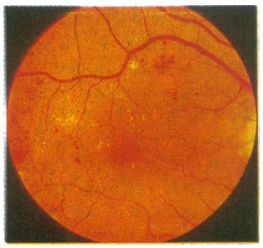
GENERAL MEDICAL DISORDERS AND THE EYE

A R Elkington, P T Khaw

- Diabetes mellitus
- Hypertension
- Dysthyroid eye disease
- Rheumatoid arthritis
- Other arthritides
- Rosacea
- Sarcoid
- Congenital rubella
- AIDS

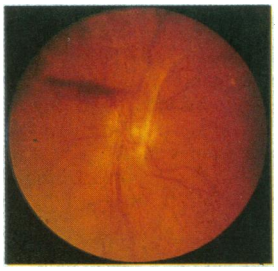
There are few serious medical conditions that do not affect the eye. It is important to know the ocular manifestations of systemic diseases because, firstly, screening is required to detect early ocular changes that may require treatment to prevent blindness. A good example is a diabetic with new vessels on the optic disc, which signal an exceptionally high risk of visual loss unless treatment is given in time. Secondly, knowledge of the ocular complications of other diseases may help in the diagnosis of an ocular problem. A red, locally injected, and tender eye in a patient with rheumatoid arthritis suggests scleritis, which may progress to perforation of the eye. Iritis should be strongly considered in a young man with ankylosing spondylitis who presents with a red eye. Thirdly, the ocular symptoms may suggest the systemic disease—for example, prominent eyes and lid lag in hyperthyroidism—or confirm it—for example, the Kayser-Fleisher ring of copper in Wilson's disease. Lastly, the ocular signs may have prognostic value. If cotton wool spots occur in the eyes of an otherwise asymptomatic patient with AIDS the prognosis is particularly poor.

Diabetes mellitus



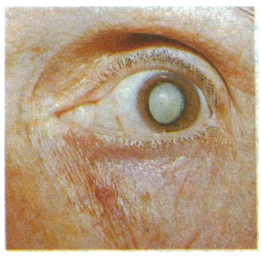
Background retinopathy: hard exudates, microaneurysms, and haemorrhages.

Diabetes mellitus is the commonest cause of blindness among people of working age in the Western world. Two per cent of the diabetic population are blind, many of them in the younger age groups. Much of this eye disease can now be prevented by treatment, which makes early identification and referral crucial. What are the treatable causes of visual loss in diabetics, and how can they be detected early enough to be effectively treated?



Proliferative retinopathy: new vessels, fibrosis, and haemorrhage.

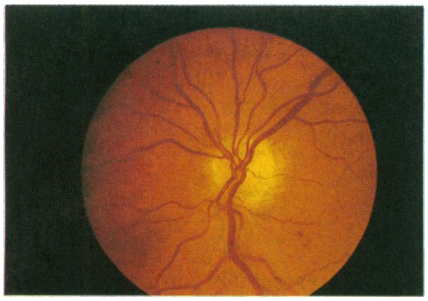
Cataract and chronic open angle glaucoma are more common in diabetic than in non-diabetic patients. Cataract can be treated by surgical removal, and chronic open angle glaucoma may be treated by drugs and operations that lower the intraocular pressure. They can often be detected by viewing the red reflex and examining the optic disc, respectively. It is only too easy to forget to look for glaucomatous cupping of the disc when looking for the signs of diabetic retinopathy.



Cataract in diabetic patient.

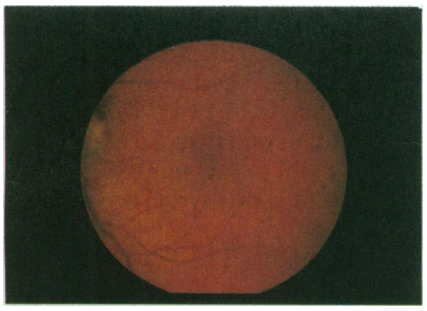
Blinding diabetic retinopathy occurs in both insulin dependent and non-insulin dependent diabetics and affects all age groups. The longer the duration of the diabetes, the more likely the patient is to have retinopathy (about 80% are affected after 20 years). Again this applies to all categories of diabetics. Diabetics should have their pupils dilated yearly with tropicamide 1% and the fundi examined if important physical signs are not to be missed. There are two main clinical types of retinopathy that cause blindness in diabetics, and these need to be identified and the patients referred for early treatment.

Types of diabetic retinopathy.

