

PRACTICING OPHTHALMOLOGISTS
CURRICULUM 2014-2016

Glaucoma

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Practicing Ophthalmologists Curriculum

Authors and Financial Disclosures

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

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The Academy gratefully acknowledges the contributions of the American Glaucoma Society.

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Background on Maintenance of Certification (MOC)

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

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

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

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

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

Background on the Practicing Ophthalmologists Curriculum (POC)

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

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

Organization of the POC

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

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

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

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

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

Creation of the POC

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

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

Revision Process

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

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

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

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's Canal →
7. Collector Channels→
8. 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 ml/min
- C. 1% turnover / minute
- D. Varies diurnally and decrease during sleep
- E. Decreases with:
 - 1. Age
 - 2. Trauma
 - 3. Inflammation
 - 4. Certain systemic drugs

- a. Anesthetics
- b. Systemic hypotensive agents
- 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. $P_0 = (F/C) + P_v$**
 - 1. P_0 = IOP (mm Hg)
 - 2. F = Aqueous formation (ul /min)
 - 3. C = Facility of outflow (ul / min /mm Hg)
 - 4. P_v = Episcleral venous pressure (mm Hg)
 - 5. R (resistance to outflow) = $1/C$
 - 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**

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**
- 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, 2013-2014.

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. Flat anterior chamber

II. Pre-procedure evaluation

A. 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

B. Pachymetry to measure corneal thickness

C. Assess patient for things that might affect tonometry accuracy

1. Abdominal or thoracic obesity
2. Tight collar or necktie
3. High astigmatism
4. Breath holding or Valsalva
5. Contact lens
6. Extraocular muscles acting on restricted globe
7. Lid squeezing
8. Narrow intrapalpebral fissure

III. Alternative techniques to measure IOP

A. Goldman tonometry

1. Most widely used in clinic and research
2. Measures IOP by applanating (flattening) a 3.06mm diameter area of the cornea
3. Based on Imbert-Fick Law: internal fluid pressure (P) acting on a thin membrane sphere is equal to the force (F) needed to flatten a small area (A) of the surface $P=F/A$
 - a. Assumes the cornea is infinitely thin and is perfectly elastic
 - b. Assumes force to bend the cornea is uniform across the population and is balanced by the attractive force of the surface tension of the tears
 - c. Assumes central corneal thickness is uniform across the healthy population
 - d. Calculates IOP by flattening the cornea without accounting for its biomechanical properties, which may affect measurement accuracy
 - i. Corneal curvature: eyes with higher corneal power cause falsely higher IOP measurements
 - ii. Central corneal thickness (CCT)
 - i) A thicker normal cornea is harder to applanate and may cause falsely high IOP readings. A thinner cornea is easier to applanate and may cause falsely low IOP readings
 - ii) A thicker cornea due to edema is soft and easier to applanate and may cause falsely low readings
 - iii. Corneal material properties
 - i) Stiffness: a stiff cornea is harder to applanate than a soft cornea, regardless of its thickness (e.g. corneal scar, band keratopathy)
 - iv. Viscoelasticity

B. Perkins: handheld applanation tonometer similar to the slit-lamp mounted Goldman with a 3.06mm tip with a bi-prism

1. Helpful in obese patients and those patients in whom IOP must be measured upright away from slit lamp or supine

C. Tono-Pen: handheld tonometer that contains a strain gauge and produces an electrical signal as the tip applanates a very small area of the cornea

1. Helpful in patients with nystagmus and corneal irregularity
2. Underestimates in eyes with higher IOP and overestimates in eyes with lower IOP compared with Goldman

D. Pneumotonometer: handheld tonometer with a pressure-sensing device consisting of a gas-filled chamber covered with a silastic diaphragm

1. Helpful in patients with nystagmus and corneal irregularity
2. Readings tend to be higher than Goldman 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 appanate 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 tonometer which 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. Useful in uncooperative patients
4. May be useful for screening and home tonometry

IV. 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 until appanation occurs
 - a. Appanation 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. IOP in mmHg = 10 (grams 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

B. Perkins tonometry

1. Hand-held counterbalanced appplanation 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. Pneumotonometer

1. One drop of topical anesthetic is administered
2. Tonometer tip is touched to the central corneal until measurement 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. Despite contact with the cornea, topical anesthetic is not needed

V. Complications of the procedure, their prevention and management

A. Complication of contact tonometry (Goldmann, Perkins, Tono-Pen, Pneumotonometer, DCT)

1. Corneal abrasion
 - a. Prevent by slow careful appplanation and encouraging patient to maintain steady head and eye position
 - b. Most appplanation induced abrasions heal overnight without treatment
2. Antiseptic toxicity to the epithelium
 - a. Tonometer tip should be allow to fully dry between patients
3. Anesthetic toxicity to epithelium
4. Potential for transmission of infection

VI. Considerations in interpretation of this procedure

A. Central corneal thickness may affect IOP measurements by Goldmann, Perkins and Tono-Pen

1. Pachymetry measurements should be used to help interpret IOP measurements, but not to "convert" measurements. In general, Goldmann IOP measurements in eyes with CCT>600 should be considered falsely high, and CCT<500 falsely low
 - a. Relationship between CCT and IOP is non-linear and is inconsistent between trials comparing tonometry to intracameral pressure by manometry
 - b. There is a large variability in CCT measurements in health and disease
 - c. OHTS study indicated thinner corneas are more likely to progress to glaucoma
 - d. 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 appplanation tonometry reduces IOP readings

Additional Resources

1. AAO Basic and Clinic Science Course, Section 10. Glaucoma, 2013-2014.
2. Allingham, RR et al. Shields Textbook of Glaucoma 6th Edition 2011
3. Tonometry in Adults and Children. Eisenberg, et al. Ophthalmology 1998; 105:1173-1181
4. Positive Correlation between Tono-Pen Intraocular Pressure and Central Corneal Thickness.

Dohadwala et al. Ophthalmology 1998; 105 1849-1854

5. Intraocular Pressure difference in Goldmann Applanation tonometry vs Perkins Hand-held Applanation tonometry in Overweight Patients. Gonzaga dos Santos et al. Ophthalmology 105; 2260-2263
6. Sources of Error with Use of Goldmann-type Tonometers. Whitacre et al. Survey of Ophthalmology 1993; 38(1):1-30
7. Intraocular Pressure Measurement Precision with the Goldmann Applanation, Dynamic Contour, and Ocular Response Analyzer Tonometers. Kotecha, et al. Ophthalmology 2010; 117:730-737.

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. Angle recession
 - b. Foreign bodies
 - c. Abnormal pigmentation
 - d. Tumors
 - e. Angle neovascularization
 - f. 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., Koepe 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. Comfortable 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. Treatment of angle
 - 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

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, 2013-2014.
2. AAO, Atlas: Color Atlas of Gonioscopy, 2nd ed. 2008.
3. Robert L. Stamper, Marc F. Lieberman and Michael V. Drake, editors: Becker-Shaffer's Diagnosis and Therapy of the Glaucomas, 8th Ed. St. Louis: Mosby: 2009.
4. Ritch R, Shields MB, Krupin T, eds. The Glaucomas. 2nd Ed. St Louis: Mosby; 1996.
5. AAO, Focal Points: Gonioscopy in the Management of Glaucoma, Module #3, 2006.
6. <http://www.gonioscopy.org/> (<http://www.gonioscopy.org/>)

Examination of the optic nerve head by direct ophthalmoscopy

I. List the indications/contraindications

- A. Qualitative, clinical evaluation of the optic disc and retinal nerve fiber layer**
 - 1. Estimate size of disc (compare disc to smallest aperture size $\approx 1.5\text{mm}$)
- B. Useful to estimate media clarity**
- C. Does not provide stereoscopic view of optic disc**
- D. Provides less detail than stereoscopic techniques**

II. Describe the pre-procedure evaluation

- A. Pupillary dilation is not necessary**

III. List the alternatives to this procedure

- A. Indirect ophthalmoscope**
- B. Slit-lamp biomicroscopic techniques**
 - 1. High power concave contact (Goldmann) or noncontact (Hruby) lens
 - 2. High power convex (60, 66, 78, or 90 diopter) lens
- C. Fundus photography**
 - 1. Stereo disc photography
 - 2. Red free nerve fiber layer photography
- D. Optic nerve imaging analyzers**
- E. Viewing the optic nerve head with a direct ophthalmoscope through the center of a Koeppel lens may help in patients with small pupils, nystagmus, and involuntary lid closure**
 - 1. There must be a high + lens in the direct ophthalmoscope to visualize the disc

IV. Describe the instrumentation and technique

- A. Position patient and physician at approximately similar eye level so that both are comfortable.**
- B. Instruct patient to look at a distant target and try and continue to focus on this spot during the examination. Inform patient that it is okay to blink during the examination**
- C. Dim room lights**
- D. Ask patient to remove glasses (if pertinent)**

- E. The examiner should hold the direct ophthalmoscope with the right hand while examining the patient's right eye and with the left hand while examining the patient's left eye**
- F. Dial in the patient's refraction on the ophthalmoscope**
- G. Slowly move direct ophthalmoscope closer to patient and locate a blood vessel on fundus**
- H. Adjust power of lens in direct ophthalmoscope as needed to focus on blood vessel**
- I. Follow blood vessel to locate optic disc and evaluate (See VI-6 below)**

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

- A. Transient photopsias with prolonged examination of the fovea**

VI. Describe the considerations in interpretation for this diagnostic procedure

A. Considerations

- 1. Provides a virtual, upright image of the fundus with about 15X magnification
- 2. Inexpensive, portable, and simple to use
- 3. Advantages include its excellent optics and high magnification
- 4. Major disadvantage is the lack of stereopsis
- 5. Retinal nerve fiber layer best visualized with red-free filter
- 6. Look for signs of glaucoma
 - a. Generalized
 - i. Large optic cup vs. large disc diameter
 - ii. Asymmetry of the cups
 - iii. Progressive enlargement of the cup
 - b. Focal
 - i. Narrowing (notching) of the rim
 - ii. Vertical elongation of the cup
 - iii. Cupping to the rim margin
 - iv. Regional pallor
 - v. Splinter hemorrhage
 - vi. Nerve fiber layer loss
 - vii. Laminar dot sign
 - c. Less specific
 - i. Exposed lamina cribrosa
 - ii. Nasal displacement of vessels

