Systemic and topical medications
G. ETOH transiently decreases
H. Cannabis transiently decreases
I. Higher reclined than vertical
J. Increases with age
K. Higher in first degree relatives of POAG patients

XVII. Diurnal variation

A. IOP varies 2-6mmHg over 24hr period
B. Higher IOP associated with wider fluctuations in IOP
C. Diurnal fluctuation >10mmHg suggestive of glaucoma
D. Peak IOP usually during early morning hours
E. Impact of IOP fluctuation on optic nerve remains unknown
F. Systemic hypotension during sleep ➔ decreased optic nerve perfusion! optic nerve damage

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Tonometry: clinical measurement of intraocular pressure (IOP)

I. Indications and contraindications

A. Indications
   1. Measurement of IOP for screening and monitoring of glaucoma

B. Relative contraindications
   1. Corneal laceration
   2. Corneal ulcer or abrasion

C. Pre-procedure evaluation
   1. Assess patient for things that might affect tonometry accuracy
   2. Obesity or straining to reach slit lamp
   3. Tight collar or necktie
   4. Breath holding or Valsalva
   5. Contact lens
   6. Extraocular muscles acting on restricted globe
   7. Lid squeezing or narrow interpalpebral fissure
   8. High corneal astigmatism
   9. Pachymetry and other biomechanical properties

D. Slit lamp biomicroscopy to evaluate cornea for abnormalities that may affect IOP measurement accuracy
   1. Corneal edema
   2. Corneal scars
   3. Band keratopathy
   4. Epithelial irregularity or defects
   5. Keratoconus
   6. Corneal prosthesis
   7. Prior corneal surgery

II. Techniques to measure IOP

A. Goldmann tonometry
   1. Most widely used in clinic and research
   2. Measures IOP by applanating (flattening) a 3.06mm diameter area of the cornea using a split-image prism
   3. Based on Imbert-Fick principle: Internal fluid pressure (P) acting on a thin membrane sphere is equal to the force (F) needed to flatten its surface divided by the area of flattening (A), P=F/A
   4. Is most accurate when the central corneal thickness (CCT) is 520 um
      a. Increased CCT may give an artificially high IOP measurement
      b. Decreased CCT may give an artificially low IOP measurement
   5. Corneal properties can affect measurement
      a. Corneal edema predisposes to falsely low IOP measurement
b. Corneal stiffness, i.e. scarring predisposes to falsely high IOP measurement

c. Corneal astigmatism may require prism adjustment

6. Scleral rigidity may affect measurement

a. Scleral buckling predisposes to falsely low IOP

B. Perkins tonometer

1. Similar to Goldmann tonometry but is portable and can be used in upright or supine position

C. Tono-Pen

1. Handheld tonometer that contains a strain gauge and produces an electrical signal as the tip applanates a very small area of the cornea
2. Helpful in patients with nystagmus and corneal irregularity
3. Underestimates in eyes with higher IOP and overestimates in eyes with lower IOP compared with Goldmann.

D. Pneumatonometer

1. Handheld tonometer with a pressure-sensing device consisting of a gas-filled chamber covered with a silastic diaphragm
2. Helpful in patients with nystagmus and corneal irregularity
3. Readings tend to be higher than Goldmann tonometry

E. Non-contact tonometry (NCT)

1. Determines IOP by measuring the time necessary for a given force of air to flatten a given area of the cornea.
2. Topical anesthetic is not needed.
3. Often over-estimates IOP
4. Useful for screening programs

F. Ocular response analyzer (ORA)

1. A non-contact tonometer which uses a pulse of air to applanate the cornea and measures IOP with consideration of the biomechanical properties of the cornea
2. Corneal compensated IOP is calculated based on hysteresis and a corneal resistance factor

G. Dynamic contour tonometer (DCT)

1. Slit lamp-mounted, non applanation, contact tonometer that measures IOP with a concave tonometer tip that matches the curvature of the cornea.
2. Some studies indicate no significant effect of CCT, corneal curvature or astigmatism on measurement

H. Digital palpation

1. May be useful in in uncooperative patients
2. With keratoprosthesis, useful to compare with other eye
3. Intraoperatively

I. Rebound tonometry

1. Handheld tonometer in which a light weight probe makes momentary contact with the cornea
2. The higher the IOP, the shorter the contact time with the cornea and the faster the probe decelerates after contact
3. No topical anesthetic is required
4. Useful in less cooperative patients and children
5. May be useful for screening and home tonometry

III. Tonometry instrumentation and technique
A. Goldmann tonometry
   1. Slit lamp mounted tonometer.
   2. One drop of fluorescein/topical anesthetic solution is administered. Fluorescein highlights the tear meniscus at the margin of contact
   3. Cornea and tonometer tip are illuminated with a cobalt blue light
   4. The patient is instructed to relax, keep the eye still and lids open and avoid breath-holding
   5. Tonometer tip is brought flush with cornea and dial turned from a starting point of 1 gram until applanation occurs
      a. Applanation is defined as when the inside edges of the prism-split circular meniscus just touch at the midpoint of their pulsations.
   6. Grams of force to applanate are read from the tonometer dial, then multiplied by 10. (10 mmHg = 1 gram of force to applanate)
   7. Tonometer tip is cleaned with antiseptic solution
   8. In eyes with high astigmatism, the biprism should be rotated until the dividing line between the prisms is 45 degrees to the major axis, or an average may be take of horizontal and vertical readings, or the red line is aligned with the steep axis of the cornea

B. Perkins tonometry
   1. Hand-held counterbalanced applanation tonometer that can be used with the patient supine or upright
   2. Technique is similar to Goldmann tonometry

C. Tono-Pen tonometry
   1. Digital hand-held tonometer than can be used with the patient supine or upright
   2. Tonometer tip is covered with rubber cover
   3. One drop of topical anesthetic is administered (proparacaine)
   4. Tonometer tip is touched to the central cornea repeatedly until 6-10 measurements are taken by the instrument and the average read from the digital display.
   5. Tonometer tip cover is discarded.

D. Pneumatonometer
   1. One drop of topical anesthetic is administered
   2. Tonometer tip is touched to the central corneal until the measurement and standard deviation is read from the digital display

E. Non-contact tonometer
   1. Topical anesthetic is not needed

F. Ocular response analyzer
   1. Topical anesthetic is not needed

G. Dynamic contour tonometer
   1. Digital slit-lamp mounted tonometer
   2. One drop of topical anesthetic solution is administered
   3. Tonometer tip cover is discarded

H. Rebound tonometer
   1. Six readings are taken and averaged after discarding the highest and lowest value
   2. No topical anesthetic is required

IV. Complications of the procedure, their prevention and management
   A. Complication of contact tonometry (Goldmann, Perkins, Tono-Pen, Pneumatonometer, DCT)
1. **Corneal abrasion**
   a. Prevent by slow careful applanation and encouraging patient to maintain steady head and eye position.
   b. Most abrasions caused by applanation heal overnight without treatment.

2. **Antiseptic toxicity to the epithelium**
   a. Tonometer tip should be allowed to fully dry between patients

3. **Potential for transmission of infection**

4. **Allergic reaction to topical anesthetic**

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**V. Considerations in interpretation of this procedure**

A. **Central corneal thickness and other biomechanical properties of the cornea may affect IOP measurements by any method of tonometry**
   1. Pachymetry measurements should be used to help interpret IOP measurements, but not to "correct" measurements.
      a. Relationship between CCT and IOP is non-linear
      b. No validated correction factor exists
      c. There is a large variability in CCT measurements in health and disease and after corneal surgery
      d. OHTS study indicated subjects with thinner corneas are more likely to progress to glaucoma
      e. CCT should be re-measured after corneal surgery

B. **A single IOP measurement is only a random sample of a dynamic picture. IOP fluctuates throughout the day and night**

C. **Technician or clinician influences on IOP measurement**
   1. Pressure from fingers holding lids may be transmitted to globe and elevate IOP
   2. Excess fluorescein (thick mires) may cause overestimation of IOP
   3. Inadequate fluorescein (thin mires) may cause underestimation of IOP
   4. Improper vertical alignment of mires may cause overestimation of IOP
   5. Inadequate tonometer calibration
   6. Repeated applanation tonometry reduces IOP readings

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**Additional Resources**

5. Intraocular Pressure difference in Goldmann Applanation tonometry vs Perkins Hand-held Applanation tonometry in Overweight Patients. Gonzaga dos Santos et al. Ophthalmology 105; 2260-2263
Gonioscopy

I. List the indications/contraindications

A. Overcomes problem of total internal reflection to see angle structures

B. Indirect gonioscopy (e.g., Goldmann or four mirror lens)
   1. Should be performed as part of the initial evaluation of all patients able to cooperate with the test and repeated periodically
   2. Essential diagnostic tool in glaucoma (viewing the iridocorneal angle)
      a. Most common cause of incorrect diagnosis is omission of gonioscopy
         i. Overlooking of secondary glaucomas and other glaucomas
         ii. Periodically performed can detect emergence of angle closure in a previously open angle
   3. Identification of
      a. Narrow angle
      b. Angle recession
      c. Foreign bodies
      d. Pigmentation/Abnormal pigmentation
      e. Tumors
      f. Angle neovascularization
      g. Angle synechiae
   4. Glaucoma treatment in the angle
      a. Laser trabeculoplasty
      b. Goniosynechialysis
      c. Treatment and evaluation of trabeculectomy or non-penetrating filtering surgery site
      d. Gonioplasty/iridoplasty

C. Direct gonioscopy (e.g., Koepppe style or Swan-Jacob style lens)
   1. Examine for angle recession
   2. Glaucoma surgery
      a. Goniotomy
      b. Goniosynechialysis
      c. Angle surgery

D. Contraindications
   1. Inability of patient to cooperate
   2. Corneal disease precluding application of corneal lens

II. Describe the pre-procedure evaluation

A. Indirect gonioscopy
   1. Comfortably seated patient
   2. Well lubricated cornea
   3. Topical anesthesia applied prior to application of mirror mounted in contact lens
   4. View angle with slit-lamp biomicroscope
B. Direct gonioscopy
   1. Recumbent patient
   2. Anesthetized cornea
   3. Cornea coupled to Koeppe or Barkan lens or surgical gonioprism with saline or viscous solution
   4. View angle through hand-held binocular magnifier or surgical microscope

III. List the alternatives to this procedure
A. Ultrasound biomicroscopy
B. Anterior segment optical coherence tomography

IV. Describe the instrumentation and technique
A. Indirect gonioscopy
   1. Produces inverted image 180° away from origination
   2. Two types of gonio lenses are in common use
      a. 4 mirror type
         i. Different types (Zeiss, Posner, Sussman)
            i) Rests solely on cornea/ tear film
            ii) Requires only drop of anesthetic
         ii. Indentation gonioscopy can be performed
            i) Technique to differentiate appositional and synechial angle-closure
            ii) Helpful in assessment of iridodialysis
            iii) Searching for cyclodialysis cleft
      b. Goldmann type
         i. Goldmann lens requires clear fluid to fill space between cornea and goniolens
         ii. Lens is brought toward patient's eye and tipped forward quickly enough to trap the clear fluid
         iii. Suction cup effect is obtained keeping lens centered on cornea
            i) Beneficial for laser trabeculoplasty
            ii) Disadvantage due to inability to perform dynamic gonioscopy
      iv. Methylcellulose may interfere with subsequent perimetry
   3. Important to control pupil illumination and perform in darkened room with a very small beam of light illuminating only the angle, to prevent inadvertently opening an occludable angle with ambient light

B. Direct gonioscopy
   1. Produces natural view of angle
   2. Koeppe contact lens inserted while patient lies supine
   3. Advantages
      a. Provides ease of view of various parts of the angle with comparison to contralateral eye when Koeppe lens placed in both eyes
   4. Necessary for angle based surgery
      a. Goniosynechialysis
      b. Goniotomy
      c. Ab interno trabeculotomy
V. List the complications of the procedure, their prevention and management

A. Corneal abrasion
   1. Prevention: moist cornea, topical anesthesia, minimize movement of lens on cornea
   2. Management: artificial tears, bandage contact lens, topical antibiotics

VI. Describe the considerations in interpretation of this diagnostic procedure

A. Normal angle landmarks (best viewed with parallelepiped or corneal wedge method)
   1. Anterior to posterior
      a. Cornea
      b. Schwalbe line
      c. Non-pigmented trabecular meshwork
      d. Pigmented trabecular meshwork
      e. Scleral spur
      f. Ciliary band
      g. Iris root

B. Different methods of grading or evaluating angle depth and configuration versus description of structures
   1. Shaffer, Scheie or Spaeth system

C. Pathologic examples
   1. Developmental abnormalities
   2. Peripheral anterior synechiae
   3. Abnormal pigmentation
   4. Recession
   5. Cyclodialysis cleft
   6. Angle neovascularization
   7. Pseudoexfoliation material
   8. Inflammatory cellular deposits
   9. Tumor
   10. Foreign body

VII. Describe the follow-up care

A. Baseline gonioscopy on each patient

B. Periodic reassessment to look for change
   1. Interval depends on patient's anatomy, diagnosis and clinical course

VIII. Describe appropriate patient instructions

A. Indirect gonioscopy
   1. Explain importance of technique and need for periodic reassessment
   2. Comfortably seated, upright, head fully in slit-lamp biomicroscope
3. Gaze in direction requested by examining physician

B. Direct gonioscopy

1. Explain importance and need for procedure and possible need for reassessment or additional treatment (in case of therapeutic gonioscopy)

2. Coordinate effort necessary with examiner and patient (if patient able to cooperate, i.e., not under general anesthesia, infant)

3. Patient comfortable and recumbent

4. Gaze in direction requested by examining physician

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


Clinical examination of the optic nerve

I. Most common methods
   A. Direct ophthalmoscopy
   B. Slit-lamp biomicroscopy with handheld lens
   C. Indirect ophthalmoscopy

II. List the indications/contraindications
   A. Indications
      1. To qualitatively examine the optic nerve head for clinical signs of glaucoma
      2. Can also be used to examine the optic nerve for evidence of other optic neuropathy
      3. Also used to examine the nerve fiber layer, macula and posterior pole
   B. Contraindications
      1. No absolute contraindications
      2. Difficult to use in cases of very small pupils and dense media opacities
      3. Poor patient cooperation

III. Describe the pre-procedure evaluation
   A. Evaluate pupil function and check for afferent pupillary defect
   B. Evaluate anterior segment
   C. Evaluate angle by gonioscopy
   D. Generally, requires dilation; do not dilate if angle is occludable

IV. List the alternatives to these procedures
   A. Other slit-lamp biomicroscopic techniques
      1. High power concave contact lens (Goldmann)
      2. Noncontact high power concave lens (Hruby)
   B. Fundus photography
      1. Stereo disc photography
      2. Red free nerve fiber layer photography
   C. Optic nerve imaging analyzers

V. Instrumentation and technique
   A. Direct ophthalmoscope
      1. Position the patient and examiner at similar eye levels
         a. Hold the direct ophthalmoscope with the right hand when examining the right eye and with the left hand when examining the left eye
b. Dial the patient’s refraction on the ophthalmoscope

c. Slowly move the ophthalmoscope closer to the eye and locate a blood vessel in the fundus

d. Adjust the power of the lens in the direct ophthalmoscope as needed to focus the blood vessel

e. Follow the blood vessel to locate the optic disc and evaluate

2. Considerations

a. Monocular view

b. The optic disc appears upright and not inverted

c. Estimate disc size by comparing it to the smallest light aperture size, corresponding to approximately 1.5 mm diameter.

d. Difficult to judge contour of the optic nerve head

B. Slit-lamp biomicroscope with hand held lens

1. A 60-, 66-, 78-, or 90-diopter lens is used in conjunction with the slit-lamp biomicroscope to provide a stereoscopic view of the disc

a. With a 66-diopter lens, the height of the slit beam indicated on the scale reading equals the vertical disc diameter in millimeters

2. Considerations

a. Stereoscopic view

b. The optic disc appears upside down and laterally inverted

c. The best view is through a dilated pupil, but with experience one can see the optic nerve through an undilated pupil, though usually monoscopically

d. Provides excellent illumination, high magnification, and a sense of contour of the optic nerve head

C. Indirect ophthalmoscopy

1. Head-mounted ophthalmoscope

a. +12, +20, +25, +28, or +30 diopter lens

2. Considerations

a. Stereoscopic view

b. The optic disc appears upside down and backward

c. View usually underestimates cupping and pallor as compared to slit-lamp biomicroscopy

d. Magnification is often inadequate for detecting subtle or localized optic nerve head changes

e. Not recommended for routine evaluation of the optic nerve head

f. Useful when examining young children, uncooperative patients, high myopes and eyes with significant media opacities

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

A. Transient photopsias with prolonged examination of the fovea

VII. Describe the considerations in interpretation for this diagnostic procedure

A. Evaluate the appearance of the optic cup and neuroretinal rim for signs of glaucoma

1. Generalized

a. Large optic cup vs large disc diameter

b. Asymmetry of the cups in the presence of symmetrical disc diameters

c. Progressive thinning of the neuroretinal rim

d. A non-glaucomatous optic neuropathy is considered if the degree of neuroretinal rim pallor appears
more prominent than the extent of cupping

2. Focal
   a. Narrowing (notching) of the rim
   b. Vertical elongation of the cup
   c. Cupping to the rim margin
   d. Regional thinning of the rim
   e. Splinter hemorrhage
   f. Nerve fiber layer loss

3. Less specific
   a. Exposed lamina cribrosa
   b. Nasal displacement of vessels
   c. Baring of circulinear vessels
   d. Peripapillary crescent or atrophy

VIII. Describe appropriate patient instructions

   A. The vision will be blurry and an after image may be seen
   B. Signs and symptoms of acute angle-closure glaucoma in those that might be predisposed after pupil dilation should be discussed
   C. Provide disposable sunglasses or request patient bring sunglasses or a driver if the patient is dilated

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
2. AAO EyeWiki, Examination of the Optic Nerve at the Slit-Lamp Biomicroscope with a Handheld Lens. 2014
Optic nerve head and retinal nerve fiber layer imaging

I. List the indications and contraindications

A. Indications - documenting and quantifying the optic nerve and retinal nerve fiber layer (RNFL) to allow qualitative and quantitative evaluations over time
   1. Diagnosis of glaucoma and other optic nerve disease
      a. Glaucoma causes thinning of the RNFL as early as 6 years before visual field loss
      b. RNFL thickness determinations may help in the early diagnosis of glaucoma.
      c. Macular thinning may be associated with glaucoma
      d. Glaucoma preferentially affects the ganglion cells, and the macula is defined anatomically as that region of the retina where the ganglion cell layer is more than one cell thick
   2. Interval follow up to detect structural change over time

B. Contraindications
   1. Absolute
      a. No view to the posterior pole
      b. Patient is physically unable to position self at machine
   2. Relative
      a. Poor image quality due to patient's inability to hold the eye still and focus on a target
      b. Poor image quality due to media opacity

II. Describe the pre-procedure evaluation

A. Ensure that the patient can understand and follow instructions
B. Enter patient demographics into the imaging machine because RNFL thickness values are compared to age-matched normals as the RNFL normally thins with aging
C. Ensure that there are no physical limitations to performing the test
   1. Seat the patient comfortably
   2. Check the head and eyelid position
D. Dilation is not necessary but is recommended for stereoscopic optic nerve photos and some OCT exams
E. The technician should ensure that the camera is centered on the optic nerve head or the patient is focused on the appropriate target

III. List alternatives to this procedure

A. An eye examination with red-free (green) light or RNFL photography may detect glaucomatous changes in the RNFL
B. A different imaging platform
   1. Heidelberg Retinal Tomography (HRT 2 or 3)
   2. Optical Coherence Tomography (Time Domain or Spectral Domain OCT)
   3. GDx with variable or enhanced corneal compensation
IV. Describe the instrumentation and technique

A. Optic nerve

1. Photography
   a. Color photograph of the nerve, best taken stereoscopically
   b. Can be analog or digital
   c. Quality of photo is operator dependent
   d. Non-mydriatic photography is possible, but the stereo quality is often sub-optimal so dilation is encouraged
   e. Images can be printed as slides, Polaroid or projected onto a computer screen digitally
   f. Baseline documentation and interval photos to qualitatively compare to baseline to detect change
      i. Different lighting, angle of photo and magnification may make comparison difficult
      ii. Relative position of blood vessels helpful to detect structural change
   g. Can be quantified using special software
   h. Unlike other methodologies, optic nerve photos are not subject to changing technology
      i. Optic nerve photos will always be interpretable while other scanning devices may have new versions that are not comparable to existing data sets

2. Confocal scanning laser ophthalmoscopy (HRT)
   a. Newer models auto detect disc boundary
   b. Performs serial cross-sections through the optic nerve head
   c. Two-dimensional scans are then reconstructed as a three dimensional topographic map
   d. The HRT then determines as “rim” all tissue superficial to a reference plane 50 microns below the retinal surface at the papillo-macular bundle and “cup” as tissue that is deep to the reference plane
   e. The amount of rim and cup within six sectors is then compared to a normative database
   f. The patient does not have to be dilated
   g. The spherical equivalent is dialed on the camera lens
   h. K readings are helpful
   i. The patient needs to be able to fixate on a target
   j. The machine will acquire three scans which are used to determine image quality
   k. Tracks changes in rim status over time

3. Ocular coherence tomography (TD-OCT)
   a. Performs radial line scans centered over the optic nerve head
   b. Analyzes the cup and disc areas which are divided by a reference line that is 150 microns above the retinal pigment epithelium
   c. The borders of the optic nerve head are determined by where the retinal pigment epithelium ends

B. Retinal nerve fiber layer (RNFL)

1. Red-free (green light) or RNFL photography
   a. Provides qualitative documentation
   b. The affected glaucomatous RNFL has a decreased light reflex and will appear darker due to enhanced visualization of the RPE
   c. The affected glaucomatous RNFL has a decrease in striated texture, which becomes flatter or smoother in appearance
   d. Larger retinal blood vessels, which may normally be buried within the RNFL, may be shown in relief as the RNFL thins and exposes these vessels

2. RNFL thickness values are affected by age, disc size and tilt and axial eye length
3. GDx based on scanning laser polarimetry (SLP)
   a. Technology has been replaced by OCT
   b. GDx gave a RNFL thickness estimate that was proportional to the true RNFL
   c. Measured the amount of phase retardation of polarized light as it traveled through the birefringent RNFL
   d. Phase retardation is proportional to RNFL thickness

4. Optical coherence tomography (OCT)
   a. OCT is similar to ultrasound, however it uses a low coherence, near infrared diode light source instead of sound
   b. OCT creates images based on the different reflectivity and backscattering properties of different structures in the eye
   c. There are two versions available
      i. Time Domain OCT (TD-OCT)
         i) Yields an axial resolution of about 10 microns
      ii. Spectral Domain OCT (SD-OCT)
         i) Generates many more A-scans in a shorter period of time, further improving axial resolution to about 5 microns
   d. Each eye is tested individually
   e. RNFL thickness scans
      i. All OCT machines provide a direct measurement of the RNFL thickness along a circle circumscribing the optic nerve
      ii. Some SD-OCT machines also evaluate the optic nerve head and macular thickness
      iii. Plotted against a normative database
      iv. SD-OCT machines can register images allowing change analysis

5. HRT is based on confocal scanning laser ophthalmoscopy
   a. HRT best measures optic disc surface topography.
   b. HRT measures the RNFL thickness indirectly
   c. The RNFL value given is the distance between the surface of the retina and a reference plane, which is 50 microns below the surface of the retina temporal to the optic nerve head (papillomacular bundle)
      i. The papillomacular RNFL is thought to be the last to change in thickness
      ii. Relative changes in the RNFL thickness can theoretically be detected if the retinal surface temporal to the disc does not change from scan to scan

C. Macular retinal thickness measurements (OCT)
   1. The scan is composed of six 6mm radial line scans centered over the macula
   2. Macular retinal thickness values are obtained and compared to a normative database
   3. Some SD-OCT machines can distinguish between outer and inner portions of the retina to provide thickness measurements more reflective of the ganglion cell and NFL.

V. Describe the considerations in interpretation for this procedure
   A. Assess the patient and test reliability
   B. Disc photos
      1. Patient needs to be able to fixate
      2. Stereo photos greatly help interpretation
         a. Allows for better evaluation of neuroretinal rim thinning
b. Allows for better evaluation of optic nerve blood vessel changes, which can suggest neuroretinal rim tissue loss

3. Disc hemorrhages can be detected and followed, which are not picked up well by any other automated technique for disc imaging

4. Red free photos, now easily acquired with newer digital cameras, can be used to detect and follow peripapillary RNFL loss

C. HRT

1. The standard deviation should be as low as possible. Ideally less than 20, but less than 40 is probably acceptable

2. A very irregular cross-section on the height profile graphs can mean poor image quality

3. On older machines, the operator must make sure that the contour line is drawn correctly. Areas of peripapillary atrophy can be challenging. Newer models automatically draw the contour line.

4. Review the regression analysis that flags regions of the neuroretinal rim that are outside the normal predicted range for disc size

5. Change can be monitored
a. Red or green pixels on the topography image (red means sinking away from the camera, green means moving toward the camera) can indicate thinning or thickening
b. Differences in clinical parameters such as neuroretinal rim area and volume or cup shape measure can be followed serially

6. The HRT measurements have not yet been proven to be better than a good clinical examination with stereo disc photos

D. GDx technology replaced by OCT

E. OCT

1. Assess the image quality

2. Signal strength grades are specific for each manufacturer and are provided by the company

3. Review the RNFL thickness graph
a. RNFL thickness values are given in microns and correspond to the RNFL along a circle centered around the optic nerve head
b. The letters T, S, N, and I indicate RNFL thickness values temporal, superior, nasal, and inferior to the optic nerve head
c. The patient's RNFL thickness graph is drawn by a single line for each eye
d. In an eye with normal RNFL, the graph has "double-hump" pattern in the superior quadrant and a single hump in the inferior quadrant indicating the quadrants with the thickest measurements
e. The green shaded area of the RNFL thickness graph indicates RNFL thickness values consistent with 95% of age-matched normals. The RNFL thickness values in the yellow shaded area are consistent with 5% of age-matched normals. The red area is consistent with 1% of age-matched normals

4. Review the optic nerve head scan
a. Optic nerve head topography can be assessed by some SD-OCT machines
b. Cup and disc ratios and areas can be assessed by some SD-OCT machines

5. Review the macular scan
a. Macular thinning is associated with glaucoma
b. The Ganglion Cell Complex (nerve fiber layer-ganglion cell layer-inner plexiform layer thickness) includes the retinal layers that are affected more by glaucoma and can be assessed by some SD-OCT machines
c. This may be used if there are no other pathologies that affect the macula

6. Correlate test results with anatomy as optic disc changes and RNFL thinning should correspond to the clinical exam and to visual field loss

7. Compare results with prior tests but there aren't longitudinal studies to assess what is considered "normal"
RNFL thinning due to aging alone

8. OCT measurements have not yet been proven to be better than a good clinical exam with stereo disc photos

VI. Describe the follow up care

A. Regular exams are needed to monitor for change in the optic nerve appearance

B. Be wary about making clinical decisions based on optic nerve imaging alone, especially if the imaging does not correlate with the clinical examination and visual field test.

C. No single imaging device outperforms the rest in distinguishing glaucoma from normal

Additional Resources

1. AAO. Basic and Clinical Science Course. 2015-2016; Section 10: Glaucoma.


Standard automated static perimetry

I. List indications/contraindications
   A. Diagnosis of disease
      1. Suspected diagnosis of glaucoma (suspicious disc, ocular hypertension)
      2. Neurologic vision loss
      3. Subjective visual field loss
      4. Macular/retinal disease
      5. Detection of malingerers
   B. Monitoring of disease process
      1. Interval follow-up of suspected or established visual field (VF) loss to detect progression
   C. Disability determination
   D. Testing for motor vehicle license

II. Describe the pre-procedure evaluation
   A. Ensure patient can understand and follow instructions
   B. Assess for refractive error
   C. Note pupil size
   D. Ensure there are no physical limitations to performing test
      1. Seat patient comfortably
      2. Check head and eyelid position
   E. Input accurate demographic information as normative database is age specific

III. List the alternatives to this procedure
   A. Confrontation visual fields
   B. Amsler grid (only tests central visual field)
   C. Kinetic Goldmann perimetry
   D. Tangent screen
   E. Frequency doubling perimetry
   F. Macular perimetry

IV. Describe the instrumentation and technique
   A. Bowl perimeter
      1. Threshold: stimulus intensity at which a patient responds 50% of the time
      2. Tests differential light sensitivity of specific points in VF to determine threshold values.
      3. Checks for localized and diffuse VF loss
   B. Each eye tested individually
   C. Stimulus size
1. Goldmann size III (4 mm²) stimulus
   a. Used in majority of tests
2. Goldmann size V (64 mm²) stimulus
   a. Used in advanced cases where size III not visualized in order to define less sensitive areas of field
   b. Used with poor visual acuity

D. Stimuli presented 6-degrees apart in central 30-degrees of field (30-2 test) or 2-degrees apart in central 10-degrees of field (10-2 test). 24-2 test is similar to 30-2, measuring central 24 degrees, except in the nasal field where 30 degrees is measured

E. Bracketing/staircase technique used to determine threshold at each point

F. Fixation losses, false positive (patient makes a response when no stimulus is present) and false negative responses (patient does not respond to brighter stimulus at a previously tested location) monitored to assess patient reliability

G. Perimetrist should note patient performance on print-out (e.g., good, fair, poor fixation; sleepy, agitated, inattentive)

H. White-on-white threshold perimetry

1. Standard full threshold program used for baseline and follow-up examinations
2. Humphrey Swedish Interactive Testing Algorithm (SITA) and Octopus Dynamic algorithms
   a. Results comparable to older threshold algorithms with shorter test times
   b. SITA testing takes less time and is more reliable than longer full threshold strategy
3. Fast algorithms (SITA Fast, FastPac) should be reserved for patients unable to perform standard tests because of physical or mental limitations

I. Short-wavelength automated perimetry/SITA SWAP available

1. Also called blue-on-yellow perimetry
2. Uses a yellow background and blue size V stimulus
3. Tests a smaller subpopulation of ganglion cells than white-on-white perimetry
4. May detect field loss 3-5 years sooner than white-on-white threshold perimetry
5. Useful in primary open angle glaucoma suspects who have not yet manifested field loss on standard testing
6. Useful for confirming field loss in areas of fluctuation in white-on-white perimetry
7. The standard test takes longer than white-on-white perimetry and has more limitations with cataracts/media opacities. The SITA SWAP algorithm reduces test time

V. Describe the considerations in interpretation for this diagnostic procedure

A. Confirm accuracy of patient demographics

B. Assess patient reliability

1. Computer monitors false positive responses, false negative responses and fixation losses. Perimetrist monitors fixation loss

C. Review threshold values, global indices, total and pattern deviation plots

D. Correlate test results with anatomy

1. Patterns of glaucomatous field loss (scotoma = area of relative or absolute loss of retinal sensitivity)
   a. Paracentral scotoma
      i. Occurs within 10 degrees of fixation
      ii. Results from loss of nerve fibers on the temporal aspect of the disc
   b. Arcuate / Bjerrum scotoma
      i. Occurs in an arc-like pattern 10-20 degrees from fixation
ii. A complete arcuate scotoma arches from the blind spot and ends at nasal raphe, and is wider and closer to fixation on the nasal side.

iii. Results from loss of nerve fiber bundles from the superotemporal or inferotemporal disc.

c. Nasal step
i. Relative depression of the nasal region of one horizontal hemifield as compared to the other
ii. Results from loss of nerve fibers at the superior or inferior pole of the disc

d. Altitudinal defect
i. Near complete loss of superior or inferior hemifield
ii. Results from advanced loss of neurons of the inferotemporal and inferior pole of disc if there is a superior field defect, or corresponding loss of superior disc rim if inferior field defect is present.

e. Temporal wedge
i. Wedge-shaped area of loss in temporal field with its apex at the blind spot
ii. Results from localized loss on the nasal side of the disc.

f. Generalized depression
i. Total deviation plot shows depression of entire field compared to age-matched controls without corresponding focal defects on the pattern deviation plot
ii. May occur with glaucoma but more common with media opacities (e.g., cataract)

2. Optic disc and retina appearance should correspond to visual field
3. Be aware of neurological defects that could mimic glaucomatous field loss

E. Rule-out artifactual field loss
1. Lens rim artifact
2. Incorrect refractive correction used for test
3. Cloverleaf field from high false negative responses
4. Posterior staphyloma (uncorrected refractive error)
5. Ptosis
6. Miosis
7. Incorrect patient age entered

F. Compare to prior tests
1. Establish good baseline with 2-3 fields
   a. Takes into account learning effect
   b. Establishes presence of scotomas and fluctuation level of patient's responses
      i. Guidelines for determining progression.
         i) Reproducible changes in the visual field consisting of deepening of an existing scotoma
         ii) Enlargement of an existing scotoma
         iii) Development of a new scotoma
   2. Humphrey STATPAC2, SITA and Octopus Delta software contain change analysis programs
      a. First two fields performed by a patient are used as baseline for comparison of subsequent fields
   4. Important to repeat test to confirm progression

G. Alternative testing for patients with unreliable responses or who have insufficient visual acuity for standard
automated perimetry

1. Goldmann kinetic perimetry
   a. Manual perimetric technique that uses a test object of a given size and intensity, and moves it from a non-seeing area in the peripheral field inward to a seeing area
   b. The test object is moved inward along several meridians toward fixation
   c. Point where object is first seen is plotted which effectively outlines an oval on the hill of vision
   d. This technique outlines the boundaries of the hill of vision at a given height and defines isopters
      i. Isopter = horizontal slice through the hill of vision, defined by stimulus size and intensity
         i) Area inside isopter should have sensitivity greater than the boundary
         ii) Scotoma = area within isopter of less than expected sensitivity
   e. Disadvantages
      i. Although easier test for patient than automated perimetry, results not as reproducible (due to it being a technician administered rather than a computer administered test)
      ii. Difficult to detect subtle changes important for following visual fields
      iii. Difficult to quantify change in the field

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Corneal pachymetry

I. List the indications/contraindications

A. Indications
   1. Measurement of central corneal thickness (CCT)
   2. Known diagnosis of glaucoma
   3. Primary open-angle glaucoma (POAG) suspect and/or ocular hypertension
   4. Prior to and after laser in situ keratomileusis (LASIK)/refractive surgery procedures
   5. Diagnosis and follow-up of congenital glaucoma

