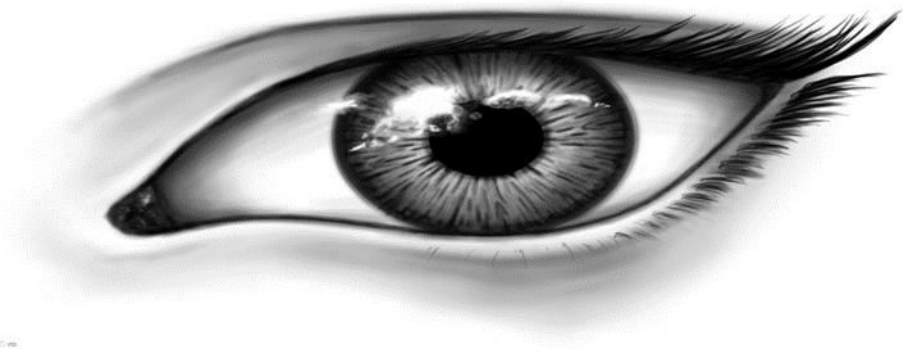
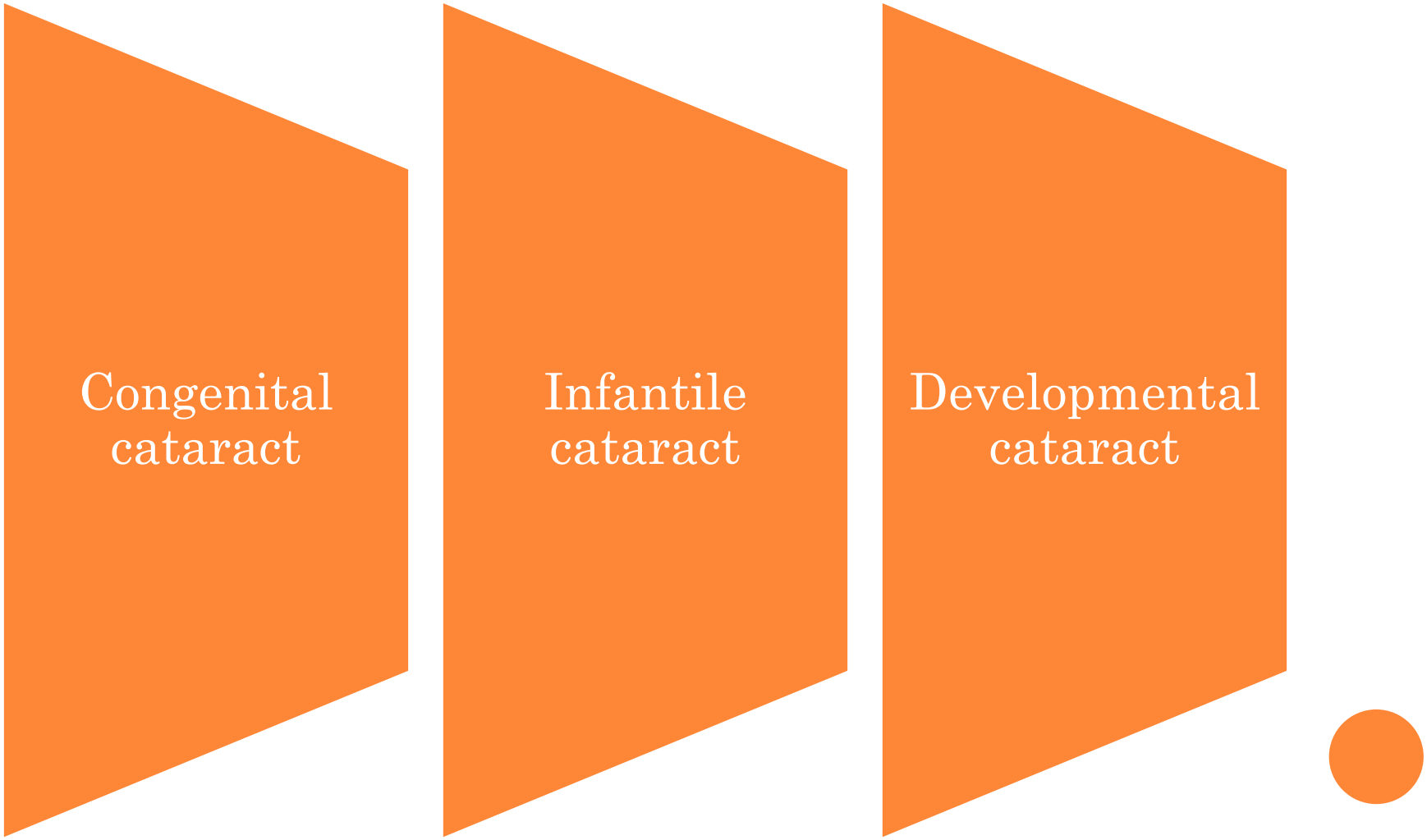
CONGENITAL CATARACT



Dr. Irfan Ullah Khattak
Pediatric Ophthalmologist
HMC

Pediatric Cataracts



Definition

- Opacity of crystalline lens matter / capsule present at birth or shortly after birth
- Congenital cataract:- at birth
- Infantile cataract:- develop during 1st year of Life..
- Both term used synonymous

CONGENITAL CATARACT

A SERIOUS CONCERN

WHY

1. One of the leading cause of blindness worldwide.

2. Increased blind person years.

3. Socioeconomic Effects.

DEMOGRAPHICS

- One of the leading causes of childhood blindness.
- The incidence ranges from **1.8 to 3.6/10,000** per year
- Median prevalence is about **1.03/10,000** children **(0.32–22.9/10,000).** [Eye (London), 2016]
- The prevalence is higher in low-income economies compared to that of high income economies.

CONTD…….

- There is no difference in the prevalence based on gender or laterality.
- Approximately 1 to 15/10,000 children worldwide, accounting for 5%-20% of childhood blindness
- Globally, about 200,000 children are blind from bilateral cataracts

(Foster et al ; Medsinge et al, 2015)

CONTD…….