- iii. Baring of circumlinear vessels
- iv. Peripapillary crescent or atrophy

VII. Describe appropriate patient instructions

- A. The patient is instructed to fixate at distant target straight ahead**
- B. The patient may have an afterimage after the examination**

Additional Resources

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

Clinical examination of the optic nerve

I. Most common methods

- A. Slit-lamp biomicroscope with handheld lens
- B. Indirect ophthalmoscope

II. List the indications/contraindications

A. Indications

1. To 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. May be used to examine the 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
- E. Instruct patient to look toward physician's ear and not straight ahead when evaluating optic nerve

IV. List the alternatives to these procedures

- A. Direct ophthalmoscopy
- B. Other slit-lamp biomicroscopic techniques
 1. High power concave contact lens (Goldmann)
 2. Noncontact high power concave lens (Hruby)
- C. Fundus photography
 1. Stereo disc photography
 2. Red free nerve fiber layer photography

D. Optic nerve imaging analyzers

V. Instrumentation and technique

A. 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 60-diopter lens, the height of the slit beam indicated on the scale reading equals the vertical disc diameter in millimeters
 - b. With a 78-diopter lens, the scale reading is multiplied by 1.1
 - c. With a 90-diopter lens, the scale reading is multiplied by 1.3
2. Considerations
 - a. The best view is through a dilated pupil, but with experience one can see the optic nerve through an undilated pupil, though usually monoscopically
 - b. Provides excellent illumination, high magnification, and a sense of contour of the optic nerve head

B. Indirect ophthalmoscopy

1. Head-mounted ophthalmoscope
 - a. +12, +20, +25, +28, or +30 diopter lens: +30 diopter lens gives the smallest
2. Considerations
 - a. Resulting view usually underestimates cupping and pallor as compared to slit-lamp biomicroscopy
 - b. Magnification is often inadequate for detecting subtle or localized optic nerve head changes
 - c. Not recommended for routine evaluation of the optic nerve head
 - d. 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/possible foveal burns 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

- b. Asymmetry of the cups in the presence of symmetrical disc diameters
 - c. Progressive thinning of the neuroretinal rim
- 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 circumlinear vessels
 - d. Peripapillary crescent or atrophy

B. Remember that the image with a 90-, 78-, 66- or 60-diopter lens is upside down and backwards

VIII. Describe appropriate patient instructions

- A. The vision will be blurry and they may see an after image**
- 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, 2013-2014.
- 2. AAO, Optic Nerve Disorders, 1996; 26-27.
- 3. AAO, Preferred Practice Pattern Guidelines, Primary Open-Angle Glaucoma. 2010.

Optic nerve head and retinal nerve fiber layer imaging

I. List the indications and contraindications

A. List the indications - documenting and quantifying the optic nerve and retinal nerve fiber layer (RNFL) to allow qualitative and quantitative evaluations over time

1. Optic nerve
 - a. Instruments to document the optic nerve
 - i. Optic nerve photos
 - i) Best taken stereoscopically
 - ii) Document the nerve for baseline purposes
 - iii) Can be quantified using special software
 - iv) Unlike other methodologies, optic nerve photos are not subject to changing technology
 - (i) Optic disc photos will always be interpretable while other scanning devices may have new versions that are not comparable to existing data sets
 - ii. Confocal scanning laser ophthalmoscopy (HRT)
 - i) Quantifies the area and volume of the neuroretinal rim using a reference plane 50 microns below the temporal surface of the retina
 - ii) Auto detects the disc boundary
 - iii) The user needs to define the nerve boundary for the Moorefield regression analysis
 - iv) Compares rim status to a normative database
 - v) Tracks changes in rim status over time
 - iii. Optical coherence tomography (OCT)
 - i) Quantifies the area and volume of the neuroretinal rim
 - ii) The optic nerve head boundary is automatically defined by where the retinal pigment epithelium ends
 2. Retinal nerve fiber layer analysis (RNFL)
 - a. Glaucoma causes thinning of the RNFL as early as 6 years before Humphrey visual field loss
 - b. RNFL thickness determinations may help in the early diagnosis of glaucoma, which is associated with RNFL thinning. RNFL thickness measurements and assist with monitoring

disease progression, which is associated with progressive RNFL thinning

- c. Areas of RNFL thinning can be correlated with visual field defects
- d. Instruments to document the RNFL
 - i. Red-free (green) light or RNFL photography
 - i) provides qualitative documentation
 - ii) Clinically, the affected glaucomatous RNFL has a decreased light reflex and will appear darker due to enhanced visualization of the retinal pigment epithelium (RPE)
 - iii) Clinically, the affected glaucomatous RNFL has a decrease in the striated texture of the RNFL, which becomes flatter or smoother in appearance
 - iv) Clinically, larger retinal blood vessels, which may normally be buried within the RNFL, may be shown in relief as the RNFL thins and exposes these blood vessels
 - ii. Scanning laser polarimetry (GDxVCC)
 - i) The GDx printout gives an "RNFL thickness" estimate that is proportional to the true RNFL thickness
 - ii) GDx does not image the optic nerve head or give optic nerve head topography
 - iii. Confocal scanning laser ophthalmoscopy (HRT)
 - i) The HRT measures RNFL thickness indirectly
 - ii) The posterior boundary of the RNFL is defined by a reference line drawn 50 microns below the surface of the retina at a point temporal to the optic nerve head, because the temporal area RNFL is thought to be last to change in thickness
 - iii) Relative changes in the RNFL thickness can theoretically be detected if the retinal surface temporal to the optic nerve head does not change from scan to scan
 - iv. Ocular Coherence Tomography (OCT)
 - i) OCT calculates RNFL thickness directly
 - ii) Some SD-OCT machines also evaluate the optic nerve head
 - iii) Evaluates macular retinal thickness
 - (i) Macular thinning may sometimes be associated with glaucoma
 - (ii) 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

B. List the 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**
 - 1. Clinically, the affected glaucomatous RNFL has a decreased light reflex and will appear darker due to enhanced visualization of the retinal pigment epithelium (RPE)
 - 2. Clinically, the affected glaucomatous RNFL has a decrease in the striated texture of the RNFL, which becomes flatter or smoother in appearance
 - 3. Clinically, larger retinal blood vessels, which may normally be buried within the RNFL, may be shown in relief as the RNFL thins and exposes these blood vessels
- 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. Stereoscopic color photograph of the nerve

- b. Can be analog or digital
 - c. Non-mydratic photography is possible, but the stereo quality is often sub-optimal so dilation is encouraged
 - d. Images can be printed as slides, Polaroid or projected onto a computer screen digitally
- 2. Confocal scanning laser ophthalmoscopy (HRT)
 - a. Performs serial cross-sections through the optic nerve head
 - b. Two-dimensional scans are then reconstructed as a three dimensional topographic map
 - c. The user then defines the edge of the disc at the disc-retina junction
 - 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. The 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
- 3. Ocular coherence tomography (TD-OCT)
 - a. This is composed of radial line scans centered over the optic nerve head
 - b. This 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 RPE ends

B. Nerve fiber layer

- 1. GDx is based on scanning laser polarimetry (SLP)
 - a. Scanning laser polarimetry is possible, because the RNFL has an orderly arrangement of axons and microtubules and, therefore, exhibits birefringence
 - b. Polarized light is directed into the eye
 - c. The GDx measures the amount of phase retardation of the polarized light as it travels through the birefringent RNFL
 - d. Phase retardation is proportional to RNFL thickness
 - e. Intrinsic corneal and macular birefringence affects RNFL thickness calculations
 - i. The GDX-VCC has a built-in variable corneal compensator that automatically subtracts the effect of the intrinsic corneal birefringence.
 - f. Each eye tested individually
- 2. OCT is based on optical coherence tomography

- a. OCT is similar to ultrasound, however it uses a super luminescent diode (SLD) light 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 RNFL thickness measurement along a circle circumscribing the optic nerve
 - ii. Plotted against a normative database
- 3. HRT is based on confocal scanning laser ophthalmoscopy
 - a. HRT best measures surface topography.
 - b. 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

C. Macular retinal thickness measurements (OCT)

- 1. This scan gives macular retinal thickness values compared to a normative database
- 2. 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.
- 3. The scan is composed of six 6mm radial line scans centered over the macula

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. Make sure that the contour line is drawn correctly. Areas of peripapillary atrophy can be tricky
4. Review the regression analysis that flags regions of the neuroretinal rim that are outside the normal database
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

1. Quality factor should be 8 or better
2. Review the "double hump" RNFL thickness graph
 - a. The GDx RNFL layer thickness values correlate to the RNFL along a circle/ellipse around the optic nerve head
 - b. The letters T, S, N, and I indicate the RNFL thickness values temporal, superior, nasal, and inferior to the optic nerve head
 - c. The "double hump" pattern exists, because the superior and inferior RNFL normally are thicker
 - d. The patient's RNFL thickness graph is drawn by a single, dark line
 - e. The patient's RNFL thickness line is plotted against a lighter-colored shaded area, which indicates the range of RNFL thickness values for 95% of the age-matched normal population
3. Review nerve fiber analysis values
 - a. Thickness values are given in microns
 - b. RNFL thickness values are given for areas superior and inferior to the optic nerve head since these areas are usually first affected in early glaucoma
 - c. "The Number" or the nerve fiber indicator (NFI) is an experimental number from 1 to 100 indicating the likelihood that glaucoma is present. Patients scoring under 30 have a low likelihood of having glaucoma and patients scoring above 70 have a high likelihood of having glaucoma

4. Correlate test results with anatomy as areas of RNFL thinning by GDx should correspond to the clinical exam and eventually to visual field loss
5. Compare with prior tests to assess for progressive glaucomatous RNFL thinning. More longitudinal studies need to be done to assess what is considered "normal" RNFL thinning due to aging alone.
6. The GDx measurements have not yet been proven to be better than a good clinical examination with stereo disc photos

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 layer thickness values are given in microns and correspond to the RNFL along a circle/ellipse 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 with the 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.

