# Paediatric Eye Disease & Assessment:

# Red flags and common complaints


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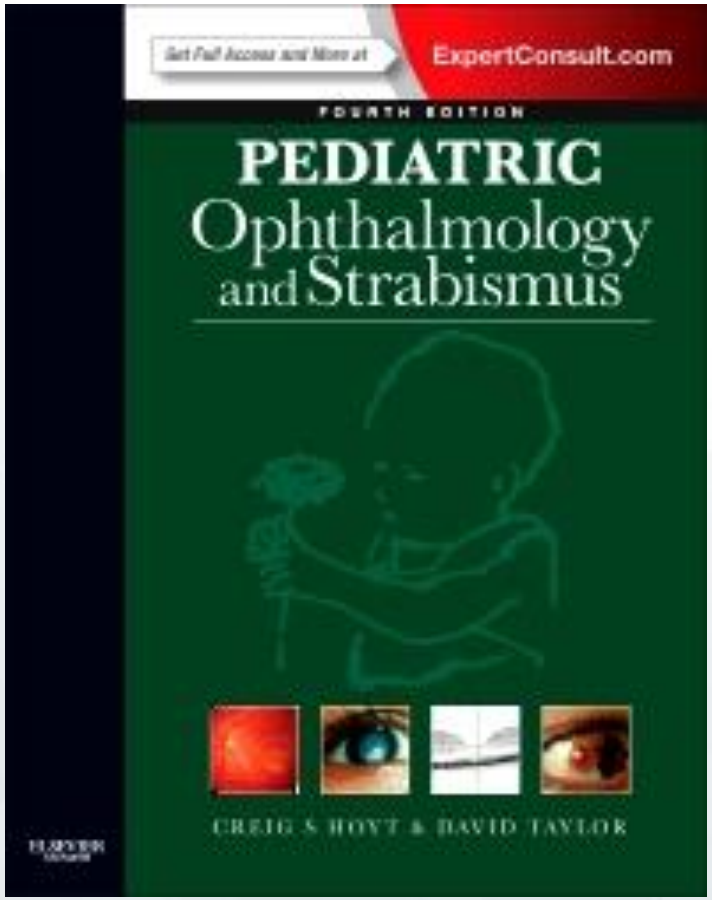
# Overview

**BRIEF:**

- Most common paediatric clinical presentations, treatments what to look for

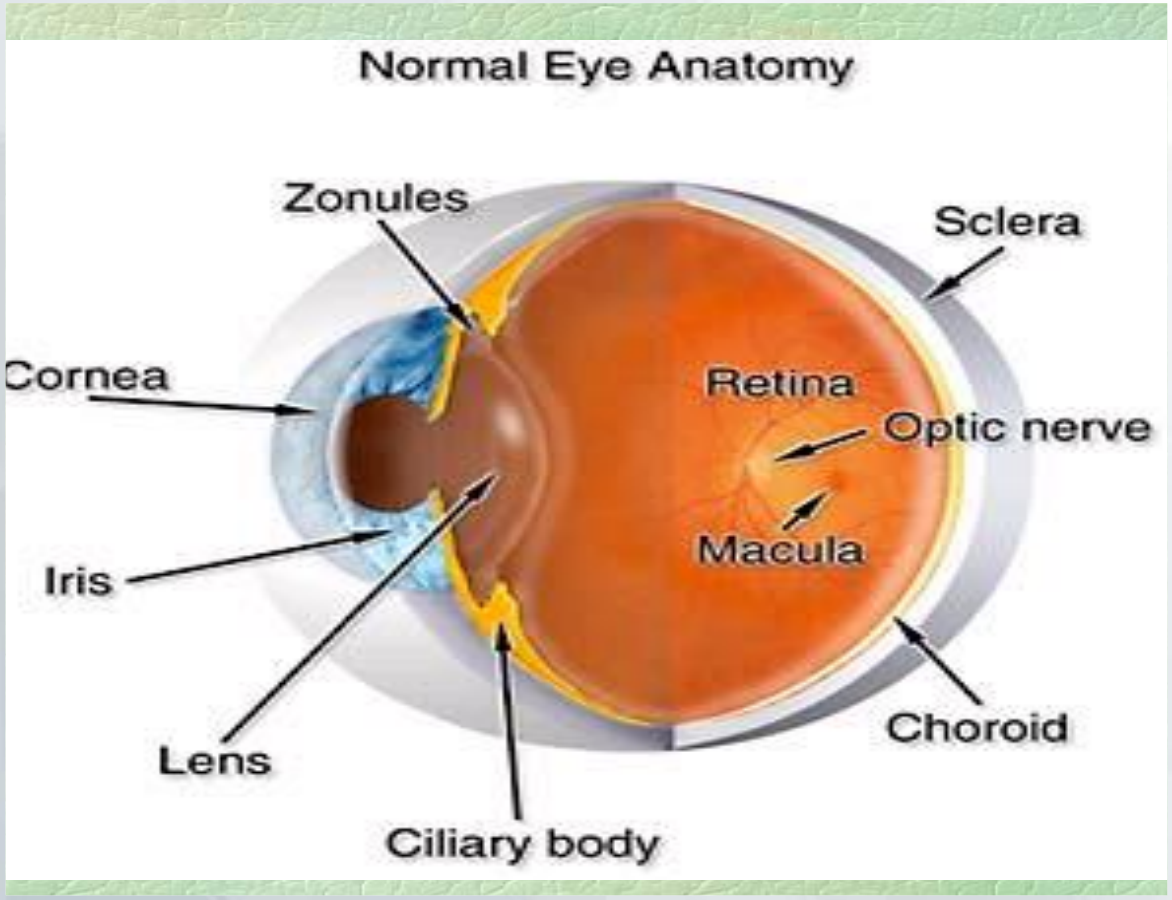
  - Obvious – benign
  - Obvious – serious / visually significant
  - Obvious – benign or serious?
  - Less obvious – serious & important



# Basic eye anatomy

## Anterior segment – 1/3

- Adnexa
  (lids/brow/lacrimal apparatus)
- Cornea
- Sclera
- Iris
- Posterior chamber
- Ciliary body
- Lens




## Posterior segment – 2/3

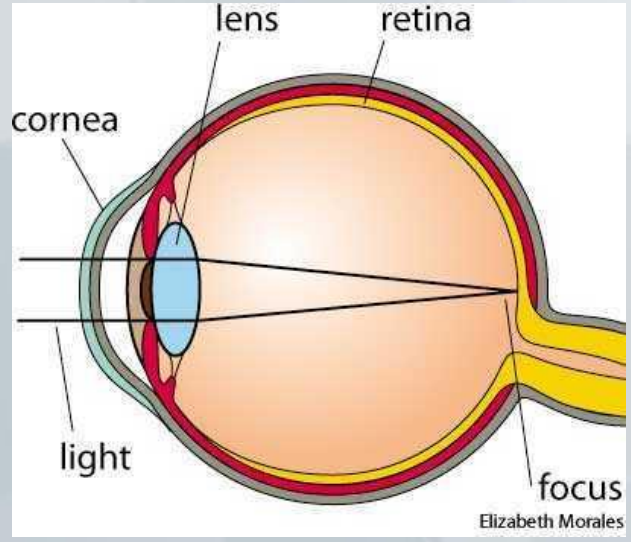
- Vitreous
- Retina
- Optic Nerve

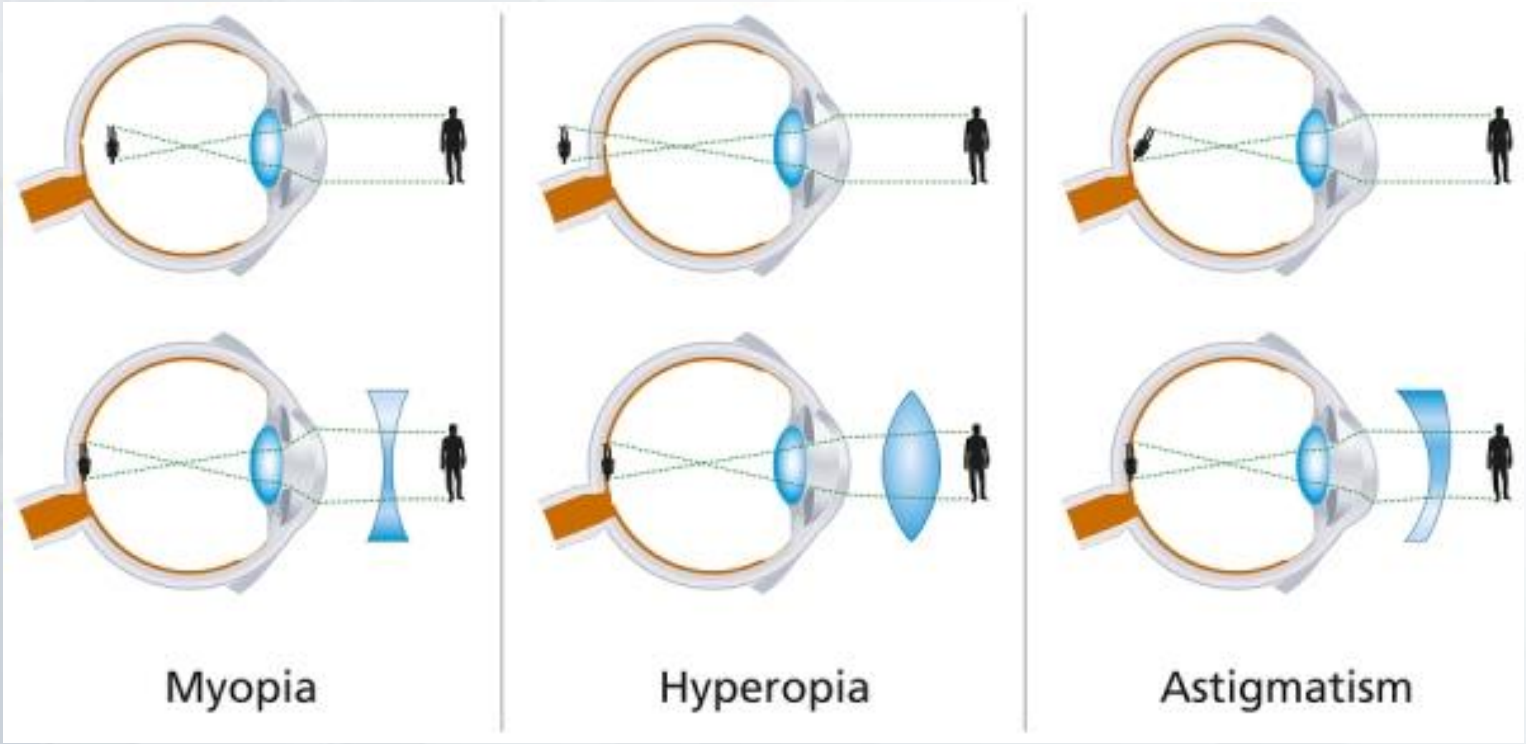
**Posterior pole*

- b/w macula and optic nerve

# Refractive error

**Emmetropia:** No refractive error




Short sighted
'larger' eye
Too long

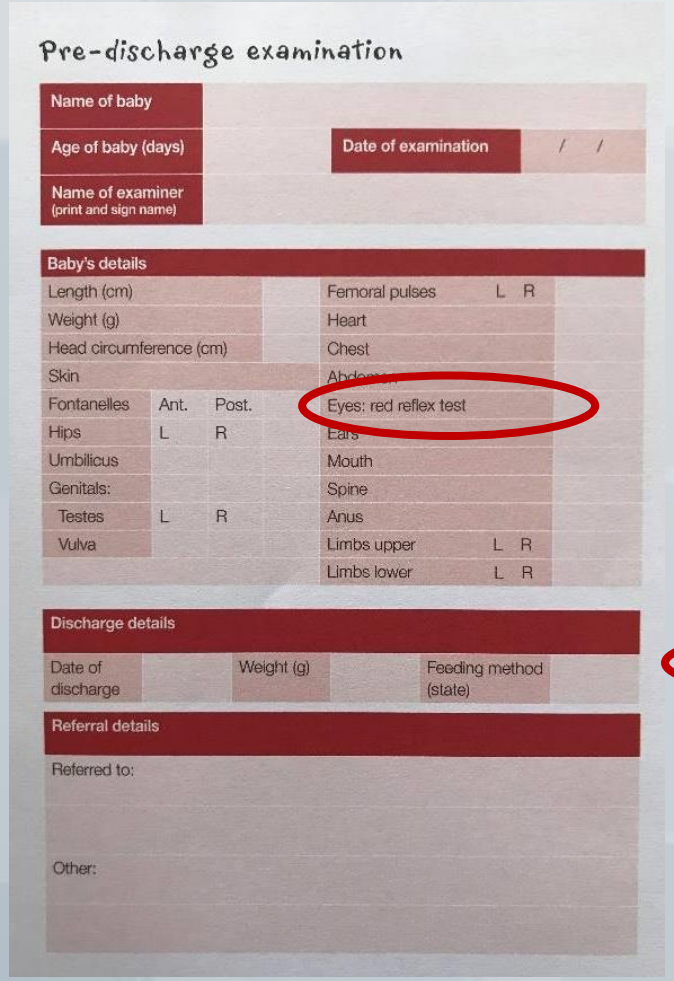
Long sighted
'smaller' eye
Too short

Asymmetrical cornea
'football'

