Ocular emergencies 2: non-traumatic

Introduction

This pair of articles aims to familiarize physicians with acute ophthalmic conditions requiring urgent referral and attention. They primarily focus on conditions that pose a direct threat to patients' vision and/or life, as well as on problems seen frequently in clinical practice.

A wide range of ocular emergencies, conditions and manifestations of systemic illness are encountered regularly in the primary care setting. Prompt recognition and appropriate referral of ocular emergencies are essential when the outcome may depend on timely management. This article covers non-traumatic ocular emergencies.

Non-traumatic ocular emergencies

Acute visual loss

Acute loss of vision is the most common pattern of visual loss seen in casualty. It can occur painlessly in an uninflamed eye, but can also be associated with pain, as in optic neuritis and arteritic anterior ischaemic optic neuritis, or with a red inflamed eye as in corneal ulcers and acute glaucoma. Those conditions that chiefly present with dramatic visual loss without a red eye are considered first and are summarized in *Table 1*.

Table 1. Ocular emergencies presenting mainly with sudden loss of vision

Central retinal artery occlusion
Central retinal vein occlusion
Optic neuritis
Non-arteritic anterior ischaemic optic neuropathy
Arteritic ischaemic optic neuropathy
Retinal detachment
Vitreous haemorrhage
'Wet' age-related macular degeneration

Retinal artery occlusions

Arterial occlusions occur as a result of complete interruption of the vessels by embolic, thrombotic or vasospastic diseases and are mainly seen in elderly people. Central retinal artery occlusions present with a sudden-onset, severe loss of vision in one eye. Visual acuity is severely diminished and pupillary examination always reveals a relative afferent pupillary defect.

The retinal arteries are extremely narrowed and the retina appears pale and oedematous. In contrast, as the fovea is very thin and receives its blood supply from the choroidal circulation, it retains its normal colour and appears as a cherry-red spot against the pale, infarcted retina (*Figure 1a*). Branch retinal artery occlusions usually present with visual field defects and the central vision may be preserved if the fovea is not affected.

Management involves urgent referral to an eye unit, where the intraocular pressure may be lowered to help restore the ocular circulation. Despite best management, visual recovery is usually very poor (Atebara et al, 1995). Cardiological assessment is mandatory to diagnose underlying cardiac or carotid disease. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) measurement is also indicated in all patients over the age of 50 years as arterial occlusion may be caused by an underlying giant cell arteritis.

Amaurosis fugax is a painless, transient, monocular loss of vision that can last from a few minutes to a few hours and may be repeated. During the episode, visual loss can be sectorial or total and results from temporary cessation of the ocular blood flow. Although retinal emboli can be sometimes seen during fundoscopy (*Figure 1b*), examination is usually unremarkable and the diagnosis is mainly based on history taking. Medical work up of patients with amaurosis fugax is essential as they are at risk of developing central retinal artery occlusion and stroke (Nguyen et al, 1999).

Retinal vein occlusions

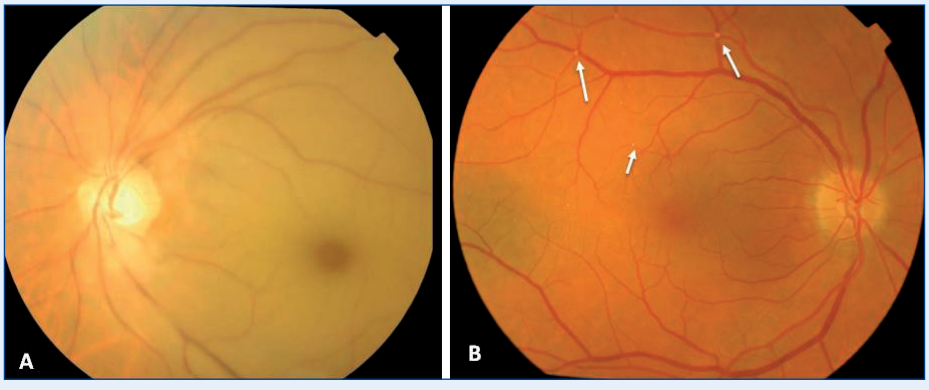
Patients with retinal vein occlusions typically present with acute visual loss that ranges from 6/9 vision down to hand movements, with the level of visual impairment dependent on the severity of the vascular blockage and its proximity to the fovea. Severe cases are usually associated with an relative afferent pupillary defect.