Additional Resources

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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 loss

C. Disability determination

D. Testing for motor vehicle license

II. Describe the pre-procedure evaluation

A. Ensure patient can understand and follow instructions

B. 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 appropriate demographic information

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 technology

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 or 2-degrees apart in central 10-degrees of field

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 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

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. Assess patient reliability

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

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

C. 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

D. 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

E. 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
2. Guidelines for determining progression
 - i. Reproducible changes in the visual field consisting of:
 - ii. Deepening of an existing scotoma
 - iii. Enlargement of an existing scotoma
 - iv. Development of a new scotoma
3. 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. Various algorithms used in multi-center clinical trials to assess change (Advanced Glaucoma Intervention Study, Collaborative Initial Glaucoma Treatment Study, Ocular Hypertension Treatment Study, Collaborative Normal Tension Glaucoma Study, Early Manifest Glaucoma Trial)
5. Important to repeat test to confirm progression

F. 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
 - 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, 2013-2014.
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3. AAO, Focal Points: Glaucoma and Automated Perimetry, Module #9, 1993.
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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

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. Partial coherence interferometry
- C. Slit-scan imaging
- D. Optical coherence tomography
- E. Optical low-coherence reflectometry
- F. Scheimpflug imaging

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. Goldmann applanation tonometry measurements may be more affected by CCT**
- H. Alternative methods (e.g. Tono-Pen, pneumatonometer, dynamic contour tonometer, ocular response analyzer) may be less affected by CCT**
- I. 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, 2013-2014.
- 2. Shah S, Chatterjee A, Mathai M, et al. Relationship between corneal thickness and measured intraocular pressure in a general ophthalmology clinic. *Ophthalmology* 1999;106:2154-60.
- 3. Whitacre MM, Stein RA, Hassanein K. The effect of corneal thickness on applanation tonometry. *Am J Ophthalmol* 1993;115:592-6.
- 4. Brandt JD, Beiser JA, Kass MA, Gordon MO. Central corneal thickness in the Ocular Hypertension Treatment Study (OHTS). *Ophthalmology*. 2001;108:1779-88.
- 5. Herndon LW, Weizer JS, Stinnett SS. Central corneal thickness as a risk factor for advanced glaucoma damage. *Arch Ophthalmol*. 2004 Jan;122(1):17-21.
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Primary open-angle glaucoma

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. Stresses importance of direct compression of optic nerve fibers against collagenous laminar beams of the lamina cribrosa with interruption 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 1.7%
 - c. Prevalence in African-Americans 3 times higher
 - d. Prevalence in Hispanics > 40 years old 2.0 - 4.7%
 - e. Prevalence increases with age
 - i. 5-6 times higher prevalence in 70's compared to 40's
 - f. Over 120,000 bilaterally blind in the US
 - g. 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. Refractive state
7. Medications
8. Rule out secondary causes (i.e., corticosteroids)
9. Previous eye injury and surgery

D. Describe pertinent clinical features

1. Usually insidious onset
2. Slowly progressive visual loss without symptoms
3. Painless
4. Usually bilateral, can be asymmetric
5. Central acuity 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 or Bjerrum 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 drawing and 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 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 (3.7 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)

B. Weaker evidence

- 1. Diabetes mellitus
- 2. Myopia
- 3. Migraine (for NTG)
- 4. Low diastolic perfusion pressure

III. List the differential diagnosis

A. Congenital

1. Large optic nerve head with physiologic cupping
2. Congenital disc anomalies

B. Optic nerve coloboma

C. Congenital optic nerve pit

D. Tilted disc syndrome/myopic disc

E. Anterior ischemic optic neuropathy

F. Compressive optic nerve lesions

G. 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 surgical therapy options

1. Laser trabeculoplasty
2. Trabeculectomy (with or without antimetabolites)
3. Non-penetrating filtration procedures
4. Angle surgery
5. Glaucoma drainage implants
6. 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.eyecareamerica.org/eyecare/>)
- 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, 2013-2014.
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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. Immune-related diseases may be more common
3. Possible association with systemic hypotension particularly nocturnal hypotension
4. Possible association with sleep apnea
5. 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. Imaging may also be warranted if optic nerve pallor is out of proportion to degree of cupping, visual field defects greater than expected based on amount of cupping, or unilateral progression despite equal IOP in both eyes. In such cases, medical and neurologic evaluation should be considered, including tests for anemia, heart disease, 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)

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. Consider systemic medications

1. Calcium channel blocker therapy has been advocated by some because of the possible beneficial effects of increasing capillary perfusion of the optic nerve head

D. If patient is on systemic hypotensive medications, consider recommending that the patient's internist not prescribe therapy that might lower nocturnal blood pressure

E. Recognize effects of nocturnal systemic hypotension

F. Consider laser trabeculoplasty

G. Consider filtering surgery

H. 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
3. Free glaucoma medications are available for those who cannot afford it. (further information at <http://www.eyecareamerica.org/eyecare/> (<http://www.eyecareamerica.org/eyecare/>))

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, 2013-2014.
2. Sommer A. Glaucoma: facts and fancies. *Eye* 1996;10:295-301.
3. Schuman JS, Massicotte EC, Connolly S, et al. Increased intraocular pressure and visual field defects in high pressure resistance wind instrument players. *Ophthalmology* 2000;107:127-133.

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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
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 (3.7 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)

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)**

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. Adjustment made for CCT
 - i. Tonopen or pneumotonometry should be used to measure IOP when corneal scarring is present
 - b. Diurnal IOP assessment
 - c. 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% less)
 - 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 - hypotensive lipids
 - b. Describe surgical therapy options (exceptionally rare instances)
 - c. Laser trabeculoplasty (with argon, diode, or selective laser surgery techniques)
 - d. 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

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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. There are sequence changes 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. In Scandinavian countries this accounts for approximately 50% of open-angle glaucoma (OAG)
3. Low prevalence in African-Americans
4. Actual prevalence most likely much higher than that visible on clinical examination
5. Hereditary pattern uncertain, autosomal dominant inheritance reported

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 - 3 zones
2. Deposits of XFM on pupillary margin, corneal endothelium, zonules, ciliary processes, and inferior anterior chamber angle
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
10. Large fluctuations of intraocular pressure (IOP)
11. Exfoliation syndrome with glaucoma differs from primary open angle glaucoma in that it is often monocular or asymmetric in onset

II. Define the risk factors

A. Risk of developing glaucoma cumulative over time

B. Glaucoma in exfoliation syndrome

1. IOP often higher and more difficult to control
2. Greater optic nerve and visual field damage at time of diagnosis
3. Poorer response to medications
4. More frequent need for surgical intervention
5. May present with acute IOP elevation.
6. May present unilaterally or asymmetrically

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, normal IOP - yearly examinations

B. Exfoliation syndrome without glaucoma, elevated IOP - exams every 6-12 months

C. 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

- D. Possible elevated serum homocysteine levels
- E. 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

D. Cataract surgery

- 1. Vitreous loss
 - a. Zonular dialysis
 - b. Lens dislocation
 - c. Capsular rupture
- 2. Poor pupillary dilation
 - a. Posterior synechialysis
 - b. Pupillary enlargement by retractor hooks, expansion ring or bimanual stretching
- 3. Intraocular lens decentration/subluxation
 - a. Intraoperative
 - b. Spontaneous subluxation may occur even many years after surgery
- 4. Postoperative inflammation and IOP rise
- 5. Consider early cataract surgery, while the zonules have some integrity and the lens isn't too hard

VI. Describe disease-related complications

- A. Exfoliative material has been found elsewhere in the body
- B. 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

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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 30%-50% of PDS
4. Accounts for 1-1.5% of glaucoma in the western world
5. Autosomal dominant inheritance with incomplete penetrance

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

- b. Anterior zonule can be covered with pigment particles
- 5. Gonioscopy
 - a. Angle recess very wide, usually approx. 45-degrees
 - b. Heavy dark homogeneous pigmentation of TM
 - c. May have pigmentation on or anterior to Schwalbe's line, heaviest inferiorly
- 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 20-45 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

- 1. Present in approximately 90% of cases
- 2. Aggravated by deeper AC, especially with increasing degree of myopia
- 3. The greater the degree of myopia, the earlier the onset of glaucoma
- 4. Patients with asymmetric PDS often have asymmetric AC depth

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 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. Trabeculectomy
 - b. Aqueous shunt
 - c. Angle surgery
 - d. Non-penetrating procedures

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**

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Lens-induced glaucoma

I. Describe the approach to establishing the diagnosis

A. List the pertinent elements of the history

1. Phacolytic
 - a. Phakic status
 - b. Elderly, long-standing poor vision, sudden onset of pain, conjunctival hyperemia
2. Lens particle
 - a. Following disruption of lens integrity
 - i. Surgical: following cataract extraction (CE) or Nd:YAG
 - ii. Non-surgical: following lens injury
 - iii. Usually within weeks but may be months or years later
3. Phacoanaphylaxis
 - a. Following CE with or without vitreous loss
 - b. Following CE in one eye, with subsequent CE or leaking hypermature cataract in other involved eye
 - c. After traumatic or spontaneous rupture of lens capsule
4. Ectopia lentis
 - a. Visual disturbances due to lens subluxation
 - b. Family or personal history
 - i. Simple ectopia lentis
 - ii. Ectopia lentis et pupillae
 - iii. Marfan Syndrome
 - iv. Homocystinuria
 - i) Thrombotic vascular occlusions
 - ii) Mental retardation
 - v. Weill-Marchesani syndrome
 - c. Trauma
 - d. Prior ocular surgery
 - e. Pseudoexfoliation syndrome: (See Exfoliation syndrome (Pseudoexfoliation))
5. Phacomorphic
 - a. Elderly, long-standing decreased vision