# Screening for paediatric eye disease

The Royal Children's Hospital Melbourne

## RED-REFLEX TEST



Pre-discharge examination

| | |
|---|---|
| Name of baby | |
| Age of baby (days) | Date of examination / / |
| Name of examiner (print and sign name) | |

**Baby's details**

| | | | | |
|---|---|---|---|---|
| Length (cm) | | | Femoral pulses | L R |
| Weight (g) | | | Heart | |
| Head circumference (cm) | | | Chest | |
| Skin | | | Abdomen | |
| Fontanelles | Ant. | Post. | Eyes: red reflex test | |
| Hips | L | R | Ears | |
| Umbilicus | | | Mouth | |
| Genitals: | | | Spine | |
| Testes | L | R | Anus | |
| Vulva | | | Limbs upper | L R |
| | | | Limbs lower | L R |

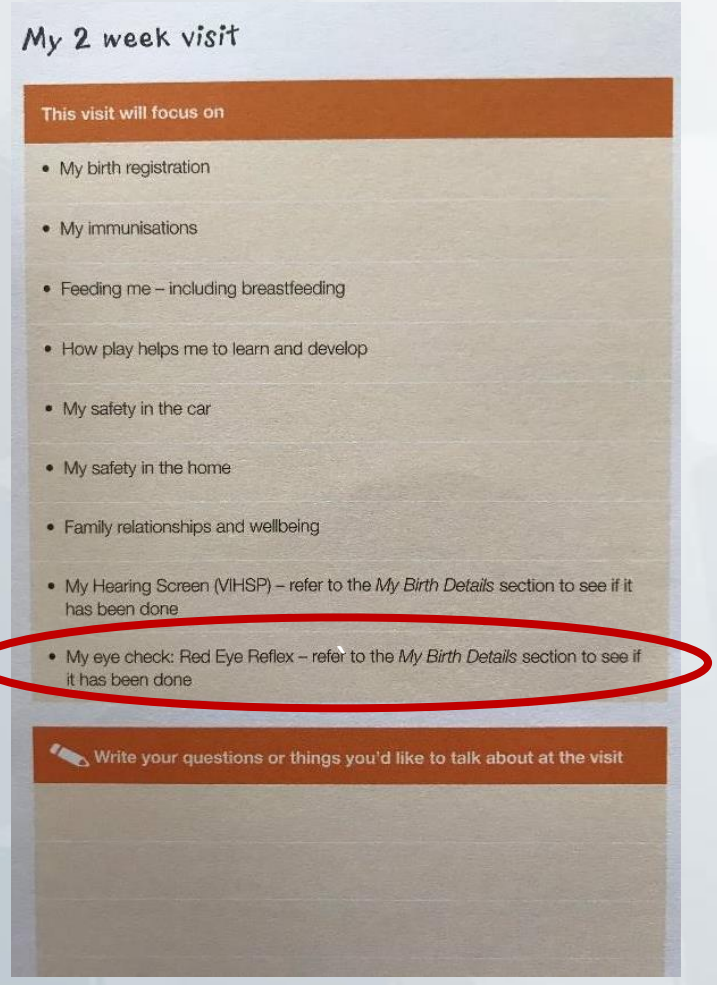
**Discharge details**

| | | | | | |
|---|---|---|---|---|---|
| Date of discharge | | Weight (g) | | Feeding method (state) | |

**Referral details**

Referred to:

Other:



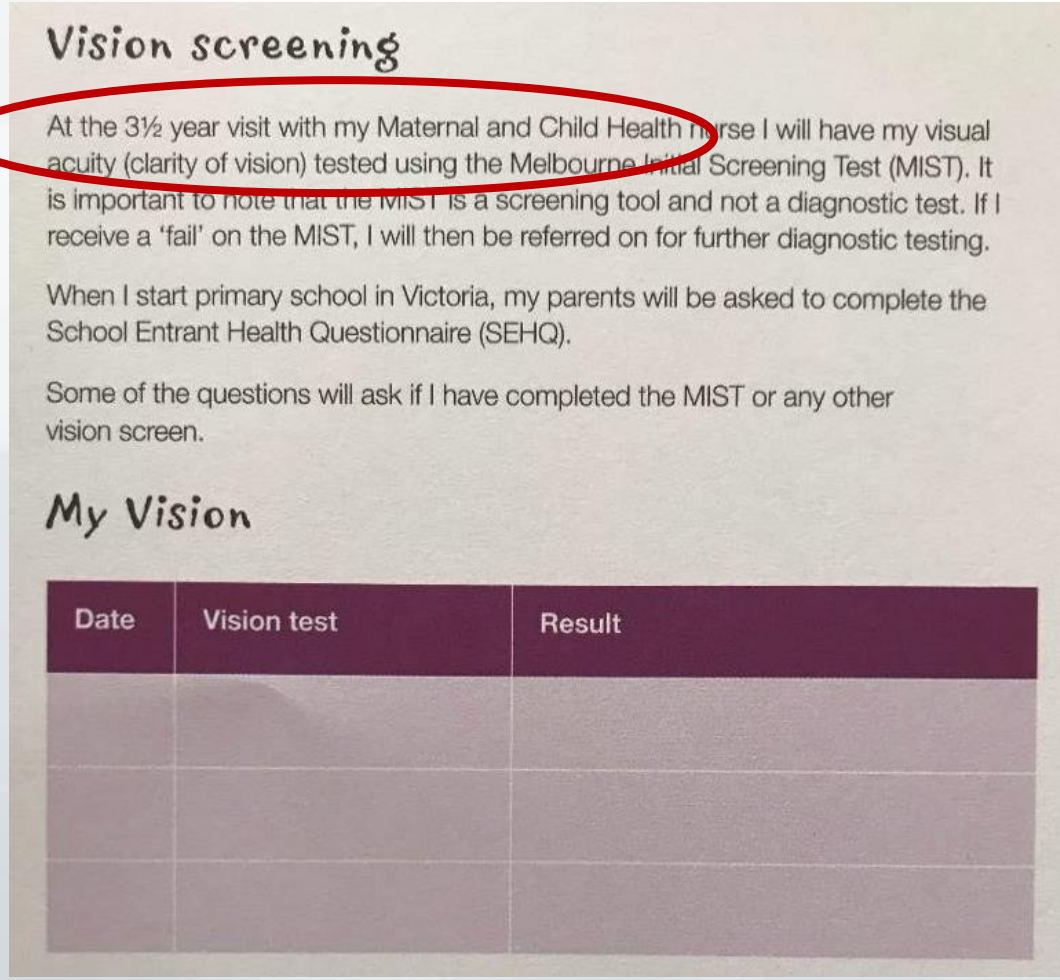
My 2 week visit

**This visit will focus on**

- My birth registration
- My immunisations
- Feeding me – including breastfeeding
- How play helps me to learn and develop
- My safety in the car
- My safety in the home
- Family relationships and wellbeing
- My Hearing Screen (VIHSP) – refer to the *My Birth Details* section to see if it has been done
- My eye check: Red Eye Reflex – refer to the *My Birth Details* section to see if it has been done

Write your questions or things you'd like to talk about at the visit

## VISUAL ACUITY



Vision screening

At the 3½ year visit with my Maternal and Child Health nurse I will have my visual acuity (clarity of vision) tested using the Melbourne Initial Screening Test (MIST). It is important to note that the MIST is a screening tool and not a diagnostic test. If I receive a 'fail' on the MIST, I will then be referred on for further diagnostic testing.

When I start primary school in Victoria, my parents will be asked to complete the School Entrant Health Questionnaire (SEHQ).

Some of the questions will ask if I have completed the MIST or any other vision screen.

My Vision

| Date | Vision test | Result |
|---|---|---|
| | | |
| | | |
| | | |

# Detecting paediatric eye disease




Congenital

Developmental

BIRTH

3 ½ y

7-8 y

Red Reflex
0-6/52

MIST

# What do *you* need to know?

- NORMAL **V** ABNORMAL EYES
  - NORMAL visual behaviour
  - NORMAL ocular alignment
  - NORMAL eye movement
  - NORMAL basic eye structure
- IDENTIFY ‘AT RISK’ CHILDREN – FHx eye disease

**Not all problems have SYMPTOMS – but there will be SIGNS!**

- Obvious – benign
- Obvious – serious / visually significant
- Obvious – benign or serious?
- Less obvious – serious and important

# Obvious - benign

## Stye  or Chalazion

- Variable severity
- Annoying – not painful
- May resolve spontaneously
- May require Rx:
  - ? warm compress
  - Topical or Oral Antibiotics
  - I&C under GA
  - Mx by GP initially

- Most *unlikely* to impact on vision

- Obvious – benign
- Obvious – serious / visually significant
- Obvious – benign or serious?
- Less obvious – serious and important

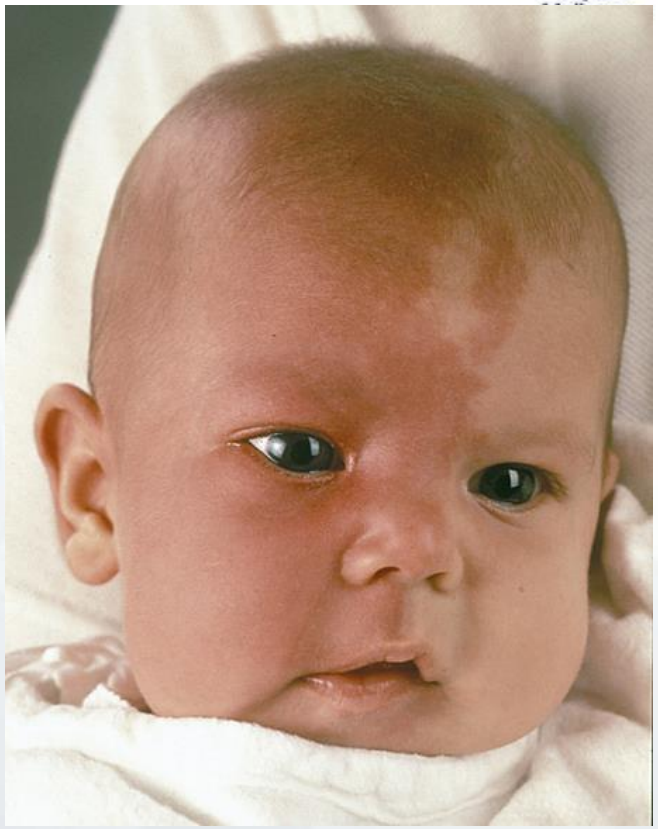
# Obvious – serious / visually significant

**Capillary haemangioma**

- Occlude visual axis
- Induces astigmatism
- Amblyopia

- Treatment – *conservative*
  - Refractive error / amblyopia

- Treatment – *active*
  - Topical/systemic beta-blockers
  - Local/systemic steroids
  - Sx excision
  - Radiation
  - Laser
  - Injection sclerosing agents



- Sturge-Weber syndrome
- 2nd Glaucoma
- Long-term surveillance for glaucoma and Rx PRN

# Obvious – serious / visually significant

**Ptosis (drooping eyelid)**

- Visual axis
- Head posture (AHP)
- Induces:
  - Astigmatism
  - Amblyopia

- Treatment – *conservative*
  - manage refractive error & amblyopia
  - +/- Sx when older PRN

- Treatment – *active*
  - Surgery
    - if visual axis occluded
    - AHP – interferes with motor development