- About 20,000 to 40,000 new cases of bilateral congenital cataract are diagnosed each year

  (Apple et al, 2000)

- In India, approximately 10% of childhood blindness is caused by cataract.

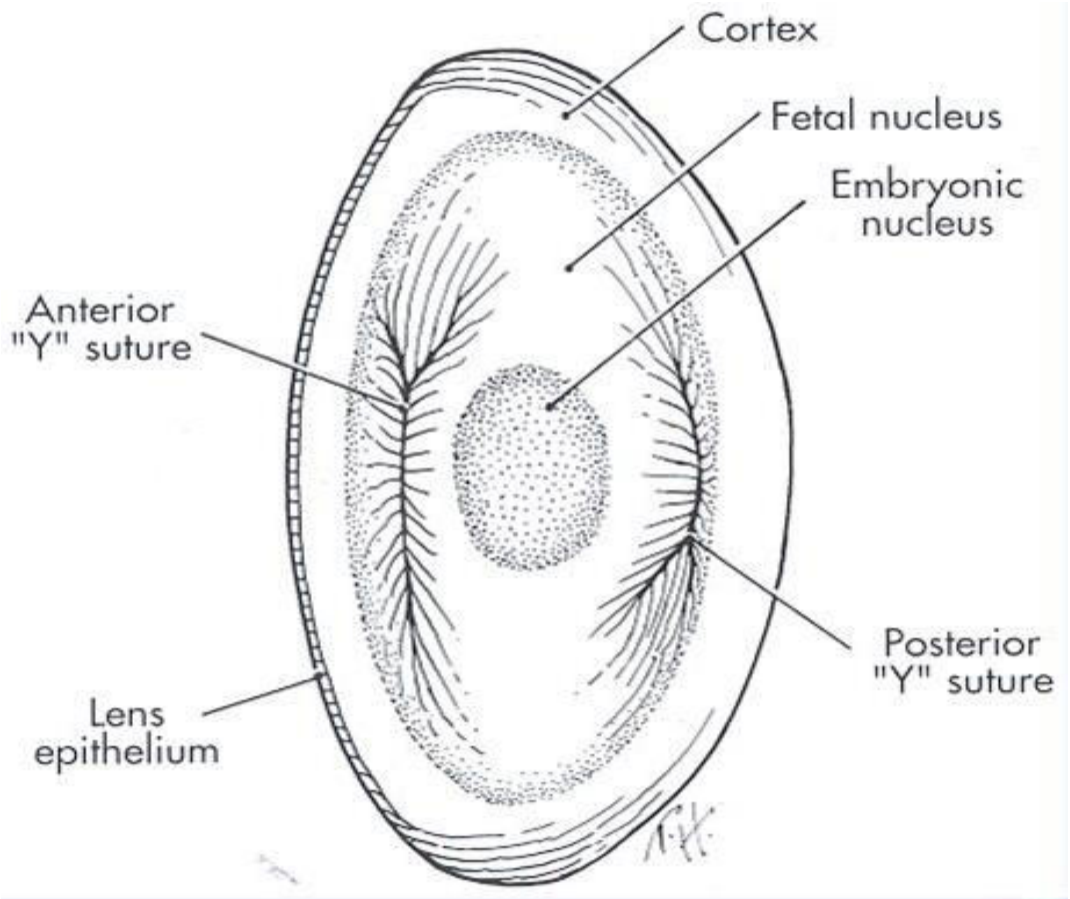
- Non traumatic cataracts(88.4%) and 11.6% were traumatic cataracts.
- (aao.org)

GLOBAL BLINDNESS

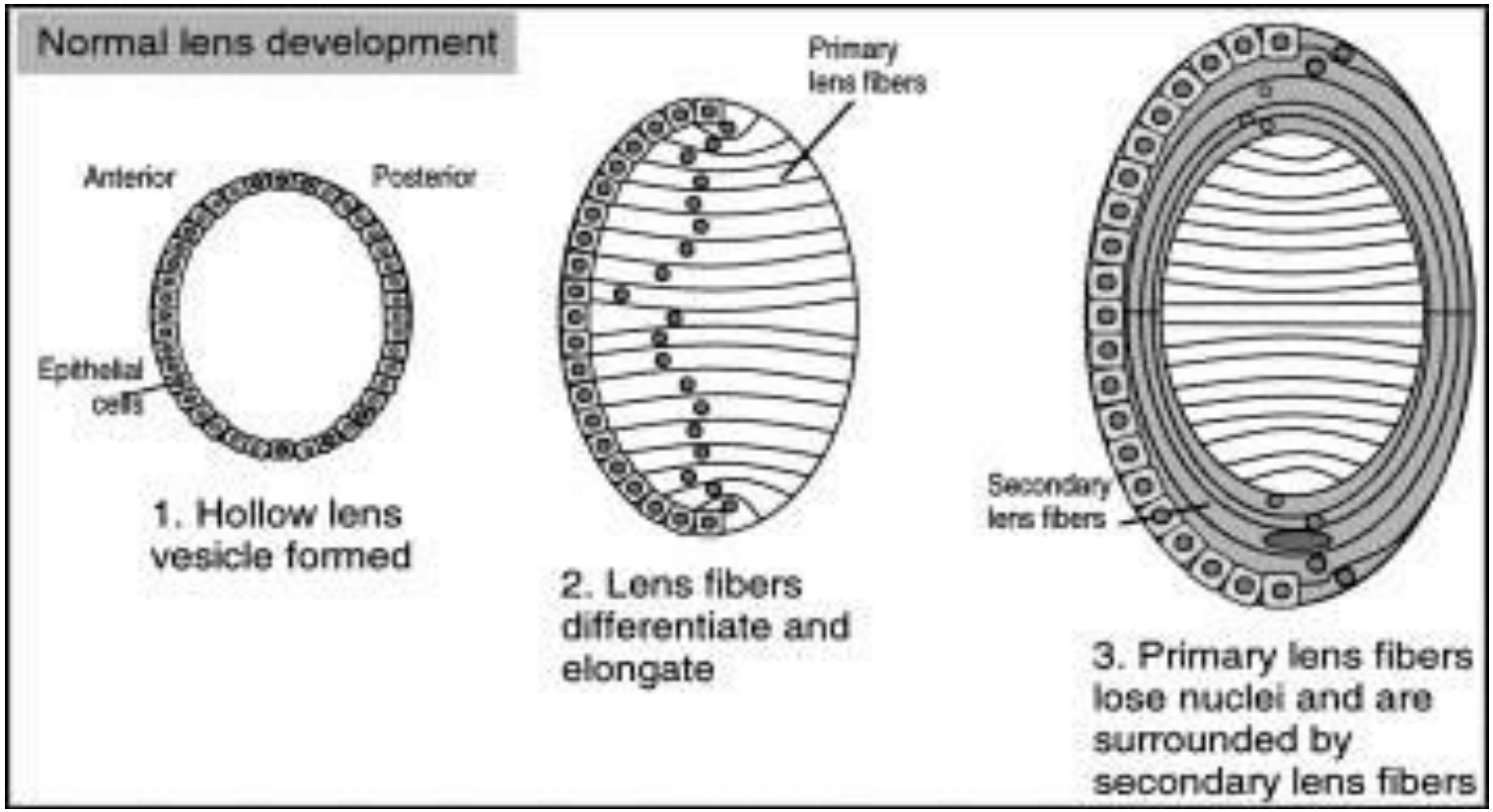
- Childhood blindness is only second to adult cataract as a cause of blind-person-years.