B. Contraindications
   1. Corneal abrasions
   2. Corneal infections

II. Describe the pre-procedure evaluation

A. No prior corneal manipulation (e.g., gonioscopy)
B. Adequate tear film (e.g., encourage blinking)
C. Probe perpendicular to pupil
D. Comfortably seated patient

III. Describe the instrumentation and technique

A. Ultrasound pachymeter (gold standard)
B. Optical coherence tomography (OCT)
C. Other techniques

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

A. Corneal abrasion
B. Corneal infection

V. Describe the considerations in interpretation for this diagnostic procedure

A. Confirm that pachymetry reading taken from central cornea
B. Consistent pachymetry values (at least 3 measurement values)
C. Intraocular pressure (IOP) should not be adjusted based on pachymetry readings. In eyes with little or no corneal edema, thinner corneas underestimate IOP while thicker corneas overestimate IOP
D. In eyes with significant corneal edema, thicker corneas may underestimate IOP
E. Ocular Hypertension Treatment Study
   1. Thinner corneas are a predictive risk factor for development of glaucoma in ocular hypertensive patients, independent of the potential effect of a thinner CCT to underestimate “true” IOP
   2. This finding suggests that a thinner CCT may be a possible marker for genetic susceptibility for optic nerve damage at any IOP
F. Reanalysis of target IOP (based on CCT reading)

G. Glaucoma patients with thinner CCT are more likely to be found among those with advanced stage of disease, normal-tension glaucoma, and black African ancestry

VI. Describe the follow up care

A. Baseline reading and possible remeasurement of CCT over time

VII. Describe appropriate patient instructions

A. Explanation of how CCT could alter course of glaucoma treatment

Additional Resources

1. AAO, Basic Clinical and Science Course, Section 10, Glaucoma, 2015-2016.
7. AAO, Preferred Practice Patterns Committee, Glaucoma Panel Primary Open Angle Glaucoma Preferred Practice Pattern, 2010.
I. Describe the approach to establishing the diagnosis

A. Describe the etiology of the disease
   1. Intraocular pressure (IOP) which is too high for an individual optic nerve is currently the principal modifiable risk factor
   2. Theories of glaucomatous damage
      a. Mechanical
         i. IOP-induced stress and strain may lead to deformation and remodeling of the lamina cribrosa resulting in axonal damage and disruption of axoplasmic transport
      b. Vascular theory
         i. Stresses the possible effects of IOP on the blood supply to the nerve
      c. Autoregulation
         i. A disturbance in the optic nerve vessel's ability to maintain vascular tone, thereby preventing constant blood flow to the nerve
         ii. Possibly independent of IOP
      d. Other theories currently being investigated leading to ganglion cell death
         i. Excitotoxicity, apoptosis, neurotrophin deprivation, molecular biologic abnormalities, autoimmunity

B. Define the relevant aspects of epidemiology of the disease
   1. Significant public health problem
      a. Estimated that 3 million Americans have glaucoma (approximately 50% do not know they have the disease)
      b. Prevalence in whites > 40 years old estimated at 1-2%
      c. Prevalence in African Americans and Hispanics 4 times higher than whites
      d. Prevalence increases with age
      e. Over 120,000 bilaterally blind in the US
      f. Most frequent cause of blindness in Hispanic and African-Americans

C. List the pertinent elements of the history
   1. Age and sex
   2. Race
   3. Symptoms
      a. Usually asymptomatic until late in disease
   4. Family history
   5. Cardiovascular disease, diabetes
   6. Myopia
   7. Medications
   8. Rule out secondary causes (i.e., corticosteroids)
   9. Previous eye injury and surgery

D. Describe pertinent clinical features
   1. Usually insidious onset
   2. Slowly progressive visual loss without symptoms
3. Painless
4. Usually bilateral, but often asymmetric
5. Central acuity usually unaffected until late in the disease
6. Elevated IOP
   a. Can be intermittent (diurnal fluctuation)
   b. 30-50% have IOP measurement < 21 mmHg on single reading
   c. Subset who never have high IOP (normal-tension glaucoma)
7. Open and normal angle by gonioscopy
8. Optic disc appearance
   a. Asymmetry of the neuroretinal rim area or cupping
   b. Focal thinning or notching of the neuroretinal rim
   c. Optic disc hemorrhage
   d. Any acquired change in the disc rim area or the surrounding retinal nerve fiber layer
   e. Large optic disc, large cup/disc ratio, peripapillary atrophy
9. Visual fields
   a. Defects can precede visible optic nerve damage in up to 20% of patients
   b. Visual field defects may not be detectable by standard perimetry until a significant number of nerve fibers are damaged
   c. Typical glaucoma defects
      i. Paracentral scotoma
      ii. Arcuate scotoma
      iii. Nasal step
      iv. Altitudinal defect
      v. Temporal wedge
      vi. Central island in far advanced cases
E. Describe appropriate laboratory/diagnostic testing for establishing the diagnosis
1. Clinical perimetry
   a. Automated static (most useful for glaucoma diagnosis)
   b. Manual kinetic and static
2. Measurement of corneal pachymetry
3. Optic nerve photography or detailed description (as per the POAG Preferred Practice Patterns)
   a. Stereo photography particularly useful
4. Optic nerve head and nerve fiber layer imaging systems
   a. Confocal scanning laser ophthalmoscopy
   b. Optical coherence tomography
   c. Scanning laser polarimetry

II. Define the risk factors
A. Strongest evidence
1. Older age
2. Race
3. Elevated IOP
4. Positive family history
   a. High prevalence for siblings and offspring of patients with glaucoma
      i. Baltimore Eye Survey (3.7 fold higher if siblings have glaucoma)
   b. Genetic studies successful in localizing genes associated glaucoma
      i. Myocilin; (3%-5% of POAG)
5. Decreased central corneal thickness
6. Decreased ocular perfusion pressure (OPP)

III. List the differential diagnosis
   A. Congenital
      1. Large optic nerve head with physiologic cupping
      2. Congenital disc anomalies
      3. Optic nerve coloboma
      4. Optic nerve pit
   B. Tilted disc syndrome/myopic disc
   C. Anterior ischemic optic neuropathy
   D. Compressive optic nerve lesions
   E. Other glaucomas
      1. Secondary open-angle types
      2. Angle-closure types

IV. Describe patient management in terms of treatment and follow-up
   A. Describe medical therapy options
      1. Adrenergic agonists (sympathomimetics)
      2. Beta-adrenergic antagonists
      3. Carbonic anhydrase inhibitors
      4. Parasympathomimetic agents
      5. Prostaglandin analogues
   B. Describe laser therapy options
      1. Selective laser trabeculoplasty (SLT)
      2. Argon laser trabeculoplasty (ALT)
   C. Describe surgical therapy options
      1. Non-penetrating filtration procedures
      2. Angle surgery
      3. Trabeculectomy (with or without antimetabolites)
      4. Glaucoma drainage implants
      5. Ciliary body ablation

V. List the complications of treatment, their prevention and management
A. Side effects of medications
   1. Local
   2. Systemic

B. Side effects of laser trabeculoplasty

C. Side effects of incisional glaucoma surgery

D. Side effects of ciliary body ablation techniques

VI. Describe disease-related complications

A. Limitations due to visual field loss

B. End stage glaucoma and blindness

VII. Describe appropriate patient instructions

A. Discussion of medications and surgical treatments
   1. Options, side effects, risk-benefit ratios
   2. Instructions relating to compliance
      a. Appropriate drop timing
      b. Nasolacrimal occlusion, passive lid closure
      c. Prevention of washout effect by spacing drop therapy
   3. Free glaucoma medications are available for those who cannot afford it. (Further information at http://www.aao.org/eyecare-america/resources)

B. Discussion regarding quality of life issues
   1. Support groups, career issues, financial issues regarding treatment

C. Advise patient that family members are at greater risk of having glaucoma and that they should seek examination by an ophthalmologist

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


20. AAO, Preferred Practice Patterns Committee, Glaucoma Panel. Primary Open Angle Glaucoma Preferred Practice Pattern, 2015.


Normal-tension glaucoma

I. Describe the approach to establishing the diagnosis

A. Definition of this disease
   1. Characteristic features of primary open-angle glaucoma (POAG) with an intraocular pressure (IOP) in the normal range without treatment
   2. Also described as low-tension glaucoma

B. Define the relevant aspects of epidemiology of this disease
   1. Represents between 20% and 30% of all cases of POAG in the United States
   2. More common in Japanese than in European populations
   3. Most often a disease of the elderly

C. List the pertinent elements of the history
   1. Associated functional vasospasm events such as migraine headaches and Raynaud's phenomenon more common
   2. Association with systemic hypotension particularly nocturnal hypotension
   3. Association with sleep apnea
   4. History of significant blood loss requiring transfusion

D. Describe pertinent clinical features
   1. No clear difference from optic nerve cupping seen in POAG
   2. Peripapillary halos and atrophy may be more common
   3. Optic disc hemorrhages prevalent
   4. Visual field defects may be closer to fixation

E. Describe appropriate testing and evaluation for establishing the diagnosis
   1. Consider computed tomography (CT) or magnetic resonance imaging (MRI) if visual field loss is suggestive of congruous, bitemporal, or other non-glaucomatous neurologic defects
      a. Imaging may also be warranted if
         i. Optic nerve pallor is out of proportion to degree of cupping,
         ii. Visual field defects greater than expected based on amount of cupping,
         iii. Color vision defect out of proportion with degree of cupping
         iv. Unilateral progression despite equal IOP in both eyes
      b. In such cases, medical and neurologic evaluation should be considered, including tests for anemia, heart disease, carotid insufficiency, syphilis, and temporal arteritis or other causes of systemic vasculitis
   2. Diurnal IOP curve to rule out pressures above normal range
   3. Consider 24-hour blood pressure monitoring

II. Define the risk factors

1. Family history
2. Vasospasm
3. Low systemic blood pressure
4. Advanced age
5. Thin central cornea
6. Female gender (at least in the United States)
7. Japanese ethnicity

III. List the differential diagnosis

A. Undetected high-pressure glaucoma
   1. POAG with large diurnal pressure variation
   2. Intermittent elevation of IOP caused by another type of glaucoma
      a. Intermittent angle-closure glaucoma
      b. Glaucomatocyclitic crisis
      c. Corticosteroid glaucoma (steroid-induced glaucoma)
   3. Previous episode of elevated IOP
      a. "Burned-out" glaucoma (e.g., pigmentary glaucoma)
      b. Wind or brass instrument player
      c. Any prolonged periods of position with head down (yoga, certain surgeries)
   4. Decreased central corneal thickness

B. Nonglaucomatous optic nerve disease resembling glaucoma
   1. Congenital anomalies
   2. Compressive lesions of the optic nerve and chiasm
   3. Ischemic optic neuropathy
   4. Compromised ocular blood flow
      a. Shock optic neuropathy
      b. Nocturnal hypotension
      c. Sleep apnea
   5. Buried optic disc drusen

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

A. Determine the severity of the disease
B. Start with medical therapy in an attempt to lower IOP > 30%
   1. Multicenter trial has proven the benefit of significant IOP lowering in patients with normal-tension glaucoma
C. For patients on systemic antihypertensive medications, consult with other providers to avoid systemic hypotension
D. Recognize effects of nocturnal systemic hypotension
E. Consider laser trabeculoplasty
F. Consider filtering surgery
G. Tube shunt surgery may be required

V. Describe disease-related complications

A. Limitations due to visual field loss
B. End stage glaucoma and blindness
VI. Describe appropriate patient instructions

A. Discussion of medications and surgical treatments

1. Options, side effects, risk-benefit ratios
2. Instructions relating to compliance
   a. Appropriate drop timing
   b. Nasolacrimal occlusion, passive lid closure
   c. Prevention of washout effect by spacing drop therapy

B. Discussion regarding quality of life issues

1. Support groups, career issues, financial issues regarding treatment

C. Advise patient that family members are at greater risk of having glaucoma and that they should seek examination by an ophthalmologist

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Primary open-angle glaucoma suspect

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
   1. Primary open-angle glaucoma (POAG): intraocular pressure (IOP)-sensitive optic neuropathy
   2. Multifactorial disease state (e.g., genetics, age, vasospasm, etc.)

B. Define the relevant aspects of epidemiology of this disease
   1. Number of POAG suspects far exceeds the number of people actually diagnosed with glaucoma
   2. Estimated that 3 million Americans have glaucoma (approximately 50% do not know they have the disease)
   3. Estimated that 130,000 persons are blind from glaucoma in the United States
   4. Increased prevalence of POAG among certain racial and ethnic groups (e.g., African descent, Hispanic)
   5. Estimated prevalence of ocular hypertension in the U.S. is 4-7% among those older than 40 years

C. List the pertinent elements of the history
   1. Previous history of elevated IOP
   2. Previous diagnosis of glaucoma
   3. Family history of glaucoma
   4. Age
   5. Ethnicity of patient (e.g., African descent, Hispanic)
   6. Previous history of vision loss
   7. Previous medication history (e.g. topical or oral steroid use)
   8. Systemic medical history and review of systems (e.g., asthma)
   9. History of ocular injuries

D. Describe pertinent clinical features (could include one of findings 1-3 listed below associated with 4. Note: 2 or more of findings 1-3 usually supports the diagnosis of POAG; however, any patient who shows deterioration of optic nerve head status (2 or 3 below) consistent with glaucomatous damage should be diagnosed as having POAG)
   1. IOP above 21 mm Hg (i.e., ocular hypertension)
   2. Optic disc or nerve fiber layer damage suggestive of glaucoma
      a. Enlarged cup/disc ratio
      b. Cup/disc ratio asymmetry between eyes
      c. Notching or narrowing of the neural rim
      d. Disc hemorrhage
      e. Diffuse or local abnormality in the nerve fiber layer
   3. Visual fields suspicious for early glaucomatous damage
   4. Normal open angle on gonioscopy/absence of secondary causes

E. Describe appropriate testing and evaluation for establishing the diagnosis
   1. Determination of central corneal thickness (CCT) with electronic pachymeter
   2. Evaluation of optic nerve head and retinal nerve fiber layer (e.g., drawings, stereo disc photographs, optic disc and nerve fiber layer analyzers, etc.)
3. Gonioscopy
4. Visual field testing and analysis (e.g., conventional white-on-white automated perimetry, full-threshold; frequency doubling perimetry, and short wavelength automated perimetry (SWAP))

II. Define the risk factors

A. Strongest evidence
   1. Older age
   2. Race (3 times higher prevalence in African-Americans, and higher prevalence for Hispanics)
   3. Elevated IOP
   4. Positive family history
      a. High prevalence for siblings and offspring of patients with glaucoma
         i. Baltimore Eye Survey (3 to 4 fold higher if siblings have glaucoma)
      b. Genetic studies successful in localizing genes associated glaucoma
         i. Myocilin; (3%-5% of POAG)
   5. Decreased central corneal thickness
   6. Decreased ocular perfusion pressure (OPP)

B. Weaker evidence
   1. Diabetes mellitus, systemic hypertension, cardiovascular disease, vasospastic condition
   2. Myopia

III. List the differential diagnosis

A. Primary open-angle glaucoma (early)
B. Normal tension glaucoma (early)
C. Corticosteroid responder
D. Burned out pigmentary dispersion syndrome
E. Burned out corticosteroid-induced glaucoma
F. Previous history of trauma (i.e., angle recession glaucoma)
G. Previous or current inflammation (e.g., glaucomatocyclitic crisis)
H. Nonglaucomatous causes (e.g., compressive lesions, ischemic episodes)
I. Physiologic cupping/ anomalous optic nerves/ tilted discs/staphyloma

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

A. Careful examination
   1. Signs of early damage to the optic nerve and documentation of baseline findings of the optic nerve
      a. Focal notching
      b. Asymmetry of cupping
      c. Splinter disc hemorrhage
      d. Nerve fiber layer dropout
   2. Periodic documentation of optic nerve head appearance by color stereophotography or computer-based image analysis of the optic nerve head and retinal nerve fiber layer
   3. Subtle visual field defects
a. Conventional white-on-white automated perimetry
   i. Full-threshold
   ii. Swedish interactive threshold algorithm
b. Frequency doubling perimetry
c. SWAP

4. Evaluation of IOP level
   a. Consideration of CCT
   b. Tono-Pen or pneumotonometry may be more accurate when corneal scarring is present
c. Diurnal IOP assessment
d. Estimate target IOP level

5. Follow-up
   a. Identify patients at risk of developing glaucoma
   b. Document appearance of optic nerve or nerve fiber layer
c. Identify, at an early stage, patients with early glaucoma
d. Identify patients at high risk of developing glaucoma
e. Treat high-risk individuals if indicated

6. Initiation of therapy
   a. Categories of patients
      i. Clear-cut diagnosis of open-angle glaucoma
      ii. Patients with significant risk of developing glaucoma (based on assessment of all risk factors)
      iii. Patients with elevated IOP level (No clear consensus on the upper level of IOP to begin prophylactic treatment)
b. Target IOP level
      i. Estimate target IOP level (lowering of 20-30% from baseline)
      ii. Attempt to maintain IOP at or below this target level with appropriate therapeutic intervention
      iii. Ocular Hypertension Treatment Study target IOP: 20% less than mean of several baseline measurements

7. Describe medical therapy options
   a. Listed alphabetically
      i. Alpha 2 adrenergic agonists
      ii. Beta-adrenergic antagonist
      iii. Carbonic anhydrase inhibitors
      iv. Combined medications
      v. Parasympathomimetic agents
      vi. Prostaglandin analogues
   b. Describe surgical therapy options (exceptionally rare instances)
c. Laser trabeculoplasty (with argon, diode, or selective laser surgery techniques)
d. Microinvasive glaucoma surgery
e. Glaucoma filtering surgeries

V. List the complications of treatment, their prevention and management
A. Risks of therapy
   1. Complications of medical therapy options
   2. Complications of surgical therapy options
   3. Inadequate and/or ineffective therapy causing glaucomatous progression

B. Expense of therapy

C. Alterations in quality of life with therapy

VI. Describe disease-related complications
   A. Development of POAG
   B. Progression of preexisting glaucoma
   C. Development of other types of glaucoma (e.g., corticosteroid-induced glaucoma)
   D. Failure to diagnose nonglaucomatous conditions mimicking disease
   E. Psychosocial issues

VII. Describe appropriate patient instructions
   A. Need for periodic follow-up
   B. Rationale for individualized therapy
   C. Check/confirm family history for glaucoma with referral if indicated

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Glaucoma Panel. Primary Open-Angle Glaucoma Suspect Preferred Practice Pattern, 2015.


23. AAO, Focal Points: Dysfunctional Glaucoma Filtration Blebs, Module #4, 2002

24. AAO, Focal Points: Automated Perimetry, Module #10, 2002

25. AAO, Focal Points: Normal Tension Glaucoma, Module #12, 2000

26. AAO, Focal Points: Techniques of Glaucoma Filtration Surgery, Module #6, 2000

27. AAO, Focal Points: Aqueous Shunting Procedures, Module #3, 2002

28. AAO, Focal Points: Update on Glaucoma Clinical Trials, Module #9, 2003

Exfoliation syndrome
(Pseudoexfoliation)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Systemic disorder with widespread deposition of fibrillar material in many organs including the anterior segment of the eye
2. Mutations in the LOXL1 gene that, while common in controls, are dramatically more common in exfoliation

B. Define the relevant aspects of epidemiology of the disease

1. Increasing prevalence with age: rare < 50 years old, most common >70 years old
2. Present in almost all ethnic groups
   a. Accounts for approximately 50% of open-angle glaucoma (OAG) in Scandinavian countries
   b. Low prevalence in African-Americans
3. Actual prevalence most likely much higher than that visible on clinical examination
4. Hereditary pattern uncertain

C. List the pertinent elements of the history

1. Family history

D. Describe pertinent clinical features

1. Deposits of exfoliative material (XFM) on anterior lens surface in a target like pattern (centrally and peripherally with an intermediate clear area
2. Deposits of XFM on pupillary margin, corneal endothelium, and inferior chamber angle zonules, and ciliary processes
3. Transillumination defects at pupillary margin
4. Pigment deposition on iris surface, endothelium
5. Gonioscopy
   a. Patchy increased pigmentation of trabecular meshwork
   b. Inferior scalloped pigment deposition is often present anterior to Schwalbe line (Sampaolesi line)
6. Poor pupillary dilation
7. Higher incidence of narrow angles
8. Zonular weakness, phacodonesis
9. Varying degrees of asymmetry, often monocular at onset
10. Glaucoma in exfoliation syndrome differs from primary open angle glaucoma as follows:
    a. Large fluctuations of intraocular pressure (IOP)
    b. May present with acute IOP elevation
    c. May be more difficult to control
    d. Greater optic nerve and visual field damage at time of diagnosis
    e. Poorer response to medications
    f. More frequent need for surgical intervention

II. Define the risk factors
A. Increasing age
B. Scandinavian ancestry
C. LOXL-1 mutations

III. List the differential diagnosis

A. Pigmentary dispersion syndrome
B. Uveitis
C. Angle-closure glaucoma
D. Fuchs heterochromic iridocyclitis

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

A. Exfoliation syndrome without glaucoma - examination frequency based on IOP and other risk factors
B. Exfoliation syndrome with glaucoma
   1. Describe medical therapy options
      a. Stepwise approach similar to OAG
      b. Poorer response to medications
   2. Describe surgical therapy options
      a. Laser iridotomy in presence of narrow angle
      b. Laser trabeculoplasty
      c. Trabeculectomy
      d. Aqueous shunt surgery
      e. Angle surgery
      f. Non-penetrating glaucoma surgery
      g. Ciliary body ablation
C. Gonioscopy should be repeated periodically

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

A. Medical treatment
   1. Side effects of medications
B. Laser trabeculoplasty
   1. Higher risk of post-laser surgery inflammation and IOP rise
   2. Greater loss of effectiveness over time
C. Trabeculectomy
   1. Comparable results to OAG patients
   2. Cataract surgery
   3. Poor pupillary dilation
      a. Posterior synechialysis
      b. Pupillary enlargement by retractor hooks, expansion ring or bimanual stretching
   4. Vitreous loss
      a. Zonular dialysis
b. Lens dislocation

c. Capsular rupture

5. Intraocular lens decentration/subluxation
   a. Intraoperative
   b. Spontaneous subluxation may occur even many years after surgery

6. Postoperative inflammation and IOP rise

7. Consider cataract surgery at an earlier stage to mitigate risk of intraoperative complications

VI. Describe disease-related complications

   A. Risk of developing glaucoma cumulative over time
      1. Risk factor for the conversion of OHT to glaucoma
      2. Risk factor for progression of open angle glaucoma
      3. Risk factor for angle closure

   B. Exfoliative material has been found elsewhere in the body

   C. An association with increased serum homocysteine levels and vascular occlusive disease has been described

VII. Describe appropriate patient instructions

   A. Nature of a chronic disease
   B. Medication instructions
   C. Compliance issues
   D. Long term followup
   E. Advise patient of greater risk of cataract surgery

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


Pigmentary glaucoma

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

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

1. Concave peripheral iris configuration, usually in myopic eye with deep anterior chamber (AC)
   a. Posterior iris surface contacts lens zonules and with physiologic movement of pupil, pigment rubbed free from iris and dispersed in aqueous
   b. Collection of pigment within angle/trabecular meshwork occurs during normal aqueous circulation and causes obstruction to outflow and chronic intraocular pressure (IOP) elevation

2. Theory of reverse pupillary block
   a. Pressure in AC higher than posterior chamber (PC) and iris pushed posterior causing iris-zonule contact
   b. Mechanism
      i. Aliquot of aqueous introduced into AC during eye blinking
      ii. Increased AC pressure forces iris against lens
      iii. Flap valve created that maintains pressure differential between AC and PC
      iv. Relative pressure difference between the two chambers exacerbates concave peripheral iris configuration and iris-zonule contact
   c. Laser iridotomy flattens iris decreasing zonule contact and pigment dispersion (controversial and only effective in selected individuals)

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

1. Usually affects ages 20-45 years
2. Prevalence 1-2% in Caucasian populations
3. Ocular hypertension and glaucoma occur in 25%-50% of PDS
4. Accounts for 1-1.5% of glaucoma in the western world
5. Autosomal dominant inheritance with incomplete penetrance
6. More common in males

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

1. Halos, blurring and/or eye pain associated with strenuous physical exercise
   a. High impact exercise increases pigment dispersion and may have acute elevations of IOP as high as 50 mm Hg
2. Increased IOP after pharmacological pupil dilation due to increased pigment dispersion
3. Myopia
4. Family history of glaucoma

D. **Describe pertinent clinical features**

1. Classic triad of pigmentary glaucoma
   a. Krukenberg spindle
   b. Heavy 360 degree trabecular meshwork (TM) pigment
   c. Mid-peripheral iris transillumination defects.
2. Corneal findings
   a. Krukenberg spindle
      i. Accumulation of pigment on posterior surface of central cornea in vertical spindle pattern
due to the convection current of aqueous

b. May have peripheral corneal endothelial pigmentation, especially inferonasal and inferotemporal

3. Iris
a. Anterior surface pigment
   i. Pigment particles in concentric dilation furrows of iris
   ii. Rarely pigment collection so heavy it makes iris darker and results in heterochromia
b. Configuration
   i. Concavity of peripheral iris with thin stroma
c. Transillumination defects
   i. Areas where pigment has been released from ruptured posterior iris pigment epithelial cells by contact with zonular packets
   ii. Occur in radial, spoke-like pattern in mid-peripheral iris
      i) Over years, defects may increase in size and coalesce to form almost a full ring
      ii) Can be seen using slit-lamp biomicroscope
         (i) With light source coaxial to eyepieces, a short narrow slit beam with brightest illumination shown through undilated pupil; red reflex seen through iris defects
   iii. May not be seen in individuals of African descent with PDS because of thick iris stroma
d. Pupil
   i. Anisocoria may result due to asymmetric iris chafing
      i) Pupil may be large and distorted in the direction of maximal iris chafing

4. Lens/zonule
a. Ring of pigment on posterior surface of lens in region of contact between posterior hyaloid and peripheral lens capsule (best seen at the slit lamp on lateral gaze after wide dilation)
b. Anterior zonule can be covered with pigment particles

5. Gonioscopy
a. Heavy dark homogeneous pigmentation of TM
b. May have pigmentation on or anterior to Schwalbe’s line, heaviest inferiorly
c. Angle recess very wide, usually approx. 45-degrees
d. Midperipheral iris often concave in appearance

6. Retina
a. Lattice degeneration with holes in about 20% of those with PDS
b. Increased risk of retinal detachment

E. Describe appropriate testing and evaluation for establishing the diagnosis
1. No special testing needed since diagnosis based on characteristic clinical findings
a. Ultrasound biomicroscopy
   i. Peripheral iris concavity with posterior iris surface draped over the lens margin and close to/touching the zonule
   ii. Iris concavity decrease after peripheral iridotomy

II. Define the risk factors
A. Young age
1. PDS usually affects young adults 20-50 years of age
2. With age, less pigment dispersed and may move into quiescent phase
   a. Age-related pupil miosis and increased anterior-posterior diameter of lens increases physiologic pupillary block and decreases peripheral iris concavity, decreasing contact between iris and zonule and pigment dispersion

B. Male gender
C. Myopia
D. European ancestry

III. List the differential diagnosis

A. Iris transillumination defects
   1. Congenital glaucoma - usually more peripheral and not radial
   2. Pseudoexfoliation syndrome - usually at pupil border and speckled defects throughout iris
   3. Postsurgical/post-trauma - irregular patches of pigment loss
   4. Herpes zoster and simplex keratouveitis - usually sectoral

B. Pigment dispersion
   1. Circulating pigment in AC may occur after pharmacologic dilation rarely with chronic rhegmatogenous retinal detachment

C. Increased TM/corneal endothelial pigmentation
   1. Pseudoexfoliation syndrome
      a. Increased TM pigment but not as homogeneous as PDS
      b. Pigment dusting of endothelium, not in spindle pattern
      c. No pigment on posterior peripheral lens
   2. Iris or ciliary body cysts
      a. Can disperse iris pigment to TM
      b. Some endothelial pigment but not in spindle
      c. No pigment on posterior lens
   3. Melanomas (iris, ciliary body, choroid)
      a. Pigmented tumor cells or pigment-laden macrophages may cause darkening of intraocular structures
      b. No Krukenberg spindle, no iris transillumination defects
         i. Tumor usually visible
   4. Inflammation
      a. Pigment mostly in inferior angle
      b. Other signs of inflammation present
   5. Intraocular lens (IOL)
      a. IOL in contact with posterior iris and causes chafing
      b. Iris transillumination not in radial pattern

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

A. Describe medical therapy options
   1. Prostaglandin analogues
   2. Beta-adrenergic antagonists
3. Alpha-adrenergic agonists
4. Carbonic anhydrase inhibitors
5. Parasympathomimetic agents

B. Describe surgical therapy options

1. Laser trabeculoplasty
   a. IOP-lowering effect may be short-lived due to continual dispersion of pigment
   b. Amount of energy needed for treatment less due to increased TM pigment
   c. Consider stepped treatment with 180 degree or less initially in order to avoid IOP spikes
2. Laser iridotomy (controversial)
   a. Addresses reverse pupillary block component of disease
3. Incisional surgery
   a. Angle surgery
   b. Non-penetrating procedures
   c. Trabeculectomy
   d. Aqueous shunt

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

A. Medical therapy: side effects related to individual medications
   1. Parasympathomimetic agents
      a. Constrict pupil and pull iris away from zonule, decreasing pigment dispersion
      i. However, use in young people problematic because of side effects including induced myopia and ciliary spasm

B. Surgical therapy: incisional surgery (See Aqueous shunt surgery and Incisional filtering surgery for open angle glaucoma)
   1. Avoid overfiltration as incidence of hypotony maculopathy higher due to young age of patients and myopia

VI. Describe disease-related complications

A. Characteristic glaucomatous optic disc damage and visual field loss

VII. Describe appropriate patient instructions

A. Compliance with treatment regimen
B. Discuss exercise-induced symptoms
C. Retinal detachment precautions

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
I. Describe the approach to establishing the diagnosis

A. Define the relevant aspects of epidemiology of the disease

1. Phacolytic
   a. Secondary to leakage of high molecular weight lens protein, usually in eyes with mature or hypermature cataract
   b. Sudden onset of open-angle glaucoma
   c. Usually monocular, not reported in children, rare in young adulthood
   d. These patients may have long-standing poor vision in the affected eye

2. Lens particle
   a. Occurs when lens cortex material obstructs the trabecular meshwork
   b. Often delay of days to weeks between trauma and onset of glaucoma
   c. Similar form may occur years after CE with lens material freed into anterior chamber
   d. Can occur in children and adults
   e. Can occur immediately after Nd:YAG capsulotomy

3. Phacoantigenic
   a. Uncommon granulomatous inflammation in response to liberated lens material
   b. Rarely causes glaucoma, more likely to be associated with hypotony
   c. Almost always unilocular, although some reports of sympathizing reaction

4. Ectopia lentis
   a. Autosomal dominant inheritance
      i. Simple ectopia lentis
      ii. Weill-Marchesani syndrome
      iii. Marfan syndrome
   b. Autosomal recessive inheritance
      i. Simple ectopia lentis
      ii. Ectopia lentis et pupillae
      iii. Weill-Marchesani syndrome
      iv. Homocystinuria
   c. Early age of onset in inherited conditions

5. Phacomorphic
   a. Multifactorial mechanism
   b. Pupillary block often contributory
   c. Intumescent lens narrows angle, causing closure

B. List the pertinent elements of the history

1. Phacolytic
   a. Phakic status
   b. Elderly patient with long-standing poor vision who has sudden onset of pain and conjunctival hyperemia
2. Lens particle  
a. Following disruption of lens integrity  
   i. Surgical: following cataract extraction (CE) or Nd:YAG  
   ii. Non-surgical: following lens injury  
   iii. Usually within weeks but may be months or years later  

3. Phacoantigenic  
a. Following CE with or without vitreous loss  
b. Following CE in one eye, with subsequent CE or leaking hypermature cataract in other involved eye  
c. After traumatic or spontaneous rupture of lens capsule  

4. Ectopia lentis  
a. Visual disturbances due to lens subluxation  
b. Family or personal history  
   i. Simple ectopia lentis  
   ii. Ectopia lentis et pupillae  
   iii. Marfan Syndrome  
   iv. Homocystinuria  
      i) Thrombotic vascular occlusions  
      ii) Mental retardation  
   v. Weill-Marchesani syndrome  
c. Trauma  
d. Prior ocular surgery  
e. Pseudoexfoliation syndrome (See Exfoliation syndrome (Pseudoexfoliation))  

5. Phacomorphic  
a. Acute or subacute onset  

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

2. Lens particle  
a. Open-angle  
b. Open capsule  
c. Free cortical material in AC  
d. Elevated IOP  
e. Moderate AC reaction
f. Microcystic corneal edema

g. Late: development of posterior and peripheral anterior synechiae

3. Phacoantigenic

a. Variable clinical presentation

b. Most common clinical picture:
   i. Open capsule
   ii. Moderate AC reaction with KP on corneal endothelium and anterior lens surface
   iii. Low grade vitritis
   iv. Peripheral anterior and/or posterior synechiae

4. Ectopia lentis

a. Lens subluxated or dislocated (into vitreous cavity or AC)

b. Body habitus
   i. Tall, slender with joint laxity and arachnodactyly (Marfan syndrome and homocystinuria)
   ii. Short, stocky with brachydactyly (Weill-Marchesani syndrome)

c. Variable refractive error

d. Unilateral lens displacement in trauma

e. Bilateral lens displacement in inherited conditions
   i. Superotemporal in Marfan syndrome and simple ectopia lentis
   ii. Inferonasal in homocystinuria

f. Small, round lens (microspherophakia) in Weill-Marchesani syndrome
   i. Consider microspherophakia in high myopes with pupillary block

g. Trauma-related ocular injuries

h. Phacodonesis

i. Shallow AC

j. Iris bombé configuration

k. Enlarged globe with long axial length (Marfan syndrome)

l. Vitreous prolapse

m. Thin sclera (Marfan, Ehlers-Danlos syndrome)

n. Retinal detachment

5. Phacomorphic

a. Advanced cataract

b. Shallow AC

c. Angle remains narrow despite patent iridotomy

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Phacolytic
   a. Gonioscopy: AC angle open without obvious abnormalities

2. Lens particle
   a. Microscopic findings: circulating white cells represent small leukocytes and larger macrophages as well as lens material
   b. Gonioscopy: open angle and may have cortical material in angle

3. Phacoantigenic
   a. Diagnosis often not made until pathologic examination of enucleated eye
Microscopic findings: granulomatous inflammation including epithelioid and giant cells; circulating lens protein or particles absent from aqueous humor