- Obvious – benign
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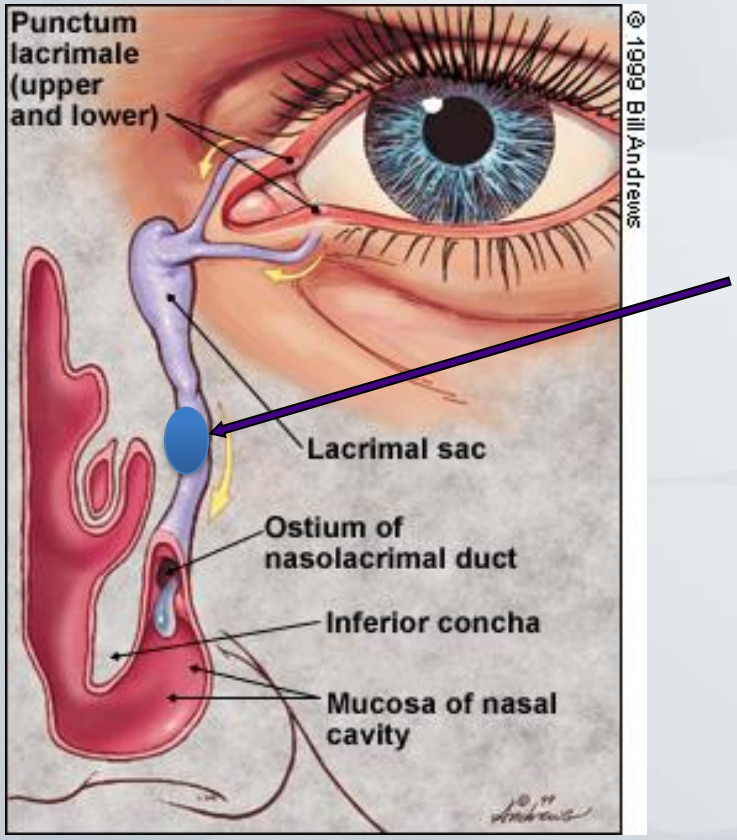
# Watery Eyes

# Obvious – benign – nasolacrimal duct obstruction (NLDO)




- ~ 20% of infants
- Epiphora
- +/- mucopurulent discharge

# Obvious – benign – nasolacrimal duct obstruction (NLDO)

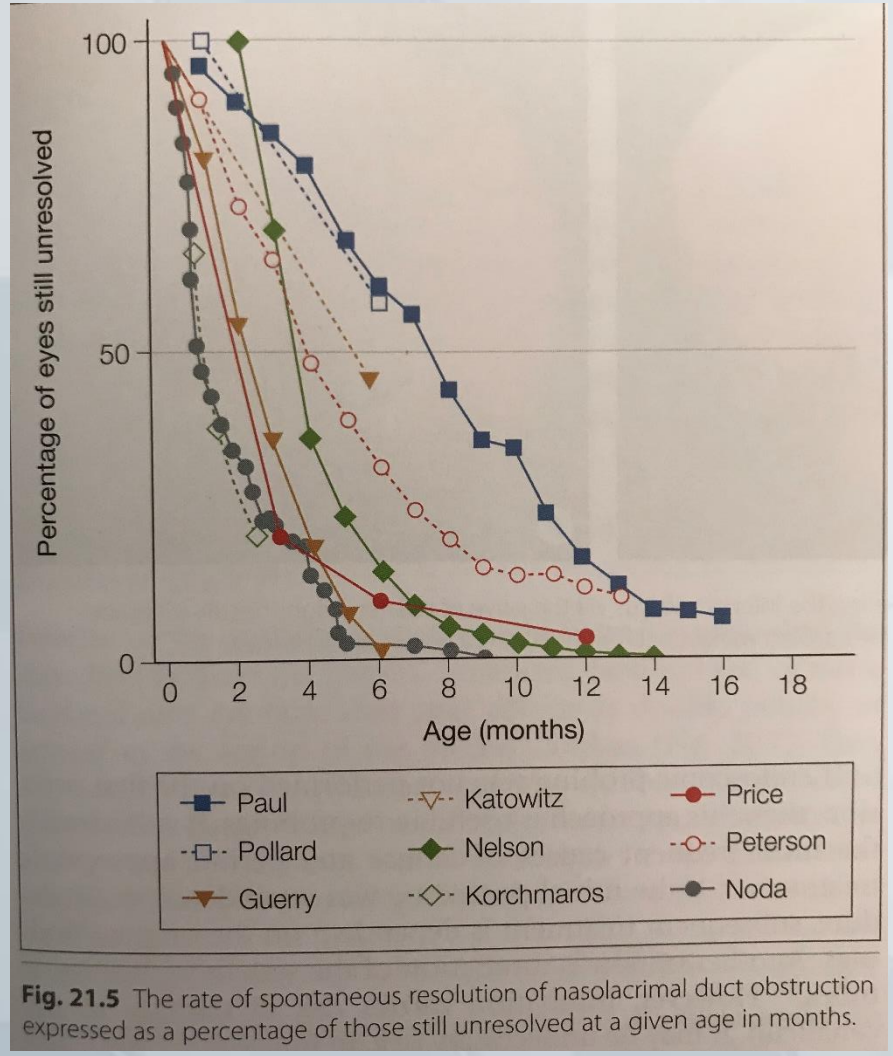



**Fig. 21.5** The rate of spontaneous resolution of nasolacrimal duct obstruction expressed as a percentage of those still unresolved at a given age in months.

- Treatment – *conservative*
  - ~ 12/12 of age
  - Eye toilet – saline; dry
  - Massage
  - +/- g/oc antibiotic for local infection
  - NOT conjunctivitis
- Treatment – *active*
  - Probe & syringe (Dx and Tx)
  - Intubation – Crawford tube
  - Dacryocystorhinostomy
- Treatment – *indications*
  - Unresolved epiphora
  - *social

**Fluorescein dye disappearance test**

- Dye normally disappears by 5 minutes
- Retained dye = obstruction
- Mucocoeles – pressure on lacrimal sac produces reflux of fluorescein stained mucous

*Hoyt & Taylor 2013 Ped. Oph. & Strab. 4th Ed.*

# Obvious – benign/serious – cong. dacryocystocoele

- Tense, bluish swelling below the medial canthus
- Obstruction – breathing difficulties

- Treatment – *conservative*
  - 1st 2 weeks of life – watch & wait
  - Most spontaneously resolve

- Treatment – *active*
  - Endoscopic drainage
  - +/- excision nasal mucosa over dacryocystocoele

- Treatment – *indications*
  - Breathing difficulties
  - Acute dacryocystitis

# Obvious – benign/serious – epiblepharon

- Tight lower lids
- Soft newborn lashes V course adult lashes
- Epiphora/rubbing/+/- photophobia
- Ethnic variation

- Treatment – *conservative*
  - Watch & wait
  - +/- ocular lubricants
  - Spontaneous resolution ~ 5-6 yo

- Treatment – *active*
  - Surgical – Quickert sutures

- Treatment – *indications*
  - Corneal ulceration/scarring

# …but when do I worry?

- ***RED EYE***
- ***LIGHT SENSITIVE***
- ***Unsettled baby/pain***
- ***‘sick’***

# Obvious – serious – preseptal cellulitis

- 5 x more common than orbital cellulitis, esp. under 5-6 years
- Associated with:
  - Lid & cutaneous infections (stye, varicella, dacryocystitis HSV
  - URTI and sinusitis
  - Lid trauma
- Generally ‘unwell’, febrile

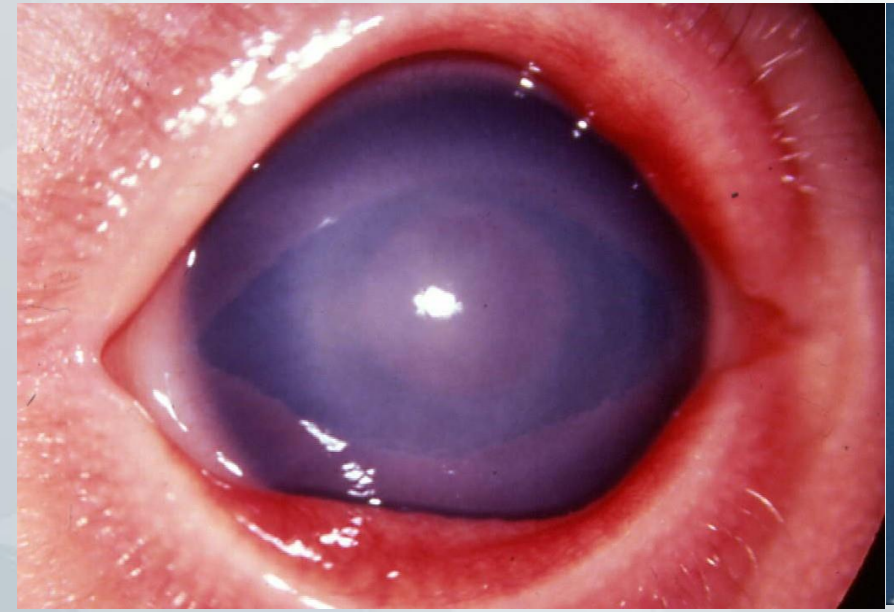
- Treatment – *conservative*
  - Oral antibiotics

- Treatment – *active*
  - IV antibiotics
  - ? CT – assess orbital/sinus/brain involvement

- *RAPIDLY* progresses ORBITAL CELLULITIS
  - ➢ *Urgent*
  - ➢ *Blinding*

# Obvious – serious – congenital/infantile glaucoma

- Epiphora / photophobia
- Opaque cornea
- Buphthalmos
- Unsettled/vomiting



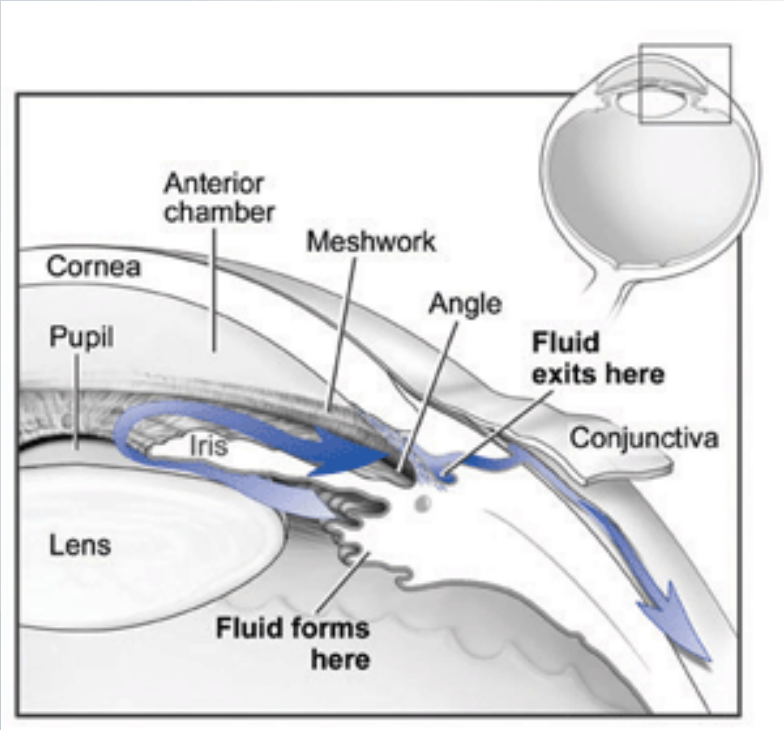
Descemet's membrane splits

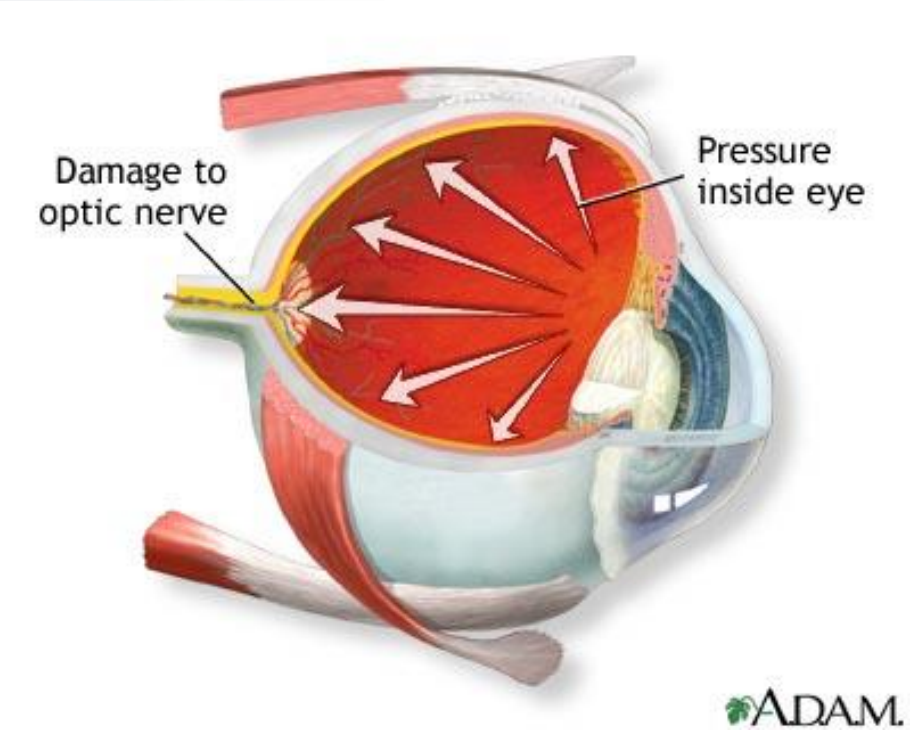
- Influx of aqueous into corneal stroma

Buphthalmos – "ox" [large] eye

- Infant sclera stretches

# Pathophysiology - glaucoma




Types:

