Treascher Collins syndrome with novel ophthalmic findings and visceral anomalies

Treascher Collins syndrome (TCS) (mandibulofacial dysostosis (MFD) or zygauroman-dibular dysplasia) is one of a group of congenital malformation syndromes that have in common maldevelopment of the first and second branchial arches. Clinical features typically include hypoplasia of the mandible and zygoma; a complex variety of ear abnormalities including malformed pinnae, atresia of the external auditory canals and anomalies of the middle ear ossicles; cleft palate; receding chin; and sinus and choanal atresia. We present a pair of twins that possessed not only classic features of TCS but also unusual visceral and previously unreported ophtalmic pathology.

Case reports
A 32 year old, gravida 3 para 2-0-0-2 woman underwent elective abortion after fetal death and major fetal malformations were diagnosed during pregnancy. Ultrasound examination at 20 weeks of gestation displayed monoyzotic, diarnniotic twins. Fetus A, noted alive at 16 weeks, was dead and surrounded by minimal amniotic fluid. Fetus B was alive with several anomalies including bilateral infraorbital cystic masses, protruding tongue, and large median facial cleft. Amniocentesis revealed a 46XY karyotype without numerical or structural chromosomal abnormalities. The parents denied any significant medical history, family history, of congenital anomalies, or use of drugs or alcohol. Their two other children, ages 17 and 33 months, were normal. No family members exhibited features of TCS on visual examination.

Necropsy measurements were consistent with 16 weeks of gestational age for twin A and 21 weeks for twin B. Both twins had bilaterally hypoplastic zygomas, maxillae, and related muscles; severely hypoplastic and misplaced pinnae; bilaterally agenic external auditory canals; bilaterally lateral facial clefts, and severe micmagnathia (Fig 1). Twin A exhibited left sided choanal atresia, agenesis of the hard and soft palate, and multiple visceral anomalies, including dual superior vena cava, bilobed right and unilobed left lungs, bilateral renal and ureteral agenesis, rudimentary urinary bladder, and absent epididymides. Twin B was noted to have right sided choanal atresia, soft palate aplasia, hard palate hypoplasia, and a left sided cleft; twin B had no visceral anomalies. Ocular pathologi-cal dissection of twin A illustrated bilateral microphthalmia; corneal sclerisation; and maldevelopment of the uvea, lens, and retina. Ocular dissection of twin B revealed microphthalmia, aniridia, congenital cataracts, and bilateral vascularisation of the corneas.

Comment
These twins had multiple features characteristic of TCS, most notably hypoplastic zygo-mas, maxillae, and related muscles—perhaps the most characteristic features of TCS. In contrast, visceral anomalies, such as those of twin A, are rare. Only two cases have been previously reported: one with tracheosopho-gastrial fistula, rectovaginal fistula, and anal atresia; another with achiadals. Renal agenesis, found in twin A, has not been previously described in association with TCS. While ophthalmological features in TCS are often extensive, they seldom involve the intraocular structures. Common findings include a defective inferior lateral angle of the orbit, caudal displacement of the superolateral orbit, true and pseudococlobomas of the lids, lateral canthal dystopia, orbital lipodermoids, corneoscleral dermoids, and microphthalmos. Cataracts, lacrimal duct atresia, pupillary ectopia, distichiasis, and uveal colobomas have been reported less commonly. Interaocular involvement in these twins is rare in TCS. Furthermore, aniridia, corneal sclerisation, and uvea, lens, and retinal maldevelopment are previously unreported.

TCS is an autosomal dominant disorder affecting one in 50 000 live births. The disorder appears to have arisen in these twins with no relevant family history, as occurs in 60% of cases. Expressivity is highly variable, ranging from clinically undetectable to peri-natal death secondary to airway compromise; the disease severity that resulted in the fetal death of twin A is highly unusual. The responsible gene, TCOF1, has been mapped to 5q32–33.2 and the structure of its protein product, treacle, elucidated.

