

Paediatric ophthalmology - What every GP should know

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Abstract

At birth one does not know for sure that a baby's eyes are normal and fully functional. The general practitioner's role is to identify the risk of inherited disease and to perform the relatively simple screening examinations for conditions that require referral for specialist attention. These depend on the child's age and serve to identify different conditions at different ages. Acquired conditions are most commonly allergic or infective in origin. The present review will highlight some of these conditions.

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Introduction

Evaluation of the eye and of vision is an integral part of each patient contact with a doctor. This is in reality a screening by history and examination, in order to identify those that must be referred to an ophthalmologist. Acquired conditions can best be described according to the age of onset.

Screening and indications for referral:

A family history of congenital cataract, congenital glaucoma, retinoblastoma, metabolic or genetic disease. Such infants should be referred for a comprehensive pediatric medical eye evaluation as early as possible. Other familial conditions that cause or are associated with eye or vision problems include strabismus, amblyopia or glasses in early childhood.

Identified risk factors such as prematurity (gestational age of <32 weeks and/or a birth mass of < 1.250 Kg) or other perinatal complications. Evaluation is required at birth and after 6 months. Patients with neurological disorders, neurodevelopmental delay, juvenile rheumatoid arthritis or syndromes known to be associated with ocular manifestations should be referred upon diagnosis. Patients with diabetes mellitus should be referred 5 years after onset and yearly thereafter, as should patients on chronic systemic steroids or other therapy (eg hydroxychloroquine).

Screening Examination includes the following:

- external inspection to detect structural ocular abnormalities: refer urgently if a problem is identified;
- examination of the red reflex: an absent or asymmetric red reflex denotes abnormalities of the ocular media such as cataract and needs urgent referral;
- the corneal light reflex (Hirschberg) test,
- cover testing for ocular alignment and motility (after 6 months of age, when babies are evaluated for fixation preference and ocular malalignment). Strabismus occurs in ~2% of babies and needs referral if still present after 3 months of age.
- testing of visual acuity from age 3 years; refer the following:
 - Unable to perform vision screening at age 3-3 1/2 years or older
 - Visual acuity 6/15 or worse, or a 2 line difference in a 3-year-old
 - Visual acuity 6/12 or worse, or a 2 line difference in a 5-year-old
 - Visual acuity 6/9 or worse, or a 2 line difference in a 6-year-old or older child

Photo screening and auto refractive devices may be helpful adjuncts but they should not replace standard clinical screening techniques

Complete Ophthalmological examination

- if abnormalities are identified on

routine screening as discussed above

- if signs or symptoms of eye disease are present, such as
 - defective ocular fixation or visual interactions;
 - abnormal light reflex;
 - ocular alignment or movement abnormality;
 - nystagmus;
 - persistent tearing or discharge;
 - persistent redness;
 - persistent light sensitivity; squinting;
 - inappropriate eye closure;
 - head tilt;
 - learning disabilities.

Congenital eye problems in babies

Many developmental abnormalities are genetic in origin. Others are due to intrauterine damage from teratogenic factors such as infection and drugs.

Defects of the globe

- Anophthalmos - complete failure of the optic vesicle to develop
- Congenital cystic eye - failure of the globe to develop
- Colobomas - failure of complete closure that can affect the iris, retina or choroid
- Nanophthalmos - small eye with normal function
- Microphthalmos - small eye without normal function (e.g. cataract, coloboma, and congenital cyst).