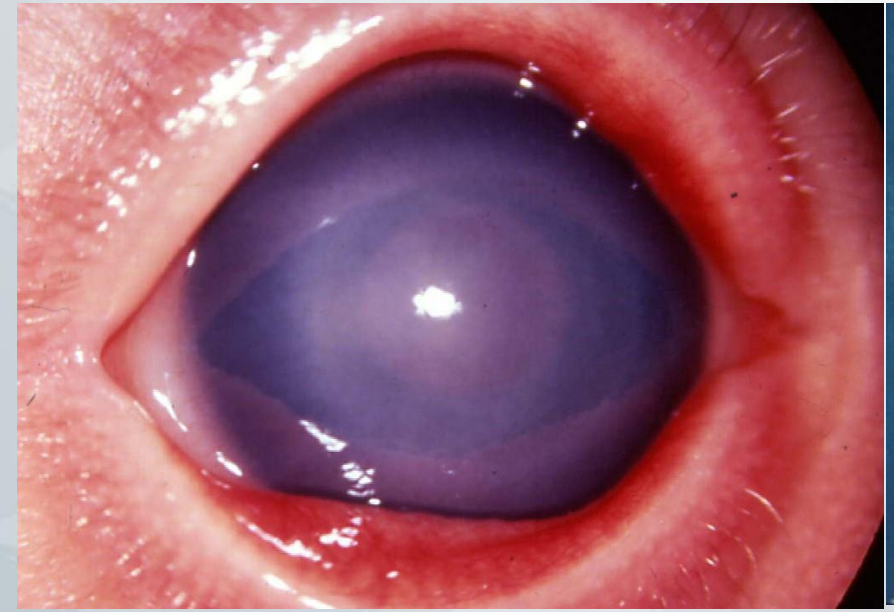
- POAG – primary open angle
- AAC – acute angle closure
- Secondary – trauma/inflammation
- *Congenital*
- Familial/hereditary

Characterised by:

- Raised intraocular pressure [IOP]
- Visual field loss
- Congenital glaucoma
  - Opaque cornea
  - Epiphora
  - Photophobia

# Obvious – serious – congenital/infantile glaucoma

- Syndromic associations
- Familial/Hereditary
- Difficult to control
  - Surgery
  - Topical eye drops
- Induces:
  - Myopia
  - Amblyopia
  - Optic nerve damage
  - Visual field defects



Descemet's membrane splits

- Influx of aqueous into corneal stroma

Buphthalmos – "ox" [large] eye

- Infant sclera stretches

# Unequal Pupils

# Obvious – benign/serious – anisocoria (unequal pupils)

Physiological anisocoria
- ~ 20% of infants
- Minimal difference
- No change in dark

Horner's syndrome
- Anisocoria – increases in dark – affected side doesn't dilate
- Ptosis
- Heterochromia

- ?? Neuroblastoma
  - Most common extracranial solid tumour
  - 9% of all childhood cancers, 33% of deaths
  - Pain/fever/weight loss
  - Cerebellar signs
  - Diarrhoea
  - Hypertension with flushing – check catecholasmines

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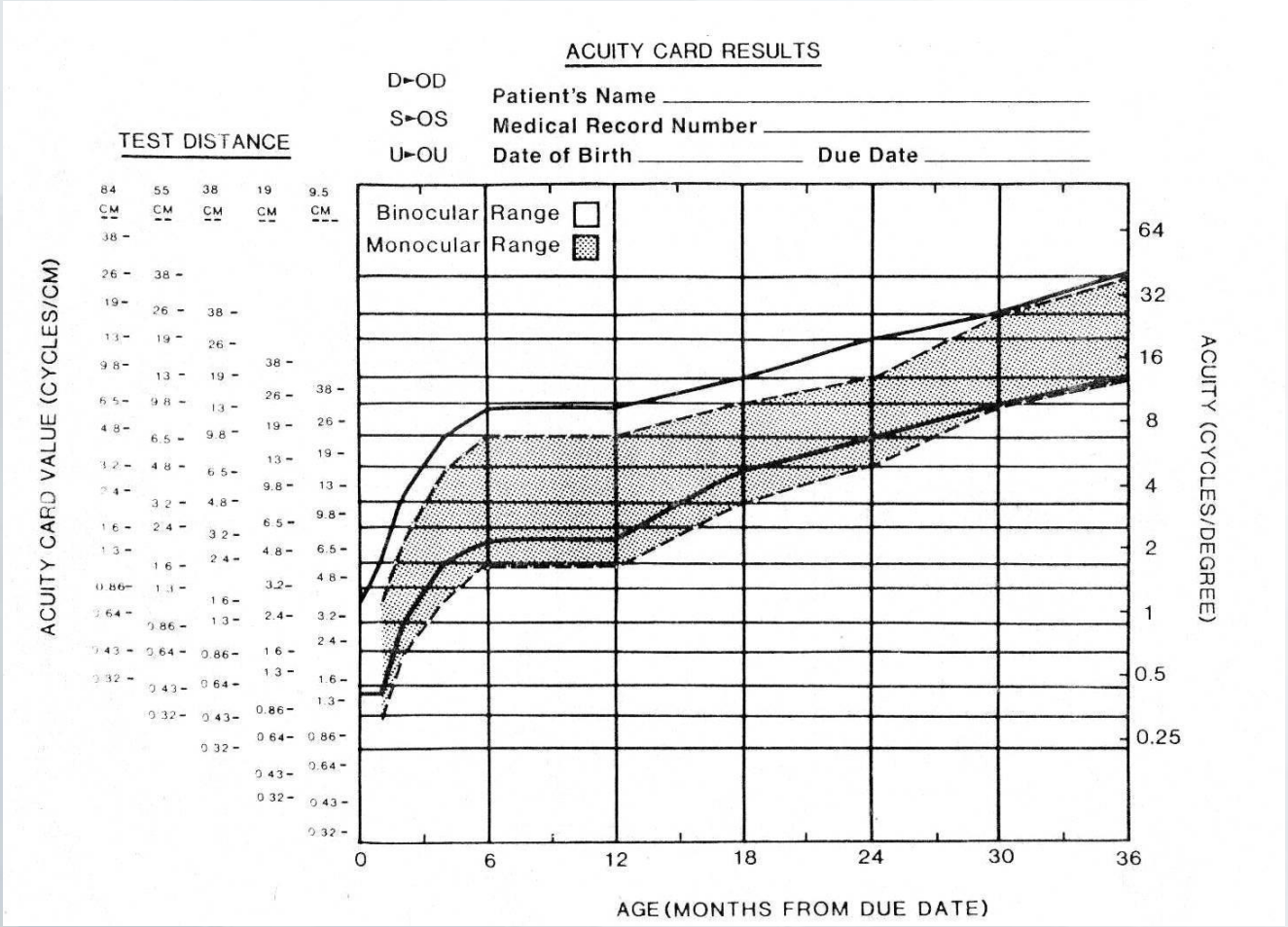
# Visual Acuity

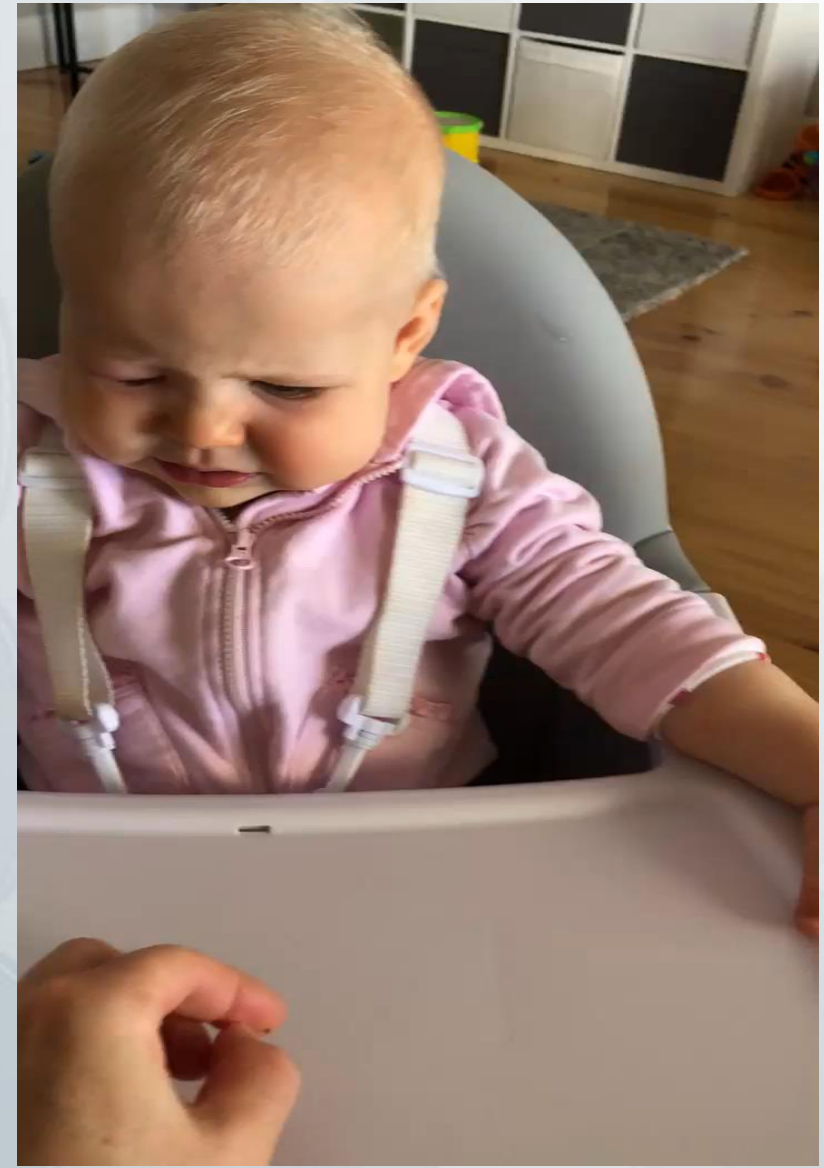
# Vision develops very quickly….




