Pediatric Cataracts
Types and Treatment

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EOF declaration

There is no conflict of interest in this presentation
Lens embryogenesis

- Starts on 4th 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

Space of Garnier

Wieger’s ligament
Diameter 8-9mm
Width 1-2mm

Space of Berger

Wieger’s ligament
Diameter 8-9mm
Width 1-2mm
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
- usually developmental and progressive
- starts after 4-6 months of age
- 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 Lenticularus
Posterior Lenticiconus

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

- always developmental
- usually related to posterior lenticonus
- 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
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)
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
Management of Pediatric Cataract

Is the cataract visually significant?

- **Neonates**
  
  Red reflex, cataract morphology, retinal view by direct ophthalmoscopy. Cataract in visual axis, larger than 3mm, posterior, with no clear zones requires surgery

- **Infants+ preverbal children** (3 months- 2 years)
  
  Fixation, preferential looking cards, VEP’s, irritability on covering good eye
Management of Pediatric Cataract

Is the cataract visually significant?

• *Verbal children*
  
  VA<3-4/10, strabismus + stereo vision assessment, family dynamics

**Timing of the surgery**

For visual significant congenital cataracts is between 4-8 weeks of age
Surgical Technique

The standard surgery for children (<8 years) is a 5 mm anterior capsulorhexis, lens aspiration with primary posterior capsulectomy of 4 mm and a deep anterior vitrectomy.

PCIOL implantation (foldable one piece PMMA) is a safe option for patients over 2 years old (1-2 years?)
Infant Aphakia Treatment Study

There was no statistically significant difference in grating visual acuity at age 1 year between the IOL and CL groups; however, additional intraocular surgeries were performed more frequently in the IOL group.

*IOL use in children is off-label
Surgical Tips

• In wound construction: scleral-tunnel, limbal or clear corneal (2,2mm) approaches

• Manual ACCC is a gold standard, but is difficult to perform. Anterior vitrectorhexis, by vitrector, plasma blade and radiofrequency diathermy anterior capsulectomy, have also been advocated

• Trypan blue?, highly viscous OVD (Healon GV or 5, Provisc, Amvisc, Disco Visk)
Surgical Tips

• No hydrodelination
• No hydrodissection in posterior polar cataracts (true disaster…)
• Lens aspiration
• PCCC manually or using vitrector
Surgical Tips

• IOL power is calculated with SRK-T formula. Target is 20% undercorrection for children <2 years and 10% for children between 2-8 years.

• If an IOL is implanted optic capture could be performed. It is predispose in greater inflammation post surgically.

• The IOL must be inserted ideally in the bag or in the sulcus (3-piece). No AC-IOL must be used in children.
Ideally performed ACCC-PCCC
Optic capture of IOL
Surgical Rules

• A good pseudophakia is probably better of a good aphakia

• A good aphakia is definitely better of a bad pseudophakia
Surgical Tips

- Good anterior vitrectomy
- Semi total removal of the OVD
- Intracameral vigamox? or cefur
- Wound closure using 10/0 vicryl or nylon suture
- Sub conj celestone-chronodose (1/2 vial < 2 years) or intracameral triamcinolone?
Post op treatment

• Combination steroid-antibiotic eye drops/oin every 2 hours for 1st week, tapering after for 1.5 month
• Carbonic anydrase inhibitors p.o. (1/4 x 2-3) or eye drops?
• Tropicamide or cyclopentolate bid?
• Systemic steroids?
• CL for aphakia or hyperopia
Post op amblyopia treatment

Finally, surgery represents a first step in the management of cataract in children and the collaboration between parents and ophthalmologist is fundamental, to achieve optimal amblyopia treatment leading to maximum visual rehabilitation.
Thank you for your attention