Retinal signs include congested tortuous veins, flame-shaped retinal haemorrhages and cotton wool spots scattered all over the fundus in case of central retinal vein occlusion or localized to a sector of the retina in branch retinal vein occlusion (*Figure 2*). Rapid ophthalmological referral is indicated to confirm the diagnosis and arrange for further follow up. Common underlying causes include hypertension, diabetes mellitus and hypercholesterolaemia. In young patients, autoimmune diseases should also be considered.

Optic neuritis

Optic neuritis is typically seen in middle-aged Caucasian patients and has a higher

Figure 1. a. Acute central retinal artery occlusion with a cherry-red spot. b. Retinal arteriolar emboli (arrows) in a patient with amaurosis fugax.



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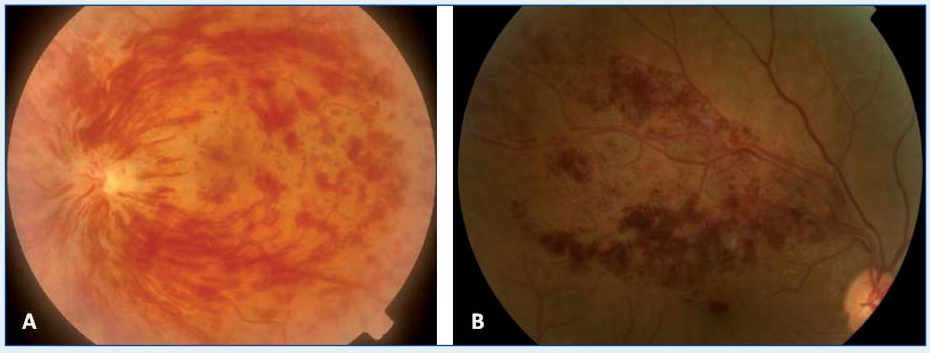


Figure 2. a. Diffuse retinal haemorrhages in a patient with central retinal vein occlusion. b. Branch retinal vein occlusion – note the localized distribution of the retinal haemorrhages.

predominance among females. It presents with blurred vision or a visual field defect and the visual acuity can range from 6/9 to no perception of light, although it is usually only moderately decreased. Ocular pain is common, being present in 90% of cases, and tends to be exacerbated with eye movements.

Assessment of colour vision is important, as it is usually markedly affected, even with a mild drop in visual acuity. The optic disc usually looks normal (retrobulbar neuritis), but may be hyperaemic and swollen. Neurological assessment needs to be considered, as optic neuritis can be a presentation of multiple sclerosis. Most cases are self-limiting and do not require treatment. Data from a large prospective randomized trial showed that intravenous steroids may have a role in speeding up recovery but do not influence the final level of visual acuity compared to observation (Beck, 1995).

Anterior ischaemic optic neuropathy

Anterior ischaemic optic neuropathy is a common cause of acquired optic nerve disease and can be classified as arteritic and non-arteritic. Non-arteritic anterior ischaemic optic neuropathy typically occurs in elderly patients who wake up with unilateral visual loss. Vision is usually moderately compromised and fundus examination reveals mild pallor and oedema of the optic nerve head that tends to be sectorial. Typically, there is an altitudinal visual field defect, with either the upper half or the lower half of the visual field being affected.