**…..from birth until *at least* 7 years of age**

# Visual Behaviour V Visual Acuity

- OBSERVING VISUAL BEHAVIOUR ***IS NOT*** SURROGATE FOR ACUITY AND FUNCTION



# Not obvious – important

## Amblyopia = *AVOIDABLE* BLINDNESS

Definition: reduction in vision that persists after any pathology is removed or corrected

Most common cause:

- unequal refractive error (anisometropia/lazy eye)
- strabismus (squint/eye turn/lazy eye)

Treatment

- Correction with spectacles
- Patching of the good eye
- Treat up to ~ 7-8 yo

**NEVER TO YOUNG, OFTEN TOO OLD!**

# “MY BABY DOESN’T SMILE AT ME!”

# Delayed Visual Maturation - DVM

- Delay in achieving normal visual milestones
  - Not fixing or following by 2-4 months
  - Normal eye examination
    - No nystagmus, normal pupil reflexes
  - Neurological development normal
- Spontaneous improvement by 6 months
- Cause unclear
- Associated with subsequent learning/motor delays

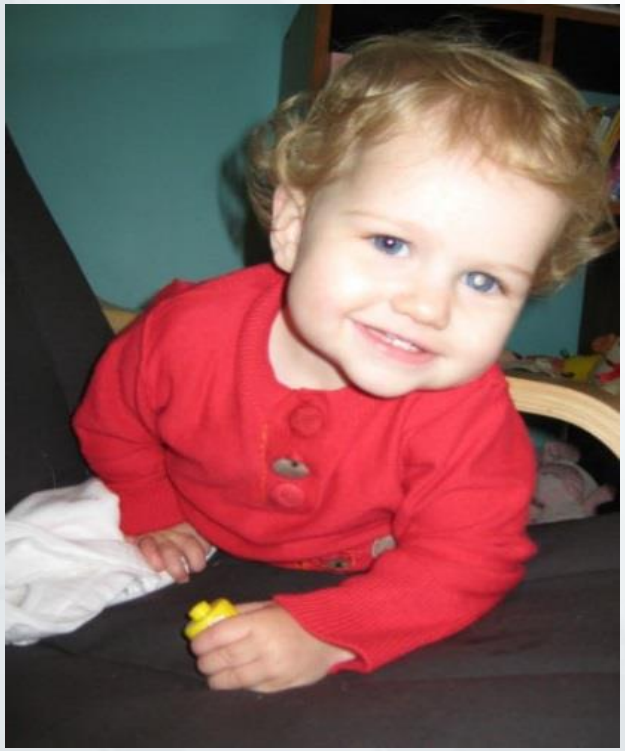
# Cortical Vision Impairment - CVI

- Loss of ‘vision’ due to cerebral insult
- Normal pupil reflexes and eye examination
- Roving eye movements
- Common causes
  - Perinatal hypoxic-ischemic insult
  - Hydrocephalus
  - Prematurity (PVH*, PVL*)
  - Non accidental injury

*PVH: periventricular haemorrhage
*PVL: periventricular leukomalacia

# Less obvious – serious & important

Strabismus



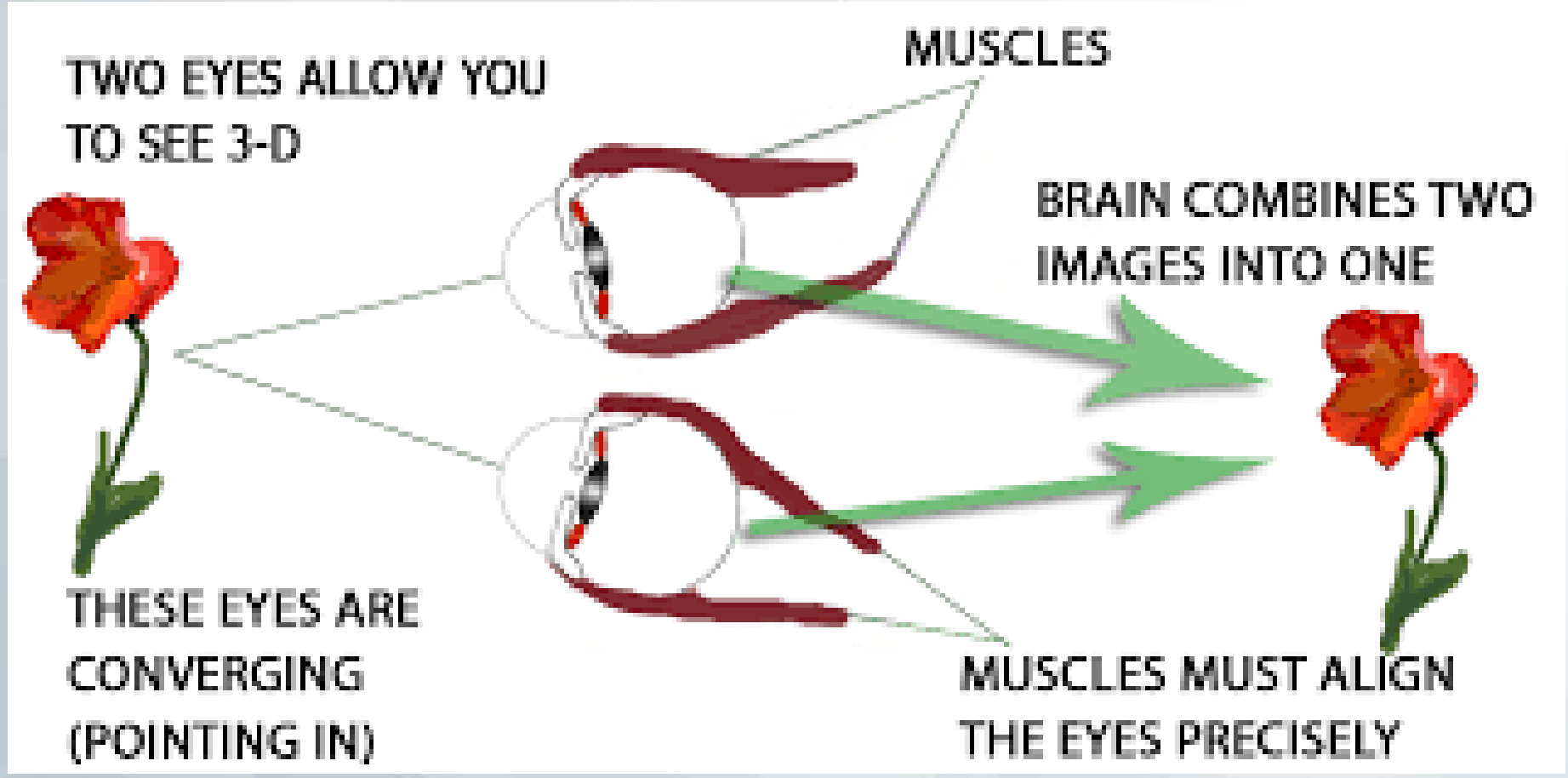
Intraocular disease

Unilateral vision loss

- Not ‘obvious’ to look at
- Infrequent/intermittent
- Child is *otherwise well* or *not complaining*
- Child appears to “see” well – functions normally ‘visually’

# Strabismus

# Binocular Vision



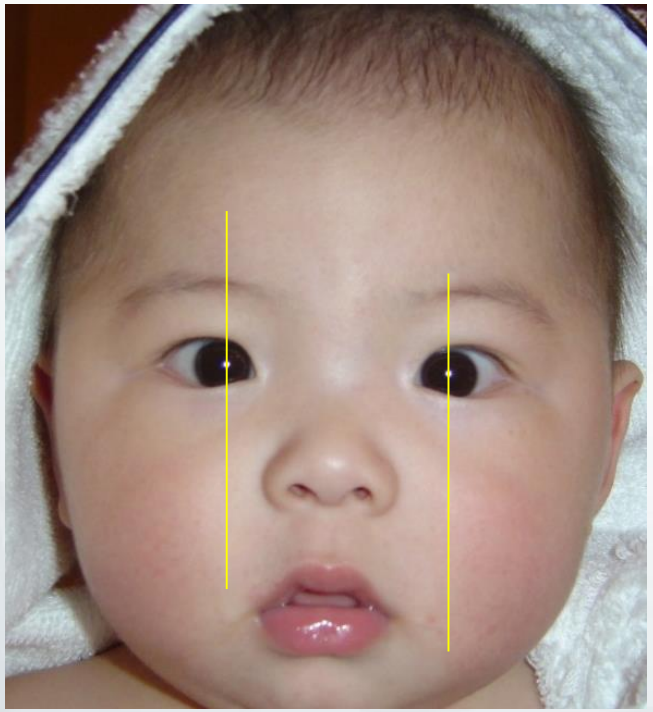

www.visioncdl.com

# Strabismus – “squint that goes away”

## Transient neonatal strabismus

- *NORMAL ocular alignment*
- intermittent
- ***Resolves by 2-4 months***[1,2]

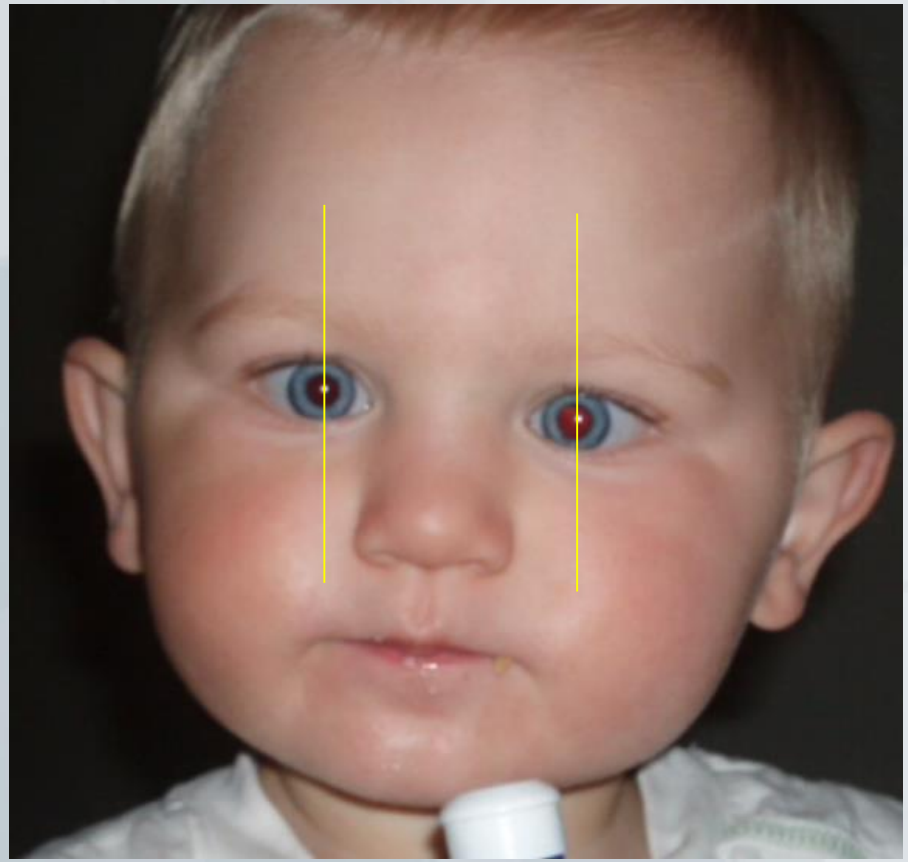
## Pseudo-strabismus: Optical Illusion

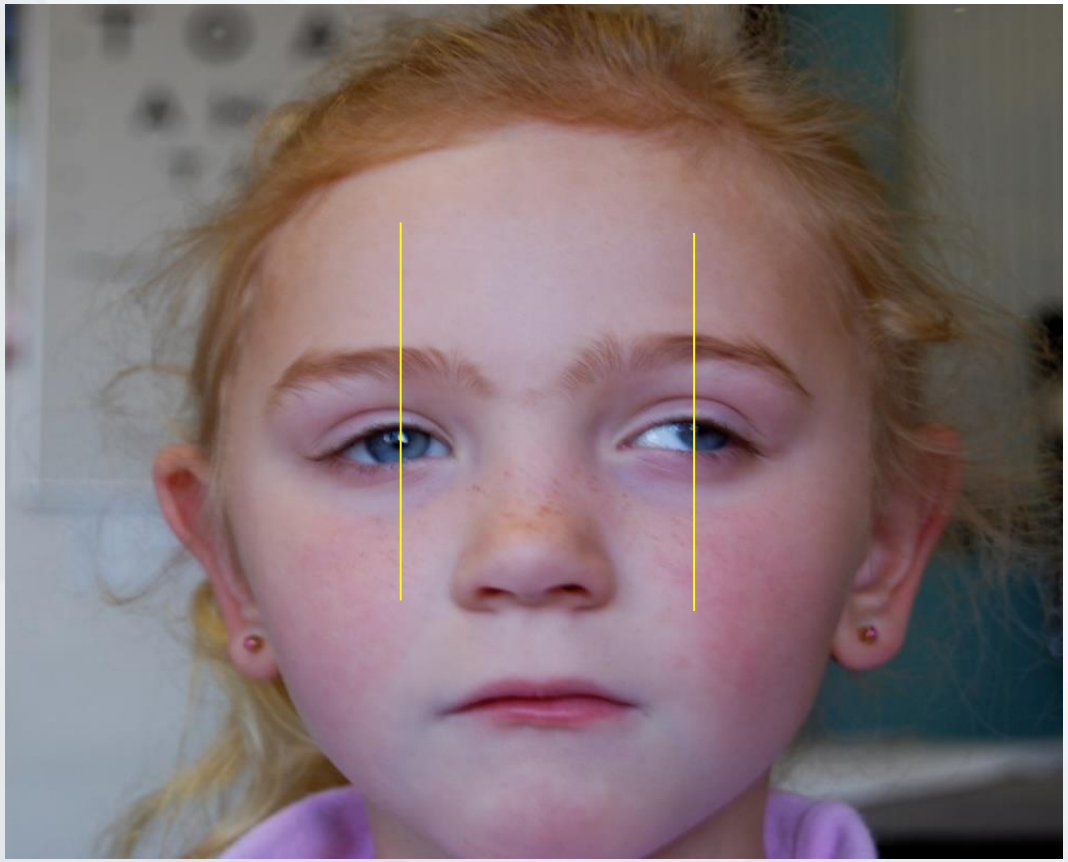


- Wide nasal fold/bridge of nose
- Intermittent – looking sideways
- “see both ears”
- Corneal light reflex - symmetry

