Paediatric ocular disease

Notes to accompany lecture to 2nd Year City University undergraduates

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Trauma

Common in childhood. Sometimes child may be reluctant to report injury.

Periorbital bruising
"Black eye" due to haemorrhage into lid and orbital tissue. May cause ptosis. Check that the eye is healthy and motility full.

Lid lacerations
May need cleaning and treatment with surgical adhesive tape or repair depending how superficial.

Subconjunctival haemorrhage
Common at birth. In child may be due to minor trauma, Valsalva, or non-accidental injury.

Corneal abrasions
Antibiotic eye ointment + cyclopentolate or homatropine (+ pressure patch ?). Do not patch vegetative abrasions. Follow up 24 hrs. DO NOT USE aspirin as analgesic in children under 16 years (Reye's syndrome)

Chemical injuries
IRRIGATE (eg 20 mins) and remove any chemical particles. Alkaline burns are AN EMERGENCY.

Corneal foreign body
Remove if epithelial (spud or needle) following instillation of anaesthetic. Cycloplegic and antibiotic eye ointment (+ pressure patch ?).

Hyphaema
Blood in anterior chamber. 5% develop secondary glaucoma. 30% have intraocular pressure rise in acute phase (first week).

Orbital blow out fracture
Possible entrapment of orbital fat/extraocular muscle. Diplopia.

Non- accidental injury
"you name it : you can get it", but most commonly bruising, sub-conjunctival haemorrhages. Retinal haemorrhages from shaking, abdominal pressure (Purtscher's retinopathy), or direct trauma. Various factors raise your index of suspicion.

The red eye

Cellulitis

Acute inflammation of orbital tissues.

Differentiating orbital/preseptal cellulitis and cavernous sinus thrombosis

	Preseptal	**Orbital**	**Cavernous sinus thrombosis**
Lid oedema	Moderate	Marked	Marked
Skin	Red	Red	Blue purple
Conjunctiva	White	Swollen	Swollen
Warmth	Yes	Yes	No
Proptosis	No	Marked	Marked
VA	Normal	Reduced ?	Reduced
Pupil	Normal	Normal	Abnormal
Motility	Full	Restricted	Paresis
Pain	No	Yes	Yes/No
Temperature	Normal	Elevated	Elevated
Disc	Normal	Normal	Disc oedema

Ophthalmia neonatorum
Commonly gonoccocal.

Bacterial conjunctivitis
Muco/purulent discharge. *Haemophilus influenzae* (gram negative). Common in children 5 weeks to 5 years. Conj. haemorrhages. Purplish discoloration of lids. Treat with aminoglycosides (eg gentamycin), chloramphenicol .
Staphylococcus (gram positive) Treat with chloramphenicol, erythromycin
Chlamydia most common cause of conjunctivitis in infants aged 5 days to 5 weeks.

Viral conjunctivitis
The most common cause of infective conjunctivitis. Hyperaemia, watering, f.b sensation, follicles, pre auricular lymph nodes. Adenovirus. Very contagious. Epidemic keratoconjunctivitis (EKC) (adenovirus 8,19). Treatment supportive, cold compresses.

Allergic conjunctivitis
(Type 1 allergy) eg associated with hay fever. **Itching**, watering, papillae, chemosis. Treatment - vasoconstrictors, mast cell stabilisers, antihistamines.

Vernal keratoconjunctivitis
Seasonal. Itching, tearing, thick ropy discharge. Giant papillae. Management - topical lubricants. Mast cell stabilisers before onset, topical steroids after. May lead to corneal ulcer.

Corneal ulcer
Many causes. Vitamin A deficiency, systemic infections (eg measles), neglected trauma, vernal KC, herpes simplex (treated with topical anti-virals).

Trachoma
Chlamydia trachomatis. Recurrent conjunctivitis. Follicles. Lid scarring, trichiasis, secondary infections.

Eyelids

Skin allergies (Dermatitis)
Type 1 hypersensitivity

Epicanthus

Epiblepharon
Prominent fold of skin on lower lid causes entropion. Resolves with growth.

Ptosis
Most often caused by congenital weakness of levator. Deprivation amblyopia. Marcus-Gunn jaw-winking (pterygoid-levator synkinesis).

