Morphology and aetiology of pediatric cataract

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EOF declaration

There is no conflict of interest in this presentation
Lens embryogenesis

- Starts on 4\textsuperscript{th} week of gestation from lens placode
- Nucleus
  - embryonic
  - fetal
- Cortex
  (outside Y sutures, mainly produced postnatally)
Important differences

• In newborns, lens has a weight of 90mg (225mg in adults) and 35 D of refractive power which decrease to 28 D in the next 6 months and to 22-24 D by the age of 5 years

• Lens is soft and flexible- no need for phaco

• Wieger’s capsulohyaloid ligament: remnant of primary vitreous, attachment between lens and vitreous, dissipates by the age of 40 years
Wieger’s capsulohyaloid ligament
General informations

➢ Incidence – 1/400 to 1/1500 live births

• 10% of childhood blindness

• Congenital or Acquired
• Visually significant or not
• Stable or Progressive
• Unilateral or Bilateral
• Partial or Complete
Morphological Classification

- Anterior cataracts
- Central cataracts
- Posterior cataracts
- Total cataracts
- Traumatic cataracts
Anterior Cataracts

Anterior Polar Cataract

Small, discrete, white opacity, ≤ 3mm, 30% bilateral, 90% sporadic, usually non progressive
Anterior Pyramidal Cataract

Anterior, conical, 2-2.5 mm, fibrotic, surrounded by cortical opacities which can progress, require surgery
Anterior Subcapsular Cataract

Opacity of anterior lens cortex

- Anterior lenticonus
- Trauma
- Alport syndrome
  (deafness, nephritis, abnormal IV collagen)
Central Cataracts

Nuclear
- between Y sutures
- congenital onset
- often combined with microphthalmia
- requires early surgery
Cortical Lamellar (zonular)

- outside Y sutures
- zones of clear + opacified cortex
- riders sometimes
- starts after 4-6 months of age
- usually developmental and progressive
Cortical Lamellar (zonular)

- good visual acuity for years
- requires surgery in late childhood
- rarely correlates with galactosemia and neonatal hypoglycemia
Cortical Lamellar (zonular) cataracts
Sutural Cataract

- opacification along Y sutures
- may be progressive
- if bilateral often autosomal dominant inheritance
Sutural Cataract
Posterior Cataracts

Posterior Lenticonus

- thinning of posterior capsule, which progress to posterior subcapsular cataract
- usually unilateral and sporadic
- the posterior bowing cause astigmatism and amblyopia despite clear-appearing lens
Posterior Lenticus
Posterior Lenticiconus

Sometimes is associated with a gap in the posterior capsule (7%-10%)
Posterior Subcapsular Cataract

- always developmental
- usually related to posterior lenticous

- can be associated with Down’s syndrome, steroid use, radiation, NF II, blunt trauma or be idiopathic
- tends to be visually significant and needs surgery
Posterior Subcapsular Cataract
PHPV
Persistent Primary Hyperplastic Vitreous

- persistence and secondary fibrosis of primitive hyaloid vascular system
- white fibrovascular membrane behind the lens
- over the time the membrane contracts, pull the ciliary processes, swallowing AC, cause SACG, cataract, RD, phthisis bulbi
PHPV

Is always unilateral, sporadic and associated with microcornea and/or microphthalmia
PHPV treatment

early removal by vitrectomy, micro scissors and intraocular cautery improved the prognosis
Posterior PHPV

Fibrovascular tissue extending from ON to retinal periphery
Total Cataracts

Diffuse Cataract

- multiple flecks
- autosomal dominant or related with congenital hypoparathyroidism, myotonic dystrophy, or Down’s syndrome (blue-dot)
True Total Cataract

- no inner layers of the lens can be visualized
- associated with:
  Down’s syndrome, autosomal dominant inheritance, TORCH, metabolic disorders (end stage)
Traumatic Cataracts

Caused by direct blunt or penetrating trauma, electricity, radiation, alkaline substances
Causes of Congenital Cataract

1. Prenatal (intra-uterine) infection e.g. rubella, cytomegalovirus, syphilis.
2. Prenatal (intra-uterine) drug exposure e.g. corticosteroids, vitamin A.
3. Prenatal (intra-uterine) ionizing radiation e.g. x-rays.
4. Prenatal / peri-natal metabolic disorder e.g. maternal diabetes.

5. Hereditary (isolated - without associated eye or systemic disorder) e.g. autosomal dominant inheritance.

6. Hereditary with associated systemic disorder or multi-system syndrome
   • Chromosomal e.g. Down's syndrome (trisomy 21), Turner's syndrome.
   • With skeletal disease or muscle disorder e.g. Stickler syndrome, Myotonic dystrophy.
   • With central nervous system disorder e.g. Norrie's disease.
   • With renal disease e.g. Lowe's syndrome, Alport's syndrome.
   • With mandibulo-facial disorder e.g. Nance-Horan cataract-dental syndrome.
   • With dermatological disorder e.g. Congenital ichthyosis, Incontinentia pigmenti
Diagnostic approach

- History, pregnancy problems, earlier photos
- Family history
- Parents and siblings ocular examination
- Complete ocular examination, A and B scans
- Pediatric evaluation
- **Lab tests** (Blood count, blood sugar, VDRL, plasma Ca and Ph, galactokinase levels, TORCH, amino acids in urine)*

*mainly in bilateral cases*
Thank you for your attention