[1]*Horwood A. 1993, JAAPOS;* [2]*Sondhi N. et al. 1988 JAAPOS*

# True strabismus – variable direction, size and frequency





**Consider**:

- CAUSE? – *secondary cause until proven otherwise*
- EFFECT ON VISION DEVELOPMENT – AMBLYOPIA

# Straightforward squint…?

**Primary strabismus**

- 2-4% population[2,3]
- Multiple associations[1]
  - FHx strabismus/amblyopia
  - Hyperopia/anisometropia
  - Prematurity
  - Down's syndrome
  - Developmental delay
  - Cerebral palsy
  - Fetal Alcohol Syndrome
  - Craniofacial syndromes
- 83% amblyopia < 3 yo[2,3]
- Stereopsis [3D vision][4]

**Treatment**

- Glasses – refractive error
- Occlusion - amblyopia
- Surgery

***1.Royal College of Ophthalmology UK: Guidelines for the Management of Strabismus in Childhood,*** *March 2012* *www.rcophth.ac.uk* ***2. Graham P.A. BJO 1974 3. Robaei et al. Ophth.2006 4. Birch et al. Optom.Vis.Sci.2009***

# Sinister sign…?

**Primary Neurological Disorder**

- Optic nerve glioma
- Medulloblastoma
- Craniopharyngioma
- Hydrocephalus

➢ **ADDITIONAL SYSTEMIC Symptoms**

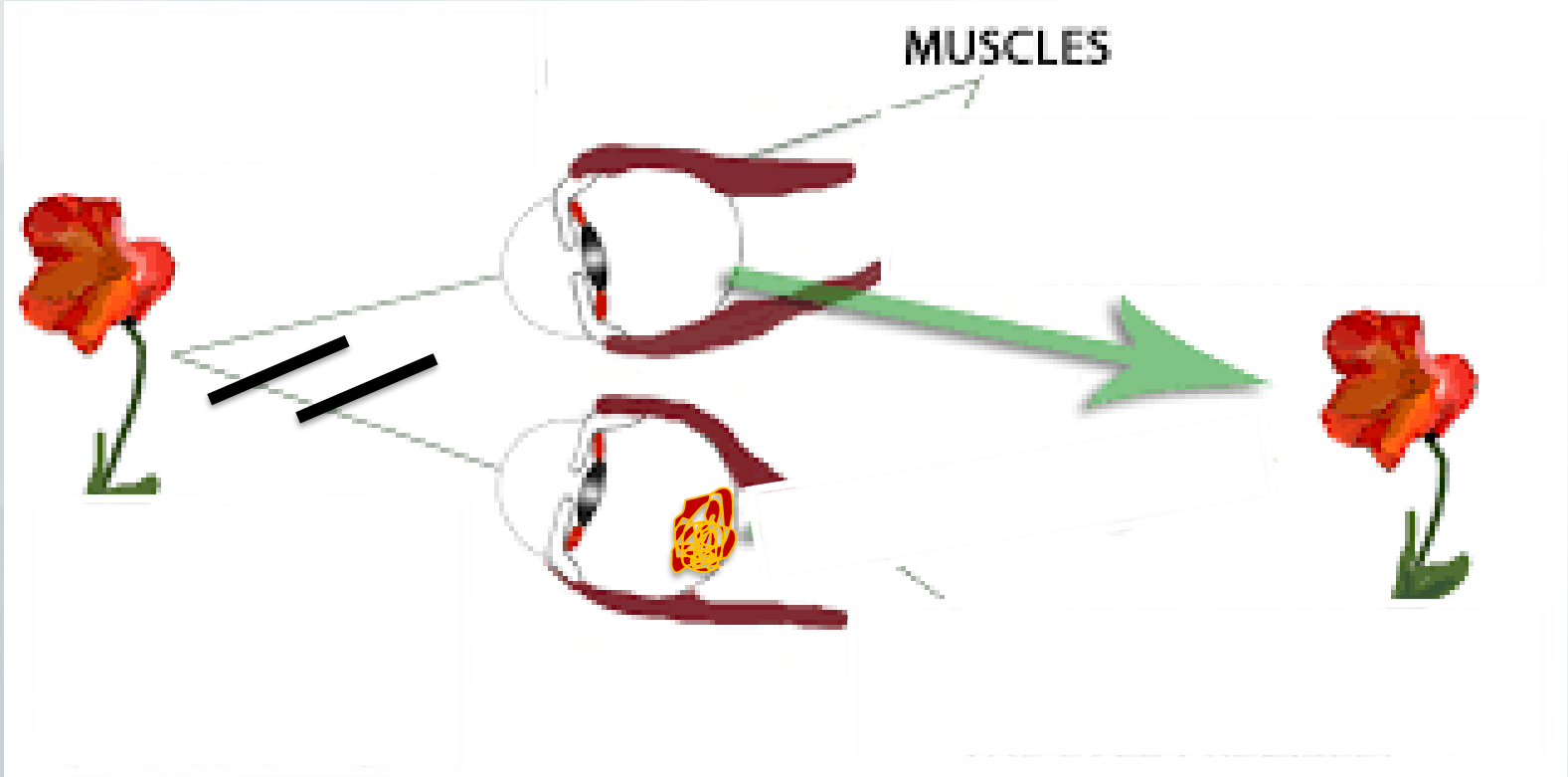
**Intraocular disease**

- Cataract
- Coat's disease
- Ocular toxocara
- PHPV
- Retinoblastoma

➢ **WELL CHILD NORMAL VISION (UNI)**

- **Lesion disrupts binocular vision**

# Intraocular disease disrupts binocular function

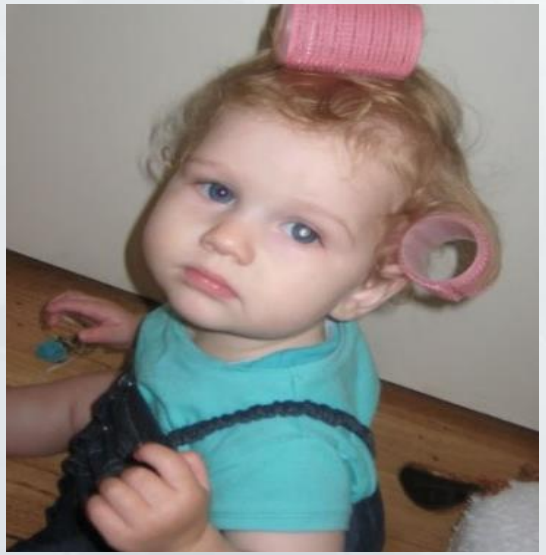



- Central vision is disrupted
- No incentive for the eyes to remain straight
- Affected eye will ‘wander’ – in or out

# Straightforward squint… or sinister sign?

# Leukocoria

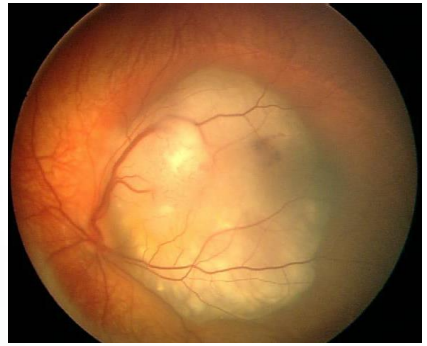
"Leuko" – white
"Coria" – pupil



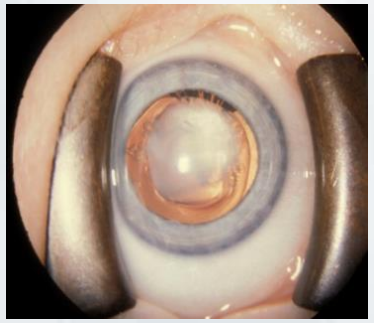
# Causes of leukocoria in children

| Cause of Leukocoria in Children | |
|---|---|
| Congenital cataract | 60% |
| Retinoblastoma | 18.2% |
| Retinal Detachment | 4.2% |
| PHPV (persistent hyperplastic primary vitreous/persistent fetal vasculature) | 4.2% |
| Coats' disease | 4.2% |
| Coloboma: iris/choroid/retinal | 2.8% |
| Infection: Ocular toxocara/Endophthalmitis/Panendophthalmitis/Posterior Uveitis | 5.6% |

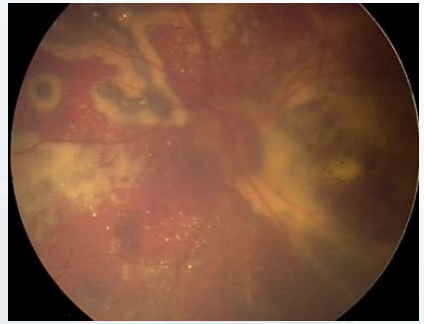
*Cataract*



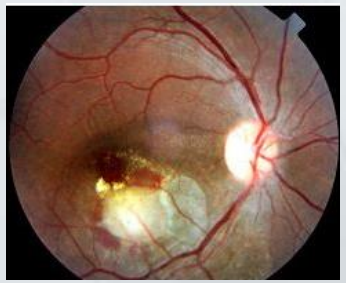
*Retinoblastoma*



*PHPV*



*Coat's disease*



*Toxocara*

*Haider et al. JAAPOS, 2008*


# Congenital cataract

- Leading cause of childhood blindness
- Congenital ***OR*** develops during early childhood
- Complete or partial
- Familial
- Unilateral or Bilateral
- Differential diagnosis – vital
- Early diagnosis – imperative
- URGENT

# Causes of cataract in children

## Intrauterine infections
- Rubella, Varicella, Toxoplasmosis, HSV

## Drug Induced
- Corticosteroids, chlorpromazine

## Metabolic Disorders
- IDDM, Galactosaemia,
- Hypocalcaemia, Hypoglycaemia

## Trauma
- Blunt/penetrating injury,
- AI/NAI, laser photocoagulation

## Radiation induced

## Inherited
- AD/AR/X-linked

## Chromosomal
- Trisomy 13, 18, 21 (Down's), Turner & Cri-du Chat Syndro

## Renal Disease
- Lowe, Alport & Hallerman-Streff-Francois syndrome

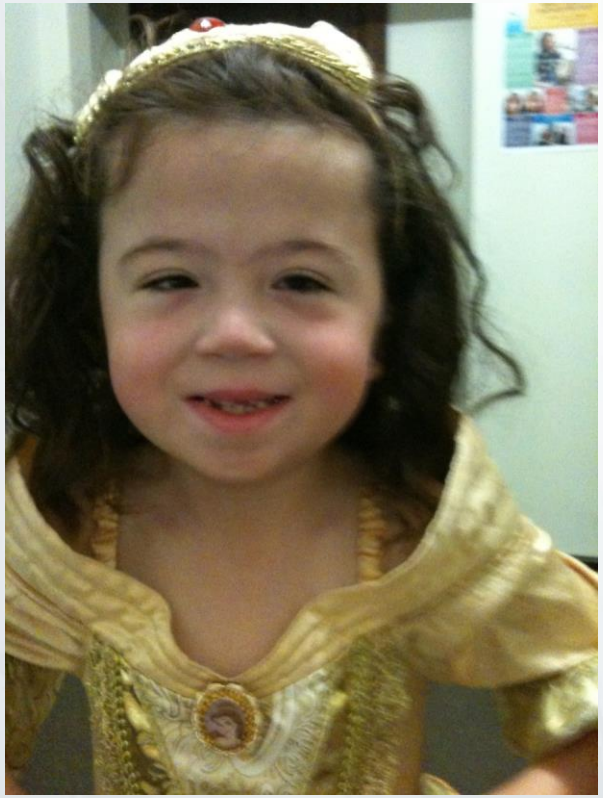
## Skeletal Disease
- Stickler, Rubenstien-Taybi, Bardet-Biedl, Conradi syndron

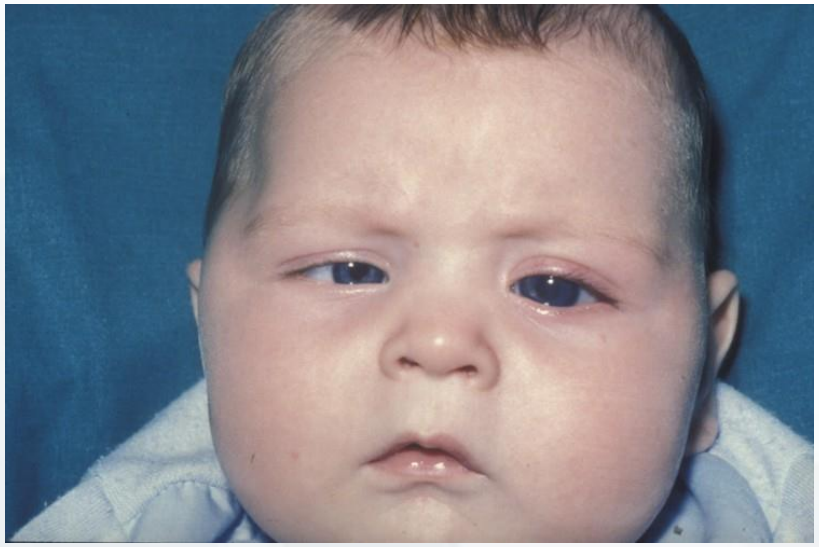
## Neurometabolic Disease
- Zellweger syndrome

## Muscular Disease

## Dermatological
- Cockayne syndrome, Incontinentia pigmenti, progeria
- Crystalline cataract & uncombable hair syndrome!