Defects of the lids

- - usually due to defective muscles

of the upper lid but also and 3rd nerve palsy

- Eyelid colobomas - often associated with specific craniofacial syndromes

Defects of the corneal clarity can be partial or complete and caused by:

- (commonest with abnormally large eye)
- Forceps damage
- Interstitial keratitis
- Endothelial development abnormalities
- Intrauterine inflammation
- Persistent attachment of lens

Defects of the iris and pupil

These include:

- Corectopia - inappropriately positioned pupils are relatively common
- Polycoria – more than 1 pupil
- Coloboma of iris - usually seen in lower part towards nose and defects may affect other parts of the eye
- Aniridia - rare genetic defect with absent iris often with secondary glaucoma. If there is no family history, it may be associated with
- Albinism - may also have poor eyesight and
- Heterochromia - irises of different colours may be associated with normal function or may occur with congenital Horner's syndrome.
- Congenital cataracts can follow rubella infection of mother early in pregnancy or be an inherited defect. Small opacities may allow adequate vision. Large opacities prevent normal sight and lead to nystagmus and amblyopia. May need surgery within weeks of birth with long-term wear contact lenses. At the age of 1-2 years intra ocular lenses can be implanted.
- Other lens and anterior segment defects include colobomas and subluxation as occur in .
- When incorrect development of the neural crest affects the anterior segment, glaucoma is an important complication

White pupils (leukocoria) can be caused by

- cataracts,
- retinoblastoma,

- stage 5 retinopathy of prematurity (retrolental fibroplasia),
- severe posterior uveitis/vitritis and
- persistent hyperplastic primary vitreous and require urgent referral

Optic nerve defects range from frequent

- Minor defects of retinal vessels, to
- More severe defects, including
- Optic nerve hypoplasia - may be uni- or bi-lateral and is a relatively common non-progressive condition causing only relatively minor visual impairment to total blindness, often associated with congenital defects of the brain and facies.

Obstruction of nasolacrimal duct causes persistent tearing (epiphoria) in up to 30% of neonates but persists in only ~6%, who need probing.

Congenital glaucoma is frequently bilateral and associated with other defects. It needs early diagnosis to avoid irreversible blindness. Signs are:

- Severe photophobia
- Corneal haze
- Corneal opacity
- Increased corneal diameter
- Increased size of eye (due to raised intraocular pressure and elastic sclera)

Acquired eye problems in babies

Ophthalmia neonatorum can be caused by a number of agents including:

- Chlamydia - common problem presenting at age 5-14 days diagnosed by laboratory studies of conjunctival scrapings. Treatment is with systemic erythromycin.
- Bacteria - infection presents at age 2-5 days. Treatment needs to be based on examination of conjunctival smears. Neisseria gonorrhoea and Pseudomonas spp. infection carry significant risk of corneal damage
- Herpes simplex - diagnosed on examination of conjunctival smears. Usually self-limiting but may require topical antiviral medication

Figure 1: Gonococcal keratoconjunctivitis - acute, profuse, purulent discharge, hyperaemia and chemosis

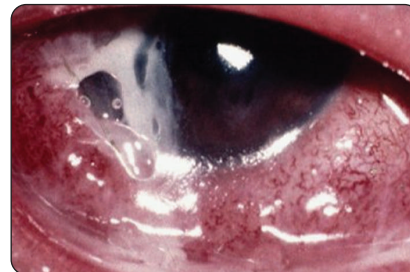


Figure 2: Complications of gonococcal keratoconjunctivitis - corneal ulceration, perforation and endophthalmitis in severe cases

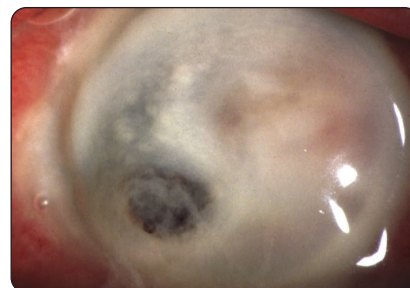


Figure 3: Herpes simplex conjunctivitis unilateral eyelid vesicles



Figure 4: Acute viral follicular conjunctivitis

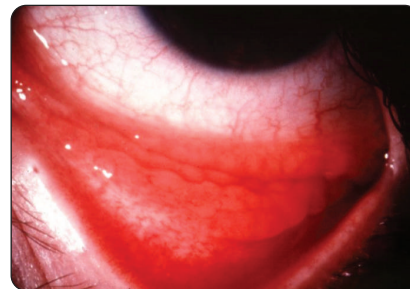


Figure 5: Neonatal chlamydial conjunctivitis - mucopurulent papillary conjunctivitis



Retinopathy of prematurity

occurs in >80% of neonates weighing less than 1kg and is increased by prolonged administration of oxygen. Develops when the vascularisation of the retina is interrupted, with active changes taking place at the line where vascular and avascular retina meet. The abnormal vascularisation clears up spontaneously in many cases but in about 4% of cases the condition progresses to blindness within the first year. Some patients require laser therapy. They should be carefully screened at 4 weeks after birth and thereafter every 2 weeks until age of 12 weeks.