Non-arteritic anterior ischaemic optic neuropathy is thought to be an ischaemic process affecting the short posterior ciliary arteries that supply the optic nerve and is frequently associated with vasculopathic risk factors such as diabetes mellitus and hypertension. Visual loss is usually stationary and, to date, no treatment has proved to be effective (Beck et al, 1997; The Ischemic Optic Neuropathy Decompression Trial, 1998).

Arteritic anterior ischaemic optic neuropathy is a severe variant of ischaemic optic neuritis seen in cases of giant cell arteritis. It presents with severe limitation of vision in one eye, and is often painful. The optic disc is characteristically very pale and may show features of vascular blockage (*Figure 3*). Eliciting the diagnosis is crucial as giant cell arteritis may be associated with rapid involvement of the other eye (Aiello et al, 1993) as well as death (Uddhammar et al, 2002). Systemic steroids should be started urgently on the basis of clinical suspicion and ESR and CRP results, and the patient then considered for a diagnostic temporal artery biopsy.

Retinal detachment

Rhegmatogenous retinal detachment is one of the most time-critical emergencies encountered in the accident and emergency department. Incidence of rhegmatogenous retinal detachment is about 1 in 12 000 population with the condition being more common in high myopes and in those with a history of ocular trauma or cataract surgery.

Retinal detachment occurs when fluid from the vitreous cavity flows through a retinal tear and dissects underneath the retina, raising it off the underling retinal pigment epithelium and choroid layers. Typically patients with retinal detachment present a with sudden visual field defect that appears peripherally as a 'black curtain' and then progresses to involve the entire visual field. The occurrence of visual field defect is commonly preceded by symptoms of floaters and flashes of light that is related to vitreous liquefaction and retinal traction.

Decreased visual acuity occurs when the retinal detachment has reached the centre of the macula – the so-called 'macula off' retinal detachment. Fundus examination is usually sufficient to establish the diagnosis of retinal detachment (*Figure 4*) and localize the retinal tears. Management is mainly surgical and the prognosis of vision depends on the extent and duration of retinal detachment (Ross and Kozy, 1998). In general, the prognosis for the return of normal vision after the repair of a 'macula on' detachment is better than in cases where the macula has already detached. In addition, macula off detachments tend to have a better visual prognosis if they are repaired within 1 week than if they are operated upon later.

Vitreous haemorrhage

Vitreous haemorrhage is an important cause of sudden visual loss. In addition to trauma and posterior vitreous detachment, vitreous haemorrhage can occur from new retinal vessels as a result of proliferative diabetic retinopathy or retinal vein occlusions.

Figure 3. Anterior ischaemic optic neuropathy with a swollen pale optic nerve.

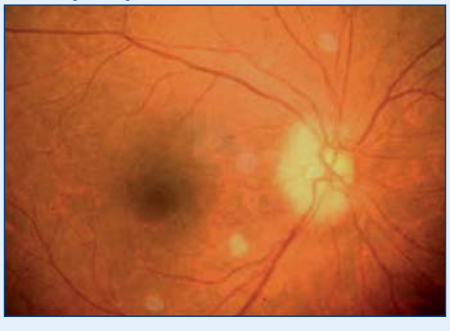
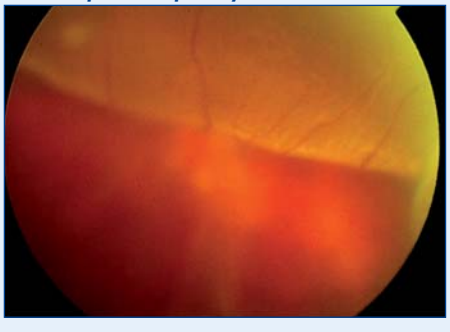


Figure 4. Retinal detachment. Note the retinal elevation present superiorly.



Patients usually complain of sudden onset floaters and reduction of vision. The visual acuity depends on the extent of the haemorrhage and can be severely decreased.