- b. Acute or subacute onset

B. Define the relevant aspects of epidemiology of the disease

1. Phacolytic
 - a. Secondary to leakage of high molecular weight lens protein, usually in eyes with mature or hypermature cataract
 - b. Sudden onset of open-angle glaucoma
 - c. Usually monocular, not reported in children, rare in young adulthood
 - d. These patients may have long-standing poor vision in the affected eye
2. Lens particle
 - a. Occurs when lens cortex material obstructs the trabecular meshwork
 - b. Often delay of days to weeks between trauma and onset of glaucoma
 - c. Similar form may occur years after CE with lens material freed into anterior chamber
 - d. Can occur in children and adults
 - e. Can occur immediately after Nd:YAG capsulotomy
3. Phacoanaphylaxis
 - a. Uncommon granulomatous inflammation in response to liberated lens material
 - b. Rarely causes glaucoma, more likely to be associated with hypotony
 - c. Almost always uniocular, although some reports of sympathizing reaction
4. Ectopia lentis
 - a. Autosomal dominant inheritance
 - i. Simple ectopia lentis
 - ii. Weill-Marchesani syndrome
 - iii. Marfan syndrome
 - b. Autosomal recessive inheritance
 - i. Ectopia lentis et pupillae
 - ii. Weill-Marchesani syndrome
 - iii. Homocystinuria
 - c. Early age of onset in inherited conditions
5. Phacomorphic
 - a. Older age
 - b. Multifactorial mechanism
 - c. Pupillary block often contributory
 - d. Intumescent lens narrows angle, causing closure

C. Describe pertinent clinical features

1. Phacolytic
 - a. Open angle
 - b. Intact capsule
 - c. Elevated intraocular pressure (IOP) (30-50 mmHg not uncommon)
 - d. Microcystic corneal edema
 - e. Cell and flare without keratic precipitates (KP)
 - f. Cellular debris or hypopyon in anterior chamber (AC)
 - g. White particles (clumps of lens protein) may be in AC or on lens capsule
 - h. Mature, hypermature or morgagnian cataract (often with wrinkled anterior lens capsule)
 - i. Microscopic findings of an AC tap: macrophages containing lens material (phase microscopy)
2. Lens particle
 - a. Open-angle
 - b. Open capsule
 - c. Free cortical material in AC
 - d. Elevated IOP
 - e. Moderate AC reaction
 - f. Microcystic corneal edema
 - g. Late: development of posterior and peripheral anterior synechiae
3. Phacoanaphylaxis
 - a. Variable clinical presentation
 - b. Most common clinical picture:
 - i. Open capsule
 - ii. Moderate AC reaction with KP on corneal endothelium and anterior lens surface
 - iii. Low grade vitritis
 - iv. Peripheral anterior and/or posterior synechiae
4. Ectopia lentis
 - a. Lens subluxated or completely dislocated (into vitreous cavity or AC)
 - b. Body habitus
 - i. Tall, slender with joint laxity and arachnodactyly (Marfan syndrome and homocystinuria)
 - ii. Short, stocky with brachydactyly (Weill-Marchesani syndrome)

- c. Variable refractive error
- d. Unilateral lens displacement in trauma
- e. Bilateral lens displacement in inherited conditions
 - i. Superotemporal in Marfan syndrome and simple ectopia lentis
 - ii. Inferotemporal in homocystinuria
- f. Small, round lens (microspherophakia) in Weill-Marchesani syndrome
 - i. Consider microspherophakia in high myopes with pupillary block
- g. Trauma-related ocular injuries
- h. Phacodonesis
- i. Shallow AC
- j. Iris bombé configuration
- k. Enlarged globe with long axial length (Marfan syndrome)
- l. Vitreous prolapse
- m. Thin sclera (Marfan, Ehlers-Danlos syndrome)
- n. Retinal detachment
- 5. Phacomorphic
 - a. Advanced cataract
 - b. Shallow AC
 - c. Angle remains narrow despite patent iridotomy

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. Phacoanaphylaxis
 - a. Diagnosis often not made until pathologic examination of enucleated eye
 - b. 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. Phacoanaphylaxis

- 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. Phacoanaphylaxis: 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. Phacoanaphylaxis

- 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. Phacoanaphylaxis

1. Submit lens material for pathology to verify diagnosis
2. If cataract in contralateral eye, consider intracapsular cataract surgery to decrease risk of immunologic reaction from residual lens protein
3. Complications associated with medications
4. 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. Phacoanaphylaxis

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, possible need for diagnostic AC tap**
- 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, 2013-2014.
2. Ritch R, Shields MB, Krupin T, eds. *The Glaucomas*. 2nd ed. St Louis: Mosby; 1996.

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 (thyrotropic ophthalmopathy)
 - 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. Selective carotid angiography
 - a. Diagnostic test of choice when one is considering carotid cavernous fistula
 - b. Helps confirm diagnosis
 - c. Provides therapeutic capability
3. Computed tomography (CT) scan of orbit
 - a. Contrast CT scan of orbit helps establish diagnosis
 - b. May demonstrate proptosis, swelling of extraocular muscles, and dilation of superior

ophthalmic vein

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 may be attempted, but is unlikely to be effective when outflow facility is normal
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, 2013-2014.
2. Yang CB, Freedman SF, Myers JS. Use of latanoprost in the treatment of glaucoma associated with Sturge-Weber syndrome. *Am J Ophthalmol*. 1998;126:600-2.
3. Weinreb RN, Karwatowski WSS. Glaucoma associated with elevated episcleral venous pressure. In: Ritch R, Shields BM, Krupin T, eds. *Glaucomas*. 2nd edn. St. Louis: Mosby, 1996:1143-55.
4. Rhee DJ, Gupta M, Moncavage MB, et al. Idiopathic elevated episcleral venous pressure and open-angle glaucoma. *Br J Ophthalmol* 2009;93:231-234

Hyphema

I. Describe the approach to establishing the diagnosis

- A. Note: The primary topic addressed here is traumatic hyphema. Other outlines, such as Fuchs heterochromic iridocyclitis, address hyphemas due to other etiologies in greater detail**
- B. Describe the etiology of this disease: Tear in the iris or ciliary body, causing bleeding from small branches of the major arterial circle**
 - 1. Trauma - blunt or lacerating
 - 2. Intraocular surgery (laser and incisional)
 - a. Especially associated with warfarin, aspirin
- C. 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
- D. 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
- E. 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 clot, inflammatory cells, erythrocytic debris
 - ii. Pupillary block secondary to clot occluding pupil
 - 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 after traumatic hyphema)
- 4. Blood in anterior chamber:
 - a. Circulating red blood cells (RBCs)
 - b. Layered hyphema
 - c. Eight-ball hyphema (clotted total hyphema)

F. Describe appropriate testing and evaluation for establishing the diagnosis

- 1. Sick 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
 - i. Vascular tufts of iris
 - ii. Juvenile xanthogranuloma
 - iii. Fuchs heterochromic iridocyclitis

- iv. Blood dyscrasias
- v. Uveitis-glaucoma-hyphema (UGH) syndrome
- vi. 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 hemoglobinopathy
 - 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. Sickle cell patients: if IOP averages 25 mmHg or greater in any 24 hour period
 - ii. Other patients: IOP remains 35-50 mmHg for 3-5 days
 - iii. Patients with pre-existing glaucoma may require earlier surgical intervention
 - b. Risk of corneal blood staining
 - i. 4 days after onset of total hyphema
 - ii. 6 or more days with IOP greater than 25mmHg with a 50% or greater hyphema
 - c. Risk of synechiae formation

- i. Greater than 50% hyphema for 8 or more days
 - 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. 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 (erythrocyte/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. Initial approach
 - i. Medical treatment with aqueous suppressants
 - c. Irrigation of AC, pars plana vitrectomy and/or trabeculectomy if medical therapy fails

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, 2013-2014.
2. Walton W, Von Hagen S, Grigorian R, Zarbin M. Management of Traumatic Hyphema. *Surv Ophthalmol.* 2002; 47:297-334.
3. Spraul C, Grossniklaus H. Vitreous Hemorrhage. *Surv Ophthalmol.* 1997; 42:3-37.
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6. Allingham, RR et al. Glaucomas Associated with Intraocular Hemorrhage. In: Allingham RR, *Shields Textbook of Glaucoma*, 6th Edition. Lippincott, Williams and Wilkins. Philadelphia, 2011. Pg 350-357.

Angle recession glaucoma

I. Describe the approach to establishing the diagnosis

A. List the pertinent elements of the history

1. History of blunt ocular trauma usually with hyphema (See Hyphema)
2. Traumatic event may occur months to years 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 occurs in 7-9% of those with angle recession following trauma
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 blunt trauma to anterior segment (e.g., 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 blunt 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**
- F. Unilateral chronic open angle glaucoma**
- G. Iridocorneal endothelial syndromes (Chandler syndrome particularly)**
- H. Posner-Schlossman syndrome**
- I. Contusion angle deformities (i.e., cyclodialysis cleft which may look similar but has a different clinical course)**
- J. Normal anomalous appearing angles**

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 (See Alpha adrenergic agonists) (See Prostaglandin analogues) (See 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, 2013-2014.
2. LaBorwit SE. Ocular trauma, hemorrhage and glaucoma. In: Morrison JC, Pollack IP, eds. Glaucoma: Science and Practice. New York, NY: Thieme; 2003:255-257.
3. Ritch R, Shields MB, Krupin T, eds. The Glaucomas. 2nd ed. St Louis Mosby: 1996; 1266.