- Approximately 70 million blind-person years are caused by childhood blindness of which about ***10 million blind-person years (14%) is due to childhood cataract.*** (Community Eye Health Volume 17)

Cross section of juvenile lens



EMBRYOLOGY OF LENS



EMBRYOLOGY OF LENS

- **Embryonic nucleus** develop – 6week of gestation
- Arises from primitive post lens epithelium
- **Fetal nucleus-next** to develop from lens fibers from equatorial epithelial cells
- They stretch ant & post to around embryonic nucleus
- At birth both form most of lens fibers..
- Cortical Lens fiber Mostly develop postnatal

FETAL NUCLEUS

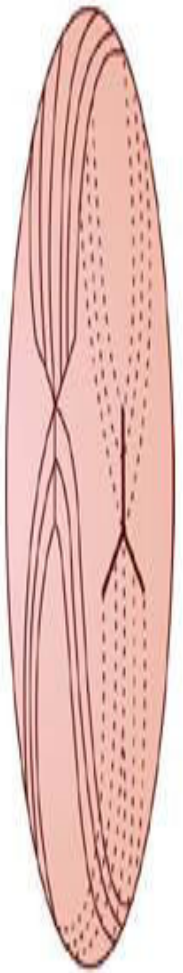
Y SUTURE is important Landmark

Fetal nucleus

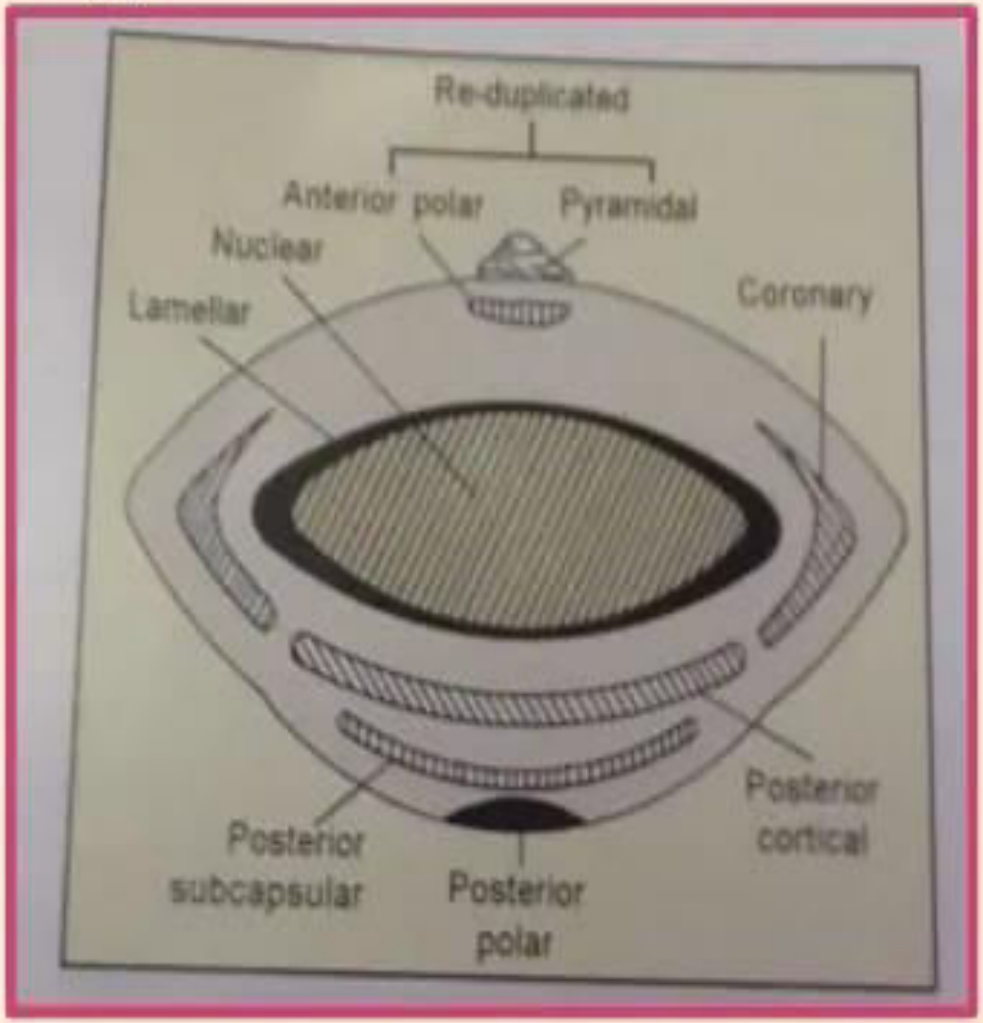
Peripheral to Y suture cortex

Within and Y SUTURE include NUCLEUS

Anterior upright Y WHILE Posterior it Inverted Y SUTURE Fibers



Morphology:



(Basak, SK., 2007)

Morphological classification

- **Anterior**:-
- Anterior polar
- Anterior pyramidal
- Anterior sub capsular cataract
- **POSTERIOR**:-
- Posterior polar cataract
- Posterior lenticonus
- Persistent fetal vasculature
- Posterior subcapsular cataract

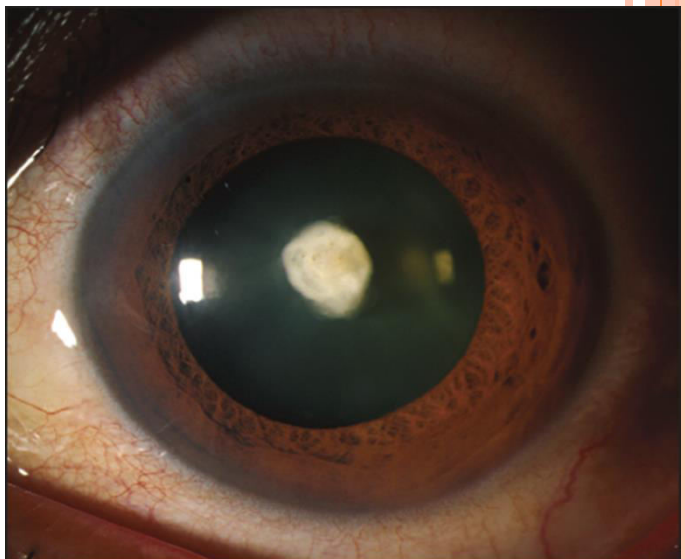
Morphological classification

- **CENTRAL** :-
- Lamellar cataract
- Sutural cataract
- Nuclear

- **DIFFUSE**:-
- Blue dot cataract(CERULEAN)
- Membranous cataract

Anterior Polar Cataract

- White opacity Locate at center anterior Capsule
- Small 1-2mm.
- Derive from abnormal separation of lens vesicle from surface ectoderm.
- 1/3 bilateral
- 90% sporadic 10% AD



Anterior Pyramidal Cataract

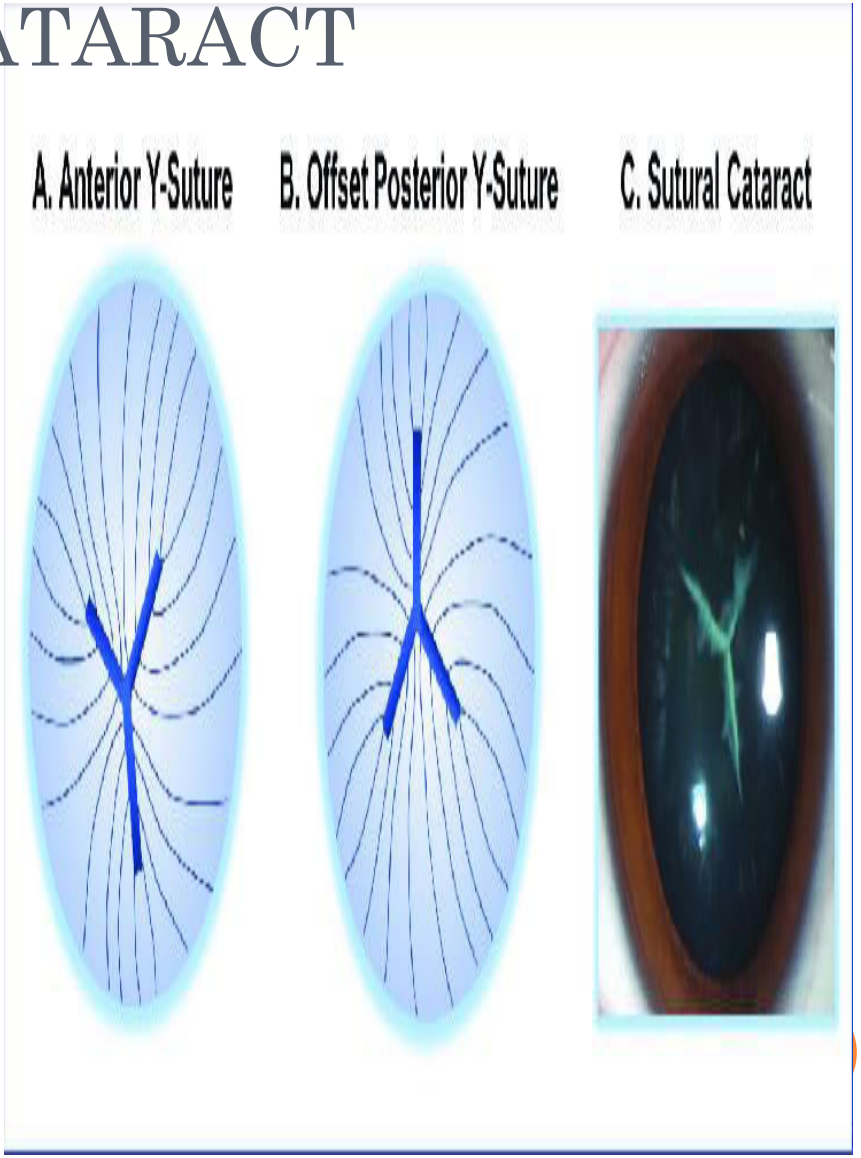
- Bilateral . Mostly Sporadic
- Type of anterior polar cataract..
- Anterior capsular fibrosis
- Conical in shape Apex projected in A/C.
- 1-2mm cone opacity.