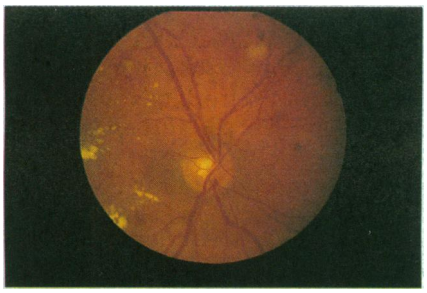


Background retinopathy with good acuity: regular review.

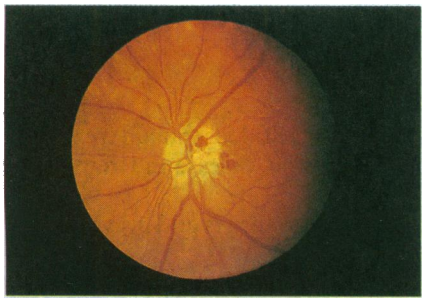
Background retinopathy with macular changes and good vision: refer.



Background retinopathy with impaired acuity: refer.



Preproliferative retinopathy. Cotton wool spots, large haemorrhages, and tortuous veins: refer.



Proliferative retinopathy: refer urgently.

Background retinopathy is typified by microaneurysms, dot haemorrhages, and hard yellow exudates with well defined edges. These changes do not have much effect on vision when they occur in the peripheral retina. When they occur in the macular area, however, central vision may be severely affected. Background retinopathy at the macula (diabetic maculopathy) is the major cause of blindness in maturity onset diabetes, but it also occurs in younger, insulin dependent diabetics. It may be amenable to focal laser photocoagulation, which may help to reduce any leakage. This is particularly true when hard exudates are a prominent feature of the maculopathy.

Proliferative retinopathy is typified by the growth of new vessels on the retina or into the vitreous cavity. This is thought to be due to the ischaemic diabetic retina producing a vasoproliferative factor that causes the growth of abnormal new vessels. These vessels may bleed causing a sudden decrease in vision due to a vitreous haemorrhage. Worse still, this blood often results in the production of contractile membranes that gradually pull off the retina, causing blindness. This may occur in any diabetic, but more commonly in the young, insulin dependent patient. The vision may be 6/6 right up to the moment of a bleed, hence the need for early detection of new vessels by adequate fundal examination. New vessels may also grow on to the iris and occlude the drainage angle of the anterior chamber causing a painful hard eye (rubeotic glaucoma).

Laser (or any other method of photocoagulation) is also used to treat proliferative retinopathy. The laser, however, is not usually used to coagulate new vessels as these may bleed or recur. When a patient has new vessels at the disc the entire retina is treated with a laser, except for the macular area, which preserves the central vision. Hence the term "panretinal photocoagulation" or "pattern bombing." This destroys the ischaemic peripheral retina and stops it producing the vasoproliferative factor that induces the growth of new vessels, and often the new vessels regress. New blood vessels on the iris that block the outflow of aqueous and cause rubeotic glaucoma may also regress. It may, however, require thousands of laser burns and repeated treatments to achieve this.

New microsurgical techniques now allow blood filled vitreous to be removed, and the membranes that are pulling off the retina to be cut, but even in the most skilful hands the risks of these procedures are high. Again this emphasises the importance of early referral for photocoagulation.

Screening

Patients may be divided into the following groups for screening purposes.

(1) Those with no retinopathy or with background retinopathy and normal vision when tested with glasses or pinhole. These patients can be reviewed yearly with dilatation of the pupils. They should be told to attend sooner if there is a change in vision that is not corrected with glasses.

(2) Those with background retinopathy and changes around the macular area. They should be referred as this may herald a blinding maculopathy.

(3) Those with background retinopathy and impaired acuity not corrected with glasses or pinhole. It may be that the patient has an oedematous or ischaemic form of maculopathy that is extremely hard to diagnose with the direct ophthalmoscope alone. The oedematous form may respond to focal laser treatment if this is given early.

(4) Those with preproliferative retinopathy. They have no new vessels, but the haemorrhages are larger, the veins are tortuous, and there are cotton wool spots. These physical signs imply that the retina is ischaemic and that there is a high risk that new vessels will subsequently form. These patients should be referred.

(5) Those with proliferative retinopathy. This is typified by new blood vessels, and sometimes cotton wool spots, fibrosis, and vitreous haemorrhages. These patients need immediate referral, particularly if there are vitreous haemorrhages.