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
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. Goldman applanation and other contact tonometers may not always give similar IOP readings
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. Describe 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 chronic use

B. Describe surgical therapy options

1. Laser trabeculoplasty
2. Surgical therapy needed in 2% to 24% of 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

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

A. Complications of treatment

- 1. Medical
 - a. Allergic reactions
 - b. 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. Possibly due to cell loss from a tube shunt in the AC, although this also is seen on a delayed basis even with the tube in the pars plana
 - b. Trabeculectomy failure

B. Prevention of complications

- 1. Oversized corneal grafts
- 2. Careful graft suturing
- 3. Consider surgical iridectomy
- 4. Consider surgical 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, 2013-2014.
2. Lee RK, Fantes F. Surgical management of patients with combined glaucoma and corneal transplant surgery. *Curr Opin Ophthalmol* 2003;14(2):95-9.
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16. Sihota R, Srinivasan G, Gupta V. Ab-externo cyclodialysis enhanced trabeculectomy for intractable post-penetrating keratoplasty glaucoma. *Eye*. 2010;24:976-9.

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**
- B. High myopes, patients with diabetes, and patients with connective tissue diseases have a predisposition to corticosteroid-induced glaucoma**

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)**

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

A. Describe medical therapy options

1. Discontinuation of the corticosteroid is the first treatment option and is often all that is required
2. 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, 2013-2014.
2. AAO, Ophthalmology Monographs 13. Glaucoma Medical Therapy: Principles and Management, 1999.
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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 ache
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
 - 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. Neutralize 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. Intermittent 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. Alpha2-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
 - 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. Laser peripheral iridotomy - neodymium yttrium-aluminum-garnet (Nd:YAG) and/or argon
2. Prophylactic iridotomy in fellow eye
3. Incisional iridectomy when laser iridotomy is not possible
4. Primary filtering surgery may be required if extensive synechial closure is present
5. Anterior chamber paracentesis may be helpful acutely to reduce IOP, relieve pain and clear cornea for later iridotomy

C. Follow-up

1. Gonioscopy following iridotomy to assess status of angle regarding synechial closure. Consider dilation challenge
2. Laser iridoplasty (gonioplasty) as indicated by angle configuration, eg. 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 laser iridotomy

1. IOP rise and inflammation should be treated with topical medications
2. Dysphotopsia (monocular diplopia and glare)

B. Posterior synechiae

1. Topical corticosteroids may prevent synechiae formation

C. Parasympathomimetics, especially strong agents, may increase pupillary block

D. Peripheral anterior synechiae

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

E. Complications from hyperosmotic use

1. Congestive heart failure
2. Tachycardia

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 ghost images or glare 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 not 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, 2013-2014.
- 2. AAO, Preferred Practice Patterns Committee, Glaucoma Panel: Primary Angle Closure Preferred Practice Pattern, 2010.
- 3. Ritch, R., M.B.Shields, T. Krupin, eds. *The Glaucomas*.
- 4. AAO, Glaucoma Medical Therapy: Principles and Management, 1999;195-198.
- 5. Lam DS, Lai JS, Tham CC, et al. Argon laser peripheral iridoplasty versus conventional systemic medical therapy in treatment of acute primary angle closure glaucoma: a prospective, randomized, controlled trial. *Ophthalmology* 2002;109:1591-6.
- 6. Yip JL, Foster PJ. Ethnic differences in primary angle-closure glaucoma. *Curr Opin Ophthalmol* 2006;17:175-80.
- 7. Salmon JF, Martell R. The role of ethnicity in primary angle-closure glaucoma. *S Afr Med J*

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Subacute angle-closure glaucoma

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 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 described as migraines or sinus
6. Frequently there are no identifiable symptoms
1. 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 synechia (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 testing
 - a. Damage varies according to the severity, frequency, and duration of the attacks
2. Consider ultrasound
 - a. Axial length, anterior chamber depth and lens thickness.
3. Consider 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
- 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

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
2. Ritch R, Lowe RF. Angle-closure glaucoma: clinical types. In: Ritch R, Shields MB, Krupin T, eds. *The Glaucomas*. 2nd ed. St. Louis, MO: Mosby; 1996: 827-829.
3. Sihota R, Mohan S, Dada T, et al. An evaluation of the darkroom prone provocative test in family members of primary angle closure glaucoma patients. *Eye* 2007; 21, 984-989.
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7. AAO, *Glaucoma Medical Therapy: Principles and Management*, 1999; 193-4,196.

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 midtrabecular 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
 - f. PAS "creep" anteriorly

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

1. Race - frequency is highest among Inuits and Asian ethnic groups (Chinese Asian, Southeast Asian, and Asian Indians)
2. Sex - higher prevalence in women
3. Refraction - typically associated with hyperopia
4. "Creeping" angle-closure is relatively uncommon in Caucasians, but much more prevalent in Asians
5. Patients of African descent with angle closure tend to have creeping 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 head 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. Define the risk factors

- A. Hyperopia
- B. Family history of angle-closure glaucoma
- C. Inuit or Asian ethnic groups
- D. Older age
- E. Female gender

III. List the differential diagnosis

- A. Neovascular glaucoma
- B. Iridocorneal endothelial syndrome (particularly Chandler syndrome)
- C. Inflammatory glaucoma
- D. Traumatic glaucoma
- E. Phacomorphic glaucoma
- F. Intermittent primary angle closure
- G. Ciliary body swelling or cysts
- 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, topiramate
- L. Retinopathy of prematurity
- M. Plateau iris
- N. Congenital glaucoma

IV. 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 aggravating pupillary block

2. Beta-adrenergic antagonists
3. Alpha-adrenergic agonists
4. Carbonic anhydrase inhibitors
5. Prostaglandin analogues

B. Describe the 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. Surgical filtering procedures
4. Peripheral laser iridoplasty
5. Goniosynechialysis

C. Cataract extraction may be indicated in selected cases and possibly could be more effective than laser or surgical peripheral iridotomy in preventing further extension of PAS

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

VI. 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

VII. 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, 2013-2014.
2. Ritch R, Lowe RF. Angle-closure glaucoma: Clinical types. In: Ritch R, Shields MB, Krupin T, eds. *The Glaucomas*, 2nd ed. St. Louis, MO: 1996; 825-827.
3. AAO, Ophthalmology Monographs 13. Glaucoma Medical Therapy: Principles and Management, 1999.
4. Aung T, Lim MC, Chan YH, and the EXACT Study Group. Configuration of the drainage angle, intraocular pressure, and optic disc cupping in subjects with chronic angle-closure glaucoma. *Ophthalmology* 2005;112:28-32.
5. AAO, Focal Points: Gonioscopy in the Management of Glaucoma, Module #3, 2006.
6. AAO, Focal Points: Angle Closure Glaucoma Update, Module #6, 2009.
7. AAO, Preferred Practice Pattern Guidelines. Primary Angle Closure Glaucoma, 2010.

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 closures

B. Define the relevant aspects of epidemiology

1. More commonly seen in younger patients (age 40 years or younger) 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
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 observation (with indentation gonioscopy)
- 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. A PI is performed to remove any component of pupillary block. 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. 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
- 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
- D. Describe surgical therapy options
 - 1. Do laser PI first to remove any pupillary block component
 - 2. Laser peripheral iridoplasty
 - 3. Glaucoma filtering surgery

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 with 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, 2013-2014.
2. Ritch R, Lowe RF. Angle-closure glaucoma: clinical types. In: Ritch R, Shields MB, Krupin T, eds. The Glaucomas. 2nd ed. St. Louis, MO: Mosby; 1996: 827-829.
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4. Ritch R, Chang BM, Liebmann JM. Angle closure in younger patients. Ophthalmology 2003; 110:

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5. Ritch R, Tham CC, Lam DS. Long-term success of argon laser peripheral iridoplasty in the management of plateau iris syndrome. *Ophthalmology* 2004; 111: 104-8.
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Secondary angle-closure glaucoma

I. Describe the approach to establishing the diagnosis

A. Describe the etiology 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
 - ii. Lens related disorders e.g. subluxed crystalline lens, spherophakia, and malpositioned intraocular lenses (IOLs)
 - b. Retinal conditions following
 - i. Central retinal vein occlusion with ciliary body edema
 - ii. Panretinal photocoagulation 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 mechanism 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 glaucoma
- v. 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
- vi. Other
 - i) Synechial 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. Describe the relative aspects of epidemiology of this disease

1. Occurs most often in association with anterior segment neovascularization
2. Can occur following glaucoma filtering and shunt procedures or following other intraocular surgeries. The mechanism may include wound leak, anterior displacement of the lens-iris diaphragm from ciliary body rotation/swelling or placement of intraocular gas/oil.
3. Secondary pupillary block from uveitis may be a common mechanism
4. May occur as a result of serous or hemorrhagic ciliochoroidal detachment following inflammation or trauma.

C. List the pertinent elements of the history

1. History of ocular surgery and/or trauma
2. Decreased vision
3. Sudden onset of pain (associated with suprachoroidal hemorrhage)
4. History of Valsalva maneuver
5. History of uveitis
6. History of diabetes, central retinal vein occlusion, carotid occlusive disease
7. Medication history

D. 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 synechial closure or neovascularization
7. Posterior synechiae 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

E. 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 (anterior segment exam) 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. Ciliary body tumor
 - a. Fine-needle aspiration biopsy
 - b. Ultrasound (See Section I.E.3)
6. Specular microscopy to diagnose corneal involvement in ICE syndrome

II. Define the risk factors

- A. Eye with markedly elevated preoperative IOP for glaucoma surgery**
- B. Diabetes, retinal vascular occlusion, carotid occlusive disease**
- C. Eyes with history of previous trauma**
- D. Eye with acute or chronic angle closure**
- E. High myopia**

- F. Filtration surgery with antimetabolites**
- G. Systemic hypertension**

III. List the differential diagnosis

- A. Ciliary block (malignant) glaucoma**
- B. Pupillary block glaucoma**

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) (See Alpha adrenergic agonists) (See 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, 2013-2014.