Examination is usually sufficient to elicit the diagnosis but ultrasonography may be needed to exclude retinal detachment in cases with extensive haemorrhage precluding retinal view. Management is mainly in the form of observation until haemorrhage clears, although vitrectomy surgery may be needed for persistent haemorrhage or coexistent retinal detachment. Rapid referral of patients with proliferative diabetic retinopathy is important, as laser retinal treatment decreases the chance of developing vitreous haemorrhage by 50% (*Figure 5*) (The Diabetic Retinopathy Study Research Group, 1981).

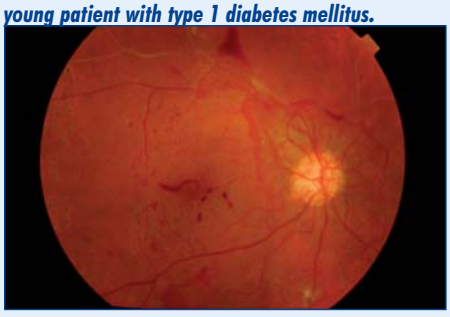
'Wet' age-related macular degeneration

So-called 'wet' age-related macular degeneration is the commonest cause of visual loss in the elderly. The disease occurs as a consequence of disturbance of the retinal pigment epithelium and growth of subretinal neovascular membrane that causes bleeding and scarring. Patients present with distortion of central vision and visual loss, which are of sudden onset. Although the disease is bilateral, it usually affects one eye at a time.

In contrast to optic nerve problems, there is usually no relative afferent pupillary defect, but ophthalmoscopy reveals subretinal blood, and hard exudates localized at the macula and sometimes the neovascular membrane can be seen as a grey patch under the macula (*Figure 6*).

Rapid referral is essential as most cases of wet age-related macular degeneration are now amenable to treatment with intravitreal injections of anti-angiogenic factors as ranibizumab (Lucentis) (Kaiser et al, 2007).

Figure 5. Proliferative diabetic retinopathy in a young patient with type 1 diabetes mellitus.



Acute red eyes

A red eye is one of the most common problems seen in the eye casualty. While most cases are self-limiting and can be fully managed by the primary care physician, others are more serious and require immediate referral. Attention to associated symptoms helps to ascertain the nature of the problem. Sight-threatening conditions are likely to be painful or associated with a decrease in visual acuity (*Table 2*), in contrast to non-sinister causes such as episcleritis and conjunctivitis.

Anterior uveitis (iridocyclitis)

Patients with anterior uveitis (iridocyclitis) usually present with ocular redness associated with pain, photophobia and blurring of vision. The visual acuity is usually normal in anterior uveitis, unless it is complicated by a particularly dense inflammatory process or macular oedema. The hallmark of anterior uveitis is the presence of cells and flare in the anterior chamber reaction resulting from disruption of the blood aqueous barrier. These inflammatory cells can be deposited on the corneal endothelium as keratic precipitates (*Figure 7*).

All patients with anterior uveitis should have a fully-dilated fundus examination to determine whether there is any concomitant posterior segment involvement, and to diagnose complications such as macular oedema. Although several syndromes can be associated with anterior uveitis, in up to 50% of cases no specific aetiology is found. Most forms of anterior uveitis need active treatment in the form of topical corticosteroids and cycloplegics.

Figure 6. 'Wet' age-related macular degeneration. Note the presence of the subretinal haemorrhage at the macula. The dark appearance of the fovea is caused by the presence of the underlying neovascular membrane.