Anterior lenticonus

- This refers to a thinned-out central anterior capsule with or without anterior cortical opacities.
- Anterior lenticonus is said to be characteristic of Alports syndrome. Spontaneous rupture of the lens can occur, resulting in a hydrated Total cataract

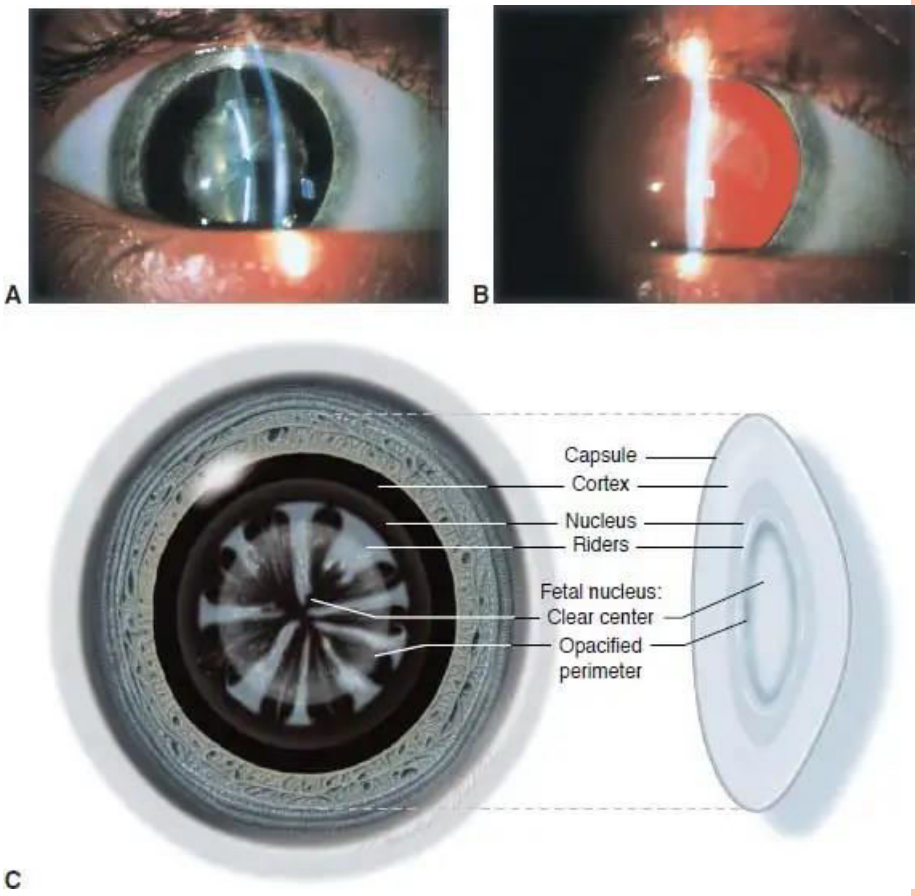
SUTURAL CATARACT

- AD
- Type of cong nuclear cataract with opacity Along Y suture in fetal nucleus.
- Progressive
- Expand into cortex and embryonic nucleus



LAMELLAR CATARACT (ZONULAR)

- Most common type
- Mostly Bilateral
- Opacification of specific zone/layer
- Layer of Opacification involving fetal nucleus surrounding clear center and surrounde in turn by layer of clear cortex

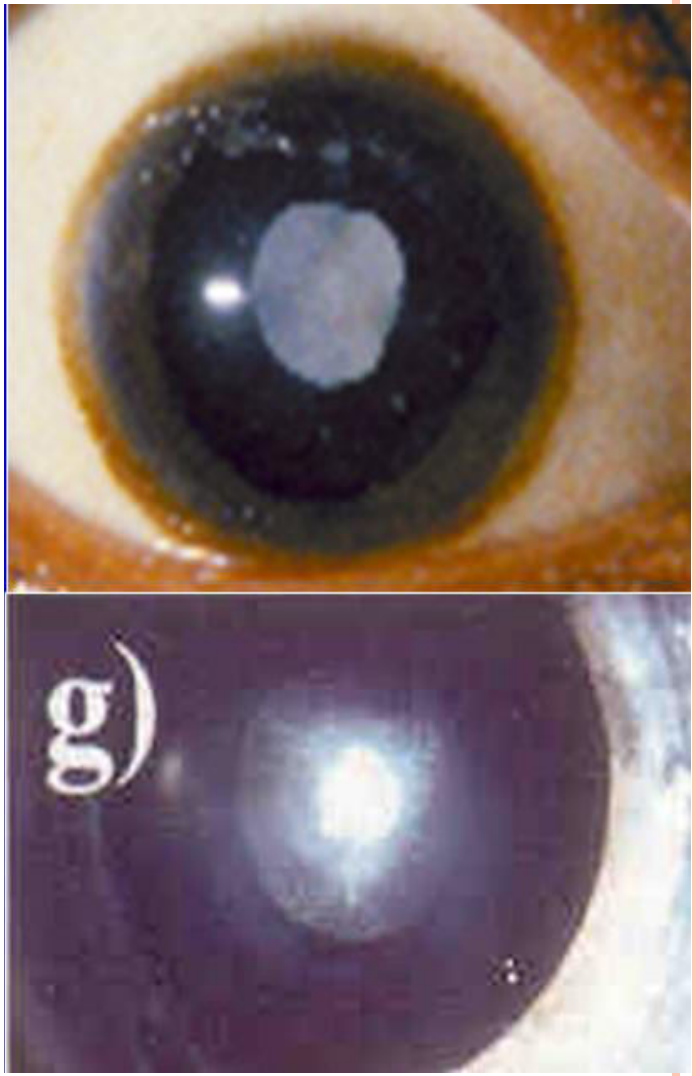


LAMELLAR CATARACT

- RIDER OPACITIES:-
  Arcuate opacity
  straddle the equator…
- Spoke of wheel

Nuclear cataract

- Opacity within embryonic nucleus or fetal nucleus..
- Mostly bilateral with AD
- Non progressive
- Congenital onset
- Common presentation intrauterine infections specially RUBELLA cataract