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7. Stamper RL, Lieberman MF, Drake MV, eds. *Becker-Shaffer's Diagnosis and Therapy of the Glaucomas*. 8th ed. St Louis: Mosby; 2009.
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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
- 5. Carotid ultrasound if indicated

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. Retrobulbar injections
3. Cyclodestruction
 - a. Laser surgery
 - b. Cryotherapy
4. Enucleation/evisceration

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

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5. Simha A, Braganza A, Abraham L, Samuel P, Lindsley K. Anti-vascular endothelial growth factor for neovascular glaucoma. Cochrane Database of Systematic Reviews 2009, Issue 3. Art. No.: CD007920. DOI: 10.1002/14651858.CD007920

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. Unusually shaped pupil (corectopia and iris holes)
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
3. Glaucoma reported in 50% to 75% of cases

4. Three forms comprise the spectrum of disease
 - a. Essential iris atrophy
 - i. More iris change than corneal change
 - ii. Iris atrophy
 - iii. Iris heterochromia
 - iv. Pupil distortion
 - v. Ectropion uveae
 - vi. Iris hole formation
 - 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. Posterior polymorphous dystrophy**
- B. Axenfeld-Rieger syndrome**
- C. Neovascular glaucoma (another cause of secondary angle-closure glaucoma with an anterior pulling mechanism)**

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
- B. Describe surgical therapy options**
 1. 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:YAG)
2. Tube shunt

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, 2013-2014.
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3. Hirst LW, Bancroft J, Yamauchi K, Green WR. Immunohistochemical pathology of the corneal endothelium in iridocorneal endothelial syndrome. Invest Ophthalmol Vis Sci 1995;36:820-7.
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13. Anderson NJ, Badawi DY, Grossniklaus HE, Stulting RD. Posterior polymorphous membranous dystrophy with overlapping features of iridocorneal endothelial syndrome. *Arch Ophthalmol* 2001;119:624-5.
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17. Doe EA, Budenz DL, Gedde SJ, Imami NR. Long-term surgical outcomes of patients with glaucoma secondary to the iridocorneal endothelial syndrome. *Ophthalmology* 2001;108:1789-95.
18. Kim DK, Aslanides IM, Schmidt CM, Jr., Spaeth GL, Wilson RP, Augsburger JJ. Long-term outcome of aqueous shunt surgery in ten patients with iridocorneal endothelial syndrome. *Ophthalmology* 1999;106:1030-4.

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
 - 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)
- 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 cases
- v. Possible etiologic role of herpes simplex virus
- vi. 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 and loss of normal iris architecture
 - 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

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. Glaucoma incidence varies and may be up to 60%
 - 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 anterior chamber 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 must start with this
 - a. History
 - i. Ocular history
 - ii. Medical history
 - iii. Social history
 - iv. Family history
 - b. Review of systems - thorough
 - c. Thorough ophthalmologic examination
 - i. Must include every part of the exam -avoid commonly missed areas such as:
 - 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 - do not miss toxoplasmosis or necrotizing herpetic retinitis - 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. Can be obtained in office or in OR at start of glaucoma procedure

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
 - 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

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, 2013-2014.
2. Shields MB. *Textbook of Glaucoma*. 4th edition. Baltimore: Williams & Wilkins; 1997.
3. AAO, Glaucoma Medical Therapy: Principles and Management, 1999; 204-205.
4. AAO, Focal Points: Management Glaucoma Secondary to Uveitis, Module #5, 1995.
5. AAO, Focal Points: Evaluation and management of elevated IOP in patients with uveitis, Module #6, 2010.

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 ocular surgery in patients with a history of angle closure or peripheral anterior synechiae
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
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 shallow anterior chamber is observed in the context of normal or elevated IOP
 - b. IOP elevation is variable, especially early in the syndrome
 - i. Clinically, misdirection can be difficult to differentiate from anterior [annular] choroidal detachment - 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. 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

E. Describe appropriate testing and evaluation for establishing the diagnosis

1. Ultrasound
 - a. B-scan to rule out ciliary body effusion or suprachoroidal hemorrhage
 - b. 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. Treat aqueous misdirection
 - a. Intensive cycloplegic therapy
 - b. Stop/avoid parasympathomimetic agents (misdirection may be aggravated by parasympathomimetic therapy)
2. 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
3. Contraindicated medications for IOP control
 - a. Parasympathomimetic agents

- i. May exacerbate inflammation
 - ii. May aggravate aqueous misdirection
- 4. 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

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. 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

Additional Resources

1. AAO, Basic and Clinical Science Course. Section 10: Glaucoma, 2013-2014.
2. Shields MB. Textbook of Glaucoma. 4th edition. Baltimore: Williams & Wilkins; 1998.
3. AAO, Focal Points: Ciliary Block Glaucoma, Module #3, 1999.
4. Ruben S, Tsai J, Hitchings R. Malignant glaucoma and its management. *Br J Ophthalmol*. 1997;81:163-167.
5. AAO, Focal Points: Diagnosis and Management of Complications of Glaucoma Filtering Surgery, Module #3, 1994.
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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. Buphthalmos
9. Epiphora

II. Define the risk factors

A. Family history of primary congenital glaucoma

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)

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
4. Be alert for apnea and hypotension with beta-adrenergic antagonists
5. Brimonidine is contraindicated in children under 3 years of age
6. Assess for possible acidosis, hypokalemia and feeding problems with carbonic anhydrase inhibitors

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
3. Aqueous shunt surgery
4. Cycloablation

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

B. Goniotomy

1. Mild to moderate hyphema, common
2. Total or "eight-ball" hyphema (rare)
3. Iridodialysis
4. Cyclodialysis
5. Peripheral anterior synechia (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. Trophic, marginal corneal ulcer
7. Subconjunctival iris prolapse
8. Descemet membrane stripping
9. Iridodialysis
10. Creation of false passage into anterior chamber
11. Creation of false passage into suprachoroidal space (producing cyclodialysis cleft)
12. Endophthalmitis
13. Zonular tears and vitreous loss
14. Incarceration of iris
15. 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, . 2013-2014.
2. Walton DS. Goniotomy. In: Thomas JV, ed. *Glaucoma Surgery*. St. Louis, MO: 1992; 107-121.
3. Shrader CE, Cibis GW. External Trabeculotomy. In: Thomas JV, ed. *Glaucoma Surgery*. St Louis, MO: 1992;123-131.
4. AAO, Ophthalmology Monographs 4. Glaucoma Surgery: Principles and Techniques, 2nd ed., 1998.
5. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
6. Walton DS, Katavounidou G. Newborn primary congenital glaucoma: 2005 update. *J Pediatr Ophthalmol Strabismus* 2005; 42: 333-341
7. AAO, Focal Points. Pediatric Glaucoma. Module #3, 2012.

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 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. Often a family history
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. Describe medical therapy options**
 - 1. Topical medication
 - a. Carbonic anhydrase inhibitors
 - b. Beta-adrenergic antagonists
 - c. Prostaglandin analogues
 - d. Alpha-adrenergic agonists are contraindicated in children younger than age 2
 - e. Sympathomimetic agents
 - 2. Oral medication - carbonic anhydrase inhibitors
- B. Describe surgical therapy options**
 - 1. Laser trabeculoplasty ineffective
 - 2. Goniotomy
 - 3. Trabeculotomy
 - 4. Trabeculectomy with or without antimetabolites
 - 5. Non-penetrating surgeries
 - 6. Tube shunts
 - 7. 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.
3. Consider tube shunts for children prone to trauma

VI. Describe disease-related complications

A. Progressive loss of vision

B. Blindness

VII. Describe appropriate patient instructions

A. Use medication

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, 2013-2014.

2. Stone EM, Fingert JH, Alward WLM, et al. Identification of a gene that causes primary open angle glaucoma. *Science* 1997;275(5300):668-70.
3. Monemi S, Spaeth G, DaSilva A, et al. Identification of a novel adult-onset primary open-angle glaucoma (POAG) gene on 5q22.1. *Hum Mol Genet* 2005;14(6):725-33.
4. Pang CP, Fan BJ, Canlas O, et al. A genome-wide scan maps a novel juvenile-onset primary open angle glaucoma locus to chromosome 5q. *Mol Vis* 2006;12:85-92.
5. Hauser MA, Allingham RR, Linkroum K, et al. Distribution of WDR36 DNA sequence variants in patients with primary open-angle glaucoma. *Invest Ophthalmol Vis Sci* 2006;47(6):2542-6.
6. Shimizu S, Lichter PR, Johnson AT, et al. Age-dependent prevalence of mutations at the GLC1A locus in primary open-angle glaucoma. *Am J Ophthalmol* 2000;130(2):165-77.
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8. Fingert JH, Stone EM, Sheffield VC, Alward WL. Myocilin glaucoma. *Surv Ophthalmol* 2002;47(6):547-61.
9. Ko YC, Liu CJ, Chou JC, et al. Comparisons of risk factors and visual field changes between juvenile-onset and late-onset primary open-angle glaucoma. *Ophthalmologica* 2002;216(1):27-32.
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13. Tsai JC, Chang HW, Kao CN, et al. Trabeculectomy with mitomycin C versus trabeculectomy alone for juvenile primary open-angle glaucoma. *Ophthalmologica* 2003;217(1):24-30.
14. Chen LJ, Tam PO, Tham CC, Liang XY, Chiang SW, Canlas O, Ritch R, Rhee DJ, Pang CP. Evaluation of SPARC as a candidate gene of juvenile-onset primary open-angle glaucoma by mutation and copy number analyses. *Mol Vis*. 2010 8;16:2016-25.
15. Gupta V, Gupta S, Dhawan M, Sharma A, Kapoor KS, Sihota R. Extent of asymmetry and unilaterality among juvenile onset primary open angle glaucoma patients. *Clin Experiment Ophthalmol*. 2011;39:633-8.
16. Khan AO. Genetics of primary glaucoma. *Curr Opin Ophthalmol*. 2011;22:347-55.
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Axenveld-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 Axenveld-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. Hypoplasia of the iris
 - b. Corectopia and polycoria which may be progressive
 - c. Posterior embryotoxon (prominent and anterior Schwalbe line)
 - d. Peripheral iris attachments to Schwalbe line
 - e. 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 (hypodontia < 6 missing teeth)
 - iii. Focal absence of teeth (oligodontia > 6 missing teeth)
- 3. Facial abnormalities
 - a. Maxillary hypoplasia
 - i. Flattening of midface
 - ii. Receding upper lip
 - iii. Prominent lower lip
 - iv. Hypertelorism and/or telecanthus
- 4. 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

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 10: Glaucoma, 2013-2014.
2. Ritch R, Shields MB, Krupin T. The Glaucomas. Edition 2, St. Louis 1996, The C.V. Mosby Company
3. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
4. AAO, Atlas: Color Atlas of Gonioscopy, 2000.

