Lens Embryology
- Surface ectoderm overlying optic vesicle
- Day 28 begins to form
- End of week 5 lens vesicle is formed
- Embryonic nucleus formed at week 7
- Weeks 12-14 anterior Y and posterior inverted Y sutures form - completing fetal nucleus
- At birth fetal and embryonic nucleus make up most of the lens volume

Lens
- 5 structures: embryonic nucleus, fetal nucleus, cortex, lens epithelium and lens capsule.
- Lens capsule size is fairly constant
- Anterior lens epithelium produces cortex
- Lens becomes more dense
- Timing of insult determined by layer of opacity
  - Nuclear (in utero) vs Lamellar (neonatal)

Cataract
- Most common lens abnormality
- 10% of visual loss worldwide
- Estimated 1 in 250 newborns
- Etiology mostly unknown
- Visual significance depends on age of onset, location and morphology

Pediatric Cataracts
- Isolated vs. part of systemic condition
- Congenital vs. acquired
- Inherited vs. sporadic
- Unilateral vs. bilateral
- Partial vs. complete
- Stable vs. progressive
Etiology

- 1/3 hereditary
- 1/3 associated with systemic syndromes
- 1/3 unknown etiology
- Bilateral – 60% idiopathic
- Unilateral – 80% idiopathic

Description of opacification

- Lamellar
- Speckled
- Membranous
- Pulverulent
- Polar
- Zonular
- Subcapsular
- Nuclear
- Total
- Lenticous/lentiglobus

Systemic

- Chromosomal abnormalities
- Systemic conditions, disease
- Infections – occur in 1st trimester
  - Rubella, HSV, mumps, toxoplasmosis, vaccinia, CMV
- Trauma, radiation
- Almost all are bilateral (the reverse is not true)
- Can have other ocular pathology: coloboma, aniridia, PFV, ant. seg. dysgenesis, etc.

Onset

- Earlier onset = more amblyogenic
- Prior to 2 months of age (fixation) may lead to nystagmus and poor vision regardless of surgery

Morphology

- Zonular
- Polar
- Total
- Membranous

Anterior Polar

- Common
- < 3 mm
- Congenital
- Sporadic
- Unilateral > bilateral
- Non progressive
- Non visually significant
- Anisometropia is common
Nuclear Cataract
- Embryonic or fetal nucleus
- Often bilateral, dense
- Microphthalmos and microcornea
- Higher risk of glaucoma
- AD inheritance common
- ≥ 3 mm

Zonular (lamellar) Cataract
- Zone of the lens
- Most common is lamellar (inner cortex)
- Usually secondary to metabolic insult (hypoglycemia/galactosemia)
- Bilateral
- ≥ 5 mm
- Occur after fixation reflex
- Better prognosis after surgery

Posterior Lenticous/Lentiglobus
- Thinning of central posterior capsule
- Oil droplet in RR
- Gradually opacify
- If capsule breaks – rapid opacification
- Unilateral, normal size eyes
- Congenital defect with late onset of opacification

Posterior Subcapsular Cataract
- Acquired and bilateral
- Progressive
- Secondary causes:
  - Steroids, uveitis, retinal pathology, radiation
- If unilateral: consider NF-2

PFV
- Fetal hyaloid vascular complex
- Retrolental membrane
  - Small or extend to ciliary processes
- Congenital, unilateral
- Microphthalmic; if not think ‘glaucoma’
- Stalk can extend to optic nerve
- Lens can push forward causing secondary glaucoma

Structural Lens pathology
- Congenital Aphakia: rare, very abnormal eye
- Spherophakia
- Coloboma: flattening of lens equator due to loss of zonules.
  - Not a true lens defect.
- Dislocated lenses
Dislocated Lenses
- Systemic (bilateral)
  - Marfans (up)
  - Homocytinurea (down)
  - Weill-Marchesani (spherical)
  - Sulfite oxidase def.
  - Syphilis
  - Ehlers-Danlos
- Ocular Conditions (bilateral or unilateral)
  - Aniridia
  - Iris coloboma
  - Trauma
  - Hereditary ectopia lentis
  - Congenital glaucoma

Exam
- 3 mm or larger is visually significant
- Nystagmus (bilateral) and strabismus (unilateral) is significant
- If no view to retina get a B scan

Work-Up
- Unilateral: none needed
- Bilateral: if family Hx is + no work up needed
- Lab W/U:
  - Urine for reducing substances (galactose metabolism), galactokinase, galactose 1-phosphate uridylyltransferase.
  - TORCH, VDRL, Varicella titers
  - Urine for amino acids
  - Serum calcium, phosphorus, glucose, ferritin
  - Consider genetic consult
  - New: +/- Evaluation for Cerebrotendinous Xanthomatosis (CTX) – progressive neuro-dengerative genetic disease that may cause early onset bilateral cataracts.

Treatment
- Glasses
- Dilation
- Amblyopia treatment
- Surgery – General anesthesia

Surgery
- General anesthesia
- Under 1 year of age, usually leave aphakic
- Need to do a posterior capsulotomy
- Aphakic contact lenses vs. intraocular lenses
- Complications:
  - Chronic aphakic glaucoma (15%)
Leukocoria (white reflex)

- Referring diagnosis
- Should be seen urgently in amblyogenic age ranges (birth-8 years)
- Differential diagnosis:
  - Cataract
  - Corneal opacity
  - Peter’s anomaly
  - Persistent fetal vasculature
  - Retinoblastoma
  - Coats disease
  - Retinal detachment
  - Anisometropia
  - Toxoplasmosis
  - Toxocariasis
  - ROP

Case Study 1

- 5 month old previously 29 week premature twins with bilateral congenital cataracts.
- Bilateral lensectomies with anterior vitrectomies on both babies on the same day and required admission to the hospital overnight due to their young age.
- Both have had intraocular pressure problems and required further surgeries.
- The boy twin required two glaucoma surgeries.
- The girl twin required a couple further surgeries for secondary membranes to clear the visual axis.
- They currently are in RGP lenses and routinely wear them but tend to lose them frequently.

Case Study 2

- 8 month old M presented with exotropia of the right eye.
- Exam revealed a unilateral total cataract in the right eye with a clear lens in the left.
- Proceeded with lensectomy/anterior vitrectomy in the right eye, no lens implanted.
- He was fitted for a contact lens and initiated amblyopia tx right away.
- At age 3, he wouldn’t wear his contact lens and only sometimes his glasses and he had a large anisometropia due to the aphakia in the right eye.
- Decided to do a secondary lens implant to give him any chance of improving his vision.
- He continues to be non compliant with patching or atropine and his vision is HM to count fingers in that eye.

Case Study 3

- 2 year old F with dense cataracts.
- Had been seen previously as an infancy because her mother had early onset cataracts. Healthy exam.
- Came back at 2 years of age because she was bumping into things.
- Had bilateral cataract extraction with intraocular lens implant.
- One Year later had to do a pars plana posterior capsulotomy OU.
- Current vision is: 20/80 20/150 in glasses.

Case Study 4

- 14 y/o M history of corneal laceration repair and retinal detachment repair OD.
- He developed a dense PSC/cortical cataract.
- Lens implant choice was complicated by corneal scar.
- Vision now 20/80 uncorrected.

Questions?