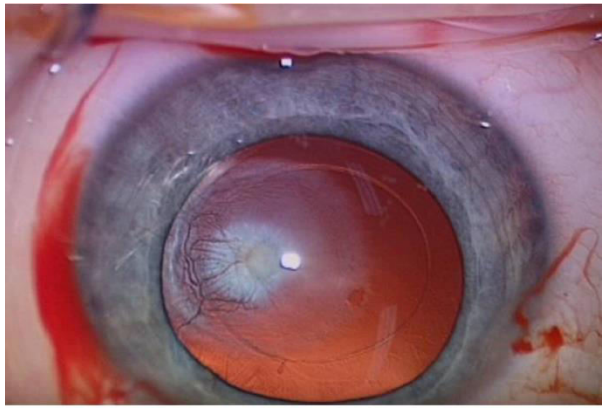


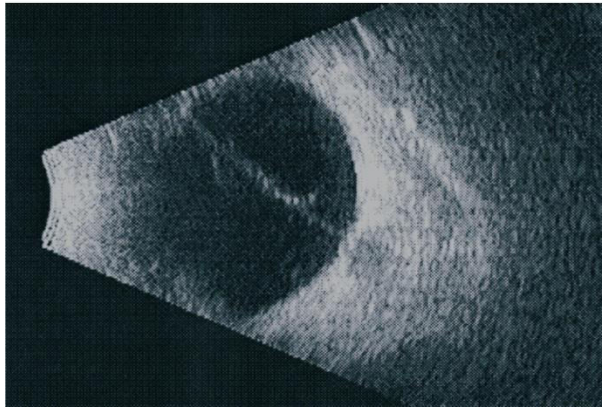
PFV
PERSISTENT FETAL VASCULATURE

- Previously (Persistent hyperplastic primary vitreous)
- MOST COMMON CAUSE OF UNILATERL CATARACT
- Isolated, sporadic
- Progressive. Anterior chamber shallowing causing Secondary glaucoma

PFV

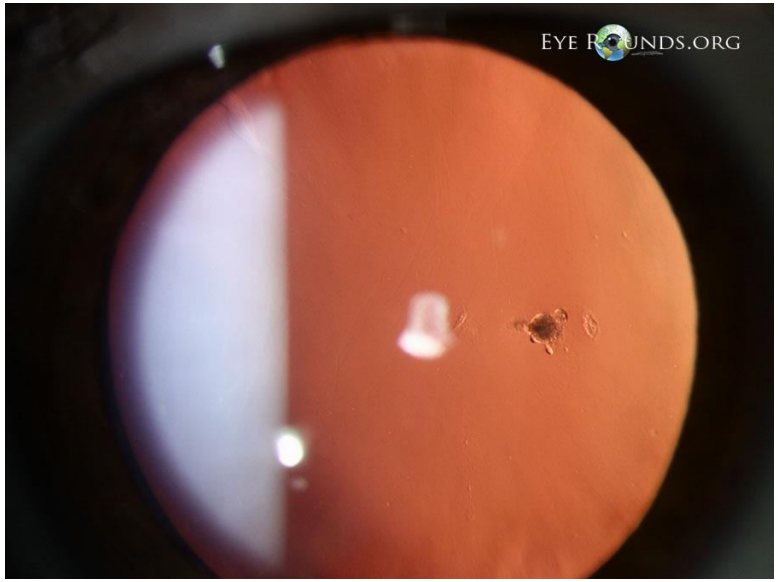
- The lens opacities in patients with PFV are generally capsular and can be associated with *shrinkage, thickening, and vascularization of the capsule*.
- There may be a posterior plaque outside or involving the lens capsule with a clear lens that must be Treated as a cataract





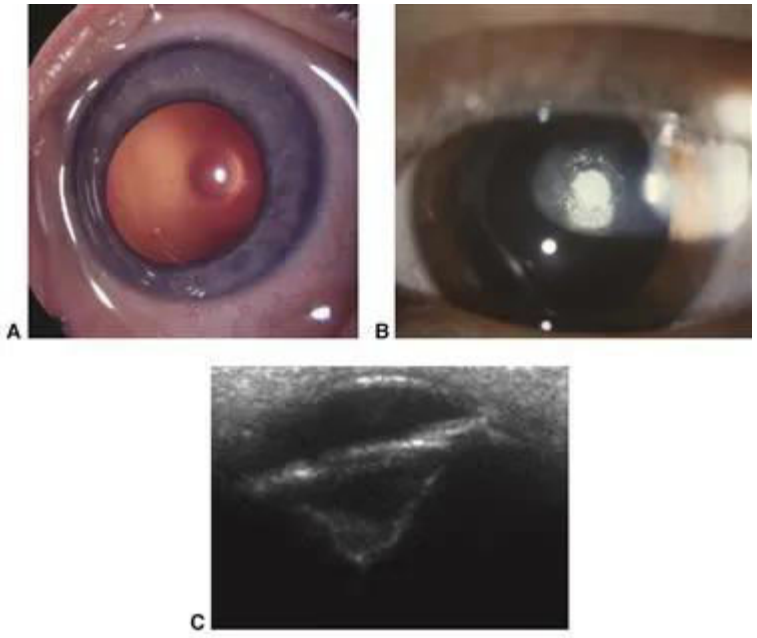
Mittendorf Dot

- Small, circular opacity on the posterior lens capsule, classically nasal in location
- Represents the anterior attachment of the hyaloid artery.
- Mildest form of PFV.
- NON Progressive, do not interfere vision