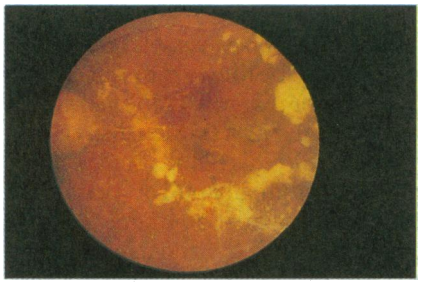
In addition to ocular treatment, blood sugar concentrations should be carefully controlled. If the blood sugar concentration is brought under control rapidly the fundus should be reviewed regularly during this period as there may be a transient worsening of the retinopathy. Hypertension and

- Control blood sugar
- Control hypertension
- Control hyperlipidaemia
- **Stop smoking**

hyperlipidaemia worsen the prognosis of retinopathy and must also be controlled. Patients should be told to stop smoking.

Diabetics are also more prone to recurrent corneal abrasions, retinal vein occlusions, and cranial nerve palsies. Aids for a diabetic with impaired vision include an audible click count syringe, and a Hypotest instrument that gives an audible signal with urinary Diastix.

Hypertension

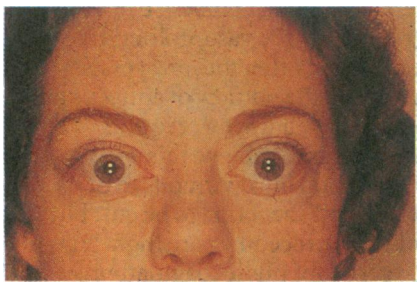


Retinopathy in malignant hypertension with macular exudates and occluded vessels; disc swelling has resolved.

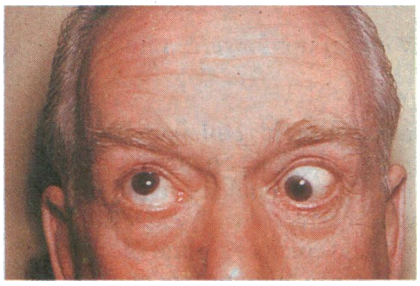
The mild fundal changes of hypertension are extremely common. "Silver wiring" of the retinal arteries and arteriovenous nipping are well known signs, but arteriolar narrowing is the most reliable fundal sign.

Malignant hypertension is classically associated with swelling of the head of the optic nerve. Any patient with hard exudates, cotton wool spots, or haemorrhages due to hypertension has a grave prognosis. A patient with these fundal signs should have his or her blood pressure checked and diabetes excluded. Urgent referral is required as not only is this combination of signs life threatening, but it may also result in blindness. Retinal vein occlusion is also more common in hypertensive patients.

Dysthyroid eye disease



Hyperthyroidism with lid retraction.



Autoimmune eye disease with restriction of ocular movements.

Choroidal folds.

Patients may have signs associated with hyperthyroidism and the consequent overactivity of the sympathetic system. These patients have retracted upper and lower lids caused by excessive stimulation of the sympathetically innervated muscles in the eyelids. This also gives rise to the well known sign of lid lag when the patient looks downwards. These features may suggest the diagnosis when the patient walks into the surgery.

If these signs are present thyroid dysfunction should be excluded. If there are no visual problems, no corneal exposure, and the eyes move normally the patient need not be referred. Patients may, however, also have evidence of autoimmune disease directed against the orbital contents, particularly the muscles. These signs may be associated with the classic signs of Graves' disease including goitre, pseudoclubbing of the fingers (thyroid acropathy), hyperthyroidism, and pretibial myxoedema. Autoimmune ocular disease may also occur on its own with no thyroid dysfunction. The clinical features include:

- *Swelling of the eyelids.*
- *Oedema (chemosis) and injection of the conjunctiva.*
- *Exposure of the cornea* because of lack of blinking and failure of the lids to cover the eye adequately.
- *Pronounced protrusion of the eyes.* The absence of this feature in association with the other features may be even more serious as it may be that a tight orbital septal wall is holding back the swollen orbital contents. This may lead to a rise in intraocular pressure as well as pressure on the optic nerve.
- *Restriction of eye movements.* This is caused by infiltration of the muscles with inflammatory cells and consequent inflammation, oedema, and finally fibrosis.
- *Optic neuropathy.* This is comparatively rare, but the fundal signs include vascular congestion and swelling or atrophy of the head of the optic nerve. There may be "folds" in the choroid caused by pressure on the globe. This should be excluded in any patient with autoimmune eye disease who experiences visual deterioration.
- These features may occur in any combination.