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. Angle closure

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
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 and serial abdominal ultrasonography**
- D. Genetic counseling**

Additional Resources

1. AAO, Basic and Clinical Sciences Course. Section 10: Glaucoma, 2013-2014.
2. Ritch R, Shields MB, Krupin T. The Glaucomas. Edition 2, St. Louis 1996, The C.V. Mosby Company
3. AAO, Ophthalmology Monographs 18. A Compendium of Inherited Disorders and the Eye, 2006.
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6. AAO, Focal Points: Childhood Glaucoma, Module #10, 1990.

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 angle closure
 - d. Hyphema
 - e. Secondary angle closure glaucoma from the air bubble migrating behind the iris
 - f. Pupillary block glaucoma
 - i. Caused by air bubble used in DSEK for donor tissue adhesion located anterior to the iris
 - 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. By increased corneal thickness after posterior corneal surgery that may lead to an artifactual elevation of IOP by Goldmann applanation or by persistent corneal edema which may lead to an underestimation of IOP due to the changes in corneal biomechanics from edema
 - b. If there is no edema, there may be no alteration in IOP despite the increased central corneal thickness

B. Define the relevant aspects of epidemiology of the disease

1. Incidence of glaucoma after DSEK ranges from up to 40% in eyes without preexisting glaucoma and up to 45% in eyes with preexisting glaucoma
2. In eyes with preexisting glaucoma, a 3 mmHg average rise in IOP after one year was noted in a recent series
3. Acute glaucomas 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. Trabeculectomy
4. Glaucoma drainage devices
5. 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. Cyclodestructive procedures

1. Cryotherapy and Nd:YAG laser associated with significant inflammation which may lead to graft rejection and failure

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

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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
 - b. For each additional 1mm Hg of IOP lowering, the risk of glaucoma progression was reduced by 10% for the population
3. 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 or
 - 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
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
- 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
- 4. 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 only seen with Baerveldt
 - c. Trend toward higher rate of reoperation for glaucoma after Trabeculectomy than after Baerveldt

Additional Resources

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3. AAO, Focal Points: Evidence-based medicine in glaucoma, Module #3, 2008.
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Beta-adrenergic antagonists

I. List the agents

A. Non-selective β_1 - and β_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. Metipranolol HCL 0.3% (OptiPranolol®)
6. Preservative-free timolol

B. Selective β_1 - antagonist

1. Betaxolol HCL, 0.25% (Betoptic-S®)

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, both systemic effects

and effects on IOP

- b. 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. β_1 - and β_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. Good corneal penetration
- 2. Peak aqueous concentration within 1-2 hours of topical dose
- 3. 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 β 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 β_1 - and β_2 - antagonists
 - a. 20%-30% IOP reduction
2. β_1 - selective antagonist
 - a. 14%-17% 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

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. Periorbital contact dermatitis
 - d. Dry eye
2. Systemic
 - a. Cardiovascular
 - i. β_1 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. β_2 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, of beta-adrenergic antagonist solution to decrease total daily dose
- 4. Viscous formulations of Timolol may have less systemic absorption
- 5. Use topical beta-adrenergic antagonists with special properties
 - a. Betaxolol - β_1 - 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

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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
- E. Osmotic agents
- F. Laser iridotomy
- G. Laser iridoplasty
- H. Laser trabeculoplasty
- I. Filtering surgery

V. Describe the technique (method 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

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. Periorcular contact dermatitis
- i. Exacerbation of visual symptoms secondary to cataract because of miosis
- 2. Less frequent
 - a. Iris pigment epithelial cysts (cholinesterase inhibitors)
 - b. Lacrimal stenosis
 - c. Pseudophthalmos
 - d. Fibrinous iritis (especially in the post op 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. 2013-2014
2. AAO, Glaucoma Medical Therapy: Principles and Management, 1999;77-93.
3. AAO, Focal Points: Medical Treatment of Open-Angle Glaucoma, Module #10, 1993.
4. AAO, Focal Points: Angle Closure Glaucoma Update, Module #6, 2009.

Carbonic anhydrase inhibitors

I. List the agents

A. Oral

1. Acetazolamide 125 mg, 250 mg, 500 mg
2. Methazolamide 25 mg, 50 mg

B. Topical

1. Dorzolamide 2%
2. Brinzolamide 1%

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
2. Prophylaxis of elevated IOP after a surgical intervention
3. Reduction of acutely elevated IOP

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 carbonic anhydrase inhibitors (CAIs) are likely to produce a life-threatening cross-reaction
3. Kidney stones
4. Aplastic anemia
5. Thrombocytopenia
6. Sickle cell disease
7. History of blood dyscrasia
8. Corneal endothelial failure

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

- A. Inhibition of carbonic anhydrase blocks aqueous production**
- B. > 90% of carbonic anhydrase must be blocked to decrease aqueous production**
- C. Dorzolamide in particular increases ocular blood flow which has been associated with preservation of visual fields**

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

- A. Complications**
 - 1. With systemic carbonic anhydrase inhibitors
 - a. Paresthesias
 - b. Malaise, anorexia
 - c. Altered taste
 - d. Kidney stones
 - e. Stevens-Johnson syndrome, if allergic

- f. Blood dyscrasias (aplastic anemia and sickle cell disease)
 - g. Hypokalemia especially if already on a thiazide diuretic
 - h. Choroidal detachment
2. With topical carbonic anhydrase inhibitors
 - a. Burning and stinging (less with brinzolamide mentioned below)
 - 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 oral CAIs
2. Consider pre-treatment blood counts with oral CAIs
3. Avoid topical CAIs for diseased corneas with impaired endothelium
4. No CAIs for a history of sulfa allergy, blood dyscrasia or kidney stones

C. Management of complications

1. Stop the medication
2. Topical toxicity
 - a. Change topical therapy
 - b. Consider brinzolamide instead of dorzolamide
 - c. Oral CAIs
3. Systemic toxicity
 - a. Decrease the dose of oral medication
 - b. Change to topical therapy
 - c. Change from acetazolamide to methazolamide
 - d. Medical consult for serious side effects
 - e. Switch to acetazolamide sequels

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. Check serum potassium and use a potassium supplement, especially if already taking a potassium-wasting diuretic for hypertension (this applies to patients taking systemic CAls)**

Additional Resources

1. AAO, Basic Clinical and Science Course. Section 10: Glaucoma, 2013-2014.
2. AAO, Focal Points: Medical Treatment of Open Angle Glaucoma, Module #10, 1993.
3. AAO, Focal Points: New Therapeutic Options for the Treatment of Glaucoma, Module #5, 1999.
4. AAO, Focal Points: Medical Treatment of Open-Angle Glaucoma, Module #2, 1983
5. AAO, Glaucoma Medical Therapy: Principles and Management, 1999, p.88, 99-111, 171-172, 181, 196, 206.
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Alpha adrenergic agonists

I. List the agents

A. Nonselective adrenergic agonists

1. Epinephrine (available as injection only)
2. Dipivefrin (not available in US)

B. Selective adrenergic agonists

1. Apraclonidine (0.5%)
2. Brimonidine tartrate (0.2%, 0.15%, 0.1%)

II. List the indications

A. Non-selective adrenergic agonists

- a. Mydriasis in cataract surgery
- b. Reduce bleeding in oculoplastic surgery
- c. Reduce bleeding in trabeculectomy
- d. Slow absorption of local anesthetics

B. Selective adrenergic agonists

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

III. List the contraindications and precautions

A. Non-selective

1. Retrobulbar anesthesia
 - a. Local
 - i. Risk of vasospasm
 - ii. Occlusion of ophthalmic or central retinal artery

- b. Systemic
 - i. Tachyarrhythmias
 - ii. Death

B. 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. Pregnancy
 - a. Category B drug
 - i. Use only if potential benefits justify potential risk

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

VI. Describe the dosing technique

- A. Nasolacrimal occlusion
- B. Passive lid closure

VII. Describe the method of action

- A. Non-selective-mixed alpha and beta adrenergic agonist
 1. Effect varies over time
 - a. Initially raising IOP slightly
 - b. Followed by reduction lasting 12-24 hours
- B. 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
 3. Controversial neuroprotective effect
 - a. Prevent demise of retinal ganglion cells due to trauma or toxins

VIII. List the complications of the ocular therapy

- A. Non-selective
 1. Systemic
 - a. Tachycardia
 - b. Arrhythmias

- c. Hypertension

B. 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
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, 2013-2014
- 2. AAO, Glaucoma Medical Therapy: Principles and Management, 1999; 47-75.
- 3. Glaucoma Prescribing Guide: Product Information. *Physician's Desk Reference*. 2011.
- 4. Thompson CD, Macdonald TL, Garst Me, et al. Mechanisms of adrenergic agonists induced allergy bioactivation and antigen formation. *Exp Eye Res*. 1997;64:767-773.
- 5. AAO, Focal Points: New Therapeutic Options for the Treatment of Glaucoma, Module #5, 1999.
- 6. Tsai JC, Forbes M. Medical Management of Glaucoma, 3rd ed. West Islip, NY, Professional Communications, Inc. 2009.