Table 2. Ocular emergencies presenting mainly with ocular redness and pain

Iridocyclitis
Anterior scleritis
Corneal ulcer
Acute glaucoma
Orbital cellulitis

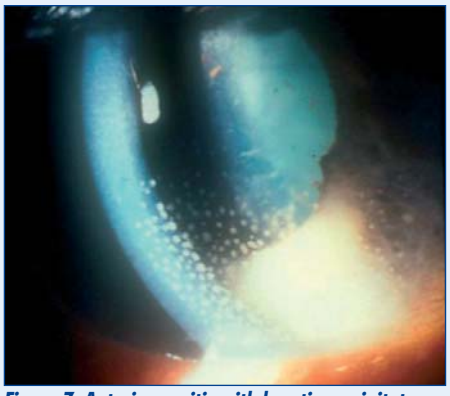


Figure 7. Anterior uveitis with keratic precipitates on the lower half of the cornea.

Anterior scleritis

Scleritis is inflammation of the sclera and can be anterior or posterior. Patients with anterior scleritis present with a painful red eye. Pain is characteristically very severe, dull aching and awakens the patient from sleep; there may also be an associated drop in visual acuity especially with posterior scleritis.

Vascular engorgement is usually extensive and deep (*Figure 8*), but may be associated with an area of blanched sclera that represents ischaemia. Scleritis can be a presenting manifestation of potentially serious systemic diseases such as rheumatoid arthritis and Wegener's granulomatosis and thus requires urgent referral (Sainz de la Maza et al, 1994).

Corneal ulcer

Corneal ulcers can be diagnosed in patients with a painful red eye and a corneal infiltrate or epithelial defect that takes up fluorescein dye. Although they can be sterile, the infective form is often seen in patients with a history of ocular trauma or contact lens wear and is usually bacterial (*Figure 9*). Other causes of infective ulcers include herpes simplex virus, which causes a characteristically dendritiform ulcer and tends to recur. Urgent referral of patients with corneal ulcers is needed for corneal scraping and the institution of antimicrobial therapy.

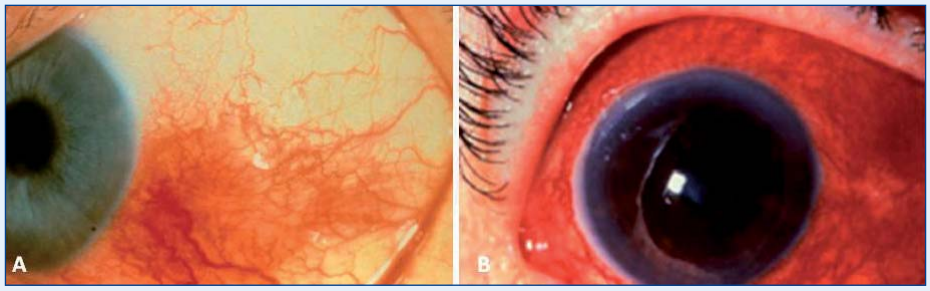


Figure 8. a. Episcleritis with localized superficial ocular redness. b. Scleritis. Note the diffuse and the deep level of ocular redness and vascular engorgement in (b).

Acute glaucoma

Acute angle closure glaucoma usually presents with a fairly dramatic onset of ocular pain, headache and sometimes vomiting. This problem is mainly encountered in elderly long-sighted patients and visual acuity is usually reduced. Examination quickly reveals a hazy cornea, shallow anterior chamber and a semi-dilated, non-reactive pupil (*Figure 10*). The diagnosis is comparatively straightforward once the intraocular pressure is measured – in the absence of specialist equipment, palpating the globe underneath closed lids can easily demonstrate a markedly raised intraocular pressure.

Figure 9. Infective corneal ulcer. Note the large corneal infiltrate and epithelial defect present.

