Shedding Light on Pediatric Cataracts

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A newborn infant presents with bilateral white cataracts. What is the best age to do the surgery?

A. 4-6 weeks of age
B. 8-12 weeks of age
C. 12-16 weeks of age
D. After 6 months of age
What is the most common etiology for pediatric cataracts?

A. Hereditary
B. Idiopathic
C. Genetic disease
D. Infection
What most limits vision most after pediatric cataract surgery?

A. Glaucoma
B. Amblyopia
C. Surgical complications
D. Poor compliance with glasses wear
In what situation is a work-up not needed for pediatric cataracts?

A. Positive family history
B. Unilateral cataract
C. Known genetic condition
D. All of the above
What is a Cataract?

A clouding or opacity of the lens of the eye
Epidemiology of Pediatric Cataracts

• Worldwide: 1 to 15 per 10,000 children
• Prevalance of blindness:
  – 1 to 4/10,000 developing countries
  – .1 to .4/10,000 industrialized countries

• Industrialized countries: 1 to 4 per 10,000
• TCH: 60-80 surgical cases a year
How Do These Patients Present?

- Positive family history
- Leukocoria: white pupil
- Strabismus: esotropia
- Known syndrome or genetic evaluation
- Funny and constant eye movements: sensory nystagmus (age 2-4 months)
Red Reflex Test: Bruckner Test

Symmetry
Absence
Disruption
Red Reflex Test

Key points:
Red Reflex Test
Nuclear cataract

Anterior polar cataract

Lamellar cataract

Persistent fetal vasculature

Posterior polar cataract

Posterior lenticonus
Abnormal Red Reflex

- Cataracts
- High refractive error
- Corneal abnormalities
- Vitreous abnormalities
- Retinal abnormalities: Retinoblastoma
Work Up: Unilateral Cataracts

• Majority idiopathic
• In general, a systemic or laboratory evaluation is not necessary
  – Obtain typical history (PMH, birth history, FH, trauma, systemic disease, medications, etc.)
• Consider TORCH titers (IgM) or VDRL/RPR only if there is anything that raises suspicion
  – Low yield
  – Intrauterine infections would be more likely to cause a bilateral cataract
Work Up: Bilateral Cataracts

- Majority idiopathic or familial, however systemic disease more likely
- Family history is important
  - Consider examining other family members
  - Marked variability can be seen between family members
- Review of systems and pregnancy history
  - Drug exposure during pregnancy
  - Maternal or congenital infection
  - Trauma
  - Congenital anomalies
  - Systemic disease
  - Associated ocular findings
- Baseline physical exam
- Developmental milestones
- Consideration of genetics evaluation if appropriate
- Consideration of laboratory evaluation
Laboratory Work Up

If family history negative and no systemic disease/syndromes being considered, can consider these labs:

- TORCH titers
- VDRL/RPR
- Serum calcium
  - Hypoparathyroidism, hypocalcemia
- Phosphorus
  - Pseudohypoparathyroidism
- Urine for reducing substances?
  - Galactosemia
Summary: Work Up

• Most congenital or infantile cataracts are idiopathic
• Bilateral cataracts: common teaching
  – 1/3 idiopathic
  – 1/3 hereditary – 75% autosomal dominant
  – 1/3 associated with disease or syndromes**

• No work up generally needed:
  – Known family history
  – Known associated syndrome or disease
  – Unilateral cataract

• In almost all systemic conditions, there is usually some other exam or historical finding to suggest the associated diagnosis
  – Evaluation targeted to patient

**PEDIG Pediatric cataract surgery outcomes registry: 5.8% with associated medical condition
Pediatric Versus Adult Cataracts

• Eye growth has implications
  – Most occurs between birth to 2 years: eye grows 30-40%
• Amblyopia: “lazy eye” due to deprivation of vision from the cataract
• Other sequelae
• Age of onset affects visual impact (congenital vs. juvenile)
Timing of Surgical Management

- Infants with visually significant cataract
  - Surgery in first 4-8 weeks of life
  - Bilateral cases
    - May be performed 1 week apart
- Older children (>5-7 years) less risk of amblyopia
Timing of Surgical Management

• Is conservative management appropriate?
• Preverbal: retinoscopic (red) reflex
• Older children:
  – Good visual acuity (no symptoms, vision 20/40 or better)
• All patients
  – Extra-axial opacities (not central)
  – Opacities < 3 mm
  – Treatable refractive error responding to treatment
  – Consider trial of patching +/- pupillary dilation in unilateral cataracts when unsure
When to Put in a Lens Implant?

• Controversial: Over 7 months
  – May wait a little longer if bilateral case
• Eye size normal
• No other significant associated ocular abnormalities
• Good lens support (capsule)
• If no lens implant: contact lenses or aphakic glasses
• Will still need glasses and bifocal if lens implant placed
Surgical Considerations

• General anesthesia
• Immediate postoperative considerations:
  – Eye drops (topical steroids, antibiotics, dilating drops)
  – No sports or swimming for 4 weeks
  – Shield wear/eye protection
  – Eye redness
Amblyopia

- Visual outcome in that patient: realistic expectations
- Patching first line of treatment
  - Atropine eye drops less or not effective
- Greatest impediment to improvement of vision
- Requires long-term commitment from parents’ standpoint
- Management until 5-7 years of age
- Adhesive eye patches (on skin)
- Patches over glasses (cloth)s
- More choices online
Refractive Management

• Very important: contact lens, aphakic glasses, glasses with bifocals
  – Lens implant versus no lens implant

• Correction of residual hyperopia, myopia, and/or astigmatism

• Significant change in refractive error over time can occur

• Initial correction for near; bifocals after 2 years of age
Strabismus

- Lens implant does not prevent development of strabismus
- Percent of patients developing strabismus over time increases
  - 24.6% (baseline) to 70.4% (12 months after surgery)
  - 60% of patients without strabismus developed it over time
- Strabismus less likely if cataract removed at an earlier age
  - Trend toward better acuity in patients without strabismus
- Esotropia most common strabismus pattern
- Surgery can be performed after amblyopia treatment initiated
Aphakic Glaucoma

• Important and frequent complication of pediatric cataract surgery
  – Mechanism unclear
• Wide range of reported frequencies
  – 15% to 21%
  – Possible risk factors: secondary membrane surgery, small cornea, family history
  – Most consistent risk factors: cataract surgery in first year of life and small corneal diameter
• Treatment with drops and/or surgery
• Long-term monitoring: challenging in kids
• Examinations under anesthesia may be required
Visual Axis Opacification or “Secondary Cataract”

- Rate high in children
  - Literature: 44-100% within first year
- Higher and earlier occurrence in younger children
- Laser versus surgical treatment
  - Nd:Yag laser capsulotomy for older, cooperative children
  - Surgical capsulotomy in younger children
Thank you for your attention!