Pediatric and Traumatic Cataract

Midwest PEENT Symposium

Shane Havens MD
Medical Students
Ophthalmology and Visual Sciences
March 22nd, 2019

Objectives

• Brief ocular anatomy and ophthalmic exam review
• Review of cataract and children
• History and Examination findings – Guide differential diagnosis
• Initial treatment and amblyopia follow-up

Disclosures

No financial interest in any of the material presented in this presentation

Ocular Anatomy

• Uvea – Iris, Ciliary body, Choroid.
• Fundus – Retina, macula, OD/OS.

The Eye “Vital Signs”

Visual Acuity – Snellen (20/20), Pocket Vision Screener (6/6)
• OD (Oculus Dexter)
• OS (Oculus sinister)
• CF (Corrected Farsightedness)
• HM (Hand Motion)
• NLP (No Light Perception)

Pupillary Examination
• IOP (Intraocular Pressure)
• Slit-Lamp, direct fundoscopy, Wood’s lamp

Lens Anatomy

Visual Acuity – Snellen (20/20), Pocket Vision Screener (6/6)
• OD (Oculus Dexter)
• OS (Oculus sinister)
• CF (Corrected Farsightedness)
• HM (Hand Motion)
• NLP (No Light Perception)

Pupillary Examination
• IOP (Intraocular Pressure)
• Slit-Lamp, direct fundoscopy, Wood’s lamp
Pupil Examination

- Inspect size and shape in dim light.
- Measure direct and consensual responses to light.
  - Parasympathetic – Constriction/miosis
  - Sympathetic – Dilation/mydriasis
- Normal response: Initial constriction followed by slow variable redilation.
- Swinging flashlight test – detect APD

Afferent Pupil Defect

Slow dilation without initial constriction.
Eponym: Marcus-Gunn Pupil
Indicates ON lesion – Optic neuropathy, optic neuritis, traumatic transection, compression (in rare cases a sign of large retinal lesion/detachment).
Red saturation test

EOM Testing

Goldman Applanation Tonometry – Slit-Lamp, fluorescein, training
Schiottz Indentation Tonometry - antiquated
Handheld digital tonometry using Tono-pen and iCare - $$$$$
We all have these and can check pressure!
“Normal” IOP 10-21

Intraocular pressure

Slit Lamp Examination

LLL – lids, lashes, lacrimal apparatus
C/S – Conjunctiva and sclera
C – Cornea - Inspect all layers, tear film
AC – Anterior chamber – depth, cells and flare.
I/L – Iris and Lens

Pediatric Cataract Introduction

Disorders of the pediatric lens
- Cataract, disorders of lens shape, size, and location
Incidence of lens abnormalities ranges from 1:4000 to 1:110,000 live births/year
Strongly Amblyogenic – especially if unilateral and early in life
Pediatric Cataract General Features

Account for 10% of all childhood vision loss

Isolated cataract or associated with systemic condition

Congenital or Acquired
- Congenital – present at birth
- Infantile – during the 1st year of life

Inherited or sporadic
- Most AD, but X-linked and AR inheritance occur

Unilateral or bilateral
- Bilateral often associated with systemic disease

Stable or progressive

Partial or complete
- Location of cataract can reflect etiology

Pediatric Cataract Evaluation

HISTORY
Newborn screening eye exam by pediatric primary care provider
- Red reflex, Ir-knee test
Assess growth, development, and systemic disorders
Family history
Evaluation of immediate family members

EXAM
Pediatric physical exam
Visual function
- Fixation reflex after 2 months in healthy infants
- Presbyopia >2 months
- Fixation behavior and preference
- Object to occlusion
Concerning features:
- Central or posterior opacities >3 mm in size
- Strabismus with unilateral cataract
- Nystagmus with bilateral cataract

Slit lamp
- Assess cataract morphology
- Dilated exam, or ultrasound if no view of retina and optic nerve
- Corneal diameter
- Iris appearance, check for coloboma

Pediatric Cataract Morphology

Location of cataract and lens morphology may reflect:
- Etiology
- Timing of onset
- Prognosis
Anterior Polar Cataract
Common
Less than 3 mm in diameter
Usually congenital and sporadic
Unilateral or bilateral
Often non-progressive, and non-significant
- if occlude pupil more concerning
Can cause asymmetric refractive error and related amblyopia

Infantile Nuclear Cataract
Involve the center of the lenses
Approximately 3 mm in diameter
Density and size can vary
Inherited or sporadic
More commonly bilateral
Usually stable, eyes may be smaller

Lamellar Cataract
Lamellar (zonular) cataract
Discrete lamellar opacity and clear cortex
Typically 5 mm or more in diameter
More often bilateral, can be unilateral
Normal eye size
Good visual prognosis – depending on density
Posterior Lenticonus
Cone-shaped deformity of the posterior lens surface
- Progressive thinning of the posterior capsule
- Oil droplet appearance on red reflex early
- Progressive opacification
Unilateral, normal eye size

Posterior Subcapsular Cataract
Usually acquired
Often bilateral
Typically progressive
Causes:
- Corticosteroid, inflammation, radiation, trauma
  - Associated with NF2

Persistent Fetal Vasculature
Most common cause of unilateral cataract
Isolated, sporadic
Smaller eye, elongated ciliary processes
Prominent hyaloid vessels remnant → dense retrolental plaque and microphthalmos
- Mittendorf dot, Bergmeister papilla
If bilateral can be associated with neurological abnormalities
Progressive
Secondary glaucoma can develop
Pediatric Cataract General Features