Angular dermoid
Benign tumour (choristoma) of a variety of embryological and histological origins. Commonest position are superior temporal brow and **outer canthus region.** Always painless. Usually stationary or slightly mobile. Surgery for cosmetic reasons only. Associated with e.g. *Goldenhar's syndrome.*

Capillary hemangioma.
"Strawberry naevus". Spontaneously involutes. If pupil covered swift intervention necessary.

Eye lid margin problems

Blepharitis
Commonly from *Staphylococcus.* Bacteria produce toxins leading to marginal keratitis, corneal infiltrates or phylctenular keratoconjunctivitis. Treatment includes "lid scrubs", topical antiobiosis and consideration of steroids for infiltrates.

Demodex follicularum very common in elderly. Rarer in children except immunologically compromised. Aracnid. Itching eye lids. Distinctive collarette. Suffocate with simple eye ointment.

Pediculosis capitis/corporis & Phthirius pubis. The latter species may infest glabella and eye lashes. Mercuric oxide ointment 1% or suffocate with simple eye ointment.

Stye
Local infection of lash follicle (Zeiss or Moll). Hot compresses and antibiosis.

Tarsal cyst

Acute = Meibomian cyst. Chronic produces fibrous chalazion. Hot compresses. If spontaneous resolution does not occur, curettage may be necessary.

Molluscum contagiosum

Small (3-5 mm) umbillicated nodule with yellow "cheesy" core which can intermittently discharge. Viral. Leave alone unless discharging in which case curettage may be carried out.

Watering eye

Nasolacrimal duct blockage

Delayed patency very common in infants. Usually spontaneously open within weeks. Management - massage and hot compresses. Refer if marked mucopurelent discharge, dacryocystitis or if not resolved after 12 months for dilation and probing under general anaesthetic.

Buphthalmos (infantile glaucoma)

May be congenital but frequently does not manifest until up to 1 year of age. Epiphora, photophobia, cloudiness of cornea, macrocornea. Goniotomy or trabeculotomy.

Primary open angle glaucoma

POAG very rare in children. Children over five can be checked with static quantitative field techniques. Keeler Pulsair great for younger children and babies.

Proptosis

Orbital cellulitis
See above.

Orbital pseudotumour
Painful proptosis, lid and conjunctival swelling, limitation of ocular motility. Non-specific granuloma.

Rhabdomyosarcoma
Commonest orbital malignancy in children (often 6-7 year olds)

Orbital dermoid cysts
The external lump may be the "tip of the iceberg" with the lesion extending into the orbit.

Optic nerve glioma

Leukocoria

Retinoblastoma
Sporadic or dominantly inherited. Usually arises before age 3 years. Presenting sign may be strabismus. Examine other members of the family. Smaller tumours can be treated with irradiation or cryotherapy. Larger tumours require enucleation of the eye and excision of optic nerve.

Retinopathy of prematurity
From mild peripheral vascular changes to advanced retinal scarring. Mild ROP can be associated with strabismus, amblyopia and myopia.. Cry- or laser treatment carried out in severe cases.

Toxocara
Eggs of these parasitic worms found in dog or cat faeces. Children may eat the eggs. Larvae have a predilection for CNS including retina. There is no effective treatment for ocular infection.

Persistent hyperplastic primary vitreous (PHPV)
Embryological remnant of hylaoid artery complex. Other end of extreme is Mittendorf dot.

Coat's disease
Extreme form of retinal ptelangiactasia.

EOM anomalies

e.g. s
Marcus Gunn jaw winking
Duane's syndrome A, B & C
Brown's syndrome

Nystagmus

Latent e.g. associated with infantile esotropia
Visual deprivation Poor development of fixation reflexes in first few months of life. Cataracts, Leber's amaurosis, ROP, cortical blindness
Primary congenital secondary to a range of conditions. Recent onset nystagmus must be referred for further investigation.

Congenital abnormalities and syndromes

Craniofacial dysotoses
Abnormal skull development may lead to abnormal orbit shape and position and hypertelorism. May lead to strabismus. . e.g. Crouzon's disease, Apert's disease.

Microphthalmos
Small eye associated with intra-uterine infections such as rubella, cytomegalovirus and toxoplasmosis. Syndrome example - Goldenhar's syndrome.