4. Ectopia lentis
   a. Systemic evaluation when appropriate

5. Phacomorphic
   a. Gonioscopy: angle closed, even with patent iridotomy

II. Define the risk factors

A. Phacolytic
   1. Elderly
   2. Poor or worsening vision
   3. Dense mature cataract

B. Lens particle
   1. Post CE
   2. Post Nd:YAG capsulotomy
   3. Post non-surgical ocular trauma

C. Phacoantigenic
   1. Following surgery or penetrating trauma

D. Phacomorphic
   1. Advanced cataract

III. List the differential diagnosis

A. Phacolytic: Primary angle closure, secondary angle closure, neovascular, uveitic, and traumatic glaucoma (ghost cell and angle recession)

B. Lens particle: Primary and secondary open-angle glaucomas (particularly uveitic varieties)

C. Phacoantigenic: Uveitis secondary to intraocular lens implants, sympathetic ophthalmia, bacterial endophthalmitis, other granulomatous uveitides

D. Ectopia lentis: primary angle closure, traumatic angle recession.

E. Phacomorphic: primary angle closure (acute or chronic)

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

A. Phacolytic
   1. Medication to control IOP
   2. Corticosteroids to reduce inflammation
   3. Definitive therapy requires cataract extraction

B. Lens particle
   1. Medical therapy to control IOP while residual lens material resorbs
      a. Aqueous suppressants
      b. Mydriatics
      c. Topical corticosteroids
   2. If glaucoma cannot be controlled, surgical removal of lens particles indicated
C. Phacoantigenic
1. Medical therapy
   a. Corticosteroids
   b. Aqueous suppressants
2. If unsuccessful, residual lens material should be surgically removed

D. Ectopia lentis
1. Medical therapy to control IOP
   a. Aqueous suppressants
   b. Hyperosmotic agents
2. Treat pupillary block
   a. Use parasympathomimetic agents with caution
   b. Cycloplegic agents may help move lens posteriorly
   c. Laser iridotomy
3. Lens removal indicated in
   a. Functionally significant monocular diplopia or uncorrectable refractive error
   b. Angle closure unresponsive to iridotomy
   c. Lens dislocation into AC

E. Phacomorphic
1. Medical therapy to control IOP
2. Treat pupillary block
   a. Avoid parasympathomimetic agents
   b. Laser iridotomy
3. Cataract extraction is definitive and may be primary treatment

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

A. Phacolytic
1. If cornea edematous
   a. Use topical glycerin to facilitate exam/surgery
2. Complications associated with medications
3. Complications associated with surgery

B. Lens particle
1. Titrate corticosteroid application to balance inflammation control and absorption of lens particles
2. Careful follow-up of Nd:YAG capsulotomy patient with early (1-2 hours post) IOP check
3. If not prompt response to medical therapy, surgery relatively early while lens particles are loose
4. Complications associated with medications
5. Complications associated with surgery

C. Phacoantigenic
1. Submit lens material for pathology to verify diagnosis
2. Complications associated with medications
3. Complications associated with surgery

D. Ectopia lentis
1. Complications associated with medications
2. Complications associated with surgery
   a. Vitreous loss
   b. Posterior lens dislocation
   c. Retinal detachment
   d. Thromboembolic events (homocystinuria)

E. Phacomorphic
   1. Complications associated with medications
   2. Complications associated with surgery
      a. Cataract surgery may be difficult due to shallow AC

VI. Describe disease-related complications

A. Phacolytic
   1. Persistent inflammation and pain
   2. Progressive elevation of IOP with disc damage

B. Lens particle
   1. Significant inflammation
   2. Peripheral anterior synechiae
   3. Increased IOP with trabecular dysfunction
   4. Open-angle glaucoma months to years later
   5. Delayed surgery may result in trapped lens material in inflammatory membrane

C. Phacoantigenic
   1. Loss of vision
   2. Inflammation and membrane formation
   3. Glaucoma
   4. Sympathetic ophthalmia

D. Ectopia lentis
   1. Chronic angle-closure glaucoma

E. Phacomorphic
   1. Chronic angle-closure glaucoma

VII. Describe appropriate patient instructions

A. Discuss disease process
B. Discuss medications, side effects
C. Discuss risks and benefits of surgery
D. Discuss risks and consequences of surgery on second eye in cases of phacoanaphylaxis

Additional Resources
1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Elevated episcleral venous pressure

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Obstruction of venous outflow
   a. Thyroid eye disease
      i. Elevated episcleral venous pressure (EVP) may occur in severe cases with marked proptosis and orbital congestion associated with an intraorbital infiltrative process
   b. Superior vena cava syndrome
      i. Lesions of the upper thorax may obstruct venous return from the head
   c. Retrobulbar tumors
   d. Cavernous sinus thrombosis

2. Arteriovenous anomalies
   a. Carotid cavernous sinus fistulas
      i. Traumatic
         i) Typical trauma is severe head injury resulting in a large fistula between the internal carotid artery and the surrounding cavernous sinus venous plexus
         ii) The internal carotid-cavernous sinus fistula causes high flow and high pressure shunting
      ii. Spontaneous (dural shunt syndrome)
         i) A small fistula is fed by a meningeal branch of the intracavernous internal carotid artery or external carotid artery
         ii) The small fistula results in low-flow, low-pressure shunting
   b. Orbital varices
      i. Characterized by intermittent exophthalmos and elevated EVP, usually associated with stooping over or the Valsalva maneuver
   c. Sturge-Weber syndrome
      i. Elevated EVP results from episcleral hemangiomas with arteriovenous fistulas

3. Idiopathic

B. Define the relevant aspects of epidemiology of the disease

1. Spontaneous carotid cavernous fistulas occur most often in middle-aged to elderly women with no history of trauma
2. The angiomata associated with Sturge-Weber syndrome are usually present at birth
3. The typical patient with idiopathic elevated EVP is elderly

C. List the pertinent elements of the history

1. History of trauma
2. Systemic hypertension
3. History of thyroid disease
4. History of head and neck tumors

D. Describe the pertinent clinical features

1. Thyroid eye disease most common cause of unilateral as well as bilateral proptosis and may be associated with severe proptosis and orbital congestion
2. Superior vena cava syndrome presents with exophthalmos, edema and cyanosis of the face and neck, and dilated veins of the head, neck, chest, and upper extremities.

3. Traumatic carotid cavernous sinus fistulas are characterized by pulsating exophthalmos, a bruit over the globe, conjunctival chemosis, engorgement of epibulbar veins, restriction of motility, and evidence of ocular ischemia.

4. Patients with spontaneous carotid cavernous fistulas present with prominent episcleral and conjunctival veins, but with minimal proptosis and no pulsations or bruit.

5. Orbital varices are characterized by intermittent exophthalmos which might be made worse by increasing venous pressure in the head and neck such as following a Valsalva maneuver or bending over.

6. Sturge-Weber syndrome presents with a port wine hemangioma of the skin within the trigeminal distribution.

   a. A prominent episcleral vascular plexus is found on the same side as the skin lesion.

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

1. Eye examination
   a. Tortuous, dilated episcleral veins
   b. Elevated intraocular pressure
   c. Blood in Schlemm canal
   d. Possible ocular ischemia, venous stasis, proptosis

2. Computed tomography (CT) scan or MRI of orbit
   a. Contrast scan of orbit helps establish diagnosis
   b. May demonstrate proptosis, swelling of extraocular muscles, and dilation of superior ophthalmic vein

3. Selective carotid angiography
   a. Diagnostic test of choice when one is considering carotid cavernous fistula
   b. Helps confirm diagnosis
   c. Provides therapeutic capability

4. Orbital ultrasound

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

A. Describe medical therapy options

1. Initially, medical therapy should be attempted

2. Theoretically, if glaucoma is entirely the result of elevated EVP, parasympathomimetic agents, such as pilocarpine, should have little effect

3. Drugs that decrease aqueous humor inflow and prostaglandin analogues are effective in some eyes

4. Medical therapy cannot lower intraocular pressure (IOP) below the abnormally high EVP unless the medication has an effect on the episcleral venous bed

B. Describe surgical therapy options

1. Laser trabeculoplasty is unlikely to be effective

2. Glaucoma filtration surgery provides more definitive lowering of IOP, but with substantially greater risks
   a. Filtration surgery provides a pathway for aqueous to bypass the hypertensive episcleral vascular bed

3. Glaucoma drainage device placement

4. Cyclophotocoagulation may provide an alternative to filtering surgery in some cases

III. List the complications of treatment, their prevention and management
A. **Intraoperative choroidal effusion or suprachoroidal hemorrhage**
   1. When EVP is elevated, the pressure differential increases significantly across the capillary membranes of uveal tissue when IOP is lowered to atmospheric pressure during surgery
      a. The increased transcapillary pressure favors the rapid transudation of fluid from the intravascular to the extravascular space
   2. Performing a posterior sclerotomy before entering the anterior chamber may allow choroidal fluid to drain.
      a. Leaving open scleral window may be helpful
   3. Tight closure of the scleral flap may lessen risk of choroidal hemorrhage

IV. **Describe disease-related complications**
   A. **Blindness if glaucoma goes untreated**

V. **Describe appropriate patient instructions**
   A. **Patients need to maintain good compliance with the medical treatment regimen**

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Traumatic hyphema

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
   1. Tear in the iris or ciliary body, causing bleeding from small branches of the major arterial circle
      a. Trauma - blunt or lacerating
      b. Intraocular surgery (laser and incisional)
         i. Especially associated with warfarin, aspirin
   2. Trauma - blunt or lacerating
   3. Intraocular surgery (laser and incisional)

B. Define the relevant aspects of epidemiology of the disease
   1. Peak incidence between ages 10-20 years
   2. Majority of patients are males - approximately 80%
   3. 70% have angle recession

C. List the pertinent elements of the history
   1. Ocular trauma or surgery
      a. Date of injury important to determine period of greatest risk of rebleed, length of treatment required
   2. Ocular or systemic disorders associated with spontaneous hyphemas
   3. Medications
   4. History of sickle cell trait or disease

D. Describe pertinent clinical features
   1. Decreased visual acuity
   2. Elevated intraocular pressure (IOP)
      a. Approximately 1/3 of hyphema patients
      b. Etiology of acute elevation
         i. Occlusion of trabecular meshwork (TM) by red blood cells (RBCs), clot, inflammatory cells, debris, and fibrin
         ii. Pupillary block secondary to clot occluding pupil
         iii. More common following recurrent hemorrhage or rebleeding
      c. Late-onset glaucoma
         i. Incidence ranges from 0-20%, days to years after injury
         ii. Etiology
            i) Damage to TM
            ii) Descemetization and fibrosis of TM
            iii) Siderosis of TM
            iv) Peripheral anterior synechiae (PAS) leading to secondary angle closure
            v) Hemolytic and/or ghost cell glaucoma if there is an associated vitreous hemorrhage
   3. Gonioscopic evidence of angle recession (tear into face of ciliary body)
   4. Blood in anterior chamber
      a. Circulating RBCs
      b. Layered hyphema
      c. Eight-ball hyphema (clotted total hyphema)
E. **Describe appropriate testing and evaluation for establishing the diagnosis**
   1. Sickle cell hemoglobin screening in all African-American patients
   2. Coagulation studies where indicated

II. **Define the risk factors**
   A. Secondary hemorrhage (rebleed) is often more severe than primary hemorrhage and is more likely to result in increased IOP, corneal blood staining and poorer visual prognosis.
      1. Average incidence of rebleeding is 5-10%
      2. Rebleeding usually occurs during the first week after initial hyphema
      3. Risk of rebleed is greater in African-Americans, larger hyphemas, delayed medical attention, sickle cell disease
   B. Larger hyphemas are associated with higher incidence of increased IOP
   C. Risk of glaucoma and other complications greater in patients with sickle cell trait or disease

III. **List the differential diagnosis**
   A. **Spontaneous causes of hyphema**
      1. Rubeosis iridis
         a. Diabetic eye disease
         b. Central retinal vein occlusion
         c. Carotid occlusive disease
         d. Chronic retinal detachment
      2. Vascular tufts of iris
      3. Juvenile xanthogranuloma
      4. Fuchs heterochromic iridocyclitis
      5. Blood dyscrasias
      6. Uveitis-glaucoma-hyphema (UGH) syndrome
      7. Tumors

IV. **Describe patient management in terms of treatment and follow-up**
   A. **Describe medical therapy options**
      1. Topical cycloplegics
      2. Topical and/or systemic corticosteroids
         a. Useful in treating associated inflammation
      3. Topical or systemic IOP-lowering medications as needed
         a. Hyperosmotics and systemic carbonic anhydrase inhibitors should not be used in patients with sickle cell trait or disease
         b. Parasympathomimetic agents contraindicated
      4. Aminocaproic acid
         a. May prevent rebleeding
         b. Contraindicated in eight-ball hyphemas
         c. Side effects include nausea, vomiting, postural hypotension
5. Analgesics and antiemetic medications as needed
   a. Avoid aspirin or nonsteroidal anti-inflammatory drugs (NSAIDs)
6. Rigid shield
7. Elevate head of bed to encourage blood to settle inferiorly
8. Bed rest or limited activity
9. Frequent examination during first week when risk of rebleed is highest

B. Describe surgical therapy options
1. Indications
   a. Persistently elevated IOP
      i. Patients with pre-existing glaucoma and or sickle cell may require earlier surgical intervention
   b. Risk of corneal blood staining
   c. Risk of synechiae formation
   d. Potential for amblyopia if hyphema obstructs vision in very young children
2. Surgical options
   a. Paracentesis
   b. Anterior chamber washout
      i. Irrigation and aspiration of blood either via paracenteses or with anterior vitrector if clot very dense
   c. Iridectomy if pupillary block present
   d. Trabeculectomy
   e. Glaucoma drainage devices

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

A. Complications associated with use of topical and systemic IOP-lowering medications
1. Avoid use where contraindicated
   a. Systemic carbonic anhydrase inhibitors may increase risk of sickling in patients with sickle cell trait or disease

B. Complications associated with trabeculectomies
C. Complications associated with glaucoma drainage devices

VI. Describe disease-related complications

A. Corneal blood staining
1. Occurs in setting of large or total hyphemas with increased IOP and/or corneal endothelial damage
2. May cause amblyopia in children
3. May require penetrating keratoplasty to clear visual axis

B. Ghost cell glaucoma
1. Occurs following vitreous hemorrhage
2. Degenerated RBC (erythroclast/ghost cells) develop in the vitreous, enter anterior chamber (AC) and obstruct outflow
3. Ghost cells
   a. Khaki-colored, spherical, rigid cells
b. Contain denatured hemoglobin adherent to the cell membrane (Heinz body)

4. Cells develop within 1-3 months following a vitreous hemorrhage
   a. Gain access to AC via disruption in anterior hyaloid face (previous surgery, trauma, spontaneous disruption)

5. Clinical findings
   a. Increased IOP
   b. History of recent vitreous hemorrhage
   c. AC filled with circulating or layered khaki-colored cells, may be mistaken for a hypopyon (or hidden by co-existing hyphema)
   d. AC taps and examination of fluid will reveal round, rigid "ghost" cells
   e. Cellular reaction out of proportion to aqueous flare
   f. Layering of ghost cells over inferior TM seen gonioscopically

6. Treatment
   a. Usually self-limited and resolves with clearing of hemorrhage
   b. Medical therapy with aqueous suppressants
      i. Initial approach
   c. Surgical therapy including AC washout, pars plana vitrectomy, and incisional glaucoma surgery
      i. If medical therapy fails
      ii. If RBCs or ghost cells persist

C. Hemolytic glaucoma
   1. Occurs following intraocular hemorrhage
   2. Hemoglobin-laden macrophages block TM
   3. Clinical findings
      a. Red-tinged cells floating in AC
      b. Reddish brown discoloration of TM
   4. Treatment as per ghost cell glaucoma

D. Angle Recession (tear in face of ciliary body)
   1. May lead to glaucoma months or years after injury
   2. Outflow obstruction is caused by adjacent angle damage resulting in proliferative or degenerative changes to meshwork or endothelialization of angle.

VII. Describe appropriate patient instructions

A. Compliance with prescribed medical and activity regimen to prevent complications
B. Quiet activity and/or bedrest
C. Avoid bending over, elevate head of bed
D. Provide safe home environment
E. Reliable follow-up
F. No aspirin or NSAIDs
G. Appropriate follow-up for angle recession and the future risk of glaucoma

Additional Resources
   1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


I. Describe the approach to establishing the diagnosis

A. List the pertinent elements of the history
   1. History of ocular trauma (often blunt trauma with hyphema)
   2. Traumatic event may occur months, years, or decades prior to development of glaucoma and may not be remembered

B. Define the relevant elements of the epidemiology of this disease
   1. 70-85% of ocular injuries occur in males
   2. The prevalence of visual impairment and blindness due to trauma is 3-4 times higher among African American men than white men
   3. Maximal risk occurs in young adults followed by those 70 years and older
   4. Up to 50% of fellow eyes may develop increased intraocular pressure (IOP) suggesting a possible predisposition to glaucoma
   5. Glaucoma may occur early or years after injury
   6. There is an increased risk of developing glaucoma when the area of angle recession is greater than 180 degrees

C. Describe pertinent clinical features
   1. Elevated IOP
   2. Optic nerve and visual field findings consistent with glaucoma
   3. Other signs of trauma to anterior segment (e.g., corneal scars, iris sphincter tears, Vossius ring; cataract or subluxed lens)
   4. Gonioscopy reveals:
      a. Angle recession (need to compare to opposite eye- since some eyes have very wide angles that appear to be recessed but are normal)
      b. Broad angle recess-wide ciliary body face
      c. Absent or torn iris processes
      d. White glistening scleral spur
      e. Depression in the overlying trabecular meshwork (TM)
      f. Localized peripheral anterior synechiae (PAS) at the border of the recession - extensive PAS occasionally mask recession

II. Define the risk factors

A. History of ocular trauma
B. Gonioscopic evidence of angle recession of 180 degrees or more

III. List the differential diagnosis

A. Pseudoexfoliation glaucoma
B. Neovascular glaucoma
C. Uveitic glaucoma
D. Glaucoma associated with tumor
E. Glaucoma associated with increased episcleral venous pressure
IV. Describe the management

A. Describe medical therapy options (Listed alphabetically)
   1. Alpha-adrenergic agonists
   2. Beta-adrenergic antagonists
   3. Carbonic anhydrase inhibitors (contraindicated in cases of hyphema associated with sickle cell anemia)
   4. Parasympathomimetics may cause a paradoxical rise in pressure
   5. Prostaglandin analogues

B. Describe surgical therapy options
   1. Laser trabeculoplasty is not usually recommended and is generally unsuccessful
   2. Trabeculectomy with antimetabolites
   3. In eyes with multiple prior procedures and scarred conjunctiva, consider a drainage device

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

A. (See Beta-adrenergic antagonists, Alpha adrenergic agonists, Prostaglandin analogues, Carbonic anhydrase inhibitors)

B. (See Incisional filtering surgery for open angle glaucoma)

C. (See Aqueous shunt surgery)

VI. Describe the disease-related complications

A. Blindness

B. Visual field loss

VII. Describe appropriate patient instructions

A. Regular IOP checks for the rest of life at least once a year

B. Appropriate eye precautions, including safety glasses, in future

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


Glaucoma and penetrating keratoplasty

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Open angle
   a. Wound distortion of the trabecular meshwork
   b. Corticosteroid-induced
   c. Inflammatory
   d. Ophthalmic viscosurgical device (OVD) (viscoelastic)
   e. Fibrous/epithelial ingrowth

2. Closed angle
   a. Chronic angle-closure
   b. Pupillary block
   c. Inflammatory
   d. Fibrous/epithelial ingrowth

B. Define the relevant aspects of epidemiology of the disease - incidence of glaucoma after penetrating keratoplasty (PK)

1. Early postoperative (immediate post-op until 3 months post-op)
   a. Up to 50% incidence of elevated IOP

2. Late postoperative (a chronic, elevated IOP more than 3 months after surgery)
   a. Up to 33% incidence
   b. More common in corneal leukomas and aphakic eyes.

C. List the pertinent elements of the history

1. Previous history of PK
2. History of pre-existing glaucoma
3. Lens status
4. Medications such as topical corticosteroid use

D. Describe the pertinent clinical features

1. Surface irregularity and astigmatism may make Goldmann tonometry inaccurate. Other methods of measuring IOP such as pneumotonometry or applanation tonometry should be considered
2. Eyes with corneal edema might have falsely low IOP readings
3. Gonioscopy to determine angle status
4. Lens status
5. Iris bombé
6. Ocular inflammation

II. Define the risk factors

A. Aphakic or possibly pseudophakic patients
B. Repeat penetrating keratoplasty
C. Pre-existing history of glaucoma
D. History of trauma
E. Undersized corneal buttons and tight sutures
F. History of inflammation
G. Incidence is low after PK for keratoconus

III. List the differential diagnosis

A. Pre-existing primary open-angle glaucoma (POAG)
B. Pre-existing corticosteroid-induced glaucoma
C. Inflammatory glaucoma
D. Aphakic glaucoma
E. Chronic angle-closure glaucoma
F. Acute angle-closure glaucoma

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

A. Carefully manage optic nerve structure and function after PK
B. Medical therapy options
   1. Topical medications include
      a. Alpha-adrenergic agonists
      b. Beta-adrenergic antagonists
      c. Cautious use of carbonic anhydrase inhibitors
      d. Prostaglandin analogues
   2. Oral medications - carbonic anhydrase inhibitors for IOP control prior to surgical management or if surgical management is contraindicated or refused
C. Surgical therapy options
   1. Laser trabeculoplasty
   2. Surgical therapy needed in some cases
   3. Surgical options
      a. Trabeculectomy with antimetabolites, although this alternative may be less desirable if a contact lens is necessary for visual rehabilitation
      b. Ex-PRESS device can be considered
      c. Tube shunt
      d. Cyclodestructive procedures
      e. Angle Surgery

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

A. Complications of treatment
   1. Medical
      a. Allergic reactions
Localized drop toxicity to the transplant

2. Surgical
   a. Graft failure more common and earlier onset for patients with glaucoma
      i. Perhaps due to more rapid endothelial cell loss from elevated IOP
      ii. Tube shunts markedly increase the risk of graft failure; this risk may be decreased by pars plana placement
   b. Trabeculectomy failure

B. Prevention of complications
   1. Oversized corneal grafts
   2. Careful graft suturing
   3. Consider surgical iridectomy
   4. Consider laser iridoplasty
   5. Consider use of topical cyclosporine A instead of topical corticosteroids for corticosteroid-responsive individuals
   6. Consider use of antimetabolites for trabeculectomy
   7. Careful position of the tube away from the endothelium (sulcus or pars plana)

C. Management of complications
   1. Tube repositioning into sulcus or pars plana
   2. Repeat PK
   3. Descemet stripping automated endothelial keratoplasty on prior graft

VI. Describe disease-related complications
   A. Corneal graft failure
   B. Progressive glaucomatous optic neuropathy

VII. Describe appropriate patient instructions
   A. Understand that intensive long-term follow-up is needed
   B. Use medications as prescribed to control IOP and prevent graft rejection
   C. Report any decline in vision

Additional Resources
1. AAO, Basic and Clinical Science Course, Section 10: Glaucoma, 2015-2016.


Corticosteroid-induced glaucoma
(steroid glaucoma)

I. Describe the approach to establishing the diagnosis
A. Describe the etiology of this disease
   1. Caused by a reduction in facility of outflow
      a. Polymerized glycosaminoglycans accumulate in the trabecular meshwork
      b. Suppression of trabecular meshwork phagocytic activity
B. Define the relevant aspects of epidemiology of the disease
   1. Approximately one-third of all patients will demonstrate some responsiveness to corticosteroids, but a smaller percentage will show a clinically significant elevation in intraocular pressure (IOP)
C. List the pertinent elements of the history
   1. May develop at any time during long-term corticosteroid administration, but IOP elevation typically occurs within a few weeks with potent corticosteroids, or in months with the weaker corticosteroids
   2. Routes of administration
      a. Topical corticosteroid therapy is more often associated with IOP rise than is the case with systemic administration
         i. May occur not only with drops or ointment applied to the eye, but also with corticosteroid preparations applied to the skin of the eyelids and scalp
      b. Periocular or intraocular injection is associated with the highest risk for elevated intraocular pressure
         i. IOP elevation may occur in response to subconjunctival, sub-Tenons, intravitreal, retrobulbar injections of corticosteroid, or intraocular corticosteroid implants
         ii. Patient’s response to earlier topical corticosteroid therapy does not always predict how that individual will respond to periocular corticosteroids
      c. Systemic administration (oral or intravenous) of corticosteroids is less likely to induce glaucoma
         i. This response does not correlate with the dosage or duration of treatment, but is associated with the degree of IOP response to topical corticosteroids
      d. Inhaled or nasal corticosteroids
D. Describe pertinent clinical features
   1. The clinical picture resembles that of primary open-angle glaucoma (POAG) with an open, normal-appearing anterior chamber angle and absence of symptoms
   2. Rarely the condition may have an acute presentation, in which IOP increases have been observed within hours after corticosteroid administration

II. Define the risk factors
A. Individuals with POAG or a family history of the disease are more likely to respond to chronic corticosteroid therapy with a significant rise in IOP

III. List the differential diagnosis
A. Primary open-angle glaucoma
B. Secondary open-angle glaucoma (pseudoexfoliation, pigmentary glaucoma, lens-induced glaucoma, ocular inflammation, elevated episcleral venous pressure)
C. Excess endogenous corticosteroids

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

A. Describe medical therapy options

1. Discontinuation of the corticosteroid or reduction in potency or frequency of administration. Consider switching to steroid-sparing agent

2. The chronic form of this disease is said to normalize in 1-4 weeks, while the acute form typically resolves within days of stopping the corticosteroid

3. In rare cases, the glaucoma may persist despite stopping all corticosteroids

4. In cases of uveitis, glaucoma medical therapy algorithm, as per treatment of POAG, although parasympathomimetic agents should be avoided and prostaglandin analogues used with caution

B. Describe surgical therapy options

1. In cases of medically uncontrollable glaucoma, laser trabeculoplasty can be considered, followed by filtration surgery if necessary

2. Occasionally may be necessary to excise a depot of periocular corticosteroid (or vitrectomy for intravitreal depot corticosteroids) if this appears to be responsible for the persistent IOP elevation

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

A. To avoid loss of vision from corticosteroid-induced glaucoma, the physician must know how to prevent or minimize the chances of occurrence. This requires close attention to the patient's history

B. All physicians who prescribe corticosteroids should be educated about the potential long-term ocular consequences of prolonged corticosteroid use

C. All patients with POAG, normal tension glaucoma, and POAG suspect should be educated on the ocular side effects of all corticosteroid medications

VI. Describe disease-related complications

A. Loss of visual function

VII. Describe appropriate patient instructions

A. Maintain compliance with physician instructions

B. Check IOP if corticosteroid use recurs

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma. 2015-2016.


4. AAO, Focal Points: Steroid Therapy for Ocular Inflammatory Disease, Module #7, 2006.


Acute primary angle-closure glaucoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
   1. Physiologic pupillary block
      a. Excessive iris-lens apposition impedes flow of aqueous from posterior chamber to anterior chamber, elevating posterior chamber aqueous pressure
      b. Secondary forward bowing of peripheral iris results in occlusion of the trabecular meshwork

B. Define the relevant aspects of epidemiology of the disease
   1. Ethnicity
      a. Lower prevalence in those of European and African descent
      b. Higher prevalence in Asians
      c. Highest known prevalence rates in Alaskan Inuits
   2. Higher prevalence in women
   3. Higher prevalence with increasing age

C. List the pertinent elements of the history
   1. Symptoms suggesting intermittent angle-closure
      a. Blurred vision
      b. Colored halos around lights
      c. Red eye
      d. Eye or brow pain or headache
      e. Nausea and vomiting
   2. Symptom onset after mydriasis due to
      a. Dim illumination
      b. Emotional stress
      c. Topical or systemic anticholinergic or adrenergic medicine
   3. Family history of acute angle-closure glaucoma

D. Describe the pertinent clinical features
   1. Symptoms
      a. Acute onset of brow ache, eye pain
      b. Blurred vision
      c. Colored halos around lights
      d. Nausea and vomiting
   2. Signs
      a. High intraocular pressure (IOP)
      b. Mid-dilated, poorly reactive pupil
      c. Corneal epithelial edema
      d. Congested episcleral and conjunctival vessels
e. Shallow peripheral anterior chamber
f. Anterior chamber inflammation
g. Appositional angle-closure
h. Iris bombé
i. Indicators of previous bouts of angle-closure glaucoma
   i. Glaukomflecken
      i) Small anterior subcapsular lens opacities
   ii. Sector iris atrophy
   iii. Posterior and peripheral anterior synechiae

E. Describe appropriate testing and evaluation for establishing the diagnosis
   1. Gonioscopy of both eyes is important
      a. Typical pupillary block angle closure should have narrow angles bilaterally
      b. Indentation gonioscopy to distinguish appositional from synechial angle closure
   2. Measure the glasses to confirm hyperopia
      a. Most patients are hyperopic
      b. If myopic, consider
         i. Anteriorly displaced or spherical lens
         ii. Plateau iris
         iii. Alternative diagnosis

II. Define the risk factors
   A. Hyperopia
   B. Family history of angle-closure
   C. Older age
   D. Female gender
   E. Age-related cataract (lens swelling) - phakic status
   F. Race

III. List the differential diagnosis
   A. Subacute primary angle-closure
   B. Chronic angle-closure
   C. Secondary Angle-closure with pupillary block
      1. Phacomorphic
      2. Lens or IOL dislocation
      3. Microspherophakia
      4. Inflammation with pupil occlusion and/or seclusion
      5. Intraocular silicone oil or expansile gas following vitrectomy
   D. Secondary angle-closure without pupillary block
      1. Ciliary body congestion and ciliochoroidal effusion secondary to:
         a. Idiosyncratic reaction to medications (e.g., sulfa, topiramate)
b. Nanophthalmos

c. Panretinal photocoagulation

d. Central retinal vein occlusion

2. Ciliary body cysts

3. Plateau iris

4. Aqueous misdirection

5. Posterior segment tumors

6. Scleral buckling procedure

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

A. Describe medical therapy options

1. Medical therapy is used to lower the IOP and allow clearing of corneal edema in preparation for laser iridotomy

   a. Beta-adrenergic antagonists

   b. Alpha 2-adrenergic agonists

   c. Carbonic anhydrase inhibitors - topical, oral, IV

   d. Parasympathomimetic agents - 1-2% pilocarpine

      i. Used once the IOP has begun to decrease

   e. Prostaglandin analogues

   f. Hyperosmotic agents

   g. Topical corticosteroids

   h. Avoid strong parasympathomimetic agents and alpha 1-adrenergic agents

2. Deformation of cornea with cotton tip applicator or indentation gonioscopy occasionally opens the angle

3. Topical glycerin or epithelial removal may be necessary to enable visualization of the chamber angle

4. Topical miotic prophylaxis of at-risk fellow eye with careful follow up.

B. Describe surgical therapy options

1. Anterior chamber paracentesis may be helpful acutely to reduce IOP, relieve pain and clear cornea for iridotomy

2. Laser peripheral iridotomy - Nd: YAG and/or argon

3. Prophylactic iridectomy in fellow eye

4. Incisional iridectomy when laser iridotomy is not possible

5. Primary filtering surgery may be required if extensive synechial closure is present or if IOP is not controlled with medical therapy

C. Follow-up

1. Gonioscopy following iridotomy to assess status of angle regarding synechial closure.

2. Laser iridoplasty (gonioplasty) as indicated by angle configuration, e.g. in plateau iris syndrome where angle may remain occludable after iridotomy

3. Medical treatment as indicated for residual stage of angle-closure

4. Goniosynechialysis, usually combined with cataract surgery, may improve aqueous outflow in eyes with residual angle-closure glaucoma

   a. The benefits of goniosynechialysis are controversial

5. Check the opposite eye
V. List the complications of treatment, their prevention and management

A. Complications of medical treatment
   1. Parasympathomimetics, esp. strong agents, may increase pupillary block by increasing iris vascular congestion and moving the lens-iris diaphragm anteriorly
   2. Alpha 1-adrenergic agonist can increase dilation and iris ischemia
   3. Complications from hyperosmotic use
      a. Congestive heart failure
      b. Tachycardia
      c. Intracranial hemorrhage

B. Complications of laser iridotomy
   1. IOP rise and inflammation should be treated with topical medications
   2. Dysphotopsia (monocular diplopia and glare)
   3. Hyphema

C. Peripheral anterior synechiae
   1. Perform laser iridotomy as soon as possible to reduce the risk of synechiae formation

D. Posterior synechiae
   1. Topical corticosteroids may prevent synechiae formation

VI. Describe disease-related complications

A. Residual angle-closure glaucoma
B. Late onset mixed mechanism open angle glaucoma
C. Corneal decompensation
D. Sectoral iris atrophy
E. Posterior synechiae
F. Cataract formation
G. Late onset optic nerve damage
H. Retinal vein occlusion

VII. Describe appropriate patient instructions

A. In situations where clinical judgment is made not to perform the prophylactic laser iridotomy at that time or the patient refuses the procedure, inform patients at risk about symptoms of acute angle-closure attacks with instructions to notify ophthalmologist immediately if they occur
B. Discuss possibility of visual disturbances such as ghost images, glare, or diplopia following iridotomy.
C. Warn patients at risk to avoid over-the-counter decongestants and any other medications carrying a warning against use in glaucoma (systemic medications with anticholinergic and adrenergic properties which may cause mydriasis)
   1. Warning may not be necessary after iridotomy
D. Stress importance of regular follow-up examinations following primary angle-closure glaucoma, especially in patients (e.g., Asians) who are prone to develop chronic angle closure despite patent iridotomies

Additional Resources
1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
2. AAO, Preferred Practice Patterns Committee, Glaucoma Panel: Primary Angle Closure Preferred Practice


I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
   1. Specific anatomic configuration of the anterior segment and the anterior chamber (AC) angle
   2. Relative pupillary block with appositional closure of a portion of the angle

B. Define the relevant aspects of epidemiology of this disease
   1. Inheritance
      a. A forward position of the lens and a greater-than-average lens thickness are determined by polygenic inheritance
   2. Race
      a. Frequency is greatest among East Asians and the Inuit and Yupik populations of Alaska, Canada, Greenland, and Russia
      b. Both intermittent and acute attacks are less common in African-Americans
   3. Sex
      a. Women affected 3 to 4 times as often as men
   4. Age
      a. Prevalence of intermittent and primary angle-closure glaucoma increases with age, peaking between 55 and 70 years then declining
   5. Refraction
      a. Associated with hyperopia most typically although it may occur in eyes with any refractive error