Management

- Associated thyroid dysfunction should be excluded, though treatment of any dysfunction may make no difference to the eye disease and may even make it worse.
- Artificial tears should be used to lubricate the cornea and prevent drying and corneal ulceration.
- If there are cosmetic or exposure problems caused by lid retraction guanethidine drops 5% may reduce the lid retraction by relaxing the

- Protect cornea
- Prevent damage to optic nerve

sympathetically controlled retractor muscles. Occasionally operation on these muscles may be required.

- If corneal exposure is threatening sight the eyelids may have to be sewn together temporarily (tarsorrhaphy).
- Prisms incorporated in the patient's glasses may help to correct any double vision.
- Operations on the muscles of eye movement may be required to realign the eyes in patients with longstanding diplopia. Recently the introduction of local injections of minute doses of botulinum toxin to "loosen" the muscles has meant that patients with restrictive muscle disease may sometimes be treated at an earlier stage.
- In serious disease with corneal problems or pressure on the optic nerve emergency treatment may be required, which may include high doses of steroids, surgical orbital decompression, and radiotherapy.

Rheumatoid arthritis

Episcleritis.

Scleritis.

Rheumatoid arthritis is another common disease in which ocular complications are frequent. The lacrimal glands are also affected by an inflammatory process with consequent inadequate tear flow. The patient complains of dry, gritty, and sore eyes. Treatment consists of replacement artificial tear drops instilled as often as necessary. Simple ointment may also help, but this will blur the vision if used during the day. If there is an aggregation of mucus, mucolytic eye drops—for example, acetylcysteine—may help, but patients should be warned that these sting.

The inflammatory process may also affect the episcleral and scleral coats of the eye causing the patient to complain of a red, uncomfortable eye. The redness is usually focal and there is tenderness over the area. Scleritis is usually much more painful than episcleritis and the injected vessels are deeper. If scleritis continues the sclera may become thin (scleromalacia) and the eye may eventually perforate (scleromalacia perforans). The patient should be referred, as systemic treatment may be indicated.

These processes may also occur in other connective tissue diseases such as systemic lupus erythematosus, scleroderma, and dermatomyositis.

Other arthritides

Chronic anterior uveitis and secondary cataract in seronegative arthritis.

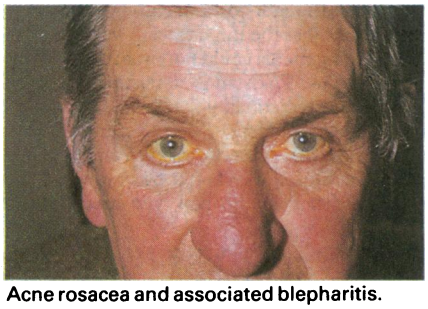
High risk groups

- Fewer than five joints affected
- Antinuclear factor positive
- Rheumatoid factor negative

The seronegative arthritides include ankylosing spondylitis, Reiter's syndrome, psoriatic arthritis, and arthritis associated with inflammatory bowel disease. Acute anterior uveitis (iritis, iridocyclitis) is much more common in these patients. If a patient with any of these conditions has a red eye anterior uveitis should be suspected. This is particularly true if the patient has had past attacks, and "experienced" patients often know when an attack is coming on. The patient should be referred for early treatment, which may prevent some of the complications of anterior uveitis.

Seronegative childhood arthritis is a particularly important cause of chronic anterior uveitis. The great danger is that the eyes in this condition can often look white and quiet and the child may not complain of any visual problems. There may also be secondary cataract that may cause irreversible amblyopia even if the cataract is removed later in life. Glaucoma secondary to the anterior uveitis may also occur. The groups of children particularly at risk are females, those with fewer than five joints affected by the arthritis (pauciarticular), and those with antinuclear antibodies in their blood. These children should be referred to an ophthalmologist.

Rosacea

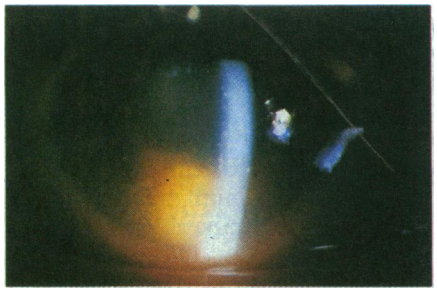


Acne rosacea and associated blepharitis.