To date, 51 disease causing mutations have been identified, nearly all resulting in a premature termination codon. Significantly, no relation has been found between any single mutation and phenotype severity. The ophthalmic pathology observed in these twins may have resulted from a focal TCOF1 mutation and a yet to be defined role treacle may have in extraocular (and renal) development. Considering the number and severity of abnormalities, the failure to identify a single “genetic hot spot,” and the novel ophthalmic features, an alternative and perhaps more likely explanation is that a second gene, itself involved in ophthalmic embryology, was affected along with TCOF1. This unidentified gene may have been disrupted from a translocation involving 5q32–33.2 or from a deletion large enough to result in a contiguous gene syndrome. A final consideration is that these twins may not have had TCS but rather a new though closely related syndrome. Further genetic investigation may shed light upon these speculations.

Acknowledgements
JLP and GB contributed equally to this work.

References


Acute postural drop in optic nerve perfusion after vitrectomy and gas in a patient with diabetic autonomic neuropathy

We report the case of an insulin dependent diabetic who suffered transient blindness as a result of a change of posture following vitrectomy surgery with injection of gas. This 34 year old woman with longstanding diabetic disease, postural hypotension, and proliferative retinopathy had undergone argon laser posterior retinal photocoagulation (PRP) to both eyes. Her right eye was amblyopic and had visual acuity of counting fingers (CF), whereas the left eye, at best, had vision of 6/12 that was frequently impaired by recurrent vitreous haemorrhage. She underwent left vitrectomy and required 30% sulphur hexafluoride (SF₆) to gas support an incidental small, inferior detachment related to a round hole. The patient posted supine overnight and remained under the care of the physicians. The following day her vision dropped suddenly and severely on her way to the eye clinic. Visual acuity was no perception of light (NPL) in the operated eye and CF in the right eye. The left pupil was already dilated and anterior segment examination was satisfactory. The view of the fundus was limited owing to the presence of gas and residual haemorrhage but the retina was flat. The intraocular pressure (IOP) was 42 mm Hg and her blood pressure (BP) was 70/40. A decision to withdraw 0.5 ml of gas from the eye was made. While the patient was lying down for this procedure she immediately noted an improvement in vision to light perception. Her best corrected visual acuity improved in the supine position to 6/18. Following removal of gas the IOP fell to 20 mm Hg and the vision improved to counting fingers. Four weeks later, with disappearance of the gas her best corrected vision had improved to 6/18 and the retina remained flat.

Comment

Judging by the severity of the visual loss we believe that it was due to obstruction of both the central retinal artery and optic nerve perfusion. The precipitating factor in this case would appear to have been the change in posture from supine to sitting. The presence of gas in the operated eye had raised the intraocular pressure to a level such that the perfusion pressure to the eye was effectively eliminated resulting in a period of no light perception. Restoring the patient to the supine position was sufficient alone to reverse the condition and improve the perfusion of the eye. When the IOP was reduced to normal she was out of this critical situation and was able to maintain the ocular perfusion, even when sitting. Collapse of the central retinal artery is known to occur in conditions of high IOP but vision does not fluctuate with posture. However, it is known that raised IOP also reduces orthograde axial transport and compromises nerve head perfusion.  Postural hypotension in diabetes is secondary to autonomic neuropathy. The pathophysiology is not clear but it seems the main factor is blunted catecholamine response to standing and failure of the lower limb vascular resistance to increase adequately. Systemic hypotension can cause optic nerve blood flow as illustrated by numerous reports in the literature of non-arteritic anterior ischaemic optic neuropathy (NA-AION) occurring following acute blood loss. Patients with NA-AION have a significantly greater postural pressure change in IOP compared with healthy subjects as shown by James and Smith. In the same study they demonstrated a higher pulsatile ocular blood flow (POBF) on standing compared with supine. In this patient with autonomic neuropathy it is likely that the normal increase in POBF on standing was inadequate.

We advise particular caution in diabetics undergoing ocular surgery in which there may be a significant postoperative pressure rise. Similar problems can occur during surgery if hypotensive anaesthesia is either deliberately or inadvertently employed in diabetics. Beware of assuming poor postoperative vision to be purely the result of the presence of haemorrhage or gas; patients may have quite poor vision immediately after vitreoretinal surgery but will rarely have NPL unless there is also optic nerve compromise. We also suggest one should aim for a lower postoperative IOP in diabetics.