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. Proven nonresponsiveness and/or allergy to agent
- 2. Active uveitis (controversial)
- 3. Active macular edema

C. Relative contraindications

- 1. Aphakia or pseudophakia with open posterior capsule, especially after complicated surgery
- 2. Recent intraocular surgery
- 3. History of herpetic keratitis
- 4. 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
 - b. Ocular irritation
 - c. Hypertrichosis and darkening of lashes
 - d. Darkening of iris and periocular skin
 - i. Secondary to increased numbers of melanosomes within melanocytes
 - ii. Risk of iris pigmentation greatest in light brown, blue-green, or two-toned irides; least in blue irides
 - e. Periorbital fat atrophy, deepening of superior lid sulcus
 - f. CME
 - g. Anterior uveitis, suspected
 - h. Exacerbation of underlying herpes simplex virus keratitis (dendrites)
 - i. 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

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, 2013-2014.
2. AAO, Glaucoma Medical Therapy: Principles and Management, 1999; 113-131,172.
3. AAO, Focal Points: New Therapeutic Options for the Treatment of Glaucoma, Module #5, 1999.
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Hyperosmotic agents

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 (Osmitol®)
 - 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 be contraindicated in patients with diabetes)
 - b. Long-term use may perturb electrolytes
2. Of limited value when blood-ocular barrier is disrupted
3. May cause rebound elevation in IOP if agent penetrates eye and reverses osmotic gradient
4. Congestive heart failure
5. Renal failure

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**
 - 1. To determine if there are medical or systemic contraindications to hyperosmotic use

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, 2013-2014.
2. Feitl ME, Krupin T. Hyperosmotic agents. In: Ritch R, Shields MB, Krupin T, eds. The Glaucomas. 2nd Ed. St. Louis, MO: Mosby; 1996: 1483-1488.
3. AAO, Focal Points: Systemic Side Effects of Glaucoma Therapy, Module #4, 1990.
4. AAO, Glaucoma Medical Therapy: Principles and Management, 1999;88:133-147,195-196,203.
5. AAO, Focal Points: Medical Treatment of Open Angle Glaucoma, Module #10, 1993.
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Laser trabeculoplasty

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. Secondary open-angle glaucoma with uncontrolled IOP (adjuvant or primary therapy)
 - a. Pseudoexfoliation glaucoma
 - b. Pigmentary glaucoma
3. Inadequacy of medical therapy
4. Intolerance of or non-compliance with medical therapy for the management of open-angle glaucomas
5. Inadequate IOP control following incisional surgical therapy of open-angle glaucoma
6. Ocular hypertension with uncontrolled IOP in high-risk selected cases
 - a. Non-responsive to medical therapy
 - b. Adjuvant therapy
7. Selective laser trabeculoplasty (SLT) after previous laser trabeculoplasty
 - a. Successful previous argon or SLT
 - b. Failed previous argon laser trabeculoplasty
8. Chronic angle closure provided at least 90 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. Traumatic glaucoma with angle recession
 - d. Uveitic and neovascular glaucoma with or without angle closure
2. Relative contraindications
 - a. Young age (< 50 years)
 - b. Failed previous laser trabeculoplasty
 - c. Less than 180 degrees of open angle

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 neodymium yttrium-aluminum-garnet (Nd:YAG) laser (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. Anesthetize eye
- 3. Perioperative apraclonidine or brimonidine most commonly used to prevent post-laser surgery IOP spike. Other topical glaucoma medications (beta-adrenergic antagonists, carbonic anhydrase inhibitors, parasymphomimetic agents) can also be used
- 4. Position patient in slit-lamp biomicroscope at laser
- 5. Place a laser gonioscope on the eye
- 6. Argon/diode laser
 - a. Apply 50 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 degrees or 360 degrees by rotating the lens to keep the laser aiming beam

in the center of the mirror

7. SLT
 - a. Fixed spot size (400 microns)
 - b. Fixed time (3 nanoseconds)
 - c. Treat the TM (large spot size covers angle)
 - d. Applications are confluent, not overlapping
 - e. Adjust power so TM does not blanch or just at bubble formation
 - f. Treat 180-360 degrees
8. Apply a drop of a topical glaucoma medication
9. 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 and haze

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

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Incisional filtering surgery for open angle glaucoma

I. List the indications/contraindications

A. Indications

1. Presence of glaucoma with high probability of continued optic nerve damage after unsuccessful laser trabeculoplasty or where laser trabeculoplasty is known not to be effective [such as chronic angle closure glaucoma (ACG), etc.] (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. Antiplatelet / anticoagulant medications should be discontinued if possible

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
 - 2. Ciliary body cyclophotocoagulation
- C. Other filtration surgery procedures**
 - 1. Glaucoma tube-shunt implants
 - 2. Nonpenetrating filtration surgery (e.g., viscocanalostomy, deep sclerectomy, canaloplasty, ab interno trabecular ,meshwork stent)

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
 - iii. Apply intraoperatively
 - 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
 - 2. Risk for failure or complication
 - a. Active anterior segment neovascularization (rubeosis iridis)
 - b. Active iritis (uveitic glaucoma)

- c. Younger patients (< 50 years of age)
- d. Patients with aphakia
- e. Previous failure of filtering surgery
- f. Episcleral fibrosis is most common cause for filtration failure and antifibrotic agents inhibit cellular proliferation and fibrosis
- g. Chronic topical glaucoma medication

D. Trabeculectomy technique

1. Exposure with corneal traction suture or superior rectus bridle suture
2. Limbus-based vs. 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. 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 AC with BSS via paracentesis
13. Possible excision of Tenon's capsule
14. Closure of Tenon's 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's 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 gonioscope 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

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management of open angle glaucoma uncontrolled with medical therapy. *Br J Ophthalmol*. 2002;86:748-54.

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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
5. Note: combined procedures are less likely to achieve a very low IOP compared to trabeculectomy alone

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., warfarin, antiplatelet drugs, Gingko biloba, Bilberry, Vitamin E, in consultation with the patient's primary care physician

III. List the alternatives to this procedure

- A. Separate 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

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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. Angle closure glaucoma not pupil block in origin (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 compression 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. 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, 2013-2014.
- 2. Ritch R, Lieberman J. Laser iridotomy and peripheral iridoplasty. In: Ritch R, Shields MB, Krupin T, eds. The Glaucomas, 2nd ed. St. Louis, MO; 1996: 1549-1573.
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Laser goniotomy or peripheral iridotomy 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 iridectomy
 - b. Once cornea clears, an iridectomy 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 irido ciliary 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. 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: 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 a laser contact lens with gonioscopy mirror on the eye
 6. Activate argon laser
 - a. Fix spot size (200-500 microns)
 - b. Set duration 0.5 second, but titrate length of burn through direct observation
 - c. Set power: 200 to 400 mW
 - 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.
 - i. The actual number of spots used depends completely on the disorder being treated
 - a. 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 within one week depending on indication for initial goniotomy
- 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, 2013-2014.
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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. Nanophthalmos
2. Predisposition to aqueous misdirection
3. Bleeding disorder

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. Microsharp blade or 30 gauge needle on TB syringe with plunger removed (aqueous will stop flowing spontaneously at IOP=10-15 mmHg) 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. Use of valved device indicated when urgent control of IOP is necessary

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

A. Infection

1. Avoid by utilizing sterile technique
2. Appropriate use of post-operative antibiotics

B. Bleeding

1. Minimize risk by grasping iris, and not ciliary body tissue
2. Discontinuation of anticoagulation pre-operatively whenever possible

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
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 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
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, 2013-2014.
2. AAO, Focal Points: Diagnosis and Management of Complications of Glaucoma Filtering Surgery, Module #3, 1994.
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8. Su WW, Chen PY, Hsiao CH, et al. Primary phacoemulsification and intraocular lens implantation for acute primary angle-closure. PLoS One 2011;6:Epub 2011.

Aqueous shunt surgery

I. List the indications/contraindications

A. Indications

1. Traditionally reserved for difficult 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. For double plate devices, intervening tube may be placed over (with division of check ligaments) or under (more difficult) adjacent rectus muscle

- d. 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)
- e. 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

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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 to the contralateral seeing 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. Endoscopic CPC
3. Transpupillary CPC
4. Transvitreal endophotocoagulation

B. Cyclocryotherapy

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 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. Endoscopic cyclophotocoagulation
 - 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 aphakic 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. Transvitreal endocyclophotocoagulation
 - a. Argon or diode laser used in conjunction with vitrectomy
 - b. Requires clear media, aphakia or pseudophakia to visualize and treat ciliary processes
5. Cyclocryotherapy
 - a. Cryosurgical instrument with nitrous oxide
 - b. Glaucoma probe placed with nearest edge 2.5mm from the limbus
 - c. Freeze 180 degrees with approximately six points for 1 minute
 - d. Topical or subconjunctival corticosteroids, atropine

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 antihypertensives 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

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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 aqueous suppressants or prostaglandin analogues in an eye with previous filtering surgery
8. Rhegmatogenous retinal detachment
9. Ocular ischemia

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
 - i. 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 ultrasound 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) tissue 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. Re-suture wound
5. Cyclodialysis cleft
 - a. Careful gonioscopy to determine location and extent of cleft

- b. Cycloplegia might close cleft
 - 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 even after hypotony is reversed 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 re-suturing 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 or pericardium 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. Ocular irritation from a large bleb**
- 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**

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