Rosacea may seriously affect the eyes. There is often associated blepharitis, which may result in recurrent chalazia and styes. The abnormal lids and lipid secretion affect the tear film and the symptoms of "dry eye" result. The cornea scars, particularly in the inferonasal and inferotemporal areas, with corneal neovascularisation. Thinning occurs and the cornea may occasionally perforate.

Treatment with tear substitutes is indicated together with treatment for any associated blepharitis. Systemic tetracycline (250 mg four times daily for up to a month, then daily for several months) may considerably improve the patient's ocular as well as facial condition.

Sarcoid



Anterior uveitis in sarcoidosis: large deposits of inflammatory white cells on posterior surface of cornea.

Sarcoid is associated with various ocular problems. Acute uveitis and chronic uveitis occur, which may result in cataract, glaucoma, and a band of calcium deposited in the cornea. The lacrimal glands may be infiltrated resulting in symptoms of "dry eye" requiring tear replacement. The granulomatous process may also affect the posterior part of the eye in the form of vasculitis and sometimes infiltration of the optic nerve.

Congenital rubella

- Cataract
- Squint
- Refractive error
- Glaucoma
- Retinopathy

The ocular manifestations of congenital rubella are extremely important. The child may be mentally retarded and deaf, thus early recognition of ocular problems and their treatment are vital. The treatable defects include cataract, glaucoma, squint, and refractive errors. The cataract may not appear until several weeks or months after birth, so the eyes should be re-examined. There may be a diffuse retinopathy ("salt and pepper" appearance).

Acquired immune deficiency syndrome (AIDS)

Cytomegalovirus retinitis in AIDS.

The ocular complications of AIDS may be blinding. Manifestations include Kaposi's sarcoma of the conjunctiva, retinal haemorrhages, and vasculitis. Cotton wool spots may appear and disappear spontaneously, and their presence signifies a poor prognosis even in a patient without symptoms. Ocular cytomegalovirus infection presents as areas of opacification with haemorrhages and exudates that proceed to severe ocular damage. Some new antiviral agents, however, have proved useful in the treatment of this severe complication.

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MULTICULTURAL MEDICINE

Fingers cause problems

Fingers are used for pointing at things. In Eastern cultures their use for eating food is well known. They are, however, also used in making gestures. A gesture can have a different meaning in different societies: what appears to be a vulgar sign in one country may well have an innocent meaning in another. Not so long ago an innocent incident almost caused a diplomatic breakdown between Pakistan and Britain.

Shakoor Rana was the umpire in a cricket match between the Pakistani and English teams. The match was televised internationally and spectators in both countries were glued to their sets. At one stage the Pakistani umpire looked Mike Gatting, the English team captain, in the eye and waving his index finger told him, "Stop, this is cheating." The English captain lost his temper in the heat of the moment and showed the umpire two fingers in a reverse of the victory sign. The match was suspended because Shakoor Rana demanded a written apology, which was refused by Mike Gatting. Each believed, as many car drivers do, that it was entirely the fault of the other person. Eventually, the Test and County Cricket Board in London ordered the English captain to give a written apology. Mike Gatting wrote an apology on a tatty piece of paper and this was shown on television world wide. This incident seriously damaged relations between the cricket teams, each believing that they were in the right. But both governments remained calm, not punishing either the umpire or the captain, confirming that both were right.

Let us analyse this interesting sequence of cultural misunderstandings. Looking someone in the eye is desired in Western cultures during a conversation, but it is a rude gesture in Eastern cultures and only used when a person is annoyed—and then it is used frequently. Waving an index finger is used for emphasis by an Asian during conversation but is considered to be extremely rude in English society, especially to a woman or the captain of a team. In cases in which an Englishman would say "it is not fair" an Asian would say "it is cheating." The word cheating is taken as an insult by a Westerner but it is a light hearted comment in the East.

Showing two fingers in a reverse of the victory sign is extremely rude—akin to a four letter word—in English society, but in Pakistan it means counting two, and no more. Demanding a written apology is an Eastern symbol of authority but is considered to be an insult in the West. Writing an apology on a tatty piece of paper is thought to be insulting by the English, but to Pakistanis it simply means that the English team was short of decent notepaper. Finally, the Pakistani media could not broadcast the meaning of the rude two finger sign because Pakistan is an Islamic state; also the sign is not mentioned in any English dictionary.

Such incidents can occur in any doctor's practice, during a medical consultation, and anywhere in the world when doctors deal with a patient or a health professional from another culture. We should be aware and tolerant of cultural differences. Nevertheless, mind how you use your fingers.

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