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Optical coherence tomography imaging of severe commotio retinae and associated macular hole

Commotio retinae results in retinal opacification following blunt trauma. Mild commotio retinae usually settles spontaneously with minimal sequelae but more severe cases are associated with visual loss. We are not aware of any previous reports describing optical coherence tomography (OCT) imaging of severe commotio retinae with an associated full thickness macular hole (FTMH).

Case report

A 13 year old boy presented 24 hours after blunt trauma from a football striking his right eye. On examination his best corrected visual acuity was counting fingers right eye and 6/6 left. Biomicroscopic examination revealed extensive commotio retinae over the posterior pole, no posterior vitreous detachment (PVD), and a FTMH. Colour photography and OCT imaging (OCT 2000 scanner, Zeiss-Humphrey) were performed (Fig 1). OCT confirms a FTMH and demonstrates extensive disruption of photoreceptor outer segments and retinal pigment epithelium (RPE).

Figure 1 (A) Right macula of 15 year old boy with extensive commotio retinae over posterior pole and an associated macular hole at 1 day after blunt injury. (B) Horizontal OCT scan through centre of macula confirms a full thickness macular hole and demonstrates extensive disruption of photoreceptor outer segment/retinal pigment epithelium layer. The optic disc is seen at the nasal edge of the scan.
He was treated conservatively with a short course of topical steroids. The colour fundus and OCT appearance at 1 month are shown in Figure 2. Despite spontaneous macular hole closure, visual acuity remained at counting fingers at 1 year follow up.

Comment
The major site of retinal trauma appeared on OCT to be at the level of the photoreceptor outer segment/RPE interface. The OCT images are consistent with fragmentation of photoreceptor outer segments and damaged cell bodies, as suggested by Sipperley et al in their study of the histological changes in commotio retinae in primate.

The exact pathogenesis of macular holes remains uncertain. Ho et al outlined the three basic historical theories regarding aetiology—traumatic theory, the cystic degeneration and vascular theory, and the vitreous theory. Of these, the latter has gathered the most support in the context of idiopathic macular holes.

In our case, the OCT imaging reveals that the edges of the macular hole are elliptical and irregular with no associated PVD, cortical vitreous condensation, or overlying prefoveal opacity. The characteristics suggest a different mechanism of hole formation from that proposed in idiopathic senile macular holes. We believe that mechanical distortion of the retina, relative to the vitreous and underlying sclera, created disruption of the photoreceptor outer segment level that the retina has the least support from Müller cells and is therefore likely to undergo greatest deformation.

In the only previous report of OCT imaging in traumatic macular hole, a case with mild commotio retinae was described in which the extensive outer retinal disruption was not observed. There have been some encouraging reports suggesting that vitrectomy can successfully close traumatic macular holes as well as improve visual function in many cases. However, it seems unlikely that cases with severe commotio retinae, and associated photoreceptor/RPE damage, as demonstrated in our cases, would gain any benefit from surgical as opposed to spontaneous closure of a traumatic FTMH. The final visual prognosis is severely limited by the extent of initial photoreceptor damage, and the excessive pigment atrophy and clumping that follows.

We believe OCT imaging provides additional information both on the pathogenesis of commotio retinae and in the assessment of outer retina disruption following ocular trauma. This information may help in the selection of patients likely to benefit from surgical intervention.

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References

Acute ocular ischaemia and orbital inflammation associated with systemic lupus erythematosus

We report a patient with systemic lupus erythematosus (SLE) who developed bilateral ocular ischaemic syndrome in association with orbital inflammation leading to devastating visual loss.

Case report
A 73 year old white woman presented with unilateral acute anterior uveitis and polyarthropathy of the hands, knees, and neck. Over the next 4 years she suffered recurrent anterior uveitis, marginal keratitis, and episcleritis, which were treated with topical steroids. She had bilateral age related cataract and underwent left cataract surgery. Postoperatively, her visual acuities were 6/60 right eye and 6/9 left eye.