Congenital cataract
Causes include autosomal dominant inheritance, prenatal infection e.g. rubella, metabolic disorders e.g. galactosaemia. 50% of patients cause is unknown. Take careful family history, blood and urine electrolytes and amino acids. Early surgery (before 3 months).

Congenital glaucoma
See above.

Corneal dystrophies
Congenital dystrophies are rare and present with cloudy corneas at birth. Two main congenital dystrophies are congenital hereditary endothelial dystrophy (CHED) and posterior polymorphous dystrophy (PPD). Other causes of cloudy corneas in infants include glaucoma and rubella. Note: Thygeson's epithelial keratitis is sometimes seen in teenagers. ***Keratoconus*** usually presents in teenagers. Increasing myopic astigmatism, conus, Fleicher's ring, atopy association (e.g. vernal conjunctivitis), stromal scarring, discomfort, watering. Treatment - optical, contact lenses, surgery.

Coloboma
Failure of fusion of choroidal fissure (seventh week). Iris, ciliary body, retina, choroid, optic nerve. Example of syndrome - Goldenhar's. Also see SB's notes on Optic Nerve Disorders.

Persistent pupillary membrane
Embryological remnant.

Aniridia
Genetic disease with absence of iris, nystagmus, reduced VA, and sometimes glaucoma and peripheral corneal scarring.

Albinism
Genetic. Reduced melanin. In oculocutaneous albinism there is little or no skin or eye pigmentation. The eyes look pink due to absence of iris, RPE and choroidal pigmentation. Photophobia, nystagmus, poorly formed foveae, reduced VA and abnormal nerve crossing at chiasma. Ocular albinism refers to milder form of albinism in which hair, skin and iris pigmentation are relatively normal but fundus pigmentation is reduced. Again, photophobia, nystagmus, poorly formed foveae, reduced VA and abnormal nerve crossing at chiasma.

Myelinated nerve fibres
Myelinated nerve fibres on retina. Also see SB's notes on Disorders of Optic Nerve.

Retinitis pigmentosa
Inherited as autosomal dominant, recessive or X-linked. The dominant form has the best prognosis and the recessive disease the worst. Underlying defect is a bilateral and irreversible degeneration of the rod and, later, the cone photoreceptors. Typically an adolescent complains of poor vision in dim illumination. In the early stages clumps of pigment seen in peripheral retina and these progress centrally. Visual field defect often mid-peripheral, often progressing to "tunnel vision" and complete loss.

Leber's congenital amaurosis
Congenital form of retinal disease. The baby is blind, develops nystagmus and the electroretinogram shows absent electrical activity. Pigment changes similar to RP develop later.

Macular dystrophies
Poor acuity and colour vision. Nystagmus. Diagnosis by appearance of fundus and electrophysiological testing. Many different disorders. *Stargardt's disease* autosomal recessive. insidious bilateral visual loss in first two decades of life. Early changes include mild pigmentary changes at the macular progressing to extensive retinal atrophy. Poor prognosis (6/60 or less). *Vitelliform dystrophy or Best's disease* autosomal dominant. Yellow cyst like sub-retinal deposit ("sunny-side up") progressing to more irregular disturbance ("scrambled egg"). Slow progress. Fairly good prognosis.

Toxoplasma chorioretinitis
Toxoplasma gondii protozoan. Transmission: oral, transplacental, transfusion, cats. Single or multiple focii of necrosis and granulomatous lesions affecting any organ. Prevention includes avoiding uncooked or "rare" cooked meat and avoiding cat litter during pregnancy. Dormant cysts persist within the lesions and occasionally reactivate during adult life. producing creamy-white fluffy opacities of the retina adjacent to the original scar. Treatment depends on severity. If threatening the macular then systemic use of e.g. sulfadiazine, clindamycin, pyrimethamine, steroids (all have side effects).

Cytomegalovirus
A Herpes virus. Commonest congenital infection in infants. At least 90% of adults in developed countries show past exposure to CMV (usually acquired during first 5 years of life). Usually sub-clinical. Manifests as CMV retinitis (e.g. in AIDS patient). Transmission - body fluids, trans-placental.