# Treatment





- Very long road
- Surgery
- Glasses/CL/IOL
- Occlusion
- NOT the same as ADULT cataract

- Amblyopia
- Strabismus (& binocular function)
- Aphakic glaucoma*
  - Timing of surgery

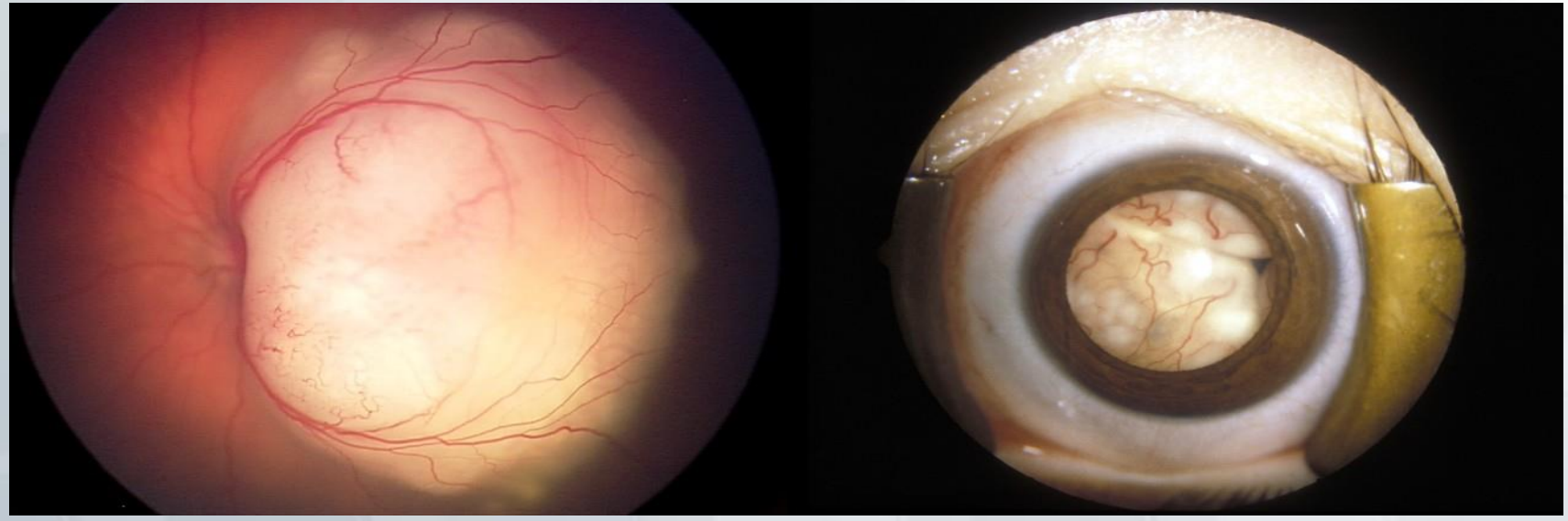
# “Isn’t it just the camera flash?”

# Retinoblastoma

- 1:15-20 000 births – VIC/TAS 1:17 500[1] (4-5 new cases/year)
- Develop from birth → 5 years of age
- all childhood cancers: 9.1% <1yr; 3% 1-4yr

…"*once uniformly fatal, now uniformly curable*…"

Grossniklaus *(LXXI Edward Jackson Memorial Lecture AJO 2014)*



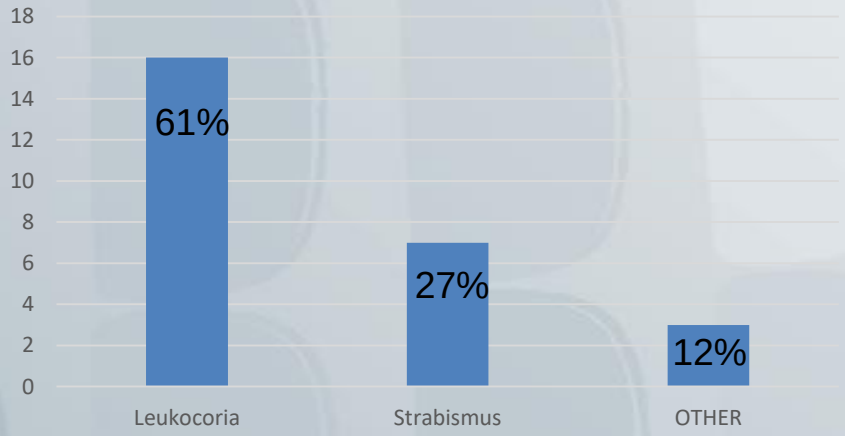
[1]Dondey J, Staffieri SE *et al. 2004 Clin.Exp.Ophth*