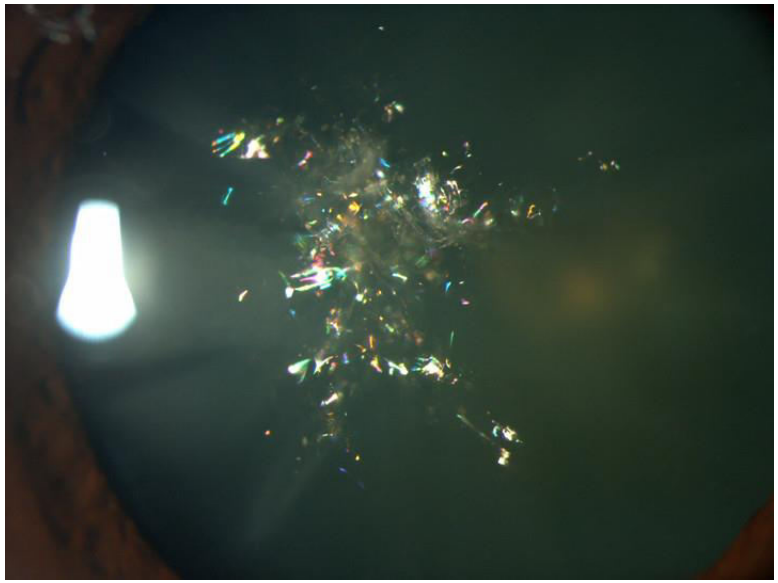
Posterior Lenticonus

- Mostly uniLateral
- Posterior capsule is thin and bulges posteriorly..
- This usually occurs at the location where the hyaloid system attaches to the eye.
- The distortion can cause a localized area of myopic refraction
- May or may not be subcapsular cortical opacification



Christmas tree cataract

- Multiple.. small flecks in cortex
- Cataract with polychromatic luster..
- Appearance of various colors
- Associated with myotonic dystropy, hypoparathyriodism



OIL DROP CATARACT

- Commonly seen in patients with galactosemia
- Bilateral
- Central aspect of Posterior lens cortex opacity with "oil droplet" appearance on retro illumination
- Restrict galactose from the diet will reverse cataract

Membranous cataract

- End stage cataract
- Lens matter absorb with ant & post capsule apposition
- TORCHS infection
- PHPV Cataract
- Long standing congenital cataract

Cerulean cataract

- Bilateral ..slowly progressing
- Scattered bluish-white opacities in cortex
- AD ..also in down syndrome
- Do not require cataract surgery

Etiology:

BILATERAL

- Idiopathic (60%)
- Hereditary (30%)
- Intrauterine infection
- Associated with ocular disorders
- Tumor
- Metabolic
- Maternal drug ingestion/ malnutrition
- Trauma

UNILATERAL

- Idiopathic (80%)
- Intrauterine infection
- Ocular abnormalities (10%)
- Trauma (9%)

(Friedman, NJ & Kaiser, PK, 2009)

Hereditary Cataract(30%)

- AD inheritance pattern is most common cause of bilateral congenital cataract
- 25% of these case represent a new AD mutation.
- AR is uncommon often associated with Consanguinity
- X-Linked cataracts are rare

Maternal Infections (3%) (TORCHS)

- Mostly Bilateral
- Dense nuclear / membranous cataracts.
- Among TORCHS RUBELLA is commonly associated with congenital cataract

TORCHS Infection

CASE STUDY
INTRO
TOXOPLASMO.
SYPHILIS
PARVO. B19
VZV
LISTERIA
RUBELLA
CMV
HSV-2
REVIEW
SUMMARY

TORCH

HIGH YIELD

* TOXOPLASMA GONDII
* OTHER AGENTS:
  - ~ SYPHILIS
  - ~ PARVOVIRUS B19
  - ~ VARICELLA ZOSTER VIRUS
  - ~ LISTERIA
* RUBELLA
* CYTOMEGALOVIRUS
* HERPES SIMPLEX VIRUS-2 (HSV-2)