C. List the pertinent elements of the history
   1. Brief episodes of blurred vision, haloes, and mild ocular pain.
   2. Complaints of ocular fatigue associated with dim light, and near work
   3. Symptoms tend to recur under provoking circumstances (movie theaters)
   4. Resolves spontaneously after cessation of inciting activity or with sleep (miosis)
   5. Patient may complain of frequent headaches confused as migraines or sinus disease
   6. Frequently there are no identifiable symptoms
   7. History of angle-closure glaucoma in family

D. Describe pertinent clinical features
   1. Anterior chamber depth shallows in the periphery
   2. Lens tends to be situated more anteriorly; the lens may be large (phacomorphic angle-closure)
   3. Gonioscopy may reveal varying degree of iris convexity (determined by degree of pupillary block) and angle narrowing
      a. On indentation, peripheral anterior synchia (PAS) may be seen
   4. Increased pigmentation on the iris close to inferior angle
   5. Mild iris atrophy
   6. Chronically dilated poorly reacting pupil
   7. Varying degree of optic nerve cupping / often asymmetric

E. Describe additional testing and evaluation for establishing the diagnosis
   1. Visual field and Optic Nerve testing
a. Damage varies according to the severity, frequency, and duration of the attacks

2. Ultrasound or optical biometry
   a. Axial length, anterior chamber depth and lens thickness.

3. Anterior segment OCT to evaluate iridotrabecular apposition and angle parameters

4. Ultrasound biomicroscopy in bright and dim illumination to demonstrate anterior rotation of lens-iris diaphragm under appropriate situation to increase pupillary block and simulate subacute angle-closure

II. Define the risk factors
   A. Family history of angle-closure glaucoma
   B. Hyperopia
   C. Alaskan and Greenland Inuits, and East Asians
   D. Female
   E. Increasing age

III. List the differential diagnosis
   A. Plateau iris syndrome
   B. Intermittent secondary angle-closure related to instability of lens, e.g Pseudoexfoliation or trauma
   C. Acute angle-closure
   D. Glaucoma associated with uveitis

IV. Describe patient management in terms of treatment and follow-up
   A. Describe medical therapy options
      1. If intraocular pressure (IOP) is elevated, glaucoma medications can be used until laser peripheral iridotomy is done
   B. Describe surgical therapy options
      1. Laser peripheral iridotomy
      2. Argon laser iridoplasty
      3. Surgical iridectomy (in patients unable to undergo laser iridotomy)
      4. Surgical iridectomy associated with glaucoma filtration surgery
      5. Cataract surgery/removal of lens in phacomorphic angle-closure; may be combined with goniosynechialysis

V. List the complications of treatment, their prevention and management
   A. Post-laser surgery IOP elevation
   B. Post-laser surgery persistent inflammation
   C. Post-laser surgery cataract
   D. Malignant glaucoma (rare)
   E. Dysphotopsia and monocular diplopia uncommon after iridotomy

VI. Describe disease-related complications
   A. Glaucomatous disc damage and visual field loss
B. Acute angle-closure glaucoma
C. Chronic angle-closure glaucoma with progressive PAS formation

VII. Describe appropriate patient instructions and follow-up

A. Symptoms of acute angle-closure glaucoma attack and need for immediate attention
B. Need for frequent and lifelong follow-up examinations
C. Possibility of residual glaucoma following laser treatment which may require further medication and/or surgery
D. Examine opposite eye for risk of angle-closure glaucoma and treat if appropriate
E. Recommend evaluation of family members

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Chronic angle-closure glaucoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Peripheral iris bowing (anterior bowing)
   a. Prolonged apposition or repeated subacute attacks lead to gradual peripheral anterior synechiae (PAS) formation
   b. PAS begin as pinpoint synechiae reaching to the mid trabecular meshwork and then gradually expand in width
   c. Circumferential closure, which begins in the most peripheral area of the angle and moves toward the cornea
   d. Occurs more commonly in eyes with darker irides
   e. Closure occurs evenly in all quadrants

B. Creeping angle closure Define the relevant aspects of the epidemiology and risk factors of this disease

1. Race
   a. Higher prevalence among Inuits and Asian ethnic groups (Chinese Asian, Southeast Asian, and Asian Indians)
   b. Chronic “Creeping” angle-closure is relatively uncommon in Caucasians
   c. Patients of African descent with angle closure tend to have creeping angle closure

2. Age - higher prevalence with increasing age

3. Sex - higher prevalence in women

4. Refraction - typically associated with hyperopia

5. Inheritance- higher prevalence with a family history of angle closure

C. List pertinent elements of the history

1. No prior symptoms (most common)

2. Prior history of acute angle-closure glaucoma

3. Prior history of subacute angle-closure glaucoma with characteristic findings of intermittent headache or brow ache associated with blurred vision and/or haloes around lights

D. Describe pertinent clinical features

1. Elevated intraocular pressure (IOP)

2. Persistent PAS on indentation (compression) gonioscopy

3. Clinical course resembles that of open angle glaucoma
   a. Modest, variable elevation of IOP
   b. Progressive cupping of the optic nerve head
   c. Glaucomatous visual field loss
   d. Lack of symptoms

4. Some eyes may eventually develop an acute attack of angle-closure glaucoma with pupillary block

5. There may be patchy, dispersed pigment granules collecting in the iridocorneal angle where the peripheral iris was in contact with the cornea, suggesting previous appositional closure

6. Glaukomflecken and/or sector iris atrophy may indicate previous attacks of angle-closure glaucoma

II. List the differential diagnosis
A. Neovascular glaucoma
B. Iridocorneal endothelial syndrome
C. Inflammatory glaucoma
D. Traumatic glaucoma
E. Phacomorphic glaucoma
F. Intermittent primary angle closure
G. Ciliary body swelling or cysts/masses
H. Aqueous misdirection
I. Posterior segment tumors
J. Scleral buckling procedures with secondary angle closure
K. Ciliochoroidal effusion—nanophthalmos, panretinal photocoagulation, central retinal vein occlusion, sulfa-based medications (e.g., topiramate)
L. Retinopathy of prematurity with shallow anterior chamber
M. Plateau iris
N. Congenital glaucoma (e.g., Axenfeld-Rieger syndrome)

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

A. Describe medical therapy options
   1. Parasympathomimetic agents—however, higher strength concentrations and prolonged use may exacerbate the condition by anterior displacement of the lens-iris diaphragm, which could aggravate pupillary block
   2. Beta-adrenergic antagonists
   3. Alpha-adrenergic agonists
   4. Carbonic anhydride inhibitors
   5. Prostaglandin analogues

B. Describe the laser and surgical therapy options
   1. Laser peripheral iridotomy—this may eliminate any element of pupillary block but may not lower IOP if PAS are widely present
   2. Surgical iridectomy if laser surgery is not possible
   3. Peripheral laser iridoplasty
   4. Surgical filtering procedures
   5. Goniosynechialysis

C. Cataract extraction with or without goniosynechialysis may be indicated in selected cases and possibly could be more effective than laser or surgical peripheral iridotomy in preventing further extension of PAS

IV. List the complications of treatment, their prevention and management (See Laser iridotomy for angle closure, Incisional surgery for angle closure)

A. Inflammation may follow all laser surgeries and surgical procedures
   1. Treat with topical corticosteroids following iridoplasty and iridotomy

B. PAS commonly reform following goniosynechialysis
   1. Topical corticosteroids may be helpful

V. Describe disease-related complications
A. Endstage glaucoma with severe visual field loss and eventual loss of central vision
B. IOP generally rises slowly or is intermittently elevated so that pain or visual symptoms are unusual complications
C. Risk for aqueous misdirection after intraocular surgeries

VI. Describe appropriate patient instructions

A. Inform the patient about the need for regular, lifelong follow up
   1. In particular, inform certain groups (Inuits and Asians) of their particularly high risk for chronic problems
   2. Inform patients that they may not have any obvious symptoms of the disease
B. Patients need to realize that laser iridotomy will probably not cure disease and that additional treatment will almost certainly be warranted
C. Appropriate preoperative instructions before laser surgery or filtering procedures should be given

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Plateau iris

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
   1. A specific anatomic configuration of the angle structures with anterior insertion of ciliary processes that results in crowding of the peripheral anterior chamber angle
   2. Two types:
      a. Plateau iris configuration
         i. Normal central anterior chamber depth with narrow or closed angle on gonioscopy
      b. Plateau iris syndrome (less common than configuration)
         i. Similar anatomic findings as above in 2a with elevated IOP and angle closure occurs with pupillary dilation.
         ii. Pupillary block does not play a role in the angle closure as it is not prevented by a patent iridotomy (PI)
   3. These are underdiagnosed entities that overlap clinically with primary angle closure

B. Define the relevant aspects of epidemiology
   1. More commonly seen in younger patients with myopia presenting with angle closure glaucoma
   2. More commonly seen in Asian eyes

C. List the pertinent elements of the history of plateau iris syndrome
   1. Eye pain or discomfort after pupillary dilation
   2. History of narrow angle and/or laser iridotomy

D. Describe pertinent clinical features of plateau iris syndrome
   1. Persistent narrow angle despite a patent iridotomy before indentation and after indentation
   2. Anteriorly positioned ciliary processes
   3. Shallow and narrow peripheral angle in setting of deep central anterior chamber (iris contour is flat centrally, not bombé)
   4. On indentation gonioscopy, "sine wave" or "double hump" configuration as iris angulates forward over region of anteriorly-displaced ciliary processes
   5. Following pupillary dilation, peripheral iris bunches up and obstructs trabecular meshwork (TM), thereby causing increased intraocular pressure (IOP)

E. Describe appropriate testing and evaluation for establishing the diagnosis
   1. Suspicion on gonioscopy
      a. Flat iris with narrow or non-visible recess
   2. Ultrasound biomicroscopy
      a. Demonstrates "draping" of peripheral iris over anteriorly-rotated ciliary processes

II. Define the risk factors

A. Family history of angle-closure glaucoma
B. Female
C. Younger age (30s to 50s)
D. Less hyperopia than patients with traditional pupillary block-associated primary angle-closure glaucoma
E. Asian ancestry
III. List the differential diagnosis
   A. Pupillary-block associated angle-closure glaucoma
   B. Aqueous misdirection or ciliary block glaucoma
   C. Iris cyst or mass (would most likely be localized)
   D. Chronic angle-closure glaucoma
   E. Angle closure from ciliary body congestion/uveal effusion, which can be seen after panretinal photocoagulation, scleral buckles, CRVO, among others

IV. Describe patient management in terms of treatment and follow-up
   A. Entails close monitoring with periodic indentation gonioscopy (after LPI and/or cataract extraction)
   B. Medical and/or surgical therapy may be necessary, depending on the status of the angle and the optic nerve. Many cases are not recognized until a patent iridotomy fails to deepen the angle
   C. Describe medical therapy options
      1. Low-dose pilocarpine in order to pull iris out of angle
      2. In cases where pupil dilation is necessary, use minimal concentration/dosage of dilating drops
      3. Consider using pilocarpine to constrict the pupil after pupil dilation
   D. Describe laser and surgical therapy options
      1. Perform a laser PI first to remove any pupillary block component. However despite a PI, eyes with plateau iris syndrome will still be predisposed to develop chronic angle closure as a result of peripheral iris anatomy and may even develop acute angle closure with dilation
      2. Laser iridoplasty can be considered to flatten and thin the peripheral iris after the PI so that with pupillary dilation the angle does not close
      3. Glaucoma filtering surgery
      4. Cataract extraction if cataract is present
      5. However despite removal of pupillary block by cataract removal, eyes with plateau iris configuration will still be predisposed to develop chronic angle closure

V. List the complications of treatment, their prevention and management
   A. Complications of pilocarpine
      1. Chronic miosis (with or without posterior synechiae)
      2. Cataract
      3. Retinal detachment (rare)
   B. Complications of laser iridotomy and/or iridoplasty
      1. Post-laser surgery IOP elevation (acute or chronic)
      2. Post-laser surgery inflammation and posterior synechiae
      3. Cataract
      4. Peripheral anterior synechiae (especially with iridoplasty)
      5. Distortion of pupil
      6. Laser iridoplasty is not a cure
         a. Creeping angle closure may still occur
         b. Periodic gonioscopy is recommended
         c. Repeat laser iridoplasty may be necessary
VI. Describe disease-related complications

A. Chronic angle-closure glaucoma (progressive closure of angle due to peripheral anterior synechiae)

B. Acute angle-closure glaucoma triggered by pupil dilation with cycloplegics or adrenergic agents
   1. This may occur despite a patent PI
   2. Post-dilation IOP measurement is indicated to identify the occurrence of this problem

VII. Describe appropriate patient instructions

A. Need for frequent follow-up exams (with indentation gonioscopy)

B. Return immediately if any symptoms of angle-closure glaucoma

C. Probable medical and/or laser therapy if IOP elevation detected

D. Probably medical and/or laser and/or surgical therapy if progressive angle closure noted

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


6. AAO, Focal Points: Angle Closure Glaucoma Update, Module #6, 2009
I. Describe the approach to establishing the diagnosis

A. Describe the etiology and epidemiology of this disease

1. Pulling mechanism
   a. Secondary angle closure due to iris membranes:
      i. Neovascularization of the iris
      ii. Epithelial downgrowth or fibrous ingrowth
      iii. Descemetization of the anterior chamber angle as in iridocorneal endothelial syndrome (ICE)
      iv. Contracture of inflammatory precipitates

2. Pushing mechanism
   a. Secondary pupillary block from
      i. Uveitis with posterior synechiae/bombe
      ii. Lens related disorders
         i) Phacomorphic glaucoma
         ii) Subluxed crystalline lens, spherophakia,
         iii) Malpositioned intraocular lenses (IOLs)
   b. Retina-related conditions following
      i. Central retinal vein occlusion with ciliary body edema
      ii. Panretinal photoagulation with ciliary body edema
      iii. Placement of a scleral buckle with ciliary body edema
      iv. Intraocular gas with forward shift of lens-iris diaphragm
   c. Specific entities
      i. Chronic serous choroidal detachment (effusion)
         i) Degree of anterior chamber shallowing influenced by the size of effusion
      ii. Hemorrhagic choroidal detachment (suprachoroidal hemorrhage)
         i) Associated with most intraocular surgical procedures (often with filtration surgery but also with other surgeries, especially vitrectomy, and surgical procedures after ocular trauma)
         ii) Most often delayed but may occur during intraocular surgery
         iii) Associated with Valsalva maneuver or change in intraocular pressure (IOP)
         iv) Usually accompanied by pain
      iii. Intraocular tumors
         i) The mechanism of secondary glaucoma is variable including forward pushing of the lens-iris diaphragm, angle infiltration by tumor causing angle closure
         ii) Tumor related secondary angle-closure glaucoma most often seen with ciliary body melanomas (ring melanoma)
         iii) Cysts of the iris pigment epithelium or ciliary epithelium (iridociliary cysts)
         iv) Other tumors of the ciliary body and peripheral retina
      iv. Retinopathy of prematurity
         i) Retrolenticular tissue contracture may result in early or later-onset angle closure
Medication-induced: sulfonamide derivatives such as topiramate. These drugs cause secondary angle-closure glaucoma by inducing idiopathic ciliary body swelling and effusion in the supraciliary space.

Other

i) Synchial angle closure following anterior segment surgery: Anterior chamber shallowing associated with either a wound leak (that may occur following corneal procedures or cataract surgery) or from over filtration (as in glaucoma filtering) may result in anterior displacement of lens-iris diaphragm, formation of peripheral anterior synechiae and result in secondary angle closure.

B. List the pertinent elements of the history

1. Ocular history: central retinal vein occlusion, uveitis, previous lasers or incisional surgery
2. Medical history: diabetes, carotid occlusive disease, trauma
3. Medication history: Topiramate, CAIs, Sulfa medications
4. Onset of symptoms
   a. Asymptomatic vs. symptomatic
   b. Acute vs. chronic
      i. History of sudden onset of pain, often with Valsalva associated with suprachoroidal hemorrhage

C. Describe the pertinent clinical features

1. Corneal edema in ICE syndrome or posterior corneal membranes in epithelial downgrowth
2. Shallow or flat anterior chamber (AC) both central and peripheral
3. Anterior displacement of lens, IOL or vitreous face
4. AC inflammation may be present
5. Iris may show presence of membrane, neovascularization or ectropion uveae
6. Angle may show varying degree of peripheral anterior synchial closure or neovascularization
7. Posterior synchiae causing a secluded pupil
8. Ectopic lens or malpositioned posterior chamber IOL
9. Serous ciliochoroidal detachment, ciliary body swelling or supraciliary effusion or hemorrhagic choroidal detachment
10. Peripheral iridociliary cysts or ciliary body tumor on gonioscopy or 3 mirror examination

D. Describe appropriate testing and evaluation for establishing the diagnosis

1. Anterior segment exam with gonioscopy
2. Indirect ophthalmoscopy
3. Ultrasound
   a. B-scan to determine if choroidal detachment or mass
   b. B-scan can determine if hemorrhagic or serous ciliochoroidal detachment
   c. Ultrasound biomicroscopy to assess for anterior rotation of ciliary body, ciliary body tumor or iridociliary cysts
   d. Carotid Doppler to identify carotid artery stenosis
4. Transillumination
   a. Hemorrhagic choroidal detachment or mass will not transilluminate
   b. Help identify ciliary body tumor
5. Fine-needle aspiration biopsy
6. Specular microscopy to diagnose corneal involvement in ICE syndrome
II. Define the risk factors

A. Risk factors for suprachoroidal hemorrhage
   1. Markedly elevated preoperative IOP
   2. Filtration Surgery with antimetabolites
   3. High myopia
   4. Systemic hypertension
   5. Advanced age
   6. Previous hemorrhage in other eye

B. Risk factors for neovascularization
   1. Diabetes
   2. Carotid occlusive disease
   3. Retinal vascular occlusion

C. Risk for lens related secondary angle closure
   1. Pseudoexfoliation
   2. Trauma
   3. Genetic conditions (e.g. Marfan Syndrome)

III. List the differential diagnosis

A. Aqueous misdirection (malignant) glaucoma

B. Primary angle closure from pupillary block

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

A. Describe medical therapy options
   1. Treatment of elevated IOP
      a. Alpha 2-adrenergic agonists
      b. Beta-adrenergic antagonists
      c. Carbonic anhydrase inhibitors (topical and oral)
      d. Prostaglandin analogs
      e. Intravenous mannitol
      f. Cessation of sulfonamide derivative if identified as the cause of secondary angle-closure glaucoma
      g. Cycloplegics to break pupillary block following uveitis, following retinal procedures, or in ciliochoroidal effusions to deepen anterior chamber and prevent synechial angle closure.

B. Describe surgical therapy options
   1. Glaucoma drainage devices may be needed to control intraocular pressure in many forms of secondary angle-closure glaucoma
   2. Laser peripheral iridotomy to treat pupillary block for some forms of secondary angle-closure glaucoma
   3. Pan retinal photocoagulation and anti-VEGF therapy in neovascular glaucoma
   4. Partial removal of intraocular gas if identified as the cause.
   5. Injection of air or ophthalmic viscosurgical device (OVD) (viscoelastic) (unlikely to be successful in case of persistent serous or hemorrhagic ciliochoroidal detachment)
6. Drainage of suprachoroidal fluid (serous or hemorrhagic) and reformation of anterior chamber
   a. If large choroidal effusions persist or are appositional ("kissing"), drainage may be necessary
   b. Flat anterior chamber with pseudophakic or phakic touch to the cornea requires prompt correction
to prevent corneal decompensation
   c. Appositional angle closure with synechial formation especially in an inflamed eye
7. Resuturing of wound and possible revision of filter
   a. Excessive filtration without wound leak may require resuturing of scleral flap
8. Goniosynechialysis
   a. May be useful in cases of secondary angle closure where IOP elevation is seen following relatively
recent post-surgical synechial angle closure
9. Repositioning of IOL or cataract surgery to treat lens or IOL related causes
10. Argon/diode laser iridoplasty for iridociliary cysts

V. List the complications of treatment, their prevention and management
   A. Intensive cycloplegic therapy
      1. Dilated pupil
      2. Blurred vision
      3. Systemic toxicity (tachycardia, fever, etc.)
   B. Complications of glaucoma medical therapy (See Beta-adrenergic antagonists, Alpha adrenergic agonists,
      Carbonic anhydrase inhibitors)
   C. Complications of surgical therapy

VI. Describe disease-related complications
   A. Corneal decompensation
   B. Cataract
   C. PAS
   D. Glaucoma surgery failure
   E. Chronic IOP elevation with subsequent required medical or surgical therapy
   F. Retinal detachment (most often associated with appositional ("kissing") hemorrhagic choroidal
detachments)
   G. Metastatic disease associated with intraocular tumors

VII. Describe appropriate patient instructions
   A. In postoperative period avoid Valsalva maneuver, use care to avoid trauma, use postoperative medications
   B. Close postoperative follow-up required
   C. Ciliary body tumor treatment management discussed elsewhere but may include radiotherapy, resection, or
enucleation

Additional Resources
1. AAO, Basic and Clinical Sciences Course. Section 10: Glaucoma, 2015-2016.
Neovascular glaucoma

I. Describe approach to establishing the diagnosis

A. List the pertinent elements of the history
   1. Pain, photophobia (usual)
   2. Markedly reduced vision (usual)
   3. Diabetes
   4. Hypertension, arteriosclerosis, carotid artery disease
   5. Previous central retinal vein occlusion (CRVO)

B. Define the relevant aspects of the epidemiology of the disease
   1. Diabetes mellitus (proliferative retinopathy)
      a. Long duration, poor glycemic control
      b. Post cataract extraction, vitrectomy, laser capsulotomy
         i. Particularly with open posterior capsule
   2. CRVO
   3. Carotid occlusive disease/ocular ischemic syndrome
      a. May have normal or low intraocular pressure (IOP)
   4. Central retinal artery occlusion
   5. Intraocular tumors
   6. Other causes (e.g., sickle cell retinopathy)

C. Describe pertinent clinical features
   1. Early
      a. Tufts of new vessels at pupillary margin (may be difficult to see in dark irides) and/or
      b. Fine vessels crossing scleral spur (onto trabecular meshwork (TM) on gonioscopy
   2. Late
      a. Very high IOP (except from carotid obstruction)
      b. Conjunctival injection
      c. Corneal edema
      d. Florid neovascularization of the iris (NVI) with ectropion uveae (except when angle closure total where NVI may be minimal)
      e. Iris ectropion
      f. Fibrovascular membrane (not clinically visible) over iris and angle structures
      g. Variable synechial angle-closure
      h. With total angle closure and with pigmented Schwalbe's line, on gonioscopy can be mistaken for chronic open angle glaucoma

D. Describe appropriate testing and evaluation for establishing the diagnosis
   1. Slit-lamp examination
   2. Gonioscopy
   3. Dilated fundus exam
   4. Fluorescein fundus angiogram
II. Define the risk factors
   A. Retinal hypoxia
      1. Vascular endothelial growth factor (VEGF)

III. List the differential diagnosis
   A. Early (iris or angle vessels)
      1. Fuchs heterochromic iridocyclitis
      2. Uveitis
   B. Late (elevated IOP, cloudy cornea)
      1. Acute angle-closure glaucoma
      2. Intraocular tumors
      3. Chronic retinal detachment
      4. Ghost cell glaucoma
      5. Iridocorneal endothelial syndrome (ICE)

IV. Describe the patient management in terms of treatment and follow-up
   A. Early
      1. Medical therapy of elevated IOP
      2. Topical corticosteroids and atropine
      3. Intraocular anti-VEGF agents
      4. Panretinal photocoagulation (PRP)
      5. Panretinal cryotherapy (when poor visualization does not permit PRP)
   B. Late
      1. Medical therapy of elevated IOP
      2. Topical corticosteroids and atropine
      3. Intraocular anti-VEGF agents
      4. PRP
      5. Surgery
         a. Filtration surgery with adjunctive antimetabolite; usually after pre-treatment with anti-VEGF
         b. Aqueous shunt
      6. Cyclodestruction
         a. Laser surgery
         b. Cryotherapy
   C. Blind, painful eye
      1. Topical steroids and cycloplegic
      2. Cyclodestruction
         a. Laser surgery
      3. Retrobulbar injections
V. List the complications of treatment

A. Complications of intravitreal injection including retinal detachment and endophthalmitis
B. Complications of PRP including loss of visual field
C. Complications of surgery (See Aqueous shunt surgery)
D. Complications of cyclodestruction including hypotony and vision loss

VI. Describe disease-related complications

A. Absolute glaucoma with blindness
B. Intractable pain
C. Hyphema

VII. Describe appropriate patient instructions

A. Medication and surgical discussion
B. Consider referral for anti-VEGF treatment, PRP, surgical intervention, and/or cyclodestruction
C. Referral to primary care physician for care related to etiology

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


Iridocorneal endothelial (ICE) syndrome (a secondary angle-closure glaucoma)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
   1. Idiopathic corneal endothelial proliferation
   2. Epithelial-like characteristics of endothelial cells
   3. Endothelial cells extend from the peripheral cornea over the trabecular meshwork and onto the iris
      a. Descemet-like membrane causes angle closure even without peripheral anterior synechiae (PAS)
      b. Contraction of the membrane causes PAS and more typical angle closure
   4. Speculated to be viral in origin
      a. Herpes simplex virus
      b. Epstein Barr virus

B. Define the relevant aspects of epidemiology of the disease
   1. More common in women
   2. Presents between ages 20-50 years

C. List the pertinent elements of the history
   1. Almost always unilateral
   2. Acquired abnormality in shape or position of pupil
   3. Iris color abnormality
   4. Decreased vision, halo and glare from corneal edema
   5. Sometimes complaints of pain from corneal edema

D. Describe pertinent clinical features
   1. All have varying amounts of corneal changes and iris abnormalities but features common to all include
      a. Hammered silver or beaten bronze endothelial appearance much finer than seen in Fuchs corneal endothelial dystrophy
      b. Iris changes
      c. Very anteriorly inserted peripheral anterior synechiae (anterior to Schwalbe's line)
   2. Classically unilateral (rarely bilateral), but fellow eye has been reported to have abnormal corneal endothelium. Specular microscopy can confirm the diagnosis. The test shows asymmetric loss of endothelial cells and atypical endothelial cell morphology
   3. Glaucoma reported in 50% to 75% of cases
   4. Three forms classically comprise the spectrum of disease, though it is often difficult to clearly distinguish them and manifestations may change as the disease progresses
      a. Essential iris atrophy
         i. More iris changes than corneal change
         ii. Iris atrophy
         iii. Iris heterochromia
         iv. Pupil distortion and corectopia
v. Ectropion uveae
vi. Polycoria
   i) Stretch holes occur in the quadrant opposite pupil distortion
   ii) Melt holes associated with iris ischemia
vii. Broad PAS
b. Cogan-Reese syndrome (iris nevus syndrome)
i. Less severe iris stromal abnormalities
ii. Iris pseudo nodules visible on the surface caused by endothelial membrane contraction pinching the iris surface
iii. Angle-closure glaucoma
c. Chandler syndrome
   i. Iris changes are not common in early disease, but the corneal changes and PAS can be seen
   ii. Often corneal edema even with normal intraocular pressure (IOP)
   iii. Classic hammered silver or beaten bronze appearance of endothelium, but may be patchy in distribution requiring a careful search

II. List the differential diagnosis

A. Corneal manifestations
   1. Posterior Polymorphous Dystrophy (bilateral, autosomal dominant)
   2. Fuchs Dystrophy
B. Pupil manifestations
   1. Axenfeld-Rieger Syndrome (congenital, bilateral, autosomal dominant)
C. Iris nodules
   1. Lisch Nodules of Neurofibromatosis
   2. Inflammatory Nodules
   3. Iris nevi and melanoma
D. Secondary angle closure glaucomas with anterior pulling mechanism
   1. Neovascular glaucoma
   2. Epithelial and fibrous downgrowth

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

A. Describe medical therapy options
   1. Aqueous suppressants
   2. Pilocarpine ineffective
   3. Prostaglandin analogues uncertain
   4. Hypertonic saline drops and ointment for corneal edema
B. Describe surgical therapy options
   1. Laser trabeculoplasty ineffective
   2. Trabeculectomy with antimetabolite
      a. Often requires repeat surgery due to failure from endothelialization of scleral fistula
      b. Endothelialization occasionally can be reopened by neodymium yttrium-aluminum-garnet laser (Nd:
IV. List complications of treatment, their prevention and management

A. Complications
   1. Glaucoma medication allergy or side effect
   2. Glaucoma surgery failure to control IOP
   3. Ultimate need for cataract or corneal surgery as a complication of surgery to control IOP

B. Prevention of complications
   1. Careful medical history to reduce drop complications
   2. Antimetabolites with trabeculectomy
   3. Consider initial tube shunt surgery

C. Management of complications
   1. Change medical therapy
   2. Repeat surgery for initial failure to control IOP

V. Describe disease-related complications

A. Corneal decompensation is frequently seen requiring penetrating keratoplasty or Descemet's Stripping Automated Endothelial Keratoplasty

B. Cosmetic issues with iris abnormalities

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


Inflammatory glaucoma (secondary open-angle and angle-closure glaucoma)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Open-angle glaucoma
   a. Intraocular inflammation in the anterior segment (anterior uveitis) leads to trabecular damage and obstruction of trabecular meshwork outflow by
      i. Edema of trabecular meshwork
      ii. Trabecular endothelial cell dysfunction
      iii. Fibrin and elevated aqueous protein from breakdown of the blood-aqueous barrier
      iv. Inflammatory cells and/or inflammatory nodules
   b. Corticosteroids used to treat inflammation may cause secondary intraocular pressure elevation and secondary open angle glaucoma.

2. Angle-closure glaucoma
   a. Intraocular inflammation involving the anterior segment can result in formation of extensive posterior synechiae and subsequent pupillary block and iris bombé or secluded pupil
   b. Inflammatory membranes can form in the angle that result in formation of peripheral anterior synechia (PAS)
      i. Due to organization of inflammatory debris in the angle or large keratic precipitates (KP) that bridge the angle
      ii. PAS are typically located in the inferior angle
      iii. PAS tend to be non-uniform in shape and height
   c. Iris/angle neovascularization ("inflammatory" not ischemic neovascularization) with subsequent contraction of vascular membrane in angle causing angle closure
   d. Uveal effusion with anterior rotation of ciliary body and narrowing of angle
   e. Posterior uveitis with exudative detachment causing anterior displacement of lens-iris diaphragm and consequent diffuse shallowing of anterior chamber

3. Specific entities
   a. Glaucomatocyclitic crisis (Posner-Schlossman syndrome)
      i. Open-angle glaucoma
         ii. Characterized by recurrent attacks of mild anterior uveitis with marked elevations of intraocular pressure (IOP), often up to 40-60 mm Hg
            i) Small, discrete, round KP may be present on corneal endothelium
            ii) Precipitates may be present on trabecular meshwork
               (i) Glaucoma may be due to inflammatory changes in the trabecular meshwork (trabeculitis)
               (ii) Microcystic corneal edema may be present
            iii. Usually involves only one eye
            iv. IOP and outflow facility usually return to normal between attacks
               i) May have chronic IOP elevation and glaucomatous disc and field damage in severe
Possible etiologic role of herpes simplex virus

Possible etiologic role of endogenous prostaglandins

b. Fuchs heterochromic iridocyclitis
   i. Open-angle glaucoma
      i) Occurs as a result of chronic inflammation
      ii) Anterior and posterior synechiae do not occur
   ii. Usually involves only one eye—the affected eye is usually the lighter eye, except in light irides, where the affected eye can appear darker
   iii. Characterized by classic triad of iris heterochromia, iritis, and cataract; other findings include angle and iris neovascularization, loss of normal iris architecture, low grade inflammation, KP, occluded/secluded pupils and posterior subcapsular cataract
   iv. Often asymptomatic; eye not red
   v. Etiology unknown
      i) Sympathetic denervation has been demonstrated in pharmacologic studies
      ii) Sympathetic disturbance leads to changes in blood vessels and atrophy of iris stroma
      iii) Possible etiologic role of rubella virus infection

B. Define the relative aspects of epidemiology of this disease

1. May be associated with systemic inflammatory or infectious disorders
   a. Sarcoidosis
   b. Ankylosing spondylitis
   c. Inflammatory bowel disease
   d. Reactive arthritis (formerly Reiter syndrome)
   e. Juvenile idiopathic arthritis (formerly known as juvenile rheumatoid arthritis)
   f. Behçet disease
   g. Herpes simplex virus keratouveitis
   h. Herpes zoster
   i. Toxoplasmosis
   j. Pars planitis
   k. Syphilis
   l. Lyme disease

2. Glaucomatocyclitic crisis (Posner-Schlossman syndrome)
   a. Usually occurs in young to middle-aged adults
   b. Many have associated systemic disorders
      i. Allergic conditions
      ii. Gastrointestinal disorders, especially peptic ulcer disease

3. Fuchs heterochromic iridocyclitis
   a. Usually occurs in middle-aged adults
   b. Men and women equally affected
   c. Increased risk for glaucoma
   d. Not considered hereditary but has been observed in families and identical twins

C. List the pertinent elements of the history
1. Pain
2. Photophobia
3. Decreased vision
4. Red eye

D. **Describe the pertinent clinical features**

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

2. **Glaucomatocyclitic crisis (Posner-Schlossman syndrome)**
   a. Low-grade AC reaction
   b. Marked IOP elevation
   c. Few, fine non-pigmented KP

3. **Fuchs heterochromic iridocyclitis**
   a. Heterochromia (usually loss of pigment is in the affected eye)
   b. Loss of normal iris architecture
   c. Low-grade AC reaction
   d. Small, circumscribed stellate KP
      i. Usually on central and inferior corneal endothelial surface, may also be diffuse
   e. Fine blood vessels in angle (not iris neovascularization)
      i. Not associated with fibrous membrane or peripheral anterior synechiae formation.
      ii. May cause anterior chamber hemorrhage either spontaneously, with gonioscopy, or with minimal trauma
   f. Posterior subcapsular cataract
   g. Anterior vitreous opacities

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

1. **History and physical - uveitis work up**
   a. History
      i. Ocular history
      ii. Medical history
      iii. Social history
      iv. Family history
   b. Review of systems - comprehensive
   c. Comprehensive ophthalmologic examination
      i. Lids (e.g. cutaneous nodules)
ii. Conjunctiva (e.g. nodules/granuloma)

iii. Anterior segment slit lamp examination (e.g. dendritic KPs [HSV, VZV, CMV, Fuchs Heterochromic iridocyclitis])

iv. Gonioscopy

v. Dilated fundus examination - toxoplasmosis and necrotizing herpetic retinitis are often associated with high IOP

2. Hematologic/serologic (always remember the big three: Sarcoid, Syphilis, TB)
   a. Angiotensin converting enzyme
   b. Purified protein derivative (PPD) w/ anergy panel
   c. Fluorescent treponemal antibody absorption
   d. Rapid plasma reagin
   e. HLA-B27
   f. Antinuclear antibodies (ANA) and Rheumatoid factor (RhF) - only order in a child with anterior uveitis when JRIA is being ruled out (note, ANA and RhF are not otherwise valuable tests in uveitis)

3. Radiologic
   a. Chest x-ray/spiral chest CT (sarcoidosis and TB)
   b. Sacro-iliac joint films (not lumbosacral joint films) for ankylosing spondylitis

4. Ultrasound
   a. B-scan to assess exudative retinal detachment, ciliary body effusion
   b. Ultrasound biomicroscopy to assess ciliary body and angle

5. Medical evaluation by primary care physician or appropriate specialist to look for systemic causes of uveitis

6. Intraocular fluid analysis
   a. Aqueous PCR for HSV, VZV, CMV, or Toxoplasmosis - if one of these agents is suspected on examination
   b. Distinguishing glaucomatocyclitic crisis from CMV is important because specific antiviral therapy for CMV is available

II. Define the risk factors
   A. Chronic, relapsing inflammation
   B. Non-compliance with prescribed therapy
   C. Chronic corticosteroid use (topical, depot, systemic) for treatment of inflammation. (See Corticosteroid-induced glaucoma (steroid glaucoma))

III. List the differential diagnosis
   A. Acute/subacute primary angle-closure glaucoma
   B. Lens-induced inflammation with secondary IOP elevation
   C. Tumor-induced inflammation
   D. Carotid insufficiency

IV. Describe patient management in terms of treatment and follow-up
   A. Describe medical therapy options
      1. Treat inflammation
         a. Topical corticosteroids
b. Topical nonsteroidal anti-inflammatory drugs (NSAIDs)
c. Cycloplegia
d. Systemic corticosteroids
e. Systemic NSAIDs
f. Systemic immunomodulatory agents (methotrexate, chlorambucil, azathioprine)

2. Treat elevated IOP
   a. Beta-adrenergic antagonists
   b. Alpha-adrenergic agonists
c. Oral and systemic carbonic anhydrase inhibitors
d. Hyperosmotic agent

3. Contraindicated medications for IOP control (may exacerbate inflammation)
   a. Parasympathomimetic agents
   b. Prostaglandins can be successfully used in uveitic glaucoma. If it is felt that inflammation is being
      exacerbated by the prostaglandin they can be stopped.