Optic nerve hypoplasia
Underdeveloped optic nerve due to reduced number of cells associated with mid-line brain defects e.g. septo-optic dysplasia with small stature. Ring of sclera visible around optic nerve head ("double ring"). Also see SB’s notes on Optic Nerve Disorders.

Optic nerve pit
A hole in the inferior temporal optic nerve head. Always juxtapositioned with the optic disc margin. 0.1 to 0.7 of disc. This is **not** related to a coloboma.

Visual field defects. Possible central serous detachment. Monitor. Also see SB's notes on Optic Nerve Disorders.

Optic nerve drusen
Hyaline spheres buried in the optic nerve. Superficial drusen are ophthalmoscopically visible. View with BIO or Volk lens. Disc appears raised. May progress. Visual field defects. Also see SB's notes on Optic Nerve Disorders.

Systemic disease and the eye

Diabetes mellitus
Chronic disorder of carbohydrate metabolism and is the most common endocrine disorder in children. Most children are IDDM. Peak onset for IDDM is between about 6 years and puberty. Frequency 1:1500 at 5 years; 1:359 at 16 years. Diabetic retinopathy in childhood is rare. Optometrists should monitor and refer maculopathy, pre- and proliferative retinopathies. Slit lamp and gonioscopic examination of iris anterior chamber angles for all patients with diabetic retinopathy because of risk of rubeosis.

Rubella
Ocular involvement is common in congenital rubella particularly if mother contracted the disease in first 4 weeks of pregnancy. Microphthalmia, cataract (usually bilateral nuclear progressive), glaucoma, corneal opacity, uveitis, small difficult to dilate pupil), pigmentary ("pepper & salt") fundus. The latter is not progressive and is compatible with good vision.

Down's syndrome
Trisomy 21. Staphylococcal blepharitis, broad epicanthal folds, keratoconus, cataract, Brushfield's spots. **Caution:** prone to idiosyncratic reactions to anti-muscarinic drugs.

Marfan's syndrome
Dominantly inherited condition. Three connective systems:- *Skeleton*: arachnodactyly and hyperextensible joints; *Eye:* lens dislocation, cataract and retinal detachment, lens induced uveitis; C*ardiovascular:* mitral valve prolapse, aortic root dilation-dissection.

Neurofibromatosis
Autosomal dominant phakomatosis ("mother spot"). Multisystem benign tumours of neuroectoderm. Café-au-lait pigmented skin patches and fibrous skin lumps. Small nodules on iris (Lisch nodules). Benign optic nerve gliomas may develop.

Sturge-Weber syndrome
A phakomatosis. "port wine stain". If upper eye lid involved glaucoma may occur. Dilated episcleral veins.

Stevens-Johnson syndrome
Iatrogenic induced allergy (commonly sulphonamides) causing conjunctival ulceration and scarring leading to dry eye and corneal disease.

Juvenile chronic arthritis (JCA)
Still's disease. Commonest cause of anterior uveitis in children. Anterior uveitis: often white painless eye. The only symptom is blurred vision. Signs include aqueous flare and cells. Always do slit lamp exam on child with reduced VA. If cells present ask about joint pain. Band keratopathy, glaucoma, cataract.

Leukaemia
Can result in retinal haemorrhages and creamy white retinal infiltrates.

Neurological disease

Cortical blindness
Reduced acuity in presence of cerebral palsy, seizures, or general developmental delay. Disease of optic radiations or occipital cortex. Normal pupil reflexes.

Delayed visual development
Can occur in an otherwise normal baby. Appears to be blind in first few months but later develops normal vision. ERG, VEP and EEG rules out major organic disease. Wait and measure.

Optic atrophy
Inherited or as a result of neonatal anoxia, trauma, compression by tumour or secondary to hydrocephalic papilloedema.

Papilloedema
Choked disc. Raised, indistinct margins, splinter/flame haemorrhages, enlarge blind spot. Normal vision and colour vision in early stages. Differential diagnoses include papillitis (may be very difficult), optic nerve drusen, hypermetropic pseudo-papilloedema. Raised intracranial pressure.

Paediatric neurodevelopmental disorders

Cerebral palsy
Non-progressive neurological disorder resulting from damage occurring to immature brain *in utero*, during or after birth. High incidence of visual anomalies including strabismus and significant refractive errors.