Amblyopia is an unilateral reduced visual acuity that cannot be corrected with lenses, with no obvious cause. Usually occurs where an eye is deprived of normal visual experience during the first 6 -8 years of life. It becomes more difficult to overcome with age. Commonest causes are strabismus, due to suppression of images from affected eye to avoid double vision, and anisometropia, a high refractive error in one or both eyes.

The red inflamed eye

A number of features help to differentiate the various causes of uncomfortable inflamed eyes. The diagnosis can usually be based on a careful history and eye examination, looking for features such as watery or purulent discharge, itch or photophobia, unilateral or bilateral disease, as well as associated conditions on history and examination: atopy, eczema, rhinitis, upper respiratory infections. Patients should be referred for a full examination (including fluorescein staining) if any of the following features are present:

- Moderate to severe pain suggests a secondary cause such as keratitis or uveitis. Keratitis often presents with a unilateral, acutely painful, photophobic, intensely injected eye.
- Marked redness. The greater the redness, the more likely that there is a serious secondary cause. Ciliary injection, which is not always obvious, occurs with inflammation of deeper structures due to a secondary cause. It is

- indicated by redness and dilated episcleral blood vessels
- Reduced visual acuity, measured with a Snellen chart may indicate a serious secondary cause of conjunctivitis.

Figure 6: Conjunctivitis usually bilateral, acute watery discharge and follicles



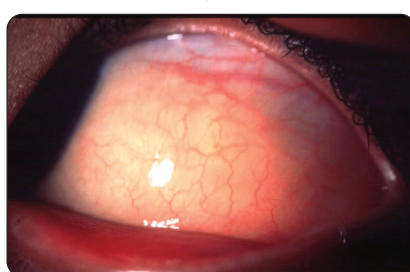
Figure 7: Subconjunctival haemorrhages and pseudomembranes if severe



Allergic Eye Disease

Presents with an intense itch or a burning sensation and with mild photophobia. There may be a history of contact with chemicals or eye drops and the history may have a seasonal time course. On examination a bilateral red eye, often with a clear watery discharge may be present. Oedema may be seen on the inside of the eyelid and lid swelling may occur.

Figure 8: Allergic conjunctivitis with transient conjunctival oedema (Usually due to hypersensitivity reaction to specific airborne antigens and frequently associated nasal symptoms)



Seasonal allergic conjunctivitis
Perennial allergic conjunctivitis: occurs all year because of

permanent exposure to the allergen, which is often house dust mites or pet dander. These conditions are often associated with a family history of asthma, eczema or allergic rhinitis.

Atopic keratoconjunctivitis is a chronic, bilateral inflammation of conjunctiva and eyelids, which is often associated with atopic eczema

Vernal keratoconjunctivitis is a bilateral chronic disease occurring in children who have a history of atopy. Black children often have the limbal (corneal margin) form. Symptoms usually peak prior to the onset of puberty and then gradually resolve over a period of five to ten years. Figure 9A-C depicts the progression of vernal conjunctivitis.

Figure 9A: Diffuse papillary hypertrophy, most marked on superior tarsus



Figure 9B: Formation of cobblestone papillae



Figure 9C: Rupture of septae - giant papillae



Giant papillary conjunctivitis is an inflammatory disorder of the superior tarsal conjunctiva. It occurs in the presence of an ocular foreign body and is often associated with

either soft or hard contact lenses.