4. Special considerations
   a. Glaucomatocyclitic crisis
      i. Excellent response to topical corticosteroids during an acute attack.
      ii. No evidence suggesting the benefits of a long-term steroid use to prevent an attack
   b. Fuchs heterochromic iridocyclitis
      i. Corticosteroids generally not effective
   c. Aqueous suppressants and alpha-adrenergic agonists agents of choice

B. Describe surgical therapy options
   1. Laser iridotomy or surgical iridectomy for angle-closure glaucoma/iris bombé
      a. More than one iridotomy may be required
         i. Inflammation may cause closure of single iridotomy
         ii. Iris bombé may be sectoral
   2. Glaucoma filtering surgery with mitomycin
   3. Glaucoma drainage tube
   4. Cyclophotocoagulation
   5. Laser trabeculoplasty is usually not successful and may be contraindicated
   6. Fluocinolone implant to control inflammation

V. List the complications of treatment, their prevention and management
   A. Corticosteroid-induced IOP elevation (See Corticosteroid-induced glaucoma (steroid glaucoma))
      1. Use minimal corticosteroid dose required to control inflammation
      2. Use corticosteroid with less of an IOP effect (rimexolone, loteprednol etabonate and fluorometholone)
      3. Use systemic corticosteroids to minimize or eliminate topical corticosteroid dose
      4. Use topical/systemic NSAID
   B. Use systemic immunosuppression to minimize or eliminate systemic or topical corticosteroid use
   C. Corticosteroid-induced cataract
      1. Try to minimize dose and duration of corticosteroid use
D. Systemic corticosteroid side effects
1. Weight gain, fluid retention, gastrointestinal symptoms, blood glucose elevation, musculoskeletal effects (i.e. avascular necrosis)
   a. Minimize dose and duration
   b. Follow patient in conjunction with primary care physician

E. Systemic immunosuppression
1. Bone marrow suppression
   a. Monitor hematologic status
   b. Follow in conjunction with primary care physician

F. Laser iridotomy
1. Thickened, inflamed iris may be difficult to penetrate with laser surgery and result in bleeding, exacerbation of anterior chamber inflammation
2. Laser iridotomy may close due to continued inflammation
3. More than one peripheral iridotomy necessary if sectoral iris bombé present
4. Surgical iridectomy/sector iridectomy may be needed to maintain patency of opening

G. Complications of glaucoma medical therapy

H. Complications of glaucoma surgery

VI. Describe disease-related complications
A. Decreased vision
1. Visual field loss associated with optic disc damage
2. Cataract
3. Miotic pupil due to posterior synechiae, occlusion/seclusion by inflammatory membrane
4. Cystoid macular edema

B. Chronic IOP elevation requiring medical or surgical therapy

VII. Describe appropriate patient instructions
A. Maintain anti-inflammatory therapy at prescribed dose to control uveitis
B. Taper anti-inflammatory therapy as instructed to minimize treatment related complications
C. Follow recommended IOP-lowering regimen (medical or surgical) to prevent permanent vision loss
D. Continued management and follow-up essential

Additional Resources
1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
3. AAO, Glaucoma Medical Therapy: Principles and Management, 1999; 204-205.
Aqueous misdirection (malignant glaucoma, ciliary block glaucoma)

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of aqueous misdirection

1. Usually follows intraocular surgery or laser procedure in patients with a history of angle closure or peripheral anterior synchiae

2. There is anterior rotation of the ciliary body postoperatively (rarely this rotation can occur spontaneously)

3. Posterior misdirection of aqueous humor into the vitreous cavity causes shallowing of the peripheral and central anterior chamber with elevation of intraocular pressure (IOP)
   a. Classically, thought to result from anterior rotation of the ciliary body and posterior misdirection of the aqueous, in association with a relative block to aqueous movement at the level of the ciliary processes, lens equator and vitreous face
   b. Recently it is proposed that primary angle closure and aqueous misdirection may result from the simultaneous presence of several factors, including a small eye, a propensity for choroidal expansion, and reduced vitreous fluid conductivity

4. It can be difficult to diagnose misdirection in its earlier stages before there is a rise in IOP
   a. One must have a high index of suspicion in post-trabeculectomy cases where a uniformly shallow anterior chamber is observed in the context of normal or elevated IOP
   b. IOP elevation is variable, especially early in the clinical course
      i. Clinically, misdirection can be difficult to differentiate from anterior [annular] choroidal detachment or hemorrhage - anterior segment ultrasound is useful to make this distinction

B. Define the relative epidemiologic aspects of this disease

1. Occurs following glaucoma surgery in < 5% of eyes (with preexisting angle closure),
2. Rarely occurs after lens extraction
3. May occur spontaneously in eyes with open angles and/or without history of surgery (rarely)
4. Fellow eye is at increased risk of developing aqueous misdirection
5. Reported with cyclocryotherapy and other laser procedures

C. List the pertinent elements of the history

1. Pain
2. Photophobia
3. Decreased vision
4. Red eye

D. Describe pertinent clinical features

1. Uniformly shallow or flat anterior chamber (both peripheral AND central flattening are seen)
2. Anterior displacement of lens, intraocular lens (IOL), or vitreous face
3. Anteriorly rotated ciliary processes
4. Normal or elevated IOP
5. Shallow anterior chamber in presence of patent iridotomy / iridectomy and absence of choroidal effusion or hemorrhage

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Ultrasound
   a. B-scan to rule out ciliary body effusion or suprachoroidal hemorrhage
Ultrasound biomicroscopy (UBM) to assess for anterior rotation of ciliary processes or supraciliary effusion

II. Define the risk factors
   A. Eye with acute or chronic angle closure
   B. Persistent shallowing of the anterior chamber in the perioperative period
   C. Small eye

III. List the differential diagnosis
   A. Acute suprachoroidal hemorrhage
   B. Pupillary block glaucoma
   C. Choroidal effusion, which can also shallow the anterior chamber, but IOP is low in this condition

IV. Describe patient management in terms of treatment and follow-up
   A. Describe medical therapy options
      1. A classic triad; intensive cycloplegics, aggressive aqueous suppression, and shrinking of the vitreous by hyperosmotic agents
      2. Treat aqueous misdirection
         a. Intensive cycloplegic therapy
      3. Treat elevated IOP
         a. Beta-adrenergic antagonists
         b. Alpha2-adrenergic agonists
         c. Carbonic anhydrase inhibitors (topical and/or oral)
         d. Hyperosmotic agents
         e. Prostaglandin analogues
      4. Contraindicated medications for IOP control
         a. Parasympathomimetic agents
            i. May exacerbate inflammation
            ii. May aggravate aqueous misdirection
      5. Medical therapy is successful in approximately 50% of cases
         a. Many days of therapy may be necessary to reverse the acute problem
         b. Indefinite therapy may be necessary to prevent recurrence
   B. Describe surgical therapy options (laser surgery and incisional)
      1. Nd:YAG laser surgery disruption of anterior vitreous face in aphakic or pseudophakic eyes
         a. Placement of hole in anterior hyaloid peripheral to the IOL may be more effective than placement in other locations
         b. If a PI is already present, lysis of the anterior hyaloid is possible through the PI
      2. Argon laser photocoagulation of the ciliary processes
      3. Pars plana vitrectomy (with or without glaucoma tube shunt) with emphasis on rupture of hyaloid face and creation of a unicameral eye with complete communication of posterior and anterior chambers via a patent iridectomy
      4. Lens extraction with posterior capsulectomy and concurrent anterior hyaloid vitrectomy
5. Diode laser cyclophotocoagulation
6. Zonulo-hyaloido-vitrectomy (anterior hyaloid vitrectomy performed via clear corneal incision through peripheral iridectomy in pseudophakic eyes)

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

A. Intensive cycloplegic therapy
   1. Dilated pupil
   2. Blurred vision
   3. Ocular discomfort
   4. Systemic toxicity (e.g., tachycardia, etc.)

B. Complications of glaucoma medical therapy (e.g., hyperosmotics)

C. Complications of glaucoma laser surgery and incisional surgery
   1. Choroidal detachment
   2. Retinal detachment
   3. Cataract
   4. Persistently elevated IOP
   5. Recurrence of malignant glaucoma
   6. Suprachoroidal hemorrhage
   7. Endophthalmitis

VI. Describe disease-related complications

A. Decreased vision
   1. Acute or chronic IOP elevation with subsequent optic disc damage and visual field loss
   2. Cataract
   3. Corneal decompensation (secondary to persistent shallowing of anterior chamber)

B. Chronic IOP elevation requiring medical or surgical therapy

C. May recur after pars plana vitrectomy especially in phakic eyes because of difficulty in removing anterior vitreous

VII. Describe appropriate patient instructions

A. May need to maintain long-term low dose of cycloplegic therapy to avoid recurrence (taper as possible)
B. Follow recommended IOP-lowering regimen (medical or surgical) to prevent permanent vision loss
C. Continued management and follow-up essential
D. The risk for aqueous misdirection in the fellow eye

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Primary congenital or infantile glaucoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
   1. Goniodysgenesis of the anterior chamber angle without other ocular or systemic abnormalities
   2. Pathophysiology theories
      a. A cellular or membranous abnormality in the trabecular meshwork (Barkan membrane)
      b. Possible developmental arrest of anterior chamber tissue in utero

B. Define the relevant aspects of epidemiology of the disease
   1. Presents within the first 3 years of life
      a. Two-thirds of cases are diagnosed by age 6 months
      b. 80% diagnosed within first year of life
   2. Approximately two-thirds of congenital glaucomas are primary
   3. Two-thirds involve males
   4. Approximately two-thirds of cases are bilateral
   5. Genetics - variable inheritance
      a. Most common inheritance pattern is autosomal recessive with complete or incomplete penetrance

C. List the pertinent elements of the history
   1. Classic triad
      a. Epiphora
      b. Photophobia
      c. Blepharospasm

D. Describe pertinent clinical features
   1. Elevated intraocular pressure (IOP)
   2. Corneal edema
   3. Haab's striae
   4. Reduced visual acuity
      a. Glaucomatous optic atrophy
      b. Amblyopia
         i. Multiple causes (i.e., corneal clouding, astigmatism, anisometropia)
         ii. A common cause of visual loss
   5. Lens dislocation
   6. Gonioscopy
      a. Deep anterior chamber
      b. High flat iris insertion
      c. Absence of angle recess
      d. Peripheral iris hypoplasia
      e. Tenting of the peripheral iris pigment epithelium
f. Thickened uveal trabecular meshwork

g. High insertion of the iris root that forms a scalloped line as a result of abnormal tissue with a shagreen glistening appearance

7. Glaucomatous optic nerve cupping—may be reversible if treated adequately and early
8. Increased corneal diameter and buphthalmos due to cornea stretching from elevated IOP before age of 3
9. Epiphora

II. Define the risk factors

A. Family history of primary congenital glaucoma
B. Consanguinity

III. List the differential diagnosis

A. Excessive tearing
   1. Nasolacrimal duct obstruction
   2. Corneal epithelial defect or abrasion
   3. Conjunctivitis

B. Corneal enlargement or apparent enlargement
   1. X-linked megalocornea
   2. High myopia
   3. Exophthalmos
   4. Shallow orbits

C. Corneal clouding
   1. Birth trauma
   2. Inflammatory corneal disease
   3. Congenital hereditary corneal dystrophies
   4. Corneal malformations (dermoid tumors, sclerocornea, Peter anomaly)
   5. Keratomalacia
   6. Metabolic disorders with associated corneal abnormalities (mucopolysaccharidoses, corneal lipidosis, cystinosis)
   7. Skin disorders affecting the cornea (congenital ichthyosis and congenital dyskeratosis)

D. Optic nerve abnormalities
   1. Optic nerve pit
   2. Optic nerve coloboma
   3. Optic nerve hypoplasia
   4. Physiologic cupping

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

A. Describe medical therapy options
   1. Limited long-term value
   2. Beta-adrenergic antagonists, prostaglandin analogues, or carbonic anhydrase inhibitors may be used to temporize and clear a cloudy cornea
3. Use nasolacrimal occlusion

**B. Describe the surgical therapy options**

1. Angle surgery
   a. Goniotomy (option for clear corneas only)
   b. Trabeculotomy (option for either clear or cloudy corneas)
   c. Best prognosis occurs when onset of symptoms occurs between ages 3-12 months, with 80% success of angle surgery
   d. When symptoms present at birth or after one year of age, the surgical prognosis is more guarded
2. Trabeculectomy with or without mitomycin-C or 5-fluorouracil- with limited success in children younger than 2 years.
3. Aqueous shunt surgery-useful for IOP control and avoiding bleb-related complications
4. Cycloablation- reserve for refractory cases

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

A. Medical therapy (See Beta-adrenergic antagonists) (See Prostaglandin analogues) (See Carbonic anhydrase inhibitors)
   1. Note: Brimonidine is contraindicated under the age of 3 years
   2. Be alert for apnea and hypotension with beta-adrenergic antagonists
   3. Assess for possible acidosis, hypokalemia and feeding problems with oral carbonic anhydrase inhibitors

B. Goniotomy
   1. Mild to moderate hyphema, common
   2. Total or "eight-ball" hyphema (rare)
   3. Iridodialysis
   4. Cycloidalysis
   5. Peripheral anterior synchia (PAS)
   6. Retinal detachment
   7. Cataract
   8. Corneal decompensation
   9. Endophthalmitis

C. Trabeculotomy
   1. Hyphema
   2. PAS
   3. Filtering blebs
   4. Choroidal detachment
   5. Cataract
   6. Subconjunctival iris prolapse
   7. Descemet membrane stripping
   8. Iridodialysis
   9. Creation of false passage into anterior chamber
   10. Creation of false passage into suprachoroidal space (producing cyclodialysis cleft)
   11. Endophthalmitis
   12. Zonular tears and vitreous loss
13. Incarceration of iris
14. Lens dislocation

D. Other surgical modalities (See Laser trabeculoplasty) (See Aqueous shunt surgery) (See Ciliary body ablation procedures)

VI. Describe disease-related outcomes

A. Blindness
B. Amblyopia
C. Strabismus
D. Dislocated lens
E. Corneal scarring
F. Cataract
G. Susceptibility of an eye with a thinned sclera to trauma
H. Recurrent glaucoma in the affected or unaffected eye many years later

VII. Describe appropriate patient instructions

A. Close and frequent follow-up examinations are required
B. Examinations under anesthesia may be required to monitor IOP
C. Correction of any refractive error is mandatory
D. Lifelong checkups will be required

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Juvenile open-angle glaucoma (JOAG)

I. Describe the approach to establishing the diagnosis

   A. Describe the etiology of the disease
      1. Unknown
      2. Myocilin mutation
      3. Mutations on other chromosomes have been identified in some pedigrees with JOAG

   B. Define the relevant aspects of epidemiology of the disease
      1. Defined as autosomal dominant open-angle glaucoma diagnosed between ages 3-30 years
      2. Associated with myocilin mutation in up to 33% of pedigrees
         a. Do not have to have mutations in the myocilin gene to have juvenile open-angle glaucoma (JOAG)
         b. Not all myocilin gene mutations are associated with JOAG
         c. Less than 5% of primary open-angle glaucoma has myocilin mutations

   C. List the pertinent elements of the history
      1. Young patient
      2. Family history is common
      3. Myopia
      4. Decreased peripheral vision

   D. Describe pertinent clinical features
      1. Often very elevated intraocular pressure (IOP) but can be normal in the unaffected eye in asymmetric cases
      2. Bland appearing angle with poorly defined angle structures or landmarks, often with peripheral iris atrophy and visible prominent vessels
      3. Characteristic glaucomatous optic nerve damage
         a. Deeper, steeper cup
         b. Concentric rim thinning
      4. Characteristic glaucomatous visual field damage
         a. More diffuse depression
         b. Symmetric superior and inferior field loss
      5. Normal appearing trabecular meshwork (TM)
         a. Ultrastructural analysis reveals excess extracellular basement-membrane-like material
         b. Similar to corticosteroid glaucoma (steroid-induced glaucoma)

II. Define the risk factors

   A. Elevated IOP
   B. Family history
   C. Myocilin mutation

III. List the differential diagnosis
A. Primary open-angle glaucoma
B. Secondary open-angle glaucoma
   1. Corticosteroid-induced glaucoma
   2. Pigment dispersion syndrome/glaucoma
   3. Angle-recession glaucoma
C. Ocular hypertension
D. Congenital glaucoma

IV. Describe the patient management in terms of treatment and follow-up
A. May need closer follow up than POAG because of larger IOP fluctuations
B. Describe medical therapy options
   1. Topical medications
      a. Carbonic anhydrase inhibitors
      b. Beta-adrenergic antagonists
      c. Prostaglandin analogues
      d. Brimonidine is contraindicated in children younger than age 2
   2. Oral medication: carbonic anhydrase inhibitors
C. Describe surgical therapy options
   1. Laser trabeculoplasty often ineffective
   2. Goniotomy, trabeculotomy and other angle procedures
   3. Trabeculectomy with or without antimetabolites
   4. Non-penetrating surgeries
   5. Tube shunts
   6. Cyclodestructive procedures

V. List the complications of treatment, their prevention and management
A. Complications
   1. Allergic reaction to medication
   2. Surgical complications (See Incisional filtering surgery for open angle glaucoma)
      a. Infection
      b. Hemorrhage
      c. Loss of IOP control
      d. Corneal injury
      e. Cataract formation
      f. Hypotony maculopathy
      g. High rate of failure of trabeculectomy even with antimetabolites in this young group, often requiring a glaucoma drainage device
B. Prevention
   1. Careful history looking for medication allergies
   2. To reduce the potential for blebitis or endophthalmitis, consider cautious use of antimetabolites for trabeculectomy. However, an antimetabolite will help prevent scarring and may help surgery survival.
VI. Describe disease-related complications

A. Progressive loss of vision

B. Blindness

VII. Describe appropriate patient instructions

A. Use medication as directed

B. Recognize and emphasize the implications of having a genetic disease (genetic counseling may be indicated)

C. Stress with parents and patient the importance and need for frequent, lifelong follow-up and compliance with treatment plan for long-term vision preservation

D. Suggest that family members such as siblings and children have a comprehensive eye exam

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


Axenfeld-Rieger Syndrome

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
   1. Abnormal development of structures that are neural crest in origin which includes structures in the anterior segment, facial bones and teeth

B. Define the relevant aspects of epidemiology of the disease
   1. Most cases are autosomal dominant
   2. Equally affects males and females
   3. 50% develop glaucoma with variable age of onset-usually not present at birth
   4. Family history of Axenfeld-Rieger syndrome

C. List the pertinent elements of the history
   1. Decreased vision
   2. Lacrimation and photophobia if congenital and associated with glaucoma
   3. Abnormal pupil
   4. Positive family history
   5. History of systemic abnormalities

D. Describe the pertinent clinical features
   1. Ocular - wide variation in phenotypic abnormalities
      a. Typically, bilateral signs
      b. Hypoplasia of the iris
      c. Corectopia and polycoria which may be progressive
      d. Posterior embryotoxon (prominent and anterior Schwalbe line)
      e. Peripheral iris attachments to Schwalbe line
      f. Microcornea/megalocornea
         i. Gonioscopic features
         ii. Prominent Schwalbe line
         iii. Iris strands from peripheral iris attached to Schwalbe line
         iv. Angle open and visible between strands
         v. Scleral spur often not visible
   2. Systemic
      a. Dental abnormalities
         i. Reduction in crown size (microdontia)
         ii. Decreased number of evenly spaced teeth
         iii. Focal absence of teeth
         iv. Facial abnormalities
      b. Maxillary hypoplasia
         i. Flattening of midface
         ii. Receding upper lip
         iii. Prominent lower lip
iv. Hypertelorism and/or telecanthus

3. Other
   a. Anomalies in region of pituitary gland
      i. Primary empty sella and growth hormone deficiency
   b. Redundant umbilical skin

II. List the differential diagnosis

A. Partial aniridia
B. Iridocorneal endothelial syndrome
C. Congenital iris hypoplasia
D. Ectopia lentis et pupillae
E. Peters anomaly
F. Congenital ectropion uveae
G. Iridoschisis

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

A. Describe medical therapy options to initially lower IOP (listed alphabetically)
   1. Alpha adrenergic agonists
   2. Beta adrenergic antagonists
   3. Carbonic anhydrase inhibitors
   4. Prostaglandin analogues
   5. Pilocarpine is not effective

B. Describe surgical therapy options
   1. Goniotomy and trabeculotomy have both been tried with limited success in infantile onset glaucoma
   2. Trabeculectomy with antimetabolite
   3. Aqueous shunt procedure
   4. Cyclodestructive procedure
   5. Laser trabeculoplasty ineffective

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

A. Complications of glaucoma medical therapy (See Beta-adrenergic antagonists) (See Alpha adrenergic agonists) (See Carbonic anhydrase inhibitors) (See Prostaglandin analogues)
B. Complications of surgical therapy (See Aqueous shunt surgery) (See Incisional filtering surgery for open angle glaucoma) (See Ciliary body ablation procedures)

V. Describe appropriate patient instructions

A. Close follow-up throughout childhood and into adulthood (it is rare in infancy) because the major concern is the development of glaucoma
B. Genetic counseling
Aniridia

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Aniridia is a genetic disorder that can be autosomal dominant (two-thirds) or rarely recessive
   a. About one-third are sporadic
2. Affects 1 in 80,000 individuals
3. Equally affects males and females
4. Glaucoma may occur in pre-adolescent or early adult years
5. Infantile glaucoma is unusual
6. The incidence of glaucoma is variable and ranges between 50 and 75% of patients
7. Elevated intraocular pressure (IOP) may be secondary to
   a. Developmental goniodysgenesis
   b. Synechial angle closure from iris stump

B. List the pertinent elements of the history

1. Photophobia
2. Nystagmus
3. Decreased vision
4. Family history of aniridia
5. Delayed milestones
6. Mental retardation
7. Gait abnormalities
8. Genitourinary abnormalities

C. Describe the pertinent clinical features

1. Usually bilateral
2. Strabismus and nystagmus
3. Iris hypoplasia with small vestigial iris stump
   a. Degree of aniridia is highly variable
4. Remnants of tunica vasculosa lentis and pupillary displacement might be seen
5. Microcornea
6. Elevated IOP which is usually not detected at birth
7. 50-75% develop glaucoma
8. Aniridic keratopathy (pannus formation and corneal scarring)
9. Cataracts (polar cataracts are common)
10. Foveal hypoplasia
11. Optic nerve hypoplasia
12. Ectopia lentis
13. Thick corneas (should be considered in setting target IOP)
14. Angle may be open with anomalies reminiscent of primary congenital glaucoma
15. Angle closure type glaucoma with peripheral anterior synechiae
16. Systemic disease associations
   a. Sporadic form
      i. Caused by large deletions in PAX gene
      ii. Associated with Wilms tumor (nephroblastoma)
      iii. 67-fold greater chance of developing Wilms tumor compared to the normal population
      iv. Children with Wilms tumor-aniridia-genitourinary malformation-retardation (WAGR) syndrome are at greatest risk for developing Wilms tumor
   b. Central nervous system (CNS) abnormalities such as absence of corpus callosum
   c. Isolated genitourinary abnormalities

II. List the differential diagnosis
   A. Traumatic or surgical aniridia associated with glaucoma
   B. Axenfeld-Rieger Syndrome (See Axenfeld-Rieger Syndrome)
   C. Microcornea with ectopia lentis
   D. Congenital iris coloboma
   E. Oculocutaneous albinism Type I

III. Describe patient management in terms of treatment and follow-up
   A. Describe medical therapy options
      1. Limited long term value, may lower IOP initially
         a. Beta-adrenergic antagonists
         b. Adrenergic agonists
         c. Prostaglandin analogues
         d. Carbonic anhydrase inhibitors
      2. Refractive error treatment options
         a. Watch for amblyopia
   B. Describe surgical therapy options
      1. Goniotomy
      2. Trabeculotomy
      3. Trabeculectomy with or without antimetabolite
      4. Aqueous shunt surgery
      5. Cycloablation

IV. List the complications of treatment, their prevention and management
   A. Complications of glaucoma medical therapy (See Beta-adrenergic antagonists) (See Alpha adrenergic agonists) (See Carbonic anhydrase inhibitors) (See Prostaglandin analogues)
   B. Complications of surgical therapy (See Aqueous shunt surgery) (See Incisional filtering surgery for open angle glaucoma) (See Ciliary body ablation procedures)

V. Describe appropriate patient instructions
A. Lifelong follow-up for treatment of glaucoma
B. Monitoring of IOP in individuals not affected by glaucoma over their lifetime
C. Sporadic cases (WAGR syndrome) must be monitored for Wilms tumor with physical examination, serial abdominal ultrasonography, and/or genetic testing
D. Genetic counseling

Additional Resources
1. AAO, Basic and Clinical Sciences Course. Section 10: Glaucoma, 2015-2016.
Glaucoma and posterior corneal transplantation

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease

1. Glaucoma may develop or be exacerbated by anatomic effects of posterior corneal surgery including endothelial keratoplasty (e.g. Descemet Stripping Endothelial Keratoplasty, DSEK)

2. Causes in the early postoperative period include:
   a. Postoperative inflammation
   b. Viscoelastic substances
   c. Wound leak with shallow anterior chamber and peripheral anterior synechiae formation
   d. Hyphema
   e. Secondary angle closure glaucoma from the air bubble migrating behind the iris
   f. Pupillary block glaucoma
      i. Caused by air bubble located anterior to the iris used in DSEK for donor tissue adhesion
      ii. Decrease risk with
         i) Intraoperative or postoperative mydriatics
         ii) Ensuring free movement of bubble in anterior chamber
   g. Preexisting glaucoma

3. Causes in the later postoperative period include:
   a. Chronic angle-closure glaucoma
   b. Preexisting glaucoma
   c. Steroid-induced glaucoma
   d. Epithelial downgrowth

4. Intraocular pressure measurements may be affected
   a. Without corneal edema, an increased corneal thickness after DSEK does not lead to an artifactually high IOP reading by Goldmann applanation. Therefore, high IOP reading by Goldmann applanation likely reflects a truly elevated IOP in DSEK eyes
   b. With corneal edema, IOP may be underestimated due to the changes in corneal biomechanics

B. Define the relevant aspects of epidemiology of the disease

1. Incidence of IOP elevation increases after DSEK, particularly in eyes with preexisting glaucoma

2. Acute IOP elevation can arise from a mixture of angle closure from air behind the iris and pupillary block from air in front of the iris

C. List the pertinent elements of the history

1. Pre-existing angle closure
2. History of glaucoma or ocular hypertension
3. History of steroid-related elevated IOP

D. Describe pertinent clinical features

1. Elevated intraocular pressure
2. Pupillary block from air in front of the iris
3. Secondary angle closure from air behind the iris
4. Typical glaucomatous optic nerve and nerve fiber layer changes

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Intraocular pressure (IOP) measurement
   a. Goldmann applanation may be difficult if surface irregular or edematous; pneumotonometer or Tono-Pen may be more accurate
   b. Epithelial defects do not preclude IOP measurement
2. Gonioscopy - may be difficult if cornea not clear
3. Determine where the air bubble is located
4. Dilated fundus examination
5. Visual field
6. Documentation of optic nerve head and nerve fiber layer

II. Define the risk factors

A. Preexisting glaucoma
B. Preexisting angle closure
C. Large air bubble

III. List the differential diagnosis

A. Steroid-induced glaucoma
B. Non air-bubble related pupillary block
C. Other causes of angle closure glaucoma

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

A. Describe the natural history, outcome and prognosis
   1. Patients with pupillary block or acute secondary angle closure glaucoma have pain which will continue until the pupillary block breaks or the angle closure is reversed
   2. Persistent elevated IOP can lead to glaucomatous optic neuropathy
B. Describe medical therapy options
   1. Topical glaucoma medications
   2. Oral glaucoma medications
C. Describe surgical therapy options
   1. Remove the air bubble
   2. Laser peripheral iridotomy
      a. Indicated in patients with air bubble-related pupillary block after endothelial keratoplasty
      b. Sometimes used prophylactically
   3. Laser trabeculoplasty
   4. Angle Surgery
   5. Trabeculectomy
   6. Glaucoma drainage devices
   7. Cyclodestructive procedures
V. List the complications of treatment, their prevention and management

A. Topical glaucoma medications

1. Topical carbonic anhydrase inhibitors may exacerbate corneal edema, especially if endothelial cell counts low
2. Prostaglandin analogues may cause or exacerbate inflammation, cystoid macular edema, and recurrent herpes simplex keratitis
3. Cholinergics may exacerbate inflammation and lead to graft rejection
4. Benzalkonium chloride may cause dose-dependent surface toxicity

B. Trabeculectomy

1. Use postoperative 5-FU with caution; may cause or exacerbate epithelial toxicity and lead to persistent epithelial defects

C. Glaucoma drainage devices (GDD)

1. Associated with high risk of penetrating graft failure but unknown risk after posterior corneal transplantation

D. Cyclophotocoagulation

VI. Describe disease-related complications

A. Glaucoma-related vision loss
B. IOP-related endothelial cell loss and/or damage, potentially leading to graft failure

VII. Describe appropriate patient instructions

A. Patients with a history of corneal surgery need continued monitoring for eye conditions including glaucoma, even if asymptomatic. This is of particular importance after posterior corneal surgery because of possible long-term steroid use and a thicker cornea and resultant artifactual elevation in IOP

Additional Resources

1. AAO, Basic and Clinical Science Course. 2015-2016.
Clinical trials in glaucoma

I. Previous thinking

A. One should lower intraocular pressure (IOP) 20-30% (the “Target IOP”) from the level of previous injury in order to stop visual field progression

B. An IOP that is consistently in the "normal range" (<21 mm Hg) is sufficient

II. Newer evidence-based thinking

A. Target IOP is no longer simply a percentage decrease from a baseline level and/or an absolute IOP number, but constitutes a range of IOP determined by:
   1. Knowledge of central corneal thickness (CCT) that leads to a better defined IOP level
   2. Extent of optic nerve and visual field damage
   3. Previous IOP levels with and without medication
   4. Age and life expectancy
   5. Family history
   6. Vascular factors (e.g., low perfusion pressures, mean arterial pressure, migraine)

III. Multi-centered randomized controlled trials

A. Ocular Hypertension Treatment Study (OHTS)
   1. Purpose
      a. To determine the efficacy of topical ocular hypotensive medication in delaying or preventing the onset of primary open-angle glaucoma (POAG) in patients with ocular hypertension
   2. Treatment group
      a. Used any available hypotensive medications to lower IOP
   3. Control group
      a. Observation
   4. Results
      a. Mean IOP reduction of approximately 20% reduced progression to POAG from 9.5% in the observation group to 4.4% in the treatment group at 5 years
   5. Risk factors for conversion to glaucoma status
      a. Identified via multivariate analysis
         i. Thinner CCT
         ii. Older age
         iii. Higher IOP
         iv. Worse pattern standard deviation (PSD)
         v. Larger cup/disc ratio
      b. Race/ethnicity and family history were not significant in a multivariate model

B. Early Manifest Glaucoma Treatment (EMGT)
   1. Purpose
      a. To determine the efficacy of glaucoma treatment in patients with newly detected early OAG
b. Treatment group
   i. Betaxolol plus argon laser trabeculoplasty (ALT)
c. Control group
   i. Observation

2. Results
   a. Mean IOP reduction of 25% reduced progression from 62% in the observation group to 45% in the treatment group at 6 years. Treatment delayed the time to progression by about 2 years

3. In a regression model, 1 mm lower IOP was associated with 10% lower chances of glaucoma progression.
   Risk factors associated with progression
   a. Higher baseline IOP
   b. Exfoliation syndrome
   c. Bilateral disease
   d. Worse mean deviation
   e. Older age
   f. Frequent disc hemorrhages

4. Complications
   a. Treatment group had an increased incidence of nuclear lens opacities

C. Collaborative Initial Glaucoma Treatment Study (CIGTS)

1. Purpose
   a. To determine the long-term effect of treating patients with newly diagnosed POAG with trabeculectomy vs. with medication

2. Treatment groups
   a. Randomized patients (not eyes) to
      i. Initial filtering surgery
      ii. Initial medical therapy

3. Results
   a. At 5 years, visual field results were comparable between the medical treatment group (which had a 38% IOP reduction) and the surgical treatment group (which had a 45% IOP reduction) with minimal progression in both groups
   b. Similar visual field outcomes between both groups were also observed at up to 9 years of follow-up
   c. The Quality of Life (QOL) impact was similar in both treatment groups. The most persistent QOL finding was an increase in local eye symptoms in the surgery group
   d. The overall rate of visual field progression was lower in CIGTS than other clinical trials, presumably the result of more aggressive IOP goals

4. Complications
   a. The surgical group had more early visual acuity loss than the medical group, but the difference converged over time
   b. The surgical group had more bothersome local symptoms related to quality of life than the medical group, but these symptoms diminished with time
   c. The surgical group had a greater rate of cataract removal than the medical group.