Spina bifida and hydrocephalus
Developmental defects caused by failure of fusion of neural tube occurring in early embryonic life. The neural tube may be affected anywhere along its length, although the thoracolumbar region is the most common.

Hydrocephalus occurs in 80% of children with spina bifida. Hydrocephalus is caused by overproduction or malabsorbtion of cerebrospinal fluid and/or obstruction of the aqueduct. Nystagmus.

Fragile X
Mental retardation occurs in 3% of population and fragile X is the most common form of familial retardation being second only to Down's syndrome in frequency among children with chromosomal abnormalities. 30% prevalence of strabismus. High prevalence of significant refractive errors.

Autism
Severe difficulties in social interaction. Cause ? May be associated with fragile X chromosome. High prevalence of squint and eye movement control difficulties. Asperger's syndrome.

Further reading

Dhillon B & Fleck B (1996) Diseases of the eye and orbit, Chapter 12 in ***Pediatric Eye Care*** Eds. Barnard S & Edgar D, Blackwell Science, Oxford

Taylor D (1990) ***Pediatric Ophthalmology***, Blackwell Science, Oxford

Multiple choice questions

1) An infant with an enlarged cornea and photophobia is most like to be suffering from:
   (a) keratoconus
   (b) keratoglobus
   (c) microcornea
   (d) buphthalmos

2) A 7 year-old child presents with multiple brown organisms attached to the base of the eye lash follicles. You suspect
   (a) *pediculosis capita*
   (b) *demodex follicularum*
   (c) *pthirius pubis*
   (d) *Arachnidia meibomianatus*

3) A four month old infant presents with a watering eye with slight mucous discharge from the inferior punctum. Correct management is the following:
   (a) dilation and irrigation
   (b) dilation and probing
   (c) topical antibiotic ointment
   (d) hot compresses and gentle massage

4) The commonest cause of bacterial conjunctivitis in young children is
   (a) *Staphylococcus aureus*

(b) *Streptococcus pneumoniae*
(c) *Haemophilus influenzae*
(d) *Pseudomonas aeruginosa*

5) Adenoviral conjunctivitis can be differentiated from bacterial conjunctivitis by
(a) the lack of mucopurelent discharge
(c) the presence of swollen pre-auricular lymph nodes
(d) the presence of follicles
(e) all of the above

6) Retinitis pigmentosa has the worst prognosis when the mode of inheritance is
(a) autosomal dominant
(b) recessive
(c) X-linked
(d) non-Mendelian

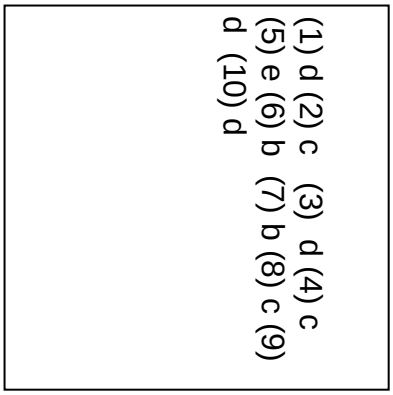
7) Hypertrophy may be best described as:
(a) a reduced number of cells
(b) an increased number of cells
(c) an increase in cell size
(d) a disturbance in cell growth involving both cell proliferation and an altered differentiation

8) The commonest congenital infection is:
(a) rubella
(b) gonococcal ophthalmia neonatorum
(c) a herpes virus
(d) toxoplasmosis

9) A 12 year-old girl presents complaining of reduced vision and joint pain. Following refraction you find her VAs to be R. 6/9 L. 6/12. Slit lamp examination shows grade 2 cells in the anterior chamber. Which of the following may be a systemic disease associated with this uveitis:
(a) syphilis
(b) tuberculosis
(c) juvenile rheumatoid arthritis
(d) all of the above may cause uveitis in a twelve year -old

(10) A 10 year-old boy presents complaining of headaches after closework. Both optic nerve heads appear swollen. Refraction show him to be R & L +3.50DS and he obtains 6./5 with each eye. You diagnose:
(a) optic nerve drusen
(b) papilloedema
(c) papillitis
(d) pseudo-papilloedema

MCQ Answers… do not view till you have attempted all questions



Any questions about this lecture ?

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