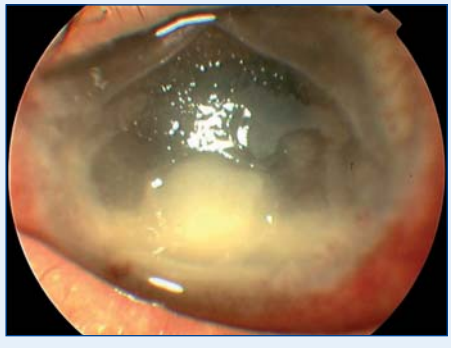
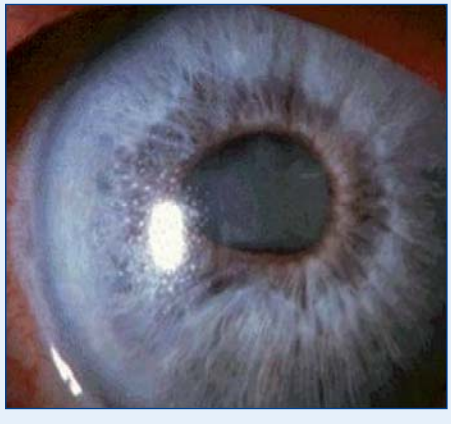


Figure 10. Acute glaucoma. Note the corneal oedema present.



Urgent referral is indicated for early introduction of intraocular pressure-lowering agents, in the form of systemic carbonic anhydrase enzyme inhibitors as well as topical drops, including miotics. Laser iridotomy can be used to abort an existing attack and to prevent further attacks from occurring in the future.

Orbital cellulitis

Orbital cellulitis results from microbial infection of the orbit and the eye lid(s). In healthy patients, the most common route of infection is spread from the adjacent sinuses or dental infection. Patients typically present with significant pain and fever. Signs include marked lid swelling, oedema and tenderness.

Proptosis, decreased ocular motility, decreased visual acuity and the presence of a relative afferent pupillary defect are important signs to look for as these differentiate orbital cellulitis from isolated lid inflammation (i.e. preseptal cellulitis), which is not a real emergency (*Figure 11*). Orbital cellulitis is a medical emergency that can lead to cavernous sinus thrombosis and blindness as well as death, and patients need immediate hospitalization and intravenous broad-spectrum antibiotics, as well as computed tomography scanning of the orbit and brain to look for abscess formation.

Figure 11. Right ocular redness and limited upward eye movement in a young adult with proptosis resulting from orbital cellulitis.

Acute neuro-ophthalmological problems

Papilloedema

Patients with papilloedema are usually referred from their GP because of persistent headaches. The hallmark of the diagnosis is the presence of bilateral swollen optic discs and normal vision, although optic nerve functions can be affected in advanced cases. Mild disc swelling can be easily overlooked and it is also sometimes difficult to differentiate between true disc swelling and a congenital condition called optic disc drusens in which the disc appears swollen as a result of the presence of buried colloid material (*Figure 12*). The observation of spontaneous venous pulsations at the disk may assist the diagnosis, as these are extremely unlikely in the presence of raised intracranial pressure.

Papilloedema signifies an increase in intracranial pressure that could be life threatening and necessitates urgent referral for brain imaging. Common causes include malignant hypertension, brain tumours and intracranial haemorrhage.

Acute diplopia

Development of double vision that disappears on covering one eye (i.e. binocular diplopia) is most likely to indicate a problem of ocular movement. If this happens dramatically, paralysis of one or more of the cranial nerves controlling the eye movements (III, IV or VI) should be considered. While vascular causes are common underlying aetiologies, an urgent neuro-ophthalmology referral is mandatory to exclude an intracranial space-occupying lesion, such as a neoplasm or aneurysm, especially in young patients with no history of long-standing vasculopathy or in pupil-involving III nerve palsies where a posterior communicating artery aneurysm must be urgently excluded.