<table>
<thead>
<tr>
<th>Cataract Morphology</th>
<th>Diagnosis</th>
<th>Other Possible Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Synciotic</td>
<td>Fabry disease</td>
<td>Cerebral edema, retinopathy</td>
</tr>
<tr>
<td>Vascular</td>
<td>Marfan syndrome</td>
<td>Hypoparathyroidity</td>
</tr>
<tr>
<td>Multicollarette</td>
<td>Diabetis insipidus</td>
<td>Elevated triglyceride level</td>
</tr>
<tr>
<td>Gross “sunflower”</td>
<td>Myotonic dystrophy</td>
<td>Characteristic facial features, “grip”</td>
</tr>
<tr>
<td>Thin dilatoriform</td>
<td>Wilson disease</td>
<td>Kayser-Fleischer ring</td>
</tr>
</tbody>
</table>

Pediatric Cataract Workup

Unilateral – lab evaluation not necessary

Bilateral
- Disorders of galactose metabolism
  - Urine reducing substances
- ID: TORCH and Varicella titers, VDRL
- Metabolic disease:
  - Urine AA (Lowe’s)
  - Serum Calcium (low in hypoparathyroid)
  - Phosphorus (high in hypoparathyroid)
  - Glucose (DM)
  - Ferritin (high in hyperferritinemia)

Pediatric Cataract Management

Surgery Timing:
Younger child – greater urgency to facilitate normal vision development
- Risk of deprivation amblyopia
Significant unilateral cataract – should be removed before 6 weeks of age
Bilateral before 10 weeks of age

Older children
- When VA 20/40 or worse

To IOL or not to IOL?
IOL placement widely accepted in children 1-2 years and older
- Relatively unpredictable refractive outcomes

Primary posterior capsulotomy at time of surgery

Post-operative care:
- topical antibiotic, steroid, +/- cycloplegics

AMBLYOPIA MANAGEMENT
- Contact lens or glasses within a few weeks of surgery
- Glasses for infant with bilateral aphakia
- CL for infants with unilateral aphakia
- Patching of better eye
- Time with patch in place
- Based on degree of amblyopia and age of child

Prognosis:
Age at diagnosis
Type of cataract
Surgical timing
Optical correction
Adherence to amblyopia therapy

Pediatric Cataract Management

Post-operative Monitoring:
Strabismus
Glaucoma rates increased with cataract surgery in infancy
Etiology of Bilateral Cataract

Structural or Positional Lens Abnormalities

Congenital aphakia
- Rare, markedly abnormal eye

Spherophakia
- Small, spherical lens, can dislocated into AC
- Usually bilateral

Lens coloboma
- Flattening or notching of lens periphery
- Can be associated with iris, CN, retinal coloboma
- Typically not progressive

Lens Dislocations

Located/ectopic – completely dislocated lens
Subluxated – partially dislocated
Familial or sporadic
Traumatic

Marfan Syndrome

Syndrome with CV, muscular, and ocular abnormalities
- Tall, arachnodactyly, scoliosis, pectus excavatum
- Enlargement of aortic root, floppy MV, risk of aortic dissection
- Life expectancy about half of general population
AD, but 15% have negative family history
80% with ocular abnormalities – mostly lens subluxation superotemporally

Flat Cornea, long eyes (myopic)
Retinal detachment risk in 20-30’s
Intact and stretched zonular fibers

Homocystinuria

Rare, 1:100,000 births
AR
Similar skeletal changes to Marfan’s
50% with CNS abnormalities, learning disabilities and seizures
Risk of arterial and venous thrombotic events with anesthesia events
Inferior subluxation of the lens more than superior
Anterior subluxation of lens
Broken Zonular fibers
Dx: Disulfides (HC) in urine
Treated with diet
- Low methionine
- Supplemental cysteine
- B6 supplementation

Weill-Marchesani

Clinical opposite from Marfan’s
Short, brachydactyly and short limbs
AD or AR, ADAMST10 gene
Microspherophakic lenses
Subluxation in the anterior chamber
**Sulfite Oxidase Deficiency**

Very rare
- Severe neurologic disorders and lens dislocation
  - Infantile hemiplegia
  - Choreoathetosis
  - Seizure
- Increased sulfite in the urine
- Absence of SO in skin fibroblasts

**Isolated Ectopia Lentis**

Superior and temporal displacement of lens
- Bilateral and symmetric
- AD inheritance
  - Some FBN1 heterozygous patients may have only ectopia lentis
- Risk of glaucoma

**Ectopia Lentis et Pupillae**

Displacement of the pupil and lens dislocation in opposite direction
- Bilateral
- Spherophakia

**Management of lens dislocation**

Optical correction sometimes possible
- Subluxed lens in visual axis → myopic astigmatism
- Lensctomy if unable to optically correct vision

- Contact lenses
- Anterior chamber IOL or scleral fixated IOLs

**Traumatic Cataract in Pediatrics**

Timing of removal
- Depend on concurrent ocular injury
  - Pupil, iris
  - Retinal injury
  - traumatic optic neuropathy

Importance of full examination of anterior and posterior ocular structures
- B-scan ultrasound if no view of retina due to cataract

Amblyogenic age
- Post-operative visual rehabilitation.