HIGH YIELD

* VERTICALLY TRANSMITTED

A. via PLACENTA

B. via BLOOD, BODY FLUIDS or BREAST MILK

Rubella Cataract

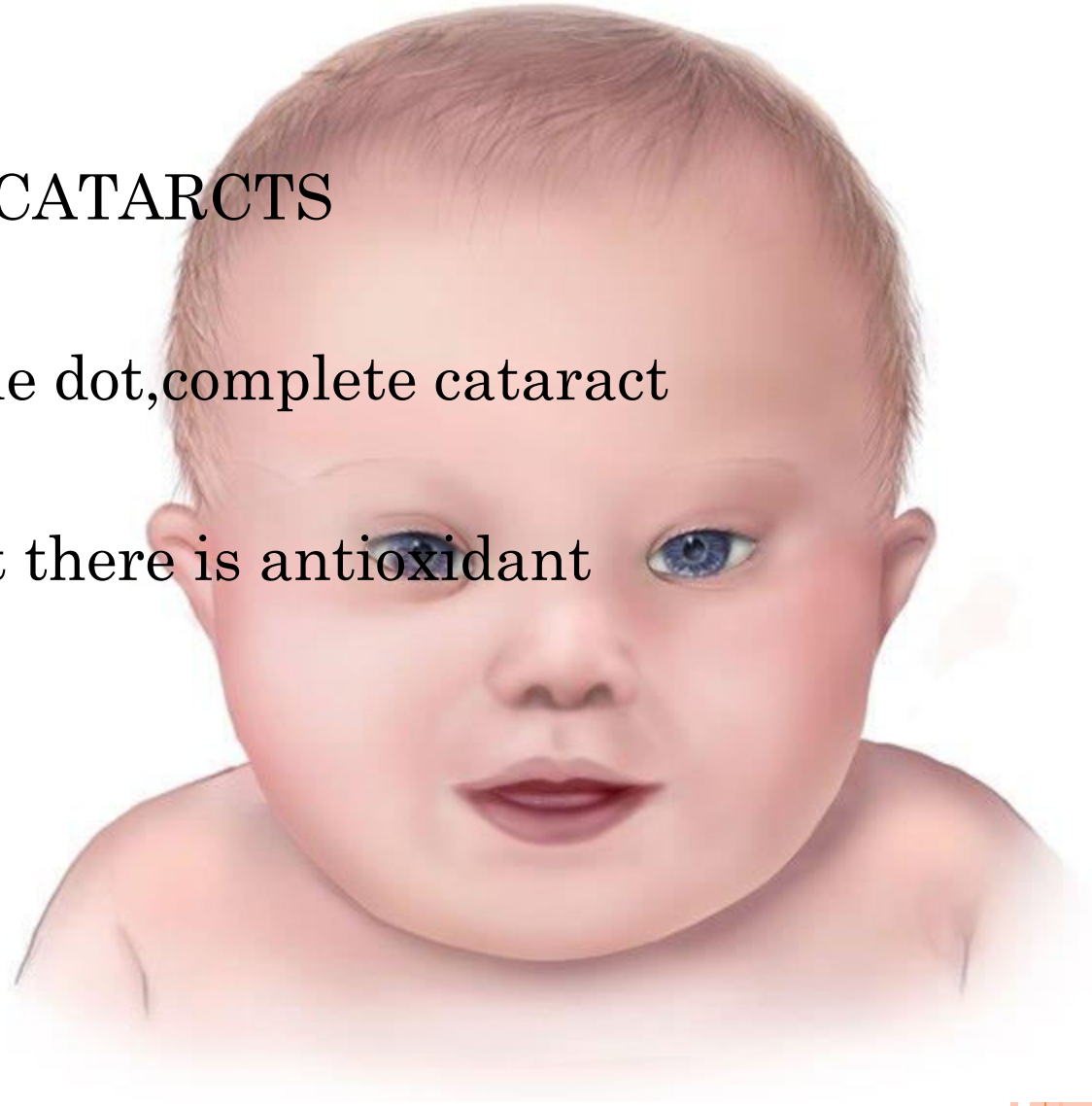
- Caused by invasion of lens by rubella virus
- 1st trimester of pregnancy
- **Ocular**:-Bil cataract, rarely unilateral
- Dense nuclear / membranous cataracts
- microphthamia, retinopathy, corneal Haze, glaucoma
- **CRS**:-Cong heart defect, hearing loss, mental retardation

Chromosomal Abnormality

- Downs syndrome (21)
- Edward syndrome (28)
- Patau syndrome (13)

DOWN'S SYNDROME

- CONG/INFANTILE CATARCTS
- Sutural ,Nuclear, blue dot,complete cataract
- In Trisomy21 patient there is antioxidant enzyme defect



METABOLIC DISEASES

- **GALACTOSEMIA**:-
- AR.
- Oil drop cataract ,mental retardation,GI distrabance
- Galactose free milk
- **FABRY SYNDROME**:-
- X-LR
- Error of glycosphingolipid
- Posterior sub capsular/ anterior subcapsular

Metabolic diseases

- Alpha **Mannosidosis**:-
- AR
- Defective degradation of lipoproteins
- Mental redardation,coarse facies
- Cataracts
- **Refsum disease:-**
- AR
- Accumulation of phytanic acid
- RP with cataract

Wilson Disease

- AR.
- DEFECCTIVE Inborn error of Cu metabolism ..into ceruloplasim
- Cu accumulation at anterior capsule..
- Sunflower cataract resolve with Treatment (penicillamine)

Sunflower cataract of Anterior capsule

Cataract with prematurity

- Transient lens opacities noted in premature infants
- Bilateral
- Some cataracts appear after argon laser for ROP
- RESOLVE spontaneously after2-4 weeks

Renal diseases

Low syndrome

- X-LR
- Oculocerebrorenal
- Congenital catarcts with glaucoma

Alports syndrome

Unilateral cataracts

- Idiopathic (80%)
- Ocular abnormalities (10%)
- A.Post Lenticonus
- B.PHPV
- C.ANT.segment dysgensis
- Masked bilateral cataract(6%)
- Traumatic (4%)(must rule out child abuse)

ASSESSMENT

- History
- Observation of fixation and following reflexes
- Forced Choice Preferential Looking
- OKN
- VEP

NON SURGICAL

- Dilating Drops
  - Small cataract
  - Partial cataract
- Optical Correction
  - Lamellar cataract

Visual assessment

- Anterior capsule opacities are not visually significant
- Posterior capsule opacities are usually visually significant
- Small axial cataract often maintain good vision if pupil is dilated continuously

INDICATIONS FOR SURGERY

- When the visual defect in the child with congenital cataract is sever enough to interfere significantly with visual development
- Dense and total cataract
- Partial cataract

Preparation

- GA fitness
- Biometry
- B. Scan
- Blood Tests

- Informed consent
- Post op care
- Follow up

Timing of surgery

- Unilateral
  - 4-6 weeks
- Bilateral
  - 6-8 weeks

Surgical management

- LMA+ PC+AV± IOL

Complications of surgery

- Operative and immediate postoperative complications
  - Hyphaema
  - Posterior capsule rapture
  - Vitreous loss
  - Endothelial cell loss
  - Endophthalmitis

Complications of surgery

- Late postoperative complications
  - Posterior capsule opacification
  - Retinal detachment (1-10%)
  - Glaucoma (30%)
  - CMO
  - Vitreo-retinal haemorrhage
  - Corneal oedema

Optical correction

- The changing environment
- Spectacles
  - Can be worn at any age
  - Not unduly expensive
  - Can be readily changed
  - Safe
  - Make microphthalmic eyes appear normal
  - May be the form of optical correction available in a community

Optical correction (cont'd)

- Contact lenses
  - Well established method for unilateral aphakia
  - Silicon hydrogel lenses for extended wear
- Primary IOLs
- Secondary IOLs
- Epikeratophakia

Optical correction (cont'd)

- Primary IOLs
  - Calculation is difficult, may be done UGA
  - Full correction /under correction
  - Choose IOL acrylic hydrophobic, heparin coated
- Secondary IOLs

Amblyopia Therapy

- Occlusion
- Penalization

Thank you