# Retinoblastoma

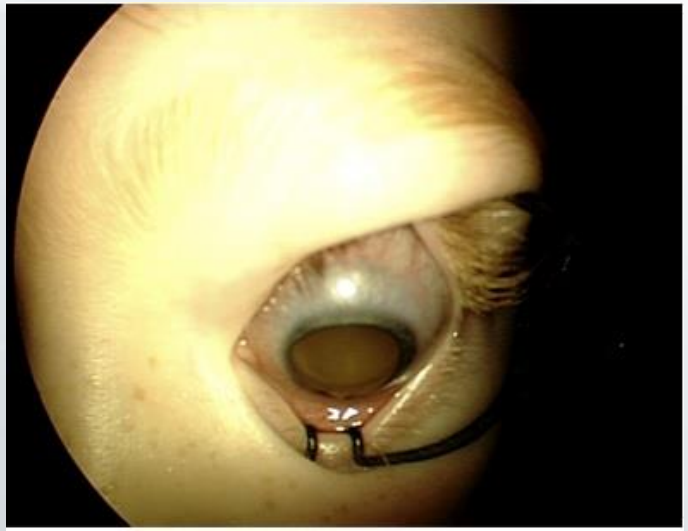
Leukocoria – white pupil

Strabismus – squint


FHx - *RB1*+ - AD - 50% risk



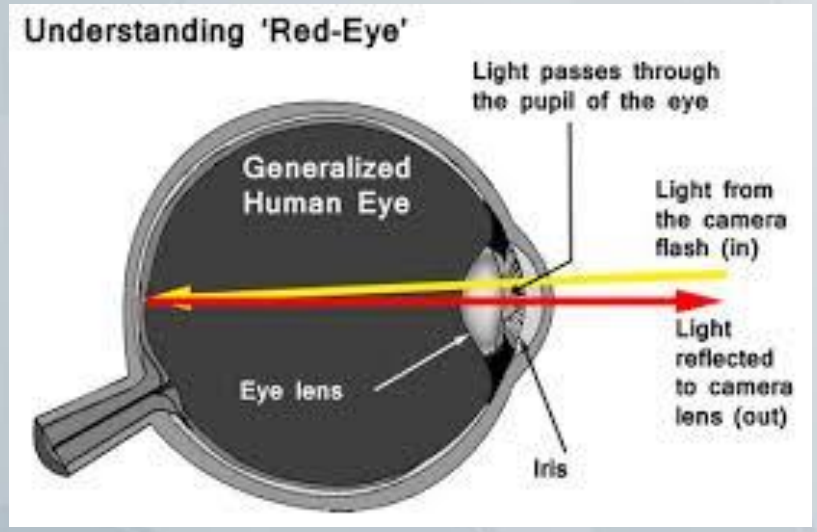

Source: Victorian RB Database DRN DB#090 *est. 1992*



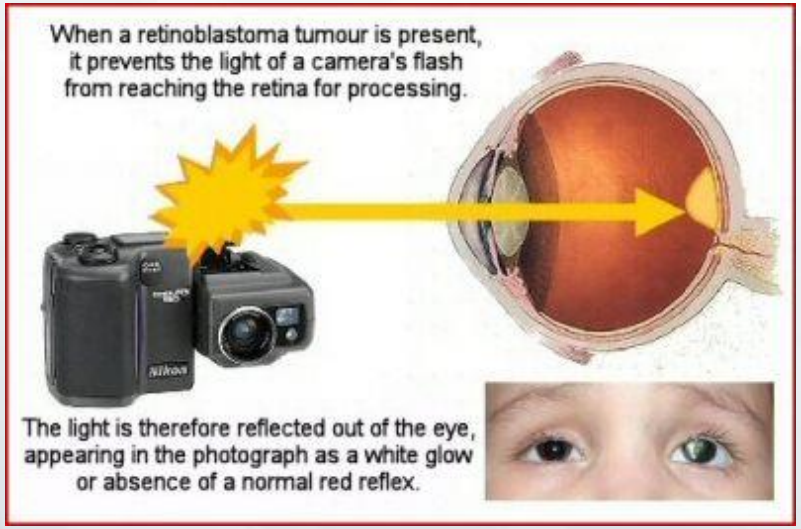
Anterior segment disease

# Retinoblastoma

- Fleeting
- Dim light
- Photograph
- Not seen with naked eye




- "glint"
- "glow"
- "hologram"
- "cat's eye reflex



# Barriers to early diagnosis - leukocoria

The Royal Children's Hospital Melbourne

## Red-eye reduction

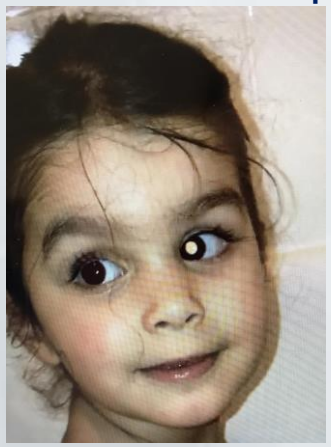

## Artefact - desensitised

## Photoshop

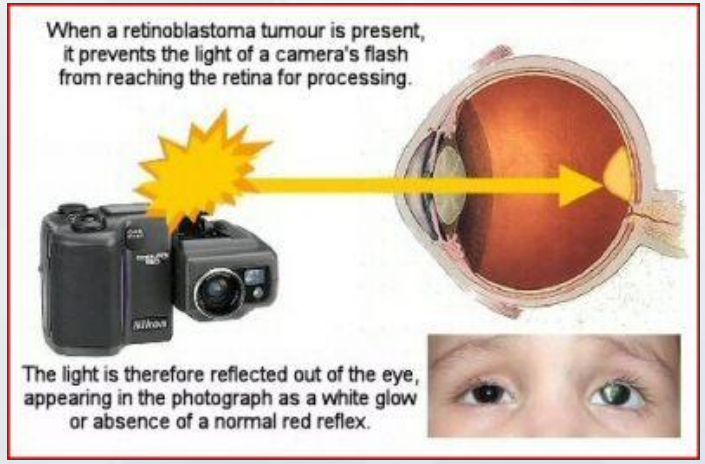

*Murphy D, et al. Lancet 2012*

## Artefact – optic nerve

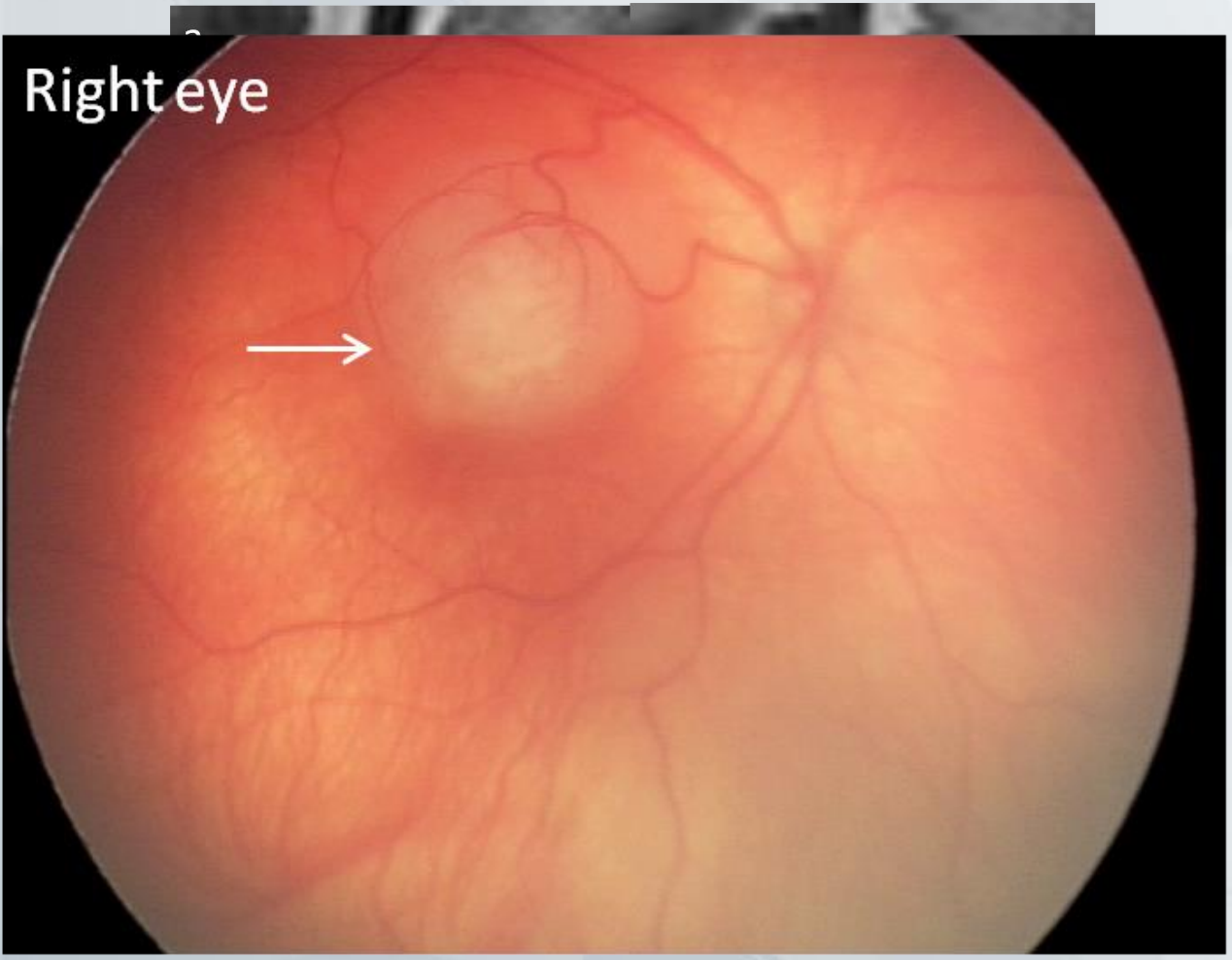
Retinoblastoma

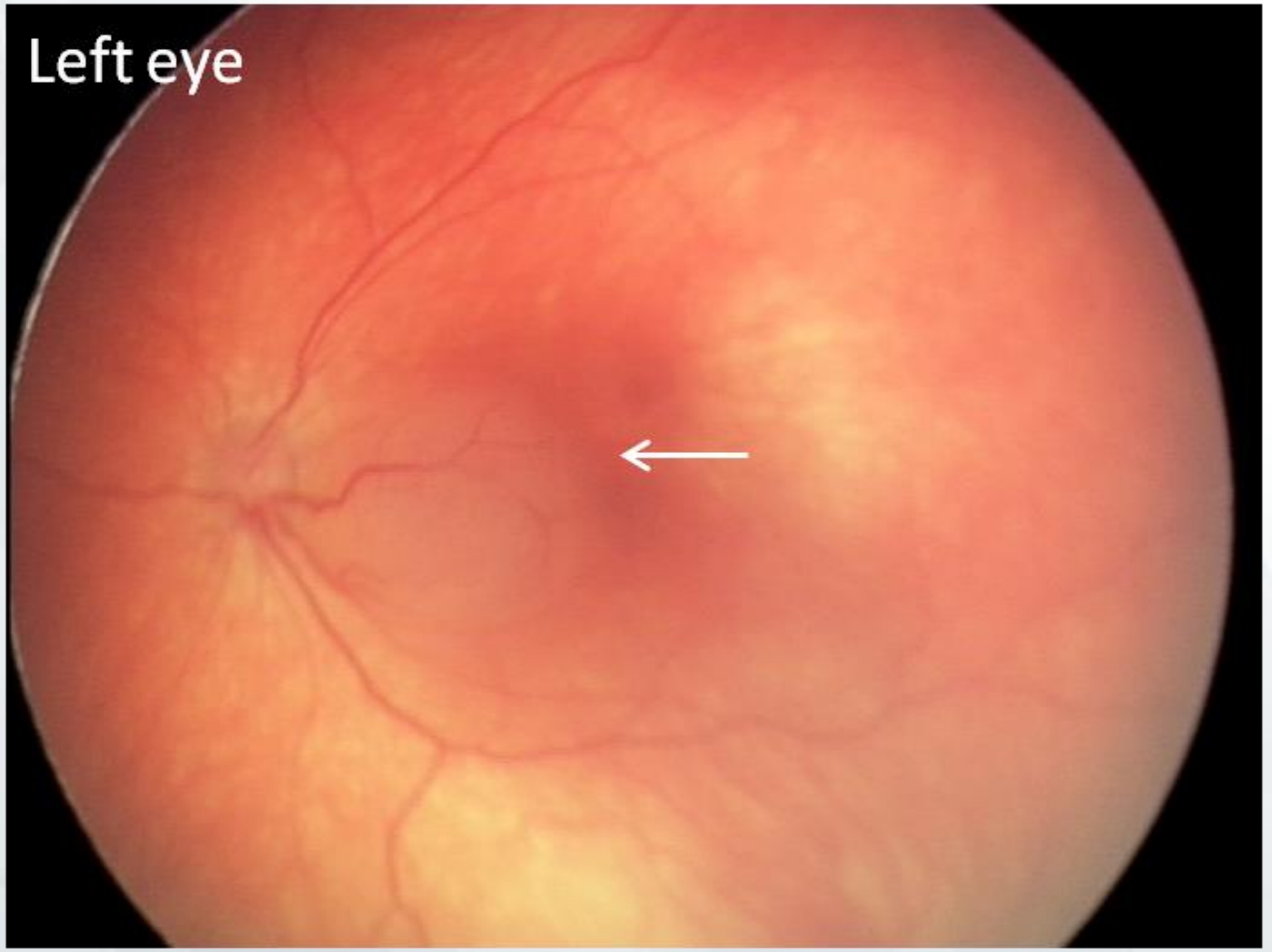


Optic nerve



# Identifying tumours early…




*Staffieri SE et al. PrenatDiag.2015*

# Early diagnosis saves eyes

**RCH 2000 - 2018**

| | Family History RB | No Family History RB |
|---|---|---|
| **Unilateral:** *Enucleated eyes* | 0/2 (0%) | 44/48 (91.7%) |
| **Bilateral:** *Enucleated eyes* | 1/22 (4.5%) | 17/34 (50%) (4 children saved BE) (4 children both eyes removed) |

## >90% of children rely on recognition of early signs of disease

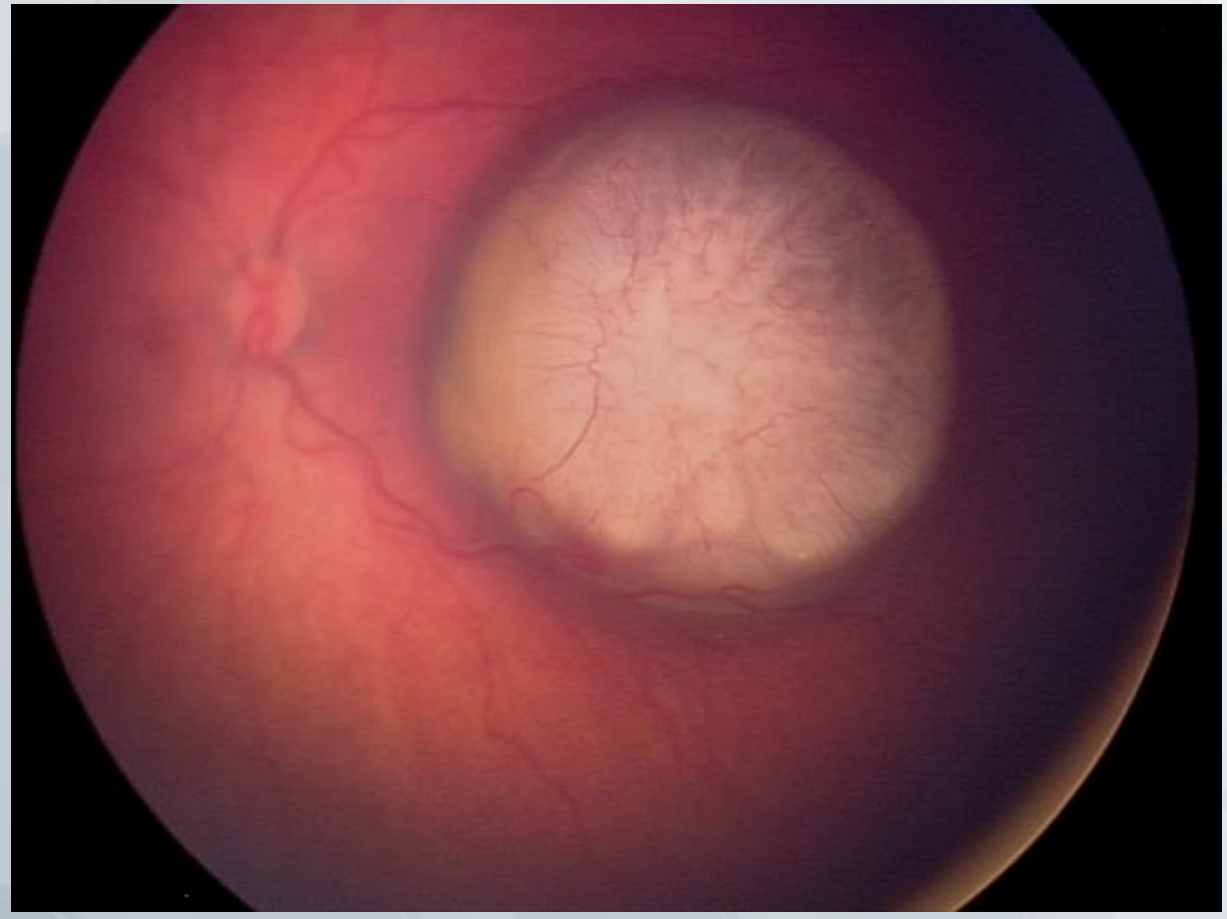
Source: Victorian RB Database DRN DB#090 *est. 1992*

# Strabismus can be a very early sign

- 4.5 month old; ex-33/40
  - (3 month - corrected)
- Several weeks Hx L intermittent strabismus
- Reassured - paediatrician
  - ‘strabismus’ was *normal* at this age
  - Risk factor – premature
  - No examination

# Strabismus can be a very early sign

- *8 day Hx leukocoria*



# Take home messages…..

- **Know what NORMAL looks like – what you notice may be critical**
- **Observe children carefully** [and their parents!]
- **Family History of disease -** [strabismus, amblyopia, cataract, glaucoma, retinoblastoma]
- **Fleeting or intermittent disease**
  - Building rapport with parent – trust & confidence to discuss their observations or concerns
- **Visual behaviour ≠ good and *equal* vision**

# Acknowledgements

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Alternate (to RCH) ophthalmologist providers

https://www.rch.org.au/uploadedFiles/Main/Content/ophthal/Alternate%20Eye%20Care%20Providers.pdf