She was admitted 7 months later with extensive erythematous, scaly plaques on sun exposed areas and a purpuric eruption over the abdomen, buttocks, and legs. Biopsy of a plaque showed changes consistent with SLE including follicular keratin plugging, epidermal atrophy, degeneration of the basal layer with colloid body formation, basement membrane thickening, myxoid change in the dermis, and periappendageal lymphocyte infiltrates. Biopsy of the purpuric eruption was consistent with a leucocytoclastic vasculitis. Further investigations revealed positive rheumatoid factor (1/100), ANA (1/25), pANCA (1/100), and anti-EN antibodies (1/100). There was reduced C3 and C4, polyclonal increase in IgA and IgM, lymphopenia, thrombocytopenia, and reduced creatinine clearance but no proteinuria. Anticardiolipin antibodies were negative. A diagnosis of SLE was made and oral prednisolone (0.8 mg/kg) was commenced.

Three weeks later she developed a rapid onset of marked bilateral conjunctival and lid oedema, subconjunctival haemorrhage, and propositis. Visual acuity deteriorated to perception of light in each eye. There was gross restriction of ocular movements. Signs of ocular ischaemia developed in both eyes with corneal oedema, iris neovascularisation, fibrovascular anterior uveitis, hyphaema, and raised intraocular pressure (Fig 1). Computed tomography and B-scan ultrasonography of the orbits showed posterior scleritis, choroidal detachment, vitreous haemorrhage, and thickening of the extraocular muscles (Fig 2).

She was treated with topical dexamethasone 0.1% and carteolol 1%, oral acetazolamide 500 mg daily, intravenous cyclophosphamide (10 mg/kg per day), and subsequently intravenous immunoglobulin (0.5 g/kg/day for 5 days). Over the following month there was regression of the propositis, conjunctival and lid oedema, corneal oedema, and fibrinuous uveitis. The intraocular pressure was controlled and the visual acuity remained perception of light in each eye. Subsequently, the patient developed pneumonia, VIIth nerve palsy, left sided hemiparesis, and died 7 weeks after admission. Postmortem examination revealed bronchopneumonia, lung abscesses, pleural effusions, fibrinous pericarditis, cardio-megaly, and jejunal mucosal haemorrhages.

Figure 1 Anterior segment of right eye showing conjunctival injection, corneal oedema, hyphaema, ruberosis iridis, and cataract

Figure 2 B-scan ultrasound of left eye showing marked posterior scleritis (crosses) and choroidal detachment (arrow).
The family did not consent to histopathological examination of the eye.

Comment

Acute orbital inflammation and orbital myositis leading to proptosis are rare manifestations of SLE. To our knowledge acute ocular ischaemic syndrome with orbital inflammation is a novel complication of SLE. The ophthalmic manifestations may have resulted from a vasculitis involving the vessels supplying the globe and extracocular muscles. The limitation of ocular movements was most probably the result of mechanical restriction. Although the proptosis, lid and conjunctival oedema was improved with immunosuppression, the visual acuity did not recover. This may have been because of irreversible visual loss from optic nerve dysfunction due to ischaemia, compression from the acute orbital inflammation, or secondary to raised intraocular pressure. Close cooperation between ophthalmologist and rheumatologist is essential in the management of these patients, in order to try and prevent blindness.

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Referen
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Chickenpox neuroretinitis in a 9 year old child

Chickenpox in children is usually thought of as a benign infectious disease with few ocular complications. Posterior segment involvement from primary varicella zoster infection has rarely been reported in children. We describe the clinical features and visual outcome of an unusual case of neuroretinitis presenting in a 9 year old child.

Case report

An immunocompetent 9 year old boy acquired primary varicella zoster virus (VZV) infection from his sibling and developed the characteristic exanthematous vesicular rash. Four days after the onset of the rash he woke with discomfort in his right eye and described his vision as being “all grey” on that side. He presented to the emergency department the same day and was found to have a visual acuity of 3/6 on the right and 3/3 on the left (logMAR). A relative afferent pupillary defect (RAPD) was present on the right. His anterior segment was quiet with no vitritis; however, he had slight macular thickening and a subtle cherry red spot on funduscopy, along with some mild peripapillary swelling and disc haemorrhage.