D. Advanced Glaucoma Intervention Study (AGIS)

1. Purpose
   a. To determine which surgical option is more efficacious, argon laser trabeculoplasty (ALT) or incisional surgery, in halting or delaying progression of glaucoma in patients with advanced disease failing medical management
   b. Main outcome measures were visual acuity and visual field, but many other secondary analyses
were performed while the study evolved over a long follow-up period

2. Treatment groups
   a. Trabeculectomy first, followed by ALT and a second trabeculectomy, as needed (TAT)
   b. ALT first, followed by first trabeculectomy and then second trabeculectomy, as needed (ATT)

3. Results
   a. Ultimately, no difference was found between treatment groups
   b. Analyses by race found that African American patients had less visual field loss if treated with ALT first. Caucasian patients had less visual field loss if treated with Trabeculectomy first

4. Risk factors
   a. Lower IOP is associated with less progression of visual field loss in a dose-response relationship
   b. Inter-visit IOP fluctuation and increased age are stronger risk factors than other variables (including mean IOP) for visual field worsening

5. Complications
   a. Approximately half of study patients developed cataracts in the first 5 years of follow-up
   b. Trabeculectomy increased the risk of cataract formation
   c. Visual function scores improved after cataract surgery. Adjustment for cataracts did not alter the AGIS findings

E. Collaborative Normal Tension Glaucoma Study (CNTGS)

1. Purpose
   a. To determine whether aggressive IOP lowering (medical and/or surgical) halts optic nerve damage and visual field loss in normal-tension glaucoma
   b. To assess risks/side effects of treatment

2. Treatment group
   a. 30% IOP lowering (medical and/or surgical)
   b. Medical treatment excluded:
      i. Systemic or topical beta-blockers
      ii. Systemic or topical adrenergic agonists
      iii. Prostaglandins and topical CAIs were not available at the time of the study

3. Control group
   a. Observation

4. Results
   a. A mean IOP decrease of 30% resulted in 12% of patients showing visual field progression versus 35% in the control group at 5 years
   b. This protective effect of IOP reduction was only evident after adjusting for the effect of cataract

5. Risk factors for progression
   a. Occurrence of disc hemorrhage
   b. History of migraine
   c. Female sex

6. Complications
   a. Cataracts especially in the filtration group

F. Glaucoma Laser Trial (GLT)

1. Purpose
   a. To determine efficacy and safety of argon laser trabeculoplasty compared to topical medication as the initial treatment in previously untreated POAG
2. Treatment groups
   a. Trabeculoplasty as initial treatment
   b. Topical medication as initial treatment

3. Results
   a. Within 2 years, initial laser trabeculoplasty was at least as effective as medication, but more than half of the eyes initially treated with laser surgery required medications to control IOP
   b. Eyes treated with laser first had lower IOP and better visual field and optic nerve outcomes over a 7 year follow up period

4. Complications
   a. Laser trabeculoplasty was associated with a transient rise of IOP in approximately 20% of patients, but there was no prophylaxis used against an increase in IOP (i.e., no perioperative adrenergic agonist was given)

G. Fluorouracil Filtering Surgery Study
1. Purpose
   a. To determine the efficacy and safety of subconjunctival 5-fluorouracil (FU) injections in patients with a poor prognosis for trabeculectomy success (i.e., those with previous conjunctival surgery)
   b. To determine risk factors for failure

2. Treatment groups
   a. Trabeculectomy with subconjunctival 5-FU injections
   b. Trabeculectomy without antimetabolites

3. Results
   a. After 5 years of follow-up, the incidence of surgical success was higher in the 5-FU group (approximately 50%) compared to the standard trabeculectomy group (approximately 25%)

4. Risk factors for failure
   a. High IOP
   b. Short time since last conjunctival incision
   c. Higher number of previous surgeries involving conjunctival incision
   d. Hispanic ancestry

5. Complications
   a. There was a higher risk of late-onset bleb leak in eyes that had 5-FU injection (9% in 5-FU group vs. 2% in standard trabeculectomy group)

H. Tube versus Trabeculectomy Study (TVT Study)
1. Purpose
   a. To compare the long-term (5-year) safety and efficacy of trabeculectomy with MMC to the Baerveldt glaucoma implant in patients with OAG having undergone prior cataract surgery, trabeculectomy or both surgeries

2. Treatment groups
   a. Trabeculectomy with MMC (0.4 mg/ml for 4 minutes)
   b. Baerveldt (350 mm²) glaucoma implant

3. Results
   a. Tube shunt surgery had a higher success rate compared to Trabeculectomy with MMC during the first 5 years
   b. Both procedures were associated with similar IOP reduction and use of adjuvant medical therapy at 5 years
   c. The incidence of postoperative complications was higher following Trabeculectomy with MMC, but most complications were transient
Complications

a. Early complications more common in Trabeculectomy group and late complications similar between groups

b. Bleb leaks and dysesthesia were more frequent with Trabeculectomies and diplopia seen more commonly with Baerveldt

c. A higher rate of reoperation for glaucoma after Trabeculectomy than after Baerveldt

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 10: Glaucoma, Section 10, 2015-2016.


22. Five-year follow-up of the Fluorouracil Filtering Surgery Study. The Fluorouracil Filtering Surgery Study


Beta-adrenergic antagonists

I. List the agents

A. Non-selective $\beta_1$- and $\beta_2$- antagonists
1. Timolol maleate 0.25%, 0.5% (Timoptic®, Timoptic XE®, Timolol GFS®, Istalol®, generic)
2. Timolol hemihydrate 0.25%, 0.5% (Betimol®)
3. Levobunolol HCl 0.25%, 0.5% (Betagan®)
4. Carteolol HCl 1.0% (Ocupress®)
5. Preservative-free timolol

B. Selective $\beta_1$- antagonist
1. Betaxolol HCl suspension 0.25% (Betoptic-S®), betaxolol HCl solution 0.5% (generic)

II. List the indications/contraindications

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

B. Contraindications
1. Proven sensitivity to agents
2. Reactive airway disease
   a. Bronchospasm
   b. Chronic obstructive pulmonary disease
3. Greater than first degree heart block

C. Relative contraindications
1. Congestive heart failure
2. Bradycardia
3. Diabetes, because the drug may mask the symptoms of hypoglycemia
4. Systemic beta-adrenergic antagonists
   a. Patients who are receiving a beta-adrenergic antagonist orally and topically should be observed for potential additive effects of beta-adrenergic blockade
5. The concomitant use of two topical beta-adrenergic agents is not recommended

III. Describe the pre-therapy evaluation

A. Review of medical history to determine whether any contraindications to beta-blocker use are present

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

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

V. Describe the method of action and pharmacokinetics
A. $\beta_1$- and $\beta_2$- receptors are on the ciliary processes. Receptor blockade reduces aqueous humor production via direct action
   1. Direct effect on non-pigmented ciliary epithelium to decrease secretion via inhibition of cyclic adenosine monophosphate
   2. Decreases local capillary perfusion to reduce ultrafiltration
B. Administration
   1. Administered once or twice daily. Timolol GFS administered once daily
   2. Most patients can be dosed once a day, first thing in the morning to blunt an early morning IOP rise while minimizing risk of systemic hypotension during sleep
   3. Good corneal penetration
   4. Peak aqueous concentration within 1-2 hours of topical dose
   5. IOP effect peaks at 2 hours and lasts at least 24 hours
      a. Short-term escape
         i. A dramatic reduction in IOP after initial administration followed by small IOP rise that plateaus within a few days
         ii. May be due to an increase in the number of $\beta$ receptors during the first few days of therapy
         iii. Wait approximately one month to evaluate therapeutic response
      b. Long-term drift/tachyphylaxis
         i. Approximately 3 months after initiating therapy, the IOP-lowering response in some patients will diminish
         ii. Some will regain responsiveness after a drug holiday
C. Efficacy
   1. Non-selective $\beta_1$- and $\beta_2$- antagonists
      a. 20%-30% IOP reduction
   2. $\beta_1$- selective antagonist
      a. 15-20% IOP reduction
   3. Decreased efficacy is possible when topical beta-blockers are used concomitantly with oral beta-adrenergic antagonists
   4. Systemic absorption may result in a small IOP lowering effect in the contralateral eye
   5. Very little effect on aqueous production and IOP during sleep

VI. Describe the complications of therapy, their prevention and management
A. Complications
   1. Ocular toxicity
      a. Burning, hyperemia
      b. Corneal anesthesia, punctate keratopathy, erosions, toxic keratopathy
      c. Periocular contact dermatitis
      d. Dry eye
2. Systemic
   a. Cardiovascular
      i. β₁ blockade slows pulse and decreases cardiac contractility
         i) May cause syncope, bradycardia, arrhythmias, heart failure, decreased exercise tolerance
      ii. Nocturnal systemic hypotension, especially in normal tension glaucoma; hence, a reason for administration in the morning
   b. Respiratory
      i. β₂ blockade produces contraction of bronchial smooth muscle
         i) May cause bronchospasm and airway obstruction, especially in asthmatics
         ii) May cause dyspnea and apneic spells, especially in young children
   c. Central nervous system
      i. Depression, anxiety, confusion, hallucinations, lightheadedness, drowsiness, fatigue, weakness, disorientation
   d. Cholesterol levels
      i. Alterations in plasma lipid profile have been reported with timolol when administered without punctal occlusion
         i) Decreases plasma high density lipoprotein and possibly increases risk of coronary artery disease
   e. Other
      i. Exacerbation of myasthenia gravis
      ii. May mask awareness of hypoglycemia in diabetics
      iii. Gastrointestinal distress
      iv. Dermatologic disorders
      v. Sexual impotence

B. Prevention of complications
   1. Avoid use of beta-adrenergic antagonists in high-risk patients
   2. Nasolacrimal occlusion
   3. Once per day administration, preferably in the morning to decrease total daily dose
   4. Viscous formations of Timolol may have less systemic absorption
   5. Use topical beta-adrenergic antagonists with special properties
      a. Betaxolol - β₁ selective antagonist
         i. Decreased incidence of respiratory side effects in patients with bronchospastic disease
      b. Carteolol - intrinsic sympathomimetic activity
         i. Adrenergic agonist effect that may partially protect against adverse effects of beta-adrenergic blockade
         ii. Has less adverse effect on plasma lipid profile

C. Management of complications
   1. Discontinuation of drug
   2. Consider switch to beta-adrenergic antagonist with special properties if indicated

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

VIII. Describe appropriate patient instructions

A. Discuss potential side effects
B. Discuss/demonstrate nasolacrimal occlusion and passive eyelid closure
C. If on more than one topical medication, instruct patient to wait at least 5 to 10 minutes between eye drop administration

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Parasympathomimetic agents

I. List the agents
   A. Carbachol
   B. Pilocarpine hydrochloride
   C. Echothiophate iodide

II. List the indications/contraindications
    A. Indications
       1. Treatment of increased intraocular pressure (IOP) in patients with at least some open filtering angle
       2. Prophylaxis
          a. For angle-closure prior to iridotomy
          b. To decrease iridozonular contact in pigment dispersion glaucoma (may not be tolerated in this younger patient population because of induced myopia)
    B. Contraindications
       1. Patients with no trabecular outflow
       2. Patients with peripheral retinal disease that predisposes them to retinal detachment
       3. Uveitic glaucoma
       4. Acute infectious conjunctivitis
       5. Proven hypersensitivity to these agents
       6. Significant lens changes with chronic use (relative contraindication)
       7. Aqueous misdirection
       8. Angle closure secondary to ciliary body edema

III. Describe the pre-therapy evaluation
    A. Comprehensive eye exam with particular attention to
       1. Gonioscopy
       2. IOP measurement
       3. Evidence of uveitis
       4. Iris examination—darker irides may require higher initial concentrations
       5. Lens examination
       6. Peripheral retinal examination
       7. Optic nerve examination

IV. List the alternatives to this therapy
    A. Prostaglandin analogues
    B. Beta-adrenergic antagonists
    C. Adrenergic agonists
    D. Carbonic anhydrase inhibitors
V. **Describe mode of action**

A. Reduces IOP by causing contraction of the ciliary muscle, which pulls the scleral spur to tighten the trabecular meshwork, increasing the outflow of aqueous humor

1. Direct-acting agents (pilocarpine and carbachol) affect the motor end plates directly in the same way as acetylcholine, which is transmitted at postganglionic parasympathetic junctions

2. Indirect-acting agents (echothiophate iodide and carbachol) inhibit the enzyme acetylcholinesterase, thereby prolonging and enhancing the action of naturally secreted acetylcholine. Of note, it however decreases uveoscleral outflow

VI. **List the complications of this therapy - ocular and systemic, their prevention and management**

A. **Ocular complications**

1. More frequent
   a. Induced myopia
   b. Brow ache—this usually abates after 1-2 weeks of use
   c. Conjunctival and intraocular vascular congestion
   d. Cataracts (especially with cholinesterase inhibitors)
   e. Paradoxical angle closure (by inducing greater lenticular-pupillary block)
   f. Posterior synechiae
   g. Corneal toxicity
   h. Periocular contact dermatitis
   i. Exacerbation of visual symptoms secondary to cataract because of miosis

2. Less frequent
   a. Iris pigment epithelial cysts (cholinesterase inhibitors) - in children
   b. Epiphora due to direct lacrimal stimulation and punctal stenosis
   c. Pseudopemphigoid
   d. Fibrinous iritis (especially in the postoperative period)
   e. Retinal detachment

3. Complications may be minimized by titrating initial dosage and starting at lower concentrations and/or frequency Compliance is probably more problematic than with other agents

B. **Systemic complications (mainly seen with indirect acting agents)**

1. Diarrhea
2. Abdominal cramps
3. Increased salivation
4. Bronchospasm
5. Enuresis
6. Nausea
7. Tremors
8. Slowing of the pulse
9. Decreased blood pressure
10. Inhibition of acetylcholinesterase by cholinesterase inhibitors, which interferes with the clearing of succinylcholine. This may prolong respiratory paralysis during general anesthesia

C. Prevention and management of complications
1. Lacrimal punctal occlusion
2. Light eyelid closure for 3 minutes
3. Warn high myopes and those with peripheral retinal pathology of the symptoms of retinal detachment
4. Do not overuse parasympathomimetic agents in cases of acute angle-closure glaucoma (an ischemic sphincter from high IOP will not constrict) and there may be a paradoxical forward shift of lens-iris diaphragm, worsening pupillary block

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

VIII. Describe appropriate patient instructions
A. Schedule patient return for follow up IOP check and to review any side effects experienced from the drug
B. Describe symptoms of retinal detachment
C. Explain possible side effects
D. Explain the need to contact the ophthalmologist if they have symptoms of angle-closure glaucoma such as halos around lights

Additional Resources
1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma. 2015-2016
2. AAO, Glaucoma Medical Therapy: Principles and Management, 1999;77-93.
Carbonic anhydrase inhibitors (CAIs)

I. List the agents

A. Oral
   1. Acetazolamide (Diamox) 125 mg, 250 mg; sustained release acetazolamide (Diamox Sequels) 500 mg
   2. Methazolamide (Neptazane) 25 mg, 50 mg

B. Topical
   1. Dorzolamide 2% (Trusopt)
   2. Brinzolamide 1% (Azopt)

C. Intravenous for acute or urgent situations
   1. Acetazolamide 250 mg to 1 gm
      a. 1-4 doses in 24 hours

II. List the indications/contraindications

A. Indications
   1. Reduction of chronically elevated intraocular pressure (IOP) in adults and children
      a. Monotherapy
      b. Additive therapy
         i. Can help lower diurnal and nocturnal IOP when added to a prostaglandin analog
         ii. Additive to topical beta-blockers, alpha2 agonists and pilocarpine
   2. Prophylaxis of elevated IOP after a surgical intervention
   3. Reduction of acutely elevated IOP
   4. Chronic use of systemic CAIs should be reserved for patients whose glaucoma is not controlled with topical therapy, who have refused surgery or in whom surgery is inappropriate

B. Contraindications
   1. Sulfa allergy that is determined to be a true allergic reaction is an absolute contraindication
   2. Self-reported sulfa allergy is a relative contraindication according to a study finding little clinical or pharmacological evidence to suggest that CAIs are likely to produce a life-threatening cross-reaction
   3. Kidney stones
   4. Aplastic anemia
   5. Thrombocytopenia
   6. Dehydration and acidosis may promote sickling in patients with Sickle cell disease
   7. History of blood dyscrasias
   8. Corneal endothelial failure
   9. Relative contraindication for methazolamide oral CAIs in patients with hepatic insufficiency and acetazolamide in patients at risk for ketoacidosis
   10. Dose of acetazolamide oral CAIs should be reduced in patients with renal insufficiency

III. Describe the pre-therapy evaluation

A. Medical history
B. Comprehensive eye examination
C. Determine if there are any contraindications

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

A. Describe medical therapy options
   1. Oral or topical carbonic anhydrase inhibitors
   2. Topical beta-adrenergic antagonists
   3. Alpha-adrenergic agonists
   4. Parasympathomimetic agents
   5. Prostaglandin analogues

B. Describe surgical therapy options
   1. Laser trabeculoplasty
   2. Trabeculectomy
   3. Glaucoma drainage devices
   4. Cyclodestructive procedures

V. Describe the method of action and pharmacokinetics

A. Inhibition of carbonic anhydrase in ciliary epithelium decreases aqueous production
   1. > 90% of carbonic anhydrase must be blocked to decrease aqueous production
   2. Acetazolamide produces metabolic acidosis which contributes to IOP lowering effect

B. Administration
   1. Topical CAIs administered three times a day lower IOP slightly more than twice daily administration
   2. Oral CAIs should be administered in the lowest dose that reduces IOP to an acceptable range. Typical oral (PO) dosages:
      a. Acetazolamide 250mg PO every 6 hours
      b. Acetazolamide 500mg sustained release capsules PO every 12 hours
      c. Methazolamide 25-50mg PO every 8-12 hours, maximum dose 100mg three times daily

C. Efficacy
   1. 15-20% IOP reduction with topical CAIs
   2. 25-50% IOP reduction with oral CAIs
   3. There is no advantage to adding a topical CAI to a patient taking an oral CAI

VI. List the complications of the therapy, their prevention and management

A. Complications
   1. With systemic carbonic anhydrase inhibitors
      a. Initial increase in urinary frequency
      b. Poor tolerance of carbonated beverages
      c. Paresthesias
      d. Malaise, depression
      e. Anorexia, weight loss
f. Altered taste

g. Kidney stones

h. Stevens-Johnson syndrome, if allergic

i. Idiosyncratic blood dyscrasias (aplastic anemia and sickle cell disease)

j. Hypokalemia especially if already on a thiazide diuretic

k. Choroidal detachment

l. Induced myopia

2. With topical carbonic anhydrase inhibitors

a. Burning and stinging (less with brinzolamide)

b. Metallic taste in mouth

c. Epithelial and endothelial corneal toxicity

d. Conjunctival injection

e. Periocular contact dermatitis

f. Choroidal detachment

B. Prevention of complications

1. Monitor blood potassium with concurrent use of oral CAIs and potassium wasting diuretics

2. Consider pre-treatment blood counts with oral CAIs

3. Avoid topical CAIs for corneas with impaired endothelial function

4. Avoid CAIs with a history of sulfa allergy, blood dyscrasia or kidney stones

5. Avoid oral CAIs in patients with hepatic insufficiency

6. Adjust dose of oral CAIs in patients with renal insufficiency

7. Recommend taking oral CAIs with meals to decrease gastrointestinal side effects

C. Management of complications

1. Stop the medication

2. Topical toxicity

   a. Change to a different class of topical medication

   b. Consider brinzolamide instead of dorzolamide

   c. Oral CAIs

3. Systemic toxicity

   a. Decrease the dose or frequency of oral medication

   b. Change to topical therapy

   c. Change from acetazolamide to methazolamide

   d. Medical consult for serious side effects

   e. Switch to sustained-release acetazolamide

VII. Describe the follow-up care

A. Consider therapeutic one-eyed trial

B. Check IOP 2-4 weeks after starting therapy to allow for IOP lowering and side effects to manifest themselves

C. Monitor side-effects

D. Follow IOP, optic nerve appearance, and visual fields
VIII. Describe appropriate patient instructions

A. Discuss possible side effects and their signs and symptoms
B. Discontinue treatment for any serious side effect
C. With oral CAIs, discuss need for potassium supplement with primary care physician

Additional Resources

1. AAO, Basic Clinical and Science Course. Section 10: Glaucoma, 2015-2016.


25. Martínez A, Sánchez-Salorio M. Predictors for visual field progression and the effects of treatment with dorzolamide 2% or brinzolamide 1% each added to timolol 0.5% in primary open-angle glaucoma. Acta Ophthalmol. 2010;88:541-52.


Alpha adrenergic agonists

I. List the agents
   A. Selective adrenergic agonists
      1. Apraclonidine (0.5%)
      2. Brimonidine tartrate (0.2%, 0.15%, 0.1%)

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

III. List the contraindications and precautions
   A. Selective
      1. Proven sensitivity to these agents
      2. Concomitant use of monoamine oxidase inhibitors (MAOI)
      3. Infants and children younger than two years old
         a. Brimonidine is contraindicated due to apnea, bradycardia, dyspnea
      4. Pediatric (age 2-7) usage reports
         a. Bradycardia
         b. Hypotension
         c. Apnea
         d. Somnolence (50-83% incidence)
            i. Less frequent (25%) in children >20 kg
         e. Brimonidine is relatively contraindicated
      5. Precaution in patients with
         a. Severe cardiovascular disease
         b. Depression
         c. Cerebral or coronary insufficiency
         d. Raynaud’s phenomenon
         e. Orthostatic hypotension
         f. Use of CNS depressants (alcohol, barbiturates, opiates, sedatives)
6. Elderly
7. Pregnancy
   a. Category B drug

IV. Describe the pre-therapy evaluation
   A. Complete history including
      1. MAOI usage
      2. History of adrenergic sensitivity
   B. Comprehensive eye examination including
      1. Gonioscopy
      2. IOP measurement
      3. Optic nerve assessment

V. List the alternatives to this therapy
   A. Prostaglandin analogues
   B. Carbonic anhydrase inhibitors
   C. Beta-adrenergic antagonists
   D. Parasympathomimetics
   E. Osmotic agents
   F. Laser trabeculoplasty
   G. Incisional glaucoma surgery
   H. Cyclodestruction

VI. Describe the dosing technique
   A. Nasolacrimal occlusion
   B. Passive lid closure

VII. Describe the method of action
   A. Selective-alpha adrenergic receptor agonist
      1. Reduction of aqueous humor production is primary mechanism of action
      2. Fluorophotometric studies suggest that brimonidine tartrate also increases uveoscleral outflow

VIII. List the complications of the ocular therapy
   A. Selective
      1. Local
         a. Ocular hyperemia
         b. Burning
         c. Stinging
d. Foreign body sensation  
e. Conjunctival follicles  
f. Allergic reaction, such as allergic conjunctivitis and associated lid dermatitis  
i. Incidence about 17%, decreased with lower concentration formulations; onset may be delayed by weeks or months  
g. Pruritus  
h. Corneal staining  
i. Ocular dryness  
j. Conjunctival blanching  
k. Granulomatous anterior uveitis (rare)  

2. Systemic  
a. Oral dryness (more common)  
b. Fatigue/drowsiness (more common)  
c. Headache  
d. Hypertension  
e. Palpation/arrhythmias  
f. Nasal dryness

IX. List the complications of the therapy, their prevention and management

A. Prevention  
1. Discussion of potential side effects with patient  
2. Nasolacrimal occlusion or lid closure  
3. Emphasis on correct dosing

B. Management  
1. Discontinue medication  
2. Symptomatic relief of side effect until resolution if applicable

X. Describe the follow-up care

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

XI. Describe appropriate patient instructions

A. Discuss possible side effects with patient to increase level of awareness  
B. Demonstrate nasolacrimal occlusion or lid closure  
C. Schedule regular follow-up visits
Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016
2. AAO, Glaucoma Medical Therapy: Principles and Management, 1999; 47-75.
Prostaglandin analogues

I. List the agents
   A. Bimatoprost 0.03% and 0.01%, Lumigan®, once daily dosing
   B. Latanoprost 0.005%, Xalatan®, once daily dosing
   C. Travoprost 0.004%, Travatan Z®, once daily dosing
   D. Tafluprost 0.0015%, Zioptan®, once daily dosing (preservative-free)

II. List the indications/contraindications
   A. Indications
      1. Initial and adjunctive therapy to lower intraocular pressure (IOP)
         a. Open-angle glaucoma and ocular hypertension
         b. Primary angle closure
         c. Secondary glaucoma
   B. Contraindications
      1. Allergy to agent
   C. Relative contraindications
      1. Proven nonresponsiveness
      2. Active uveitis (controversial)
      3. Active macular edema
      4. Aphakia or pseudophakia with open posterior capsule, especially after complicated surgery
      5. Recent intraocular surgery
      6. History of herpetic keratitis
      7. Previous history of cystoid macular edema (CME) (multiple previous surgeries/trauma)

III. Describe pre-therapy evaluation
   A. Review of ocular history to determine presence of contraindications to prostaglandin analogues

IV. List the alternatives to this therapy
   A. Beta-adrenergic antagonists
   B. Alpha-adrenergic agonists
   C. Carbonic anhydrase inhibitors (topical and oral)
   D. Parasympathomimetic agents
   E. Laser surgery
   F. Surgical therapy

V. Describe mechanism of action
   A. Agents in this class primarily increase uveoscleral outflow. Some agents may also increase trabecular
outflow

B. Efficacy

1. Maximal IOP reduction approximately 12 hours post-dosing, but maximum effect may take 4-6 weeks
2. 25-33% reduction in IOP
3. IOP lowering effect and side effects are not the same for each agent; one might work when another has not and one might not have topical side effects that another agent might cause

VI. List the complications of the therapy, their prevention and management

A. Complications

1. Ocular toxicity/side effects
   a. Conjunctival hyperemia
      i. Reversible with discontinuation
   b. Ocular irritation
   c. Hypertrichosis and darkening of lashes, trichiasis, distichiasis
      i. Reversible with discontinuation
   d. Darkening of iris
      i. Secondary to increased numbers of melanosomes within melanocytes
      ii. Risk of iris pigmentation greatest in light brown, blue-green, or two-toned irides; less common in homogenous blue irides
      iii. Irreversible
   e. Darkening of periocular skin
      i. Reversible with discontinuation
   f. Periorbital fat atrophy, deepening of superior lid sulcus
      i. May be reversible with discontinuation
   g. CME
   h. Anterior uveitis, suspected
      i. Exacerbation of underlying herpes simplex virus keratitis (dendrites)
      j. May also cause corneal pseudo dendrites

2. Systemic (systemic effects are usually rare)
   a. Headaches
   b. Muscle, joint, back pain
   c. Rash, allergic skin reactions
   d. Flu-like symptoms
   e. Periocular contact dermatitis

B. Prevention of complications

1. Avoid use in high-risk patients
2. Consider simultaneous use of topical nonsteroidal anti-inflammatory drugs (NSAIDs) in eyes at risk for developing CME

C. Management of complications

1. Discontinuation of drug
2. Consider change to prostaglandin analogue less likely to cause ocular side effects
VII. Describe the follow-up care

A. Evaluate response to therapy in 4-6 weeks or as warranted by the clinical situation
B. Inquire about side effects
C. Examine for evidence of ocular toxicity

VIII. Describe appropriate patient instructions

A. Educate patient regarding possible side effects, especially hyperemia and iris, lash, eyelid and skin pigmentary changes
B. Instruct patient to wipe excess medication off peri-ocular skin
C. Discuss shelf-life of agent
   1. Once opened, latanoprost may be stored at room temperature for up to 6 weeks. Unopened bottles should be refrigerated. Other agents are stored at room temperature
   2. Tafluprost preservative-free should be stored in the refrigerator in the original pouch. Once the pouch is opened, the single-use containers may be stored in the opened foil pouch for up to 28 days at room temperature. Unused containers must be discarded after 28 days

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
2. AAO, Glaucoma Medical Therapy: Principles and Management, 1999; 113-131,172.
I. Describe the dosing technique

A. Oral agents
   1. Glycerin
      a. Osmoglyn (commercially unavailable, but available from compounding pharmacies) 50% glycerin - 50% water solution
      b. 4-7 ounces (1-1.5g/kg or 2-3cc/kg body weight) given orally.
      c. Give solution over ice for improved tolerability. Addition of lemon juice or other tart flavoring may improve palatability

B. Intravenous agents
   1. Mannitol (Osmitrol®)
      a. 5-25% solution
      b. 1-2 g/kg body weight given intravenously over 30 minutes

II. List the indications/contraindications

A. Indications
   1. Short-term or emergency treatment of elevated intraocular pressure (IOP)
   2. Useful in acute conditions of elevated IOP (e.g., angle-closure glaucoma)
   3. Effective when elevated IOP renders iris non-reactive to agents which combat pupillary block such as the parasympathomimetic agents (e.g., pilocarpine)
   4. Used to lower IOP and/or reduce vitreous volume prior to initiation of surgical procedures

B. Contraindications
   1. Should not be used for long-term therapy (becomes ineffective with repeated dosing)
      a. Glycerin may increase blood sugar levels (may precipitate ketoacidosis in patients with diabetes)
      b. Long-term use may perturb electrolytes
   2. Of limited value when blood-ocular barrier is disrupted
   3. May cause rebound elevation in IOP if agent penetrates eye and reverses osmotic gradient
   4. Congestive heart failure
   5. Renal failure
   6. Sickle cell disease/ trait

III. Describe the pre-therapy evaluation

A. Accurate measurement of IOP
B. Slit-lamp biomicroscopic exam
   1. Pupil/iris evaluation for ischemic and non-reactive iris sphincter muscle
   2. Shallowing of anterior chamber pre-therapy (e.g., angle-closure glaucoma) with subsequent deepening of chamber after therapy (from dehydration of vitreous)
   3. Gonioscopy to evaluate for types of refractory glaucoma that may require short-term hyperosmotic therapy prior to surgery (e.g., traumatic glaucoma, neovascular glaucoma)

C. Thorough medical history and review of systems
IV. List the alternatives to this therapy

A. Aqueous suppressants (i.e., beta-adrenergic antagonists, topical and/or oral carbonic anhydrase inhibitors, alpha-adrenergic agonists)
B. Outflow enhancers (i.e., prostaglandin analogues, sympathomimetic agents, epinephrine-like agents)
C. Laser surgery procedures to correct acute glaucoma (e.g., iridotomy and/or iridoplasty for acute angle-closure glaucoma)
D. Paracentesis
E. Glaucoma surgical procedure (e.g., trabeculectomy, tube shunts, etc.)