Neonatal ocular emergencies

Opacity of the ocular media

Clarity of the ocular media is essential for normal development of visual function in infants and young children. In this age group, unlike in adults, corneal opacities and cataracts require rapid attention. Delay in referral can lead to poor development of formed vision – a condition called sensory deprivation amblyopia. Leucocoria describes the appearance of white pupillary

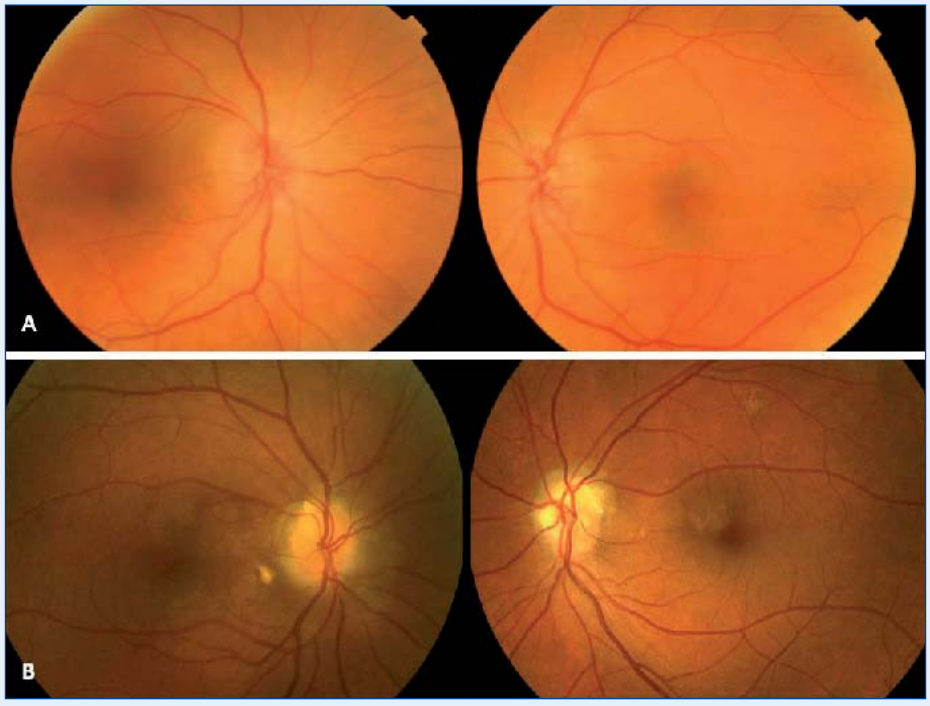


Figure 12. a. Papilloedema. b. Bilateral disc drusens. Note the lumpy nature of the discs in optic drusens that could be mistaken for swollen discs.

reflex in children (*Figure 13*). Although most cases are caused by lens opacity, leucocoria can be an early manifestation of retinoblastoma – a malignant intraocular tumour arising from the retina (Shields and Shields, 1992).

Congenital glaucoma

Congenital glaucoma should be suspected in infants who present with constant watering and photophobia of one or both eyes. Parents may also complain that their child has an unusually large eye, as children's eyes can stretch with increased intraocular pressure. Cloudiness of the cornea is seen in advanced cases. Awareness of this condition is important as early referral influences the visual prognosis.

Figure 13. Leucocoria in a child with retinoblastoma.

Conclusions

Successful management of ocular emergencies depends on prompt diagnosis and treatment by ophthalmologists as well as close collaboration with primary care physicians who frequently face the challenge of making a proper diagnosis and referral. **BJHM**

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Conflict of interest: none.

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KEY POINTS

- Acute red eye is likely to be caused by a sight-threatening condition if the eye is painful or visual acuity is decreased.
- Emergency medicine physicians must be familiar with the symptoms of retinal detachment and should be able to perform a dilated fundus examination.
- Fresh retinal detachments need urgent referral to ophthalmologists particularly if the macula is still attached.
- Early referral of patients with proliferative diabetic retinopathy is crucial as retinal laser treatment can halve the chance of developing vitreous haemorrhage and severe visual loss.
- Fundus examination should be included in the routine work up of every patient complaining of persistent headache to exclude papilloedema.
- Primary care physicians and paediatricians should be familiar with the technique of checking the red reflex in infants and have a low threshold for referring children with an abnormal reflex to ophthalmologists.