The diagnosis is usually straightforward but other causes of uncomfortable, inflamed eyes must be considered, including infective conjunctivitis, blepharitis, (iritis), orbital cellulitis and ocular herpes simplex. Episcleritis presents with a relatively asymptomatic acute onset localized redness in one or both eyes. Foreign body usually affects one eye only. Pain, severe redness and reduced visual acuity are all indicators of possible alternative diagnoses requiring referral.

Management: Cold compresses may be soothing in the acute phase. Allergen avoidance is usually not practical, but contact lenses should not be worn if conjunctivitis is present or during a course of topical therapy. The topical ocular antihistamines, antazoline, emedastine and levocabastine provide rapid relief of symptoms. Newer antihistamines include olopatadine and ketotifen. Topical antihistamines are not appropriate for prolonged use and must not be used for longer than 4 months.

Mast cell stabilizers are recommended for use throughout a period of allergen exposure. Sodium cromoglycate is usually effective but the newer agents, lodoxamide, olopatadine and ketotifen may be effective in those with an inadequate response to sodium cromoglycate.

Topical corticosteroids should never be given for an undiagnosed red eye, when visual acuity is impaired, or if there is a history of ocular herpes simplex infection. Long-term use should be avoided because this can result in cataract, glaucoma, and severe bacterial or fungal infections involving the eyelid, conjunctiva, and cornea.

Oral antihistamines provide relief of symptoms and are particularly useful when there is associated allergic rhinitis.

Complications: Rarely a severe allergic reaction can lead to corneal ulceration.

Prognosis: Prognosis is excellent with resolution over a variable time course.

Infective conjunctivitis

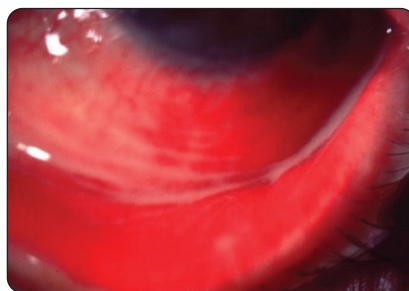
This is an inflammation of the conjunctivae caused by bacterial or viral infection.

Bacterial conjunctivitis: the most common pathogens include staphylococcus species, *Streptococcus pneumoniae*, *Haemophilus influenzae* and *Moraxella catarrhalis*. Bacterial conjunctivitis is more likely when the eyes are glued together on awakening in the morning, when both eyes are affected, there is an absence of itching and no history of previous episodes of conjunctivitis (as with Herpes simplex keratitis).

Figure 10: Simple bacterial conjunctivitis with crusted eyelids and conjunctival injection



Figure 11: Simple bacterial conjunctivitis with a subacute onset of mucopurulent discharge



Chloramphenicol is regarded as the first-line antibiotic in the treatment of bacterial conjunctivitis. It has a relatively broad spectrum of action against most Gram-positive and Gram-negative bacteria. Concerns about systemic toxicity of topical chloramphenicol, in particular aplastic anaemia, have been discounted.

Fucidic acid is an alternative to chloramphenicol, most effective against *Staphylococcus aureus*. It has very high ocular concentrations and is also effective against other pathogens.

The fluoro-quinolones are also very effective but are reserved for more serious infections like bacterial keratitis and corneal ulcers. The usual dose regimen would be 1 drop of antibiotic into the affected eye 2 hourly for 2 days then 4 x per day for 5 days. Ointment can be added at night.

Viral conjunctivitis: is commonly associated with upper respiratory tract infections, and is usually caused by adenovirus. It often presents unilaterally and has a watery discharge. A very useful clinical sign is an enlarged pre-auricular lymph node on the affected side (less than 5mm, about a half pea size).

Chlamydia presents with a chronic conjunctivitis in newborns due to maternal infection in the birth canal.

Infective conjunctivitis is contagious. Advise on measures to reduce the spread of infection: Wash hands regularly, particularly after touching the eyes or secretions. Avoid sharing towels, pillows or utensils.

Sick leave from school may be necessary. 🙅

See CPD Questionnaire, page 52

📄 This article has been peer reviewed

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