V. Describe the mechanism of action

A. Reduces vitreous volume and thus IOP by creating an osmotic gradient between the blood and vitreous
B. The larger the dose and more rapid the administration, the greater the reduction in IOP (because of increased gradient)
C. Limited effectiveness and duration of action when blood-aqueous barrier is disrupted (osmotic agent enters the eye)

VI. List complications of this therapy, their prevention and management

A. Complications: worse with mannitol
   1. Headache
   2. Backache
   3. Nausea and vomiting (oral agents)
   4. Urination frequency and retention
   5. Cardiac (chest pain, pulmonary edema, congestive heart failure)
   6. Renal impairment
   7. Neurologic status (lethargy, seizures, obtundation)
   8. Subdural and subarachnoid hemorrhages
   9. Hypersensitivity reactions
   10. Hyperkalemia or ketoacidosis (when glycerin is given to patients with diabetes)

B. Prevention
   1. Consideration of alternative therapies
   2. Cautious use in patients with known compromised cardiac or renal status
   3. Avoid use of glycerin in patients with diabetes
   4. Close observation for complications

C. Management
   1. Discontinue medication
   2. Symptomatic relief of side effect until resolution if applicable
   3. Consider urinary catheter (if IV mannitol given preoperatively)

VII. Describe the follow-up care
A. Close monitoring of IOP (to determine efficacy of hyperosmotic agents)
B. Discontinuation of therapy as soon as possible
C. Close monitoring of ocular and systemic symptoms and exam

VIII. Describe appropriate patient instructions (postoperative care, vision rehabilitation)

A. Alerting physician of any complications
B. Substitution of IOP-lowering agents when hyperosmotic agents no longer needed

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
I. List the indications and contraindications

A. Indications
   1. Primary open-angle glaucoma with uncontrolled intraocular pressure (IOP)
      a. Adjuvant therapy
      b. Primary therapy
   2. Normal tension glaucoma
   3. Secondary open-angle glaucoma with uncontrolled IOP (adjuvant or primary therapy)
      a. Pseudoexfoliation glaucoma
      b. Pigmentary glaucoma
      c. Steroid-induced glaucoma
   4. Inadequacy of medical therapy
   5. Intolerance of or non-compliance with medical therapy
   6. Inadequate IOP control following incisional surgical therapy of open-angle glaucoma
   7. Ocular hypertension with uncontrolled IOP in high-risk selected cases
      a. Non-responsive to medical therapy
      b. Adjuvant therapy
   8. Selective laser trabeculoplasty (SLT) after previous laser trabeculoplasty
      a. Failed previous argon laser trabeculoplasty (ALT)
      b. Successful previous ALT or SLT
   9. Chronic angle closure provided at least 180 degrees of trabecular meshwork is visible

B. Contraindications
   1. Absolute contraindications
      a. Acute angle-closure glaucoma
      b. No visible angle structures, with or without angle-closure glaucoma
      c. Neovascular glaucoma with or without angle closure
      d. Active anterior segment inflammation
      e. Iridocorneal endothelial syndrome
   2. Relative contraindications
      a. Developmental glaucoma
      b. Failed previous SLT

II. Describe the pre-procedural evaluation

A. Gonioscopy
B. Assess trabecular pigmentation
C. Measure IOP
D. Review other treatment options with the patient
III. List the alternatives to this treatment or therapy
   A. Medication
   B. Incisional surgery

IV. Describe the instrumentation and technique
   A. Laser surgery
      1. Argon laser trabeculoplasty (ALT)
      2. Diode laser trabeculoplasty
      3. Frequency-doubled Q-switched Nd: YAG laser trabeculoplasty (SLT)
   B. Anesthesia - topical
   C. Technique
      1. First determine if the trabecular meshwork can be lasered without also striking the iris. If the angle is narrow, an iridotomy may be needed prior to the trabeculoplasty
      2. Perioperative apraclonidine or brimonidine is most commonly used to prevent post-laser surgery IOP spike. Other topical glaucoma medications (beta-adrenergic antagonists, carbonic anhydrase inhibitors, parasympathomimetic agents) can also be used
      3. Position patient in slit-lamp biomicroscope at laser
      4. Place a laser goniolens on the eye
      5. Argon/diode laser trabeculoplasty
         a. Apply 50-75 micron spot at the border of the pigmented and non-pigmented trabecular meshwork (TM)
         b. Time of 0.1 seconds
         c. Adjust laser power until a blanching of tissue occurs
         d. If a large bubble forms, reduce the power
         e. Space the burns 2-4 burn-widths apart with the goal to obtain about 50 burns per 180 degrees
         f. Treat either 180–360 degrees by rotating the lens to keep the laser aiming beam in the center of the mirror
      6. SLT
         a. Fixed spot size (400 microns)
         b. Fixed time (3 nanoseconds)
         c. Treat the TM (large spot size covers angle)
         d. Obtain about 50 burns per 180 degrees
         e. Adjust power so TM does not blanch or just at bubble formation
         f. Treat 180-360 degrees
      7. Check IOP 30-60 minutes after laser surgery
   D. Post laser anti-inflammatory medication
      1. Topical steroids for ALT
      2. For SLT, topical anti-inflammatory meds are optional

V. List the complications of the procedure, their prevention and management
   A. Elevated IOP in the immediate post-laser surgery period
      1. Prevention
a. Pre-treat with a topical glaucoma medication
b. Do not laser eyes prone to inflammation
c. Consider a smaller treatment (180 degrees) and/or less power for eyes with heavily pigmented meshwork since such eyes are more prone to post-laser IOP spikes

2. Management
   a. Topical glaucoma medication for mild IOP elevation
   b. Oral glaucoma medication such as acetazolamide 250 mg or 500 mg for substantial pressure elevation or rarely oral hyperosmotic agents or incisional surgery
   c. Topical anti-inflammatory agents

B. No reduction, or an elevation, in IOP in the late postoperative period
   1. Prevention
      a. For an elevation - do not retreat an ALT with an ALT
      b. For no response - consider initially treating 360 degrees instead of 180 degrees and consider SLT after ALT

   2. Management
      a. For an elevation - medical or surgical therapy
      b. For no response
         i. Perform additional laser trabeculoplasty, either completing a 360-degree treatment if only 180 degrees was treated previously with an argon or diode laser
         ii. Medical or surgical therapy

C. Corneal edema
D. Corneal erosion
E. Peripheral anterior synechiae (associated with ALT)
F. Hyphema

VI. Describe the follow-up care
   A. Post-laser surgery topical anti-inflammatory agent
   B. Check IOP after the laser surgery

VII. Describe appropriate patient instructions
   A. During the laser surgery
      1. Have patient look in the direction of the mirror if needed for visualization of the angle
      2. Keep head still

   B. During the immediate post-laser surgery period
      1. Use the topical anti-inflammatory agent in addition to any other glaucoma medications
      2. Resume normal activities
      3. Call for redness, pain or change in vision

Additional Resources
   1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
   3. McIlraith I, Strasfeld M, Coleg G, Hutnik CM. Selective laser trabeculoplasty as initial and adjunctive


Incisional filtering surgery for open angle glaucoma

I. List the indications/contraindications

A. Indications
   1. Presence of glaucoma with high probability of progression of glaucomatous optic neuropathy after unsuccessful laser trabeculoplasty or where laser trabeculoplasty is known not to be effective (See Laser trabeculoplasty)
   2. Elevated intraocular pressure (IOP) on maximal tolerated medical therapy (MTMT) and after laser trabeculoplasty with significant risk of glaucomatous damage or retinal vein occlusion
   3. Clinical judgment that IOP is too high (or fluctuating too much) considering patient's functional status (e.g., life expectancy, quality of life, patient age, etc.)
   4. Poor medication compliance or adherence

B. Contraindications
   1. Blind eye, rare circumstances (relative contraindication)
   2. Eye with extensive conjunctival scarring
   3. Eye with extremely thin sclera (from extensive prior surgery or necrotizing scleritis)
   4. Eye with poor visual potential (relative contraindication)
   5. Need for contact lenses for visual rehabilitation (relative contraindication)

II. Describe the pre-procedure evaluation

A. Patient medically stable for invasive ocular procedure under local anesthesia
   1. Systemic hypertension should be controlled
   2. Consider discontinuation of antiplatelet / anticoagulant medication if there are no medical contraindications

B. Preoperative evaluation
   1. Comprehensive eye examination (including gonioscopy and baseline disc and visual field studies)
   2. Reduce IOP (slowly in stages by paracentesis intraoperatively) as close to normal levels before opening eye to minimize risk of expulsive choroidal hemorrhage

C. Patient counseling
   1. Appropriate informed consent
   2. Patient education regarding benefits, risks, alternatives of procedure

III. List the alternatives to this procedure

A. Continue MTMT with continued risk of progressive vision loss

B. Laser surgery procedures
   1. Laser trabeculoplasty (SLT or ALT)
   2. Cyclophotocoagulation (trans-scleral or endoscopic)

C. Other outflow surgery procedures
   1. Glaucoma tube-shunt implants
   2. Nonpenetrating filtration surgery (e.g., viscocanalostomy, deep sclerectomy, canaloplasty)
3. Microinvasive glaucoma surgery (e.g., ab-interno goniotomy (Trabectome), trabecular micro-bypass shunt (iStent))

IV. Describe instrumentation, anesthesia, and technique

A. Knowledge of both internal and external anatomy of limbal area

B. Anesthesia

1. Peribulbar / retrobulbar anesthesia
2. Topical / intracameral / Sub-Tenons anesthesia
3. General anesthesia

C. Use of antifibrotic agents

1. Greater success and lower IOP following surgery
   a. Potential serious postoperative complications
   b. Cautious use in primary trabeculectomy in young myopic patients (increased risk of hypotony)
   c. Cautious use in eyes with thin or friable conjunctiva or after intraoperative button holes
   d. 5-Fluorouracil
      i. Pyrimidine analogue
      ii. Concentration of 50 mg/ml for up to 5 minutes
      iii. Apply intraoperatively with pledget
      iv. Postoperative subconjunctival injections
   e. Mitomycin C
      i. Antibiotic - antineoplastic compound and antimetabolite (inhibits cellular proliferation)
      ii. Concentrations used: 0.2 - 0.4 mg/ml for 1 - 5 minutes
      iii. Apply intraoperatively with pledget or subconjunctival injection

2. Risk for failure or complication
   a. Active anterior segment neovascularization (rubeosis iridis)
   b. Active iritis (uveitic glaucoma)
   c. Young, patients
   d. Myopic patients
   e. Patients of African descent
   f. Patients with aphakia
   g. Previous failure of filtering surgery
   h. Episcleral fibrosis is most common cause for filtration failure and antifibrotic agents inhibit cellular proliferation and fibrosis
   i. Chronic topical glaucoma medication
   j. Nanophthalmos
   k. Increased episcleral venous pressure

D. Trabeculectomy technique

1. Exposure with corneal traction suture or superior rectus bridle suture
2. Limbus-based or fornix-based conjunctival flap
3. Partial thickness scleral flap (shape at discretion of surgeon)
4. Move flap dissection anteriorly in eyes of short axial length to preclude cutting into ciliary body and iris root
5. Application of antimetabolite to episcleral surface for desired amount of time, if planned
6. Copious irrigation of antimetabolite from surgical field
7. Paracentesis
8. Consideration for use of Ex-PRESS device
9. Sclerectomy
10. Consider peripheral iridectomy
11. Flap closure: adequate to guard against postoperative hypotony
   a. Permanent sutures
   b. Releasable sutures
12. Check for fluid flow through fistula by reforming the anterior chamber with balanced salt solution via paracentesis
13. Possible excision of Tenon capsule
14. Closure of Tenon capsule and conjunctiva

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

A. Intraoperative / early perioperative
1. Conjunctival tear / buttonhole
2. Scleral flap complications
   a. Dehisced flap or amputated flap
   b. Thin vs. thick flap
   c. Premature entry (into ciliary body region)
3. Intraocular hemorrhage
   a. Scleral, episcleral, subconjunctival, or intra-Tenon's
   b. Iridectomy site / ciliary processes
   c. Expulsive choroidal hemorrhage
   d. May interfere with visibility during surgery and later prevent visualization of scleral flap sutures for laser suture lysis
4. Vitreous presentation
5. Sclerectomy problems
   a. Closure due to iris adhesion to internal ostium
   b. Incomplete excision of deep scleral lamellae and Descemet membrane

B. Early postoperative
1. Infection
2. Hypotony
3. Flat anterior chamber
4. Aqueous misdirection
5. Hyphema
6. Formation or acceleration of cataract
7. Transient IOP elevation
8. Choroidal effusion
9. Suprachoroidal hemorrhage
10. Persistent uveitis
11. Dellen formation
12. Loss of vision

C. Late postoperative
1. Leakage or failure of filtering bleb
2. Cataract
3. Blebitis
4. Endophthalmitis
5. Symptomatic bleb (dysesthetic bleb)
6. Bleb migration
7. Hypotony
8. Aggravation of dry eye
9. Cystoid macular edema
10. Hypotony maculopathy
11. Induced astigmatism
12. Peripheral anterior synechiae

D. Complications of anti-fibrotic agents
1. Bleb/wound leaks
2. Blebitis
3. Endophthalmitis
4. Hypotony (which can lead to maculopathy)
5. Persistent corneal epithelial defects (especially with 5-Fluorouracil)
6. Possible ciliary body ischemia (potentially with Mitomycin C)

E. Prevention and management of complications
1. Meticulous surgical technique
2. Use minimum needed dosage and duration of antifibrotic agents intraoperatively (based on surgeon judgment)
3. Close postoperative follow-up care

VI. Describe the follow-up care

A. Postoperative follow-up
1. Careful postoperative follow-up of IOP and bleb
2. Evaluation of anterior chamber depth
3. Evaluation of possible choroidal detachment
4. Postoperative complications associated with antiproliferative agents
5. Careful follow-up of other eye's glaucoma status
6. Instruction/information sheet

B. Postoperative regimen
1. Topical corticosteroids
2. Topical antibiotics
3. Eye shield at bed time
4. Topical cycloplegic agents
5. Sub-Tenon corticosteroids / systemic corticosteroids
a. Pronounced inflammation
b. Choroidal effusion / detachment
c. Associated inflammatory disease

C. **Postoperative IOP elevation**
   1. Bleb massage by the patient, or by the physician at the slit lamp.
   2. Pull releasable suture
   3. Laser suture lysis
      a. Compress conjunctiva with Zeiss goniolens or lens designed for suture lysis (Hoskins, Ritch, Mandelkorn)
      b. Apply with caution in early postoperative period after antimetabolites due to increased risk of hypotony
   4. Modulation of healing response
      a. Discontinue topical corticosteroids
      b. Aqueous suppressant to decrease flow through bleb leak and to modulate wound healing in encapsulation
      c. Subconjunctival injections of 5-fluorouracil
      d. Change in postoperative regimen
   5. Needling of a failing or encapsulated bleb with subconjunctival injection of antimetabolites

D. **Postoperative hypotony**
   1. Intracameral injection of ophthalmic viscosurgical device (OVD) (viscoelastic)
   2. Bleb compression (e.g. pressure patch, bandage contact lens, etc.)
   3. Autologous blood injection
   4. Transconjunctival scleral flap sutures
   5. Compression suture
   6. Surgical bleb revision

**VII. Describe appropriate patient instructions**

A. A written instruction sheet to patient and family on the care of a filtered eye, including what to look for and do at first signs of an infection, and what activities to avoid
B. Need for frequent postoperative visits
C. Adherence to postoperative regimen
D. Careful attention to ocular status (e.g. pain, vision, redness, etc.)
E. In postoperative period, avoid
   1. Heavy lifting
   2. Bending
   3. Straining
   4. Contact lenses
   5. Swimming underwater without goggles
   6. Deep scuba diving
   7. Limit heavy physical activity

Additional Resources
1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


Combined cataract and filtration surgery

I. List the indications

A. Indications

1. Glaucoma that is uncontrolled, either medically or after laser trabeculoplasty, when visual function is significantly impaired by a cataract
2. Cataract requiring extraction in a glaucoma patient who has advanced visual field loss where a transient increase in postoperative intraocular pressure (IOP) could cause further damage
3. Cataract requiring extraction in a glaucoma patient requiring medications to control IOP in whom medical therapy is poorly tolerated
4. Cataract requiring extraction in a glaucoma patient who requires multiple medications to control IOP

II. Describe the pre-procedure evaluation

A. Preoperative examination

1. External examination looking for signs of conjunctival or eyelid infections
2. Visual acuity, refraction, pin hole vision, glare testing and possibly Potential Acuity Meter to determine extent of visual impairment from cataract
3. Slit-lamp biomicroscopic examination
   a. To evaluate severity of cataract and to look for evidence of conditions that might lead to complications, i.e., exfoliation, phacodonesis, other signs of trauma
   b. To evaluate conditions that might make glaucoma surgery more likely to fail, i.e., uveitis, neovascularization, signs of previous surgery or scarred conjunctiva
4. Pupil exam to determine extent of dilation and relative afferent pupillary defect
5. IOP
6. Dilated fundus examination for extent of optic nerve damage and peripheral retinal status
7. Visual field examination for severity of field loss
8. Gonioscopy to evaluate angle for neovascularization and presence and position of peripheral anterior synechiae in case the need arises for placement of an anterior chamber intraocular lens (IOL)

B. Preoperative history

1. Determine severity of visual symptoms from cataract
2. Determine how well patient is tolerating glaucoma medications
3. Investigate patient's general health to determine how well they will be able to tolerate a longer combined surgery or potentially two separate procedures
4. Take a medication history and consider discontinuing certain medications preoperatively
   a. Parasympathomimetic agents
   b. Agents that inhibit clot formation, i.e., anticoagulation agents, antiplatelet drugs, Gingko biloba, Bilberry, Vitamin E, in consultation with the patient's primary care physician

III. List the alternatives to this procedure

A. Sequential cataract and glaucoma surgery
B. Combined cataract and nonpenetrating procedures
C. Combined cataract and angle surgery
D. Combined cataract and tube shunt procedures
E. Combined cataract and endoscopic ciliary body photocoagulation
F. Cataract surgery alone
G. Laser trabeculoplasty

IV. Describe the instrumentation, anesthesia and technique

A. Instrumentation (See Incisional filtering surgery for open angle glaucoma)
   1. Standard cataract extraction instrumentation (phacoemulsification preferred over extracapsular cataract extraction, when possible)
   2. Clear corneal approach preferred for maintenance of conjunctival integrity

B. Anesthesia - preference by surgeon
   1. Topical
   2. Peribulbar
   3. Retrobulbar
   4. Intracameral
   5. General (rare)

C. Technique
   1. Two site vs. single site
      a. Two-site surgery provides slightly lower (1-3 mmHg) IOP than one-site surgery
   2. Antimetabolites mitomycin C (MMC) vs. 5 Fluorouracil (FU)
      a. The preponderance of evidence suggests a small (2-4 mmHg) benefit from the use of MMC but not from 5-FU

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

A. Cataract complications
   1. Infection
   2. Suprachoroidal hemorrhage
   3. Pupillary block from pseudophakos
   4. Corneal decompensation
   5. Increased inflammation
   6. Increased risk of trabeculectomy failure
   7. Cystoid macular edema
   8. Vitreous loss, vitreous occlusion of peripheral iridotomies and filter

VI. Describe the follow up care

A. Similar to glaucoma surgery performed alone:
   1. Increased inflammation may require additional anti-inflammatory medications
   2. Increased inflammation may require additional injections of 5-FU in the perioperative period
   3. Laser suture lysis vs. releasable sutures, as needed
   4. Care needed to prevent shallow anterior chamber and IOL corneal touch
5. Important to monitor appearance of bleb for leaks or signs of bleb failure (episcleral membrane, encapsulated bleb, tight scleral flap, fistula occlusion) and address those issues as they arise

VII. Describe appropriate patient instructions (post-op care, vision rehabilitation)

A. Avoid heavy lifting, bending or straining for several weeks
B. Stress compliance with medication regimen for optimal outcome
C. Stress compliance with frequent post-operative visits
D. Underscore that visual rehabilitation will be slower than with routine cataract surgery and that it will be longer before a stable refraction is reached
E. Stress possible need for additional outpatient manipulations in the perioperative period such as laser suture lysis, releasable sutures, and repair of potential wound leaks
F. Give patient an instruction sheet on the care of a filtered eye and signs/symptoms of blebitis

Additional Resources

Laser iridotomy for angle closure

I. List the indications/contraindications

A. Indications
   1. Pupillary block
      a. Any attack of acute angle-closure glaucoma
      b. Documented history of primary angle-closure glaucoma (acute, subacute, intermittent, chronic)
      c. Anatomically narrow angle (pupil block, plateau iris) determined to be at risk for angle-closure glaucoma
      d. To widen angle prior to laser trabeculoplasty
      e. Phacomorphic glaucoma
      f. Aphakic / pseudophakic pupil block
      g. Silicone oil pupil block (inferior placement as silicone oil rises)
   2. When pupillary block cannot be ruled out
      a. Aqueous misdirection

B. Contraindications
   1. Synechial angle closure glaucoma (e.g., neovascular, iridocorneal endothelial syndrome)
   2. Corneal edema / opacification
   3. Completely flat anterior chamber due to risk of damaging cornea
   4. Chronic angle closure with 360° of peripheral anterior synechiae (PAS)

II. Describe the pre-procedure evaluation

A. Question patient regarding symptoms of subacute / acute angle closure
B. Slit-lamp biomicroscopic evaluation of anterior chamber depth
C. Gonioscopy
   1. Determine angle anatomy, grade angle, and document appearance appropriately with a recognized grading system
   2. Distinguish appositional vs. synechial closure by dynamic gonioscopy and changes in angle width with varying pupil illumination
   3. Best to perform in a darkened room with only a small beam of light illuminating the angle to prevent inadvertently opening the angle with ambient light constricting the pupil

III. List alternatives to this procedure

A. Surgical iridectomy
B. Daily cholinergic use to constrict pupil pharmacologically and pull iris away from angle (i.e., pilocarpine 1%). Cholinergic therapy actually has potential to anteriorly displace lens-iris diaphragm and increase pupillary block

IV. Describe instrumentation, anesthesia, and technique

A. Identify appropriate area on the iris for treatment
   1. Place the PI anywhere except where it might straddle the upper lid margin
2. Select iris crypt or as thin an area as possible
3. Avoid placement near any visible iris vessels

B. Consider pretreatment with argon/diode laser in thick, densely pigmented iris, blue iris with non-compact stroma, or iris prone to bleeding

C. Pretreat with cholinergic to constrict pupil and put iris on stretch

D. Perioperative apraclonidine or brimonidine most commonly used to prevent post-laser intraocular pressure (IOP) spike. Other topical glaucoma medications (beta-blockers, carbonic anhydrase inhibitors, miotics) can also be used

E. Topical anesthesia with proparacaine or tetracaine

F. Neodymium yttrium-aluminum-garnet (Nd: YAG) or argon/diode laser may be used alone or in combination
   1. Nd: YAG most commonly used
   2. Argon/diode most commonly used for pretreatment
      a. Argon/diode pre YAG PI has the effect of coagulating blood vessels and thinning/compacting iris stroma to facilitate penetration with YAG

G. Condensing contact lens (e.g., Abraham, Wise)
   1. Used to stabilize eye and provide additional magnification and energy density at treatment site.
   2. Decreases energy delivered to cornea and retina by focusing more acutely

H. Laser settings
   1. Nd: YAG - results in mechanical disruption of tissue
      a. Power settings: 1-10 mJ, 1-3 bursts/shot. Start with powers in low range
      b. Can be used for entire procedure or pretreatment with argon/diode laser may facilitate penetration in lightly pigmented iris, thick heavily pigmented iris, or iris prone to bleeding (e.g., uveitis, anticoagulants)
   2. Argon/diode laser - results in thermal vaporization of tissue
      i. Penetrating burn
      ii. Used to vaporize tissue and create an opening
      iii. Small size, short duration burns
         i) 50-micron spot size, 0.02 - 0.2 seconds, 600-1200 mW

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

A. Intraoperative / perioperative
   1. Blurred vision
      a. Usually transient; caused by methylcellulose, corneal surface irregularities, blood, pigment dispersion
   2. Hemorrhage at iridotomy site
      a. Occurs with Nd: YAG, can be minimized with argon/diode laser pretreatment to coagulate blood vessels
      b. May interfere with visibility during treatment
         i. Compress eye with iridotomy lens to raise IOP temporarily and provide tamponade to slow a steady stream of blood
         ii. May need to take a break from treatment for a few minutes to an hour to allow blood to clot and retract or bring patient back another day to complete procedure
      c. Usually self-limited; occasionally results in small hyphema. Often worse in patients on anticoagulant or anti-platelet medication
   3. Pigment dispersion
a. Occurs to some extent in every patient

b. In thick, heavily pigmented iris, may interfere with visibility during procedure
   i. May need to take a break from treatment for a few minutes to an hour to allow pigment to clear from treatment area

4. Corneal injury

b. Causes transient epithelial / stromal whitening (argon/diode) or focal stromal disruption (yttrium-aluminum-garnet)
   i. Can interfere with delivery of laser energy
   ii. Resolves in days-weeks

b. More common if AC very shallow
   i. Contraction burns with argon/diode laser can help pull iris away from cornea then penetrating burns can be more safely delivered
   ii. May need to penetrate iris more centrally to relieve pupil block and deepen AC, then return to treat a peripheral area
   iii. Laser peripheral iridoplasty, surgical iridectomy or cataract extraction may be needed if prior maneuvers do not deepen chamber

5. Corneal epithelial irregularity / abrasion

a. Can be minimized with careful application and removal of iridotomy lens, and post-treatment lubrication with artificial tears

6. IOP spike

a. Usually occurs within 1 hour of treatment

b. Minimized by perioperative use of alpha agonist or other topical glaucoma medications

B. Post-operative

1. Blurred vision due to corneal injury, blood or pigment dispersion

a. Usually resolves within 24-48 hours

2. Inflammation

a. Due to breakdown of blood-aqueous barrier

b. Alleviated by postoperative use of topical corticosteroids and/or non-steroidal anti-inflammatory drugs (NSAID)

c. Usually resolves within one to three weeks

3. Glare / visual disturbance

a. More common with large opening that is adjacent to the lid margin and either not fully covered by the lid or not fully exposed at the 3 or 9 o’clock position

b. May result from prismatic effect of tear film at lid margin or shutter effect of lid partially covering PI

c. Symptoms often lessen with time

d. With severe symptoms, may need opaque contact lens or corneal tattoo to decrease light entering eye through PI

4. Focal lens opacity

a. More common with high energy levels and iridectomy not peripheral enough

b. Due to focal disruption of lens capsule

c. Non-progressive

5. Closure of iridotomy

a. Can occur within days/weeks due to blood and pigment dispersion and require touchup once eye is quiet

b. Closure 6-12 weeks post treatment due to proliferation of pigment epithelium, more common after argon/diode PI and occurs in up to 1/3 of patients
c. Closure common in neovascular glaucoma and uveitis due to inflammatory membranes or pigment dispersion. Frequent laser PI touchup or a surgical iridectomy may be required in the situation of inflammatory glaucoma with iris bombé

VI. Describe the follow-up care
A. Topical corticosteroids and/or NSAIDs for at least 4 days post-procedure
B. Intraocular pressure check 30 to 90 minutes after treatment
C. Follow-up visit 1-3 weeks after treatment
   1. Repeat gonioscopy post-treatment to confirm less narrowing of angle
   2. Confirm patency of iridotomy
   3. Evaluate AC reaction and continue steroids if inflammation persists
   4. Dilate eyes under careful in-office observation
   5. Consider peripheral laser iridoplasty to open angles further, in plateau iris syndrome
   6. Consider cataract surgery, when indicated, for any phacomorphic component of angle closure

VII. Describe appropriate patient instructions
A. Intraoperative
   1. Keep head stationary against forehead band of slit-lamp biomicroscope
   2. Maintain steady fixation to minimize eye movement during procedure

B. Postoperative
   1. Use topical corticosteroids and/or NSAIDs for at least 4 days post treatment
      a. May continue treatment if photophobia ensues after discontinuation
   2. Office visit 1-3 weeks post laser surgery

Additional Resources
1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Laser gonioplasty or peripheral iridoplasty for angle closure

I. List the indications/contraindications

A. Indications

1. Acute angle-closure glaucoma
   a. An attack of angle-closure glaucoma that is unresponsive to medical therapy and in which corneal edema or shallow anterior chamber precludes laser iridotomy
   b. Once cornea clears, an iridotomy can be performed

2. Plateau iris syndrome
   a. Laser iridotomy performed first to eliminate any pupillary block component
   b. Residual angle-closure following laser iridectomy is not caused by pupillary block but abnormal iris angle configuration

3. Lens-related angle-closure glaucoma
   a. Phacomorphic glaucoma (lens intumescence) with secondary angle-closure where the pupillary block is not the principal mechanism
   b. Subluxation of crystalline lens
      i. Anterior lens displacement secondary to ciliary body swelling or anterior choroidal expansion after central retinal vein occlusion, inflammation, and ciliary body swelling after sulfa medications (especially topiramate)
      ii. Following panretinal photocoagulation
      iii. Following scleral buckling procedure

4. Adjunct to laser trabeculoplasty
   a. Most of angle visible but with areas of focal narrowing due to iris irregularities

5. Nanophthalmos
   a. Persistent appositional closure following successful laser iridectomy

6. Persistent angle closure after acute attack or subacute angle-closure glaucoma to open angle; may be attempted before goniosynechialysis

7. Multiple peripheral iridociliary cysts with angle closure

B. Contraindications

1. Advanced corneal edema or opacification
2. Flat anterior chamber
3. Tumors of the ciliary body or iris
4. Uveitis
5. Extensive peripheral anterior synechiae

II. Describe the pre-procedural evaluation

A. Measure intraocular pressure (IOP)
B. Gonioscopy with and without indentation
C. Anterior segment OCT or Ultrasound Biomicroscopy
D. Review other treatment options with the patient
III. List the alternatives to this treatment or therapy

A. Medication
   1. Pilocarpine to stretch and thin the peripheral iris

B. Incisional surgery
   1. Surgical iridectomy
   2. Cataract surgery

IV. Describe the instrumentation and technique

A. Laser: argon
B. Anesthesia: topical
C. Technique
   1. Apply glaucoma medication prior to treatment
   2. Anesthetize eye
   3. Apply parasympathomimetic agent (pilocarpine) to stretch the iris maximally
   4. Position patient in slit-lamp biomicroscope at laser
   5. Place either a laser iridotomy lens or laser gonioscopy mirror on the eye
   6. Activate argon laser
      a. Fixed large spot size with long duration
      b. Titrate length of burn through direct observation
      c. Reduce power if significant bubbles or popping occurs
      d. Direct aiming beam to the most peripheral portion on iris possible
      e. Contraction effect is immediate / adjust power
      f. Treatment consists of placing approximately 24 to 36 spots over 360 degrees, leaving approximately 2 spot-diameters between each spot.
      g. Glaucoma medications recommended immediately following laser procedure and the corticosteroid treatment is continued until anterior segment inflammation is resolved
   7. Record treatment
   8. Check IOP
      a. Effective treatment may produce immediate results

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

A. Elevated IOP and associated iritis
   1. Prevention
      a. Pre-treatment with parasympathomimetic to put iris on maximal stretch
         i. Require less power to produce desired effect
      b. Do not laser eyes with contraindications (see Section I.B)
   2. Management
      a. Topical corticosteroids
      b. Topical glaucoma medications
      c. Oral acetazolamide 250-500 mg
d. Rarely, emergency incisional surgery if elevation sustained and unresponsive to conservative management

B. Corneal endothelial burns

1. Prevention
   a. In an extremely shallow peripheral anterior chamber, an initial contraction burn should be placed more centrally before placing the peripheral burn; allows for peripheral deepening
   b. Use of as low a power as possible to produce iris contraction
   c. Do not continue laser past tissue shrinkage

2. Management
   a. In virtually all cases, the endothelial burns disappear within several days and have not proved to be a major complication
   b. Topical corticosteroids for associated inflammation

VI. Describe the follow-up care

A. Post-laser topical corticosteroid, such as, prednisolone acetate, 4 times a day for 4-7 days or non-steroidal anti-inflammatory drug (NSAID)
B. IOP check 30-60 minutes following procedure
C. IOP check and gonioscopy within one week depending on indication for initial gonioplasty
D. Often temporary treatment is pending definitive treatment, especially in case of lens-induced indication
E. Long-term effectiveness is possible, but patients need to be followed closely for recurrence of angle closure (especially with plateau iris syndrome) with repeat gonioscopy at routine follow-up intervals (i.e., every 4-6 month intervals)

VII. Describe appropriate patient instructions

A. During laser surgery procedure
   1. Keep head still
   2. Keep forehead against the slit-lamp biomicroscopic band
   3. Advise patient that they may experience aching sensation because of longer duration of laser application.

B. During immediate post-laser surgery period
   1. Use topical anti-inflammatory agent in addition to other glaucoma medications given
   2. Resume normal activities
   3. Keep scheduled follow-up appointment but call sooner if pain or change in vision is noticed

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
Incisional surgery for angle closure

I. List the indications/contraindications
   A. Indications
      1. Angle-closure glaucomas that are poorly controlled despite medical and laser surgery interventions
         a. Pupillary block glaucoma
         b. Chronic angle-closure glaucoma
         c. Phacomorphic glaucoma
         d. Plateau iris syndrome
   B. Relative contraindications
      1. Use caution in nanophthalmos
      2. Use caution with systemic anticoagulation or bleeding disorders
      3. Use caution with active neovascularization

II. Describe the pre-procedure evaluation
   A. Make appropriate diagnosis by performing careful anterior segment evaluation
   B. Gonioscopy must be performed
   C. Determine urgency of surgical intervention, taking level of intraocular pressure (IOP), visual function, optic
      nerve cupping, and patient discomfort into account

III. List the alternatives to this procedure
   A. Medical treatment
   B. Laser peripheral iridotomy should be attempted prior to incisional surgery in many types of angle closure
   C. Laser iridoplasty

IV. Describe the instrumentation and technique
   A. Paracentesis
      1. Performed when extremely urgent lowering of IOP is required
         a. Procedure performed at slit-lamp biomicroscope
         b. Eye is anesthetized with topical agent
         c. Sterile preparation of eye
         d. Eyelid speculum placed
         e. Micro sharp blade or 30-gauge needle on tuberculin syringe with plunger removed (aqueous will stop
            flowing spontaneously at IOP=10-15 mmHg) is used to make full-thickness incision into limbal clear
            cornea with gentle release of a small quantity of aqueous
   B. Surgical iridectomy
      1. Performed when laser iridotomy is not possible, due to cloudy cornea, iris too thick to penetrate or poor
         patient cooperation
         a. Performed in operating room
         b. Procedure performed with local anesthesia under monitored protocol
c. Prepping of eye in usual and sterile fashion
d. Clear cornea, limbal, or scleral tunnel approach to peripheral anterior chamber
e. Peripheral iris prolapsed with posterior wound pressure or grasped with forceps and withdrawn through incision
f. Small wedge of iris tissue excised with micro scissors
g. Ensure that the wound is watertight and that there is a deep anterior chamber at the end of the case

C. Goniosynechialysis
1. Performed in cases of subacute, or chronic angle closure
   a. Performed in operating room
   b. Procedure performed with local anesthesia under monitored protocol
   c. Prepping of eye in usual and sterile fashion
   d. Clear cornea, or scleral tunnel approach to peripheral anterior chamber
   e. Two opposite incisions are usually required to treat entire angle
   f. Ophthalmic viscosurgical device (OVD) (viscoelastic) necessary to ensure adequate anterior chamber depth throughout procedure
   g. Peripheral angle reached with spatula or microforceps with gonioscopic guidance to gently open angle and to lyse peripheral anterior synechiae (PAS)
   h. May be performed with cataract surgery

D. Cataract extraction
1. Indicated for phacomorphic glaucoma
   a. Performed in operating room
   b. Procedure performed with local anesthesia under monitored protocol
   c. Prepping of eye in usual and sterile fashion
   d. Consider preoperative mannitol and carbonic anhydrase inhibitors, which may help decrease vitreous volume and IOP
   e. Planned extracapsular approach/phacoemulsification with IOL implantation

E. Pars plana vitrectomy
1. Indicated for refractory aqueous misdirection
   a. Performed in operating room
   b. Procedure performed with local anesthesia under monitored protocol
   c. Preparation of eye in usual and sterile fashion
   d. Core vitrectomy performed with special emphasis on rupture of hyaloid face and creation of a unicameral eye with complete communication of posterior and anterior chambers via a patent iridectomy

F. Trabeculectomy (See Incisional filtering surgery for open angle glaucoma)
1. Indicated in cases of chronic angle-closure glaucoma with advanced optic nerve damage, persistently elevated IOP, and significant closure of angle with PAS
   a. Performed in operating room
   b. Procedure performed with local anesthesia under monitored protocol
   c. Prepping of eye in usual and sterile fashion
   d. Fornix or limbus-based approach to conjunctival dissection
   e. Procedure may be supplemented with antimetabolite use

G. Drainage device
1. Indicated in cases of refractory angle-closure glaucoma due to active neovascular process, cases of severe inflammation, a previously failed filtering procedure or previous surgery causing conjunctival scarring
a. Performed in operating room
b. Procedure performed with local anesthesia under monitored protocol
c. Prepping of eye in usual and sterile fashion
d. Consideration of valved device when urgent control of IOP is necessary

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

A. Infection
   1. Risk reduced by using sterile technique
   2. Appropriate use of post-operative topical antibiotics

B. Bleeding
   1. Minimize risk by grasping iris, and not ciliary body tissue
   2. Discontinuation of anticoagulation and antiplatelet therapy pre-operatively if there are no medical contraindications

C. Inflammation
   1. Minimize by using topical corticosteroids

D. Cataract
   1. Avoid by utilizing OVD to maintain anterior chamber depth

E. Aqueous misdirection
   1. Minimize risk by tightly suturing scleral flap and judicious use of cycloplegia post-operatively

F. Flat anterior chamber
   1. Minimize risk by tightly suturing scleral flap and judicious use of cycloplegia post-operatively

G. Suprachoroidal hemorrhage
   1. Consider IV mannitol preoperatively for IOP > 30 mm Hg
   2. Minimize risk by releasing aqueous slowly
   3. Minimize risk by tightly suturing scleral flap

H. Decompression retinopathy
   1. Minimize risk by tightly suturing scleral flap
   2. Consider IV mannitol preoperatively for IOP > 30 mm Hg if there are no medical contraindications
   3. Minimize risk by releasing aqueous slowly

VI. Describe the follow-up care

A. Postoperative follow-up
   1. Careful postoperative follow-up of IOP and bleb
   2. Evaluation of anterior chamber depth
   3. Evaluation of possible choroidal detachment
   4. Postoperative complications associated with antiproliferative agents
   5. Careful follow-up of other eye’s angle status
   6. Instruction/information sheet

B. Postoperative regimen
   1. Topical corticosteroids
   2. Topical antibiotics
3. Eye shield at bed time
4. Topical cycloplegic agents
5. Sub-Tenon corticosteroids / systemic corticosteroids
   a. Pronounced inflammation
   b. Choroidal effusion / detachment
   c. Associated inflammatory disease

C. Postoperative IOP elevation
   1. Bleb massage
   2. Pull releasable suture
   3. Laser suture lysis
      a. Compress conjunctiva with Zeiss goniolens or lens designed for suture lysis (Hoskins, Ritch, Mandelkorn)
      b. Laser lysis of suture
      c. Avoidance in early postoperative period after antimetabolites due to increased risk of hypotony
   4. Modulation of healing response
      a. Discontinue topical corticosteroids
      b. Aqueous suppressant to decrease flow through bleb leak and to modulate wound healing in encapsulation
      c. Subconjunctival injections of 5-fluorouracil
      d. Change in postoperative regimen
   5. Needling of a failing or encapsulated bleb with subconjunctival injection of antimetabolites

D. Postoperative hypotony
   a. Intracameral injection of ophthalmic viscosurgical device (OVD) (viscoelastic)
   b. Bleb compression (e.g., pressure patch, bandage contact lens, etc.)
   c. Autologous blood injection
   2. Transconjunctival scleral flap sutures
   3. Compression suture
   4. Surgical bleb revision

VII. Describe appropriate patient instructions

A. A written instruction sheet to patient and family on the care of a filtered eye, including what to look for and do at first signs of an infection, and what activities to avoid
B. Need for frequent postoperative visits
C. Adherence to postoperative regimen
D. Careful attention to ocular status (e.g. pain, vision, redness, etc.)
E. In postoperative period, avoid
   1. Heavy lifting
   2. Bending
   3. Straining
   4. Contact lenses
   5. Swimming underwater without goggles
   6. Deep scuba diving
Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016


I. List the indications/contraindications

A. Indications

1. Traditionally reserved for refractory glaucoma cases where conventional filtering surgery has failed or has a high likelihood to fail
2. The indications for shunts have been broadening
3. Examples of appropriate clinical settings follow, although timing of shunt surgery may vary amongst surgeons
   a. No prior filtration surgery
      i. Neovascular glaucoma, after adequate treatment to reduce vascular endothelial growth factor levels
      ii. Prior corneal transplant with glaucoma
      iii. Prior intraocular surgery, including cataract surgery
      iv. Chronic uveitis with glaucoma
      v. Superior conjunctival scarring
      vi. Poor candidate for filtration surgery (high risk of bleb infection)
      vii. Patients who need to wear contact lenses
      viii. Iridocorneal endothelial syndrome
      ix. Congenital glaucoma, failed angle surgery or anatomy precludes standard angle surgery
      x. Trauma
   b. Failed prior incisional glaucoma surgery

B. Relative contraindications

1. Anterior chamber placement in eyes with endothelial dysfunction or shallow anterior chamber
2. Intraocular tumors
3. Patients unable to comply with postoperative medications and follow-up

II. Describe the pre-procedure evaluation

A. Should be similar to trabeculectomy preoperative evaluation
B. Note the status of the conjunctiva
C. Note the status of the sclera at the proposed tube placement site as well as external reservoir site
D. Close examination for any vitreous which has potential to obstruct tube
E. Examination of prior records for prior procedure (e.g., scleral buckle)

III. List the alternatives to the procedure

A. Trabeculectomy with antimetabolite
B. Angle surgery
C. Cyclodestruction

IV. Describe instrumentation, anesthesia, and technique
A. **Instrumentation**

1. Present day drainage devices include
2. Molteno (nonvalved)
3. Baerveldt (nonvalved)
4. Krupin (valved)
5. Ahmed (valved)
6. Schocket (nonvalved, tube attached to encircling element)
7. Reduction in intraocular pressure (IOP) related in part to surface area of plate
   a. Some devices have double plate model available to increase surface area (i.e., Molteno, Ahmed)

B. **Anesthesia**

1. Peribulbar or retrobulbar (most commonly)
2. General
   a. Children
   b. When concern present over intraoperative cooperation

C. **Technique for posterior shunt placement**

1. Despite hardware differences, installation is similar with most
   i. Preferred placement in superotemporal quadrant
   ii. Easier access
   iii. Reduced chance of trauma to optic nerve than superior nasal placement
   iv. Reduced chance of motility disturbance or diplopia
   b. Plate sutured between vertical and horizontal rectus muscles posterior to muscle insertion (anterior plate edge 8-12 mm posterior to limbus)
   c. Tube routed anteriorly for intraocular placement
      i. Placement in anterior chamber most common
      ii. In pseudophakic eyes, may be placed in ciliary sulcus.
      iii. Can be placed via pars plana approach (following vitrectomy/lensectomy)
   d. Different methods to cover tube (patch graft) to help prevent erosion through conjunctiva or epithelial ingrowth
      i. Under host scleral flap
      ii. Donor sclera, cornea, or pericardium
2. Non-valved devices need flow restriction until encapsulation of plate occurs (timing varies by case)
   a. Suture in lumen (externalized or subconjunctival) for postoperative removal ("rip-cord" technique)
   b. Ligation of tube with absorbable (e.g. polyglactin) suture (expect release 4-6 weeks)

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

A. **Hypotony with flat anterior chamber**

1. Risk reduced by using valved device or temporary occlusion of non-valved device
2. When tube-corneal touch is present, reformation of anterior chamber with ophthalmic viscosurgical device (OVD) (viscoelastic) should be performed
   a. If persistent serous choroidal detachments present, drainage should be considered
   b. If suprachoroidal hemorrhage present with high IOP, management may vary based on clinical setting
B. **Tube-cornea touch**
   1. Avoid by placing tube parallel to the iris away from the cornea
   2. Incorporate techniques to prevent postoperative hypotony and shallow chamber
   3. In high risk cases, consider placement via pars plana (previous or concomitant vitrectomy/lensectomy)
      a. The vitrectomy must be fastidious and complete, including special attention to the vitreous cuff at the site of the tube insertion

C. **Tube occlusion**
   1. Causes
      a. Iris
         i. Best avoided with bevel-up cut on intraocular portion of tube and parallel positioning to iris
         ii. In eyes with high peripheral anterior synechiae or shallow anterior chambers, consider tube placement in ciliary sulcus
      b. Vitreous
         i. Best avoided by vitrectomy (when applicable) prior to tube placement
      c. Blood or fibrin
         i. Usually self-limiting, can be treated with intensive topical corticosteroids or intracameral tissue plasminogen activator to dissolve clot
   2. Occlusion may be cleared with neodymium yttrium-aluminum-garnet (Nd: YAG) laser surgery or surgical intervention

D. **Tube or plate exposure or erosion**
   1. Increases risk of infection or epithelial growth
   2. Best prevented with proper closure technique (i.e., reduction of tension on tissue, use of patch graft)
   3. May necessitate surgical revision or explantation of exposed hardware
   4. Exposure of patch graft through conjunctiva also may require repair

E. **Tube migration**
   1. Best prevented with meticulous surgical technique (non-occluding mattress suture under patch graft is helpful)
   2. Can migrate further into anterior chamber or subconjunctivally
      a. Tube may be repositioned or tube extension added
      b. Fasten plate securely to underlying sclera

F. **Valve malfunction**
   1. Priming (irrigation) of valve device should be performed prior to placement
   2. May require in situ flushing of tube, removal of fibrovascular ingrowth, or removal of valve mechanism

G. **Failure by excessive bleb fibrosis**
   1. Antimetabolite usage with tube surgery does not improve success rate
   2. May consider surgical revision
   3. Consider second tube

H. **Motility disturbance**
   1. Horizontal and/or vertical deviations, often in a restrictive pattern
   2. May cause persistent diplopia
   3. Management options include observation, prism, muscle surgery, or removal of the shunt

I. **Infection of external wound or endophthalmitis**
   1. Rare; usually accompanied by exposure of tube or plate
2. Intensive antibiotic treatment, surgical revision of exposure site or explantation of hardware may be necessary

VI. Describe the follow-up care

A. Antibiotics and/or corticosteroids may be given intraoperatively and are usually continued topically after surgery
B. Atropine drops may be used postoperatively
C. IOP in postoperative period can be low or high
   1. Elevated IOP
      a. Early
         i. Non-valved implants
            i) Glaucoma medications useful until tube function established. Tube fenestrations or ‘orphan’ trabeculectomy reduce risk of pressure spike
         ii. Valved implants
            i) Ensure patency of tube and presence of bleb over reservoir; reinstitute glaucoma medications
   b. Late
      i. High pressure phase is common 2-8 weeks postoperatively
      ii. Usually treated with aqueous suppression
      iii. Usually resolves spontaneously in 1-6 months

2. Hypotony
   a. Early may be due to leakage around tube entry into anterior chamber; usually transient
   b. Transient hypotony common after tube opening in non-valved implants
   c. Late overfiltration rare but may require reduction of plate size or temporary re-ligature of tube

VII. Describe appropriate patient instructions

A. Emphasize the importance of compliance with postoperative drop regimen (antibiotics, anti-inflammatory)
B. Discuss the possible need for reinstitution of glaucoma medications or additional procedures to control IOP postoperatively
C. Discuss signs and symptoms of postoperative infection and need for immediate examination if present

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.


Ciliary body ablation procedures

I. List the indications/contraindications

A. Indications

1. Refractory glaucomas
   a. Neovascular
   b. Traumatic
   c. Aphakic
   d. Advanced developmental
   e. Inflammatory
   f. Glaucoma associated with penetrating keratoplasty
   g. Silicone oil-induced
   h. Glaucoma in eyes with conjunctival scarring
   i. Failed or poorly functioning filtration surgery or tube shunt surgery

2. Glaucoma in eyes with limited visual potential

3. Urgent situations with very high intraocular pressure (IOP)

4. Pain relief in eyes with high IOP and poor visual potential

5. When incisional procedures are refused or are not possible because of medical or ocular conditions

6. Patient refuses incisional surgery and is uncontrolled on maximal medical treatment

7. Patients with high risk of intraocular complications with standard outflow procedures

B. Contraindications

1. Relatively contraindicated in eyes with good vision because of risk of loss of visual acuity

2. Ciliary body ablation should be used with caution in blind eyes because of the small risk of sympathetic ophthalmia in the contralateral eye

II. Describe the pre-procedure evaluation

A. Review of patient history, as well as, careful ocular examination to determine underlying cause of glaucoma and likelihood of success/failure with other surgical options

B. Patient counseling regarding other surgical options, benefits and risks, including

   1. Need for retreatment
   2. Visual loss
   3. Phthisis

III. List the alternatives to this procedure

A. Filtration procedures

   1. Trabeculectomy with antifibrotic agents
   2. Glaucoma tube-shunt implants

IV. Ciliary body ablative procedures
A. Laser surgery
1. Transscleral cyclophotocoagulation (CPC) with semiconductor diode laser
2. Endocyclophotocoagulation (ECP)
3. Transpupillary CPC
4. ECP via a pars plana approach

V. Describe the instrumentation, anesthesia and technique

A. Anesthesia
1. Retrobulbar / peribulbar anesthesia
2. Sub-Tenons anesthesia
3. General anesthesia

B. Techniques
1. Semiconductor diode laser
   a. Semiconductor solid state diode laser
   b. 810 nm wavelength
   c. G - Probe: Quartz fiberoptic handpiece with polished spherical tip, centers 1.2 mm posterior to the limbus and perpendicular to the iris in eyes with normal anatomy; in buphthalmic eyes, need to move further posteriorly. Use transillumination to identify ciliary body
   d. 2 - 4 second duration
   e. Initial power setting of 1750 mW, increased by 250 mW increments until a "pop" is heard, power then decreased by 250 mW, maximum power of 2500 mW
   f. Approximately 6 spots per quadrant over 270 degrees (range of 18 to 24 spots over 180 to 360 degrees)
   g. Some surgeons prefer lower power/longer duration burns: 1250mW for 4 seconds in heavily pigmented eye, 1500mW at 3.5 seconds in lightly pigmented eye.
2. Endocyclophotocoagulation (ECP)
   a. 18 or 20 ga. handpiece with three functions
      i. Pulsed continuous-wave 810 nm diode laser
         i) Power capability of 1.2 W
         ii) Focused visually using diode laser-aiming beam
      ii. 175-watt xenon light source
      iii. Video camera with 110-degree field of view
   b. Self-sealing cataract-type incision placed in peripheral cornea or limbus in phakic, pseudophakic or aphaic eyes
   c. Pars plana incision contraindicated in phakic eyes
   d. Two incisions required for treatment of 12 clock hours
   e. Iris lifted from lens/IOL with ophthalmic viscosurgical device (OVD) (viscoelastic)
   f. Probe placed through incision and video monitor used to observe procedure
   g. Power approximately 300 mW
   h. Desired endpoint - visible whitening and shrinkage of ciliary processes without tissue disruption/explosion or bubble formation
   i. Evacuation of OVD at completion of procedure
3. Transpupillary cyclophotocoagulation
   a. Rarely used
b. Argon laser applied to ciliary processes through widely dilated pupil. Clear media required
c. May be useful in presence of aniridia, traumatically enlarged pupils, or large surgical iridectomies

4. ECP via a pars plana approach
a. Argon or diode laser used in conjunction with vitrectomy
b. Requires clear media, aphakia or pseudophakia to visualize and treat ciliary processes

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

A. Intraoperative
1. General risks of anesthetic technique used
2. Conjunctival burns with transscleral diode CPC
3. Endoscopic CPC risks
   a. Damage to lens or iris
   b. Zonular rupture
   c. General risks of intraocular procedures (i.e., retinal detachment, endophthalmitis)

B. Postoperative
1. Intraocular hemorrhage
2. Prolonged ocular inflammation
3. Hypotony
4. Pain
5. Visual loss
6. Cystoid macular edema
7. Phthisis
8. Need for retreatment
9. Sympathetic ophthalmia

C. Prevention of complications
1. Reducing degrees treated in patients judged to be at risk for hypotony
2. Reducing power to avoid audible "pops" (transscleral CPC) and visual "pops" (endoscopic CPC)
3. Using lower power and longer duration burns in heavily pigmented eyes, higher power and shorter duration burns in lightly pigmented eyes
4. Transillumination helpful for identifying location of ciliary body in enlarged or otherwise anatomically abnormal eyes

VII. Describe the follow-up care

A. Postoperative topical corticosteroids, tapered as indicated clinically
B. Postoperative cycloplegics
C. Postoperative topical antibiotics (endoscopic CPC)
D. Topical glaucoma medications continued immediately following procedure and tapered based upon IOP response
E. Cycloablative procedures may make the eyes more prone to developing cystoid macular edema, especially if they are on prostaglandins - cautious use recommended
F. Retreatment if inadequate response noted by at least 1 month postoperatively

VIII. Describe appropriate patient instructions
A. Compliance with postoperative regimen

B. Report onset of severe pain

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma2015-2016.


Ocular hypotony

I. Describe the approach to establishing the diagnosis

   A. Describe the etiology of this disease

      1. Ocular trauma - ruptured globe
      2. Postsurgical
         a. Cataract surgery wound leak
         b. Penetrating keratoplasty (PK) wound leak
         c. Glaucoma surgery wound leak
         d. Pars plana vitrectomy wound leak
      3. Excess filtration after glaucoma surgery
         a. Especially problematic after the use of adjuvant antimetabolites
         b. Full-thickness procedure or poorly closed scleral flap
         c. Recent suture lysis or suture release from scleral flap
      4. Traumatic or surgical cyclodialysis cleft
      5. Ciliary body destruction
      6. Ocular inflammation from all sources with chronic aqueous underproduction with or without previous glaucoma surgery
      7. Use of glaucoma medications in an eye with previous filtering surgery
      8. Rhegmatogenous retinal detachment
      9. Ocular ischemia
     10. Choroidal effusion

   B. List the pertinent elements of the history

      1. Young patient
      2. Myopic
      3. Ocular trauma
      4. Ocular surgery
      5. Blurred (unstable or variable vision)
      6. Pain, conjunctival injection and photophobia

   C. Describe the pertinent clinical features

      1. General
         a. Blurred vision
         b. Low intraocular pressure (IOP). Care must be taken when the chamber is flat because when the lens is against the cornea the IOP may be normal or high
         c. Vertical and horizontal corneal folds
         d. Shallow anterior chamber/flat anterior chamber
         e. Retinal or chorioretinal folds in the macula
         f. Optic disc edema
         g. Cystoid macular edema (CME)
         h. Choroidal detachments/hemorrhage
Can have hypotony without maculopathy and transient hypotony in the early post-op period can be associated with a lower long-term IOP.

2. Special situation - overfiltration following glaucoma surgery
   a. Trabeculectomy
      i. Large, high or diffuse bleb if no conjunctival leak,
      ii. Low or flat bleb if conjunctival leak present
   b. Tube shunt
      i. Non-valved shunt with no temporary occlusion (can happen with valved shunts too)
      ii. Shallow chamber, often a large bleb over the reservoir

D. Describe the appropriate testing and evaluation for establishing the diagnosis
   1. Ocular ultrasonound to evaluate choroidal detachments and retinal status
   2. Computed tomography scan for trauma
   3. Optical coherence tomography to detect macular folds and CME

II. Define the risk factors
   A. Flexible, thin sclera
      1. High myopia
      2. Buphthalmos
   B. Ocular trauma
   C. Ocular surgery
   D. Ocular inflammation
   E. Ocular ischemia

III. List the differential diagnosis
   A. Exudative retinal detachment
   B. Epiretinal membrane
   C. Papilledema

IV. Describe the patient management in terms of treatment and follow-up
   A. Describe the medical therapy options
      1. Topical corticosteroids
      2. Topical cycloplegia
      3. Discontinue glaucoma medications
   B. Describe the surgical therapy options
      1. Overfiltering trabeculectomy
         a. Ophthalmic viscosurgical device (OVD) in AC
         b. Reduce anti-inflammatory drops and observe
         c. Cryotherapy to the bleb to shrink it
         d. Autologous blood/fibrin injection to promote wound healing
         e. Bleb compression sutures
f. Re-suture trabeculectomy flap
   i. May require patch (sclera, pericardium, cornea) to reconstruct a flap

g. Consider cataract extraction if visually indicated

h. Bleb excision or revision

2. Conjunctival bleb leak
   a. Reduce anti-inflammatory drops and observe
   b. Bandage contact lens
   c. Irritating antibiotic drops
   d. Consider using aqueous suppressants
   e. Resuture or advance the conjunctiva

3. Overfiltering tube shunt
   a. Reduce anti-inflammatory drops and observe
   b. Tie-off the shunt or one plate if two plates are placed
   c. Exchange shunt for a smaller one

4. Leak from a surgical wound such as cataract incision, PK incision or ruptured globe
   a. Resuture wound

5. Cyclodialysis cleft
   a. Careful gonioscopy to determine location and extent of cleft
   b. Initial treatment is with cycloplegic agents, ex. atropine
   c. Argon laser to the cleft
   d. Surgical repair with supplementary cryotherapy over the cleft
   e. Be prepared for a large, sudden, painful IOP rise postoperatively

6. Flat chamber
   a. Shallow chamber is peripheral iris - corneal touch
     i. Flat chamber is pupillary margin - cornea touch or lens - cornea touch
     ii. Reform the chamber with ophthalmic viscosurgical device (OVD)
     iii. Use a pre-existing paracentesis if possible or a 30 G needle but with extreme caution in a phakic soft eye
     iv. Helpful tip - inject a small quantity of air into AC first to avoid injecting OVD under Descemet membrane
     v. Reform the AC but do not over-inflate because high IOPs may not occur until hours after the procedure

7. Drain choroidals
   a. Make a 3-4 mm incision 4 mm posterior to and parallel or perpendicular to the limbus in the inferonasal or inferotemporal quadrant (or both inferior quadrants)
   b. Enter the suprachoroidal space and drain the fluid
   c. Simultaneously reform the eye with an AC infusion cannula
   d. Draining and reforming the eye may need to be repeated several times

8. Persistent macular folds can be addressed with vitrectomy

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

A. Overfiltration through the trabeculectomy flap
1. Use multiple flap sutures
2. Careful laser suture lysis or removal of releasable sutures in the early postoperative period to permit gradual reduction of IOP
3. Consider using a valve-incision technique
4. Consider placing OVD in the anterior chamber at the time of surgery
5. Consider placing an AC maintaining suture (10-0 Prolene on a long, straight needle) at the time of surgery in pseudophakic eyes to prevent lens - cornea touch in the event of chamber shallowing
6. Consider non-penetrating glaucoma procedures

B. Increased IOP and trabeculectomy failure after resuturing the scleral flap
1. Prevent by careful titration of tension on the suture to allow some flow
2. Use laser suture lysis or releasable sutures to manage increased IOP
3. Use more than one suture to close the flap so future suture lysis won't result in a return to hypotony

C. Scleral flap melt
1. Much more likely after using anti-fibrotic agents
2. May require harvesting autologous sclera or donor sclera, pericardium, or cornea to reconstruct the flap

D. Tube shunt-related hypotony
1. Prevention - occlude or tie off tube with a dissolving, laserable or releasable ligature to prevent early post-op hypotony
2. Prevention - use a valved shunt or a two-stage installation of a non-valved shunt
3. Prevention - make sure that the tube is filling the sclerostomy

E. Conjunctival wound leaks
1. Prevention - meticulous closure of conjunctiva
   a. Use vascular needles
   b. Consider closing limbus-based flaps with a conjunctival closure that incorporates Tenons or that is double layered
   c. Consider closing fornix-based flaps with an interrupted or running suture techniques
2. Prevention - try to avoid placing antimetabolite on the conjunctival edge

VI. Describe the disease-related complications

A. Hypotony maculopathy
B. Blurred vision
C. Bleb dysesthesia
D. Dellen formation from a large bleb
E. Large filtering blebs migrating onto the corneal surface
F. Aggravation of pre-existing dry eye

VII. Describe appropriate patient instructions

A. No heavy lifting
B. No bending over
C. Ocular protection (protective eye wear and shield at bed time)
D. Warn of signs of choroidal hemorrhage and infections if there is a bleb leak


Angle based surgery: Ab interno trabeculotomy (Trabectome), canaloplasty, trabecular micro-bypass (iStent)

I. List the indications and contraindications

A. Indications
   1. Uncontrolled open angle glaucoma
   2. Open angle glaucoma on maximal medical therapy
   3. Secondary open angle glaucomas
   4. Ocular hypertension with open angle
   5. May be performed in conjunction with cataract surgery (iStent only FDA approved to be used in conjunction with cataract surgery)

B. Contraindications
   1. Ab interno trabeculotomy
      a. Obscuration of the trabecular meshwork
         i. CACG (i.e. peripheral anterior synechia (PAS))
         ii. NVG
      b. Anticoagulation therapy (relative contraindication)
      c. Inability of patient/microscope to allow for visualization of the angle
   2. Canaloplasty
      a. Extensive scarring at the limbus
      b. Previous angle surgery (relative contraindication)
      c. Obscuration of the trabecular meshwork
         i. CACG (i.e. peripheral anterior synechia (PAS))
         ii. NVG
      d. Prior ocular surgery precluding successful 360 degrees cannulation of Schlemm canal
   3. Trabecular micro-bypass
      a. Obscuration of the trabecular meshwork
         i. CACG (i.e. peripheral anterior synechia (PAS))
         ii. NVG
      b. Anticoagulation therapy (relative contraindication)
      c. Inability of patient/microscope to allow for visualization of the angle

II. Describe the pre-procedure evaluation

A. Medication list
B. Anticoagulation status
C. Bleeding diathesis
D. Gonioscopy
E. Lens status: Pseudophakia may make angle more open
F. Conjunctival scarring (for canaloplasty)
G. Informed consent

III. List the alternatives to this procedure

1. Medical therapy
2. Trabeculectomy
3. Glaucoma drainage devices
4. Cataract surgery, if coexisting cataract and glaucoma

IV. Describe the instrumentation, anesthesia and technique

A. Ab interno trabeculotomy

1. 19.5 gauge handpiece with irrigation, aspiration and electrocautery is connected to console; foot pedal is also connected to the console
2. Footplate of handpiece is coated with a multilayer polymer
3. Surgery performed in the operating room under sterile conditions
4. Surgery usually done with patient under monitored anesthesia care (MAC)
5. Topical, intracameral, retrobulbar, and/or general anesthesia may be used
6. Temporal clear cornea incision is made parallel to the iris plane
7. Ophthalmic viscoelastic device (OVD) may be injected in to the anterior chamber.
8. Head is rotated and microscope is positioned to allow for visualization of the nasal angle, with a surgical gonioscopy lens.
9. Trabectome handpiece introduced into the anterior chamber and advanced to the nasal trabecular meshwork (TM) 180 degrees away from incision
10. Aspiration of viscoelastics by handpiece; controlled by surgeon via the foot pedal
11. Tip of foot plate engages and is inserted through TM into canal of Schlemm
12. Foot pedal is used to activate electrocautery by foot pedal. Electrocautery used to strip and ablate TM guided by handpiece in a clockwise/counterclockwise position
13. 1-5 clock hours may be treated using one incision
14. Viscoelastic removed with aspiration
15. Handpiece withdrawn
16. Suture may be used to close the wound

B. Canaloplasty

1. Performed under sterile condition in the operating room.
2. MAC anesthesia used
3. Topical, intracameral, and/or retrobulbar anesthesia may be used
4. Limbus based conjunctival peritomy is made
5. Cautery made be used to sclera as needed to achieve hemostasis
6. Partial thickness scleral flap is made (about 4 mm in length)
7. Paracentesis used to decompress the anterior chamber
8. A second scleral flap is made underneath the initial flap such that the bed should be at the level of Descemet's membrane and exposing canal of Schlemm (SC)
   a. The second flap should leave about 1 mm on both sides of the initial flap
9. Once trabecular Descemet window is present, the second flap may be amputated
10. The iTrack 250 microcatheter is inserted in SC
11. iTrack microcatheter has a red fiberoptic light, which can been seen through the sclera, to guide the insertion into SC
12. The microcatheter is guided 360 degrees
13. A polypropylene suture is tied to the catheter
14. The microcatheter is gently removed so that the ends of the Prolene suture are on the same side
   a. Healon is injected through the catheter to dilate SC as the microcatheter is being withdrawn
15. The polypropylene suture is cut and tied in an adjustable fashion to distend SC to allow for adequate tension of SC
16. An intraoperative ultrasound biomicroscopy may be used to determine the tension of the prolene sutures
17. The initial flap is secured to the sclera with sutures
18. The conjunctiva is secured to the limbus with sutures

C. Trabecular micro-bypass
1. Performed in sterile conditions in the operating room
2. MAC anesthesia used
3. Topical, intracameral, and/or retrobulbar anesthesia may be used
4. Fill anterior chamber with OVD
5. Head is rotated and microscope is positioned to allow for visualization of the nasal angle, with a surgical goniolens
6. Introduce inserter through the phaco incision
7. Approach the upper third of the trabecular meshwork at an angle of 15 degrees
8. Engage the trabecular meshwork and advance the stent into canal of Schlemm
9. Release the stent
10. Ensure the stent is properly seated
11. Withdraw the inserter and OVD

V. List the complications of the procedure, their prevention and management
A. Intraoperative
1. Ab interno trabeculotomy
   a. Inability to visualize the angle (due to PAS, bubbles from the OVD)
   b. Hyphema
   c. Iridodialysis
   d. Shallowing of the anterior chamber
   e. Excessive ablation of the TM
   f. Descemet stripping
2. Canaloplasty
   a. Conjunctival buttonhole
   b. Perforation of Descemet membrane/ entry into the anterior chamber
c. Inability to cannulate SC

d. Entry into the anterior chamber with the microcatheter

e. Inability to place polypropylene suture through SC

f. Cheese wiring sutures into the anterior chamber

g. Inadequate closure of the conjunctiva

h. Hyphema

i. Descemet detachment

3. Trabecular micro-bypass

a. Inability to visualize the angle (i.e. due to PAS, bubbles from the OVD)

b. Hyphema

c. Iridodialysis

d. Shallowing of the anterior chamber

e. Inability to place the stent

f. Inability to visualize the stent if it becomes dislodged

g. Iris plugging the device

h. Descemet stripping

B. Postoperative

1. Ab interno trabeculotomy

a. Hyphema

b. Anterior chamber inflammation

c. PAS formation

d. Closure of the cleft

e. High intraocular pressure (IOP)

f. Low IOP

g. Retention of OVD

2. Canaloplasty

a. High IOP

b. Low IOP

c. Bleb formation

d. Hyphema

e. Anterior chamber inflammation

3. Trabecular Micro-Bypass

a. Hyphema

b. Anterior chamber inflammation

c. PAS/ iris plugging the device

d. Dislocation of the stent

e. High IOP

f. Retention of OVD

C. Prevention of complications

1. Ab interno trabeculotomy

a. Adequate preoperative assessment assessing risk factors including anti-coagulation status and sickle cell status
b. Adequate visualization of the angle

c. Prevention of bubbles may be achieved with removal of OVD in the area of ablation

d. Descemet stripping and iridodialysis may be avoided by repositioning the footplate of the handpiece

e. Removal of OVD will prevent retention of OVD

f. Hyphema/ microhyphema is a very common occurrence and will usually resolve within a few days

g. Pilocarpine post operatively may help prevent PAS formation and cleft closure

2. Canaloplasty

a. Adequate preoperative assessment including conjunctival scarring, angle surgery/ laser

b. Gently lowering IOP prior to formation of secondary scleral flap may help prevent inadvertent penetration of the Descemet membrane

c. Caution when making the secondary window may be helpful in preventing inadvertent penetration of the Descemet window

d. Redirecting the microcatheter may be helpful if a blockage should occur in one direction

e. Giving adequate OVD may help appropriately dilate SC and prevent Descemet detachment

f. “Flossing” and using releasable ties may help to give adequate tension to the prolene sutures

g. Water tight closure of the first scleral flap may help prevent bleb formation

3. Trabecular micro-bypass

a. Adequate preoperative assessment assessing risk factors including anti-coagulation status and sickle cell status

b. Adequate visualization of the angle

c. Prevention of bubbles may be achieved with removal of OVD in the area of insertion

d. Descemet stripping and iridodialysis may be avoided by careful placement of the device

e. Removal of OVD will prevent retention of OVD

f. Hyphema/ microhyphema is a common occurrence and will usually resolve within a few days

g. Adequate visualization of the angle can also help prevent misplacement of the stent

D. Management of complications

1. Ab interno trabeculotomy

a. Hyphema: monitor with topical steroids consider cycloplegia, consider washout if hyphema is large or nonresolving or if IOP is elevated

b. Anterior chamber inflammation: topical steroids/ non-corticosteroidal may be helpful

c. Retention of OVD: if IOP elevated, may manage with topical medical therapy. if IOP not controlled consider washout

d. If cleft is closing or if PAS is forming, pilocarpine may be used

e. High IOP, observation, topical medical management or consider repeat Ab interno trabeculotomy surgery or other glaucoma surgery

2. Canaloplasty

a. Inability to cannulate SC in one direction consider cannulating in the other direction

b. Cheese wiring of the polypropylene suture into the AC. May remove prolene; close first scleral flap

c. High IOP: observation, topical medical management; consider goniopuncture with Nd:YAG to Descemet window to allow for flow: consider other glaucoma surgery

3. Trabecular micro-bypass

a. Hyphema: monitor with topical steroids consider cycloplegia, consider washout if hyphema is large or if IOP is elevated

b. Anterior chamber inflammation: topical steroids/ non-corticosteroidal may be helpful

c. Retention of OVD: if IOP elevated, may manage with topical medical therapy. if IOP not controlled
consider washout

d. If stent has dislodged, removal/replacement of the stent in the operating room may be needed
e. High IOP: observation, topical medical management, consider further glaucoma surgery

VI. Describe follow up care

A. Ab interno trabeculotomy
   1. Gonioscopy to evaluate cleft
   2. Evaluate for hyphema
   3. Measure IOP
   4. Monitor inflammation

B. Canaloneplasty
   1. Measure IOP
   2. Gonioscopy to evaluate prolene suture and iris to Descemet window
   3. Monitor inflammation

C. Trabecular micro-bypass
   1. Gonioscopy to evaluate stent placement
   2. Evaluate for hyphema and corneal edema
   3. Measure IOP
   4. Monitor inflammation

VII. Describe appropriate patient instruction

A. Return if any sudden change in vision or pain
B. Head of bed elevated and avoid anticoagulation, if possible, if hyphema is present

VIII. Clinical outcomes

A. Report from the American Academy of Ophthalmology reviewed the literature regarding new or emerging surgical treatment for open angle glaucoma, including canaloplasty, Trabectome, and trabecular micro-bypass. The study concluded that these modalities are less invasive than traditional glaucoma surgery and show some promise as alternative treatments

Additional Resources

2. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2015-2016.
PRACTICING OPHTHALMOLOGIST CURRICULUM, 2017-2019

GLAUCOMA