On review in the ophthalmology clinic 2 days later his vision had reduced to 1/60 (Sheridan Gardiner singles) on the right. He had no new skin lesions and all those present had crusted. No lid lesions were present. He had a marked RAPD, red desaturation, and mild conjunctival injection. His anterior segment and vitreous remained clear. The right disc was hyperaemic with peripapillary swelling and haemorrhage. The macular area was pale and oedematous (Fig 1). Examination of the left eye was completely normal.

Considering the onset of ocular symptoms and signs following the appearance of the typical VZV skin lesions, a presumptive diagnosis of chickenpox neuroretinitis was made. He was admitted and commenced on intravenous aciclovir (250 mg × 3 per day). Confirmatory IgM titres for VZV were unfortunately not performed. No change in his acuity was observed over the next few days; however, his right disc was noted to become slightly pale after 2 days of treatment. At this point intravenous methyl prednisolone was instituted at a dose of 5 mg/kg per day. Despite a gradual resolution of the macular and peripapillary oedema over the next 5 days, his disc remained pale (Fig 2) and his acuity measured as 3/30 (logMAR) after 7 days of intravenous aciclovir and 5 days of methyl prednisolone. Systemically he remained completely well and afebrile on treatment. He was discharged with a further 3 day course of oral aciclovir and a 6 day reducing course of oral prednisolone.

Over 5 months of follow up his acuity has not improved beyond 3/30 (logMAR). The right optic disc is pale and a yellow lipid deposit is present at the macula with some reticular macular pigmentary. The left eye has been normal throughout.

Comment

Posterior segment involvement as part of primary VZV infection in children has only been reported twice to our knowledge. Copenhaver et al reported a 3 year old with bilateral papillitis and a unilateral macular lesion associated with encephalitis following varicella infection. This resulted in complete recovery of vision and resolution of the macular lesion within 3 weeks of presentation. Capone and Meredith describe a case of unilateral central visual loss in a 2 year old child caused by chickenpox retinitis. Epileptic seizures, optic neuritis resulting in a poor visual outcome. Their patient presented with an acute exotropia 24–48 hours before the onset of cutaneous VZV. Funduscopic revealed papillitis, phlebitis, and a mild disc oedema. Following hospitalisation he made a complete recovery.

Our case is particularly interesting, not only because these are the first published fundal photographs of VZV neuroretinitis in a child, but also because of the relatively mild ocular findings which have resulted in severe visual loss. The young age of the patient is atypical of ocular VZV infection. Adults who contract primary VZV infection tend to run a more severe course than children. Ocular complications in children are extremely rare.

The typical posterior segment involvement of VZV is acute retinal necrosis (ARN). The youngest case of ARN in association with chickenpox has been reported in a 4 year old. In adults, ARN is described as being less severe when presenting at the time of primary zoster infection than as a result of secondary reactivation of latent, previously acquired VZV. The changes typical of ARN were absent in this case. Unilateral papillitis and retinitis confined to the macular area were the main features. Optic neuritis has been reported by several authors in association with primary VZV. Many of these cases are bilateral and coincident with encephalitis or occurring in those who are immunocompromised. Unilateral optic neuritis has been described in an 18 year old several weeks following a varicella rash which remitted without sequelae following the administration of corticosteroid.

The mainstay of treatment of VZV retinitis is with intravenous aciclovir. Whether any advantage is gained in administering systemic steroid with the aciclovir is controversial. We do not know if a more positive visual outcome may have been achieved if intravenous therapy had been commenced on presentation. It is therefore suggested that prompt treatment of VZV retinitis with intravenous aciclovir be started in patients, particularly in a child, presenting with any posterior segment signs.

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Bilateral central retinal artery occlusion in Wegener's granulomatosis and α1 antitrypsin deficiency

Visual loss occurs in up to 8% of patients with Wegener's granulomatosis (WG) during the disease lifetime, although a small proportion of these cases are the result of central retinal artery occlusion (CRAO). We describe an unusual case of a patient with bilateral CRAO as a presenting feature of WG and α1 antitrypsin (AAT) deficiency.

Case report
A 58 year old white man presented with sudden painless loss of vision to the right eye. Vision was hand movements in the right eye and 6/6 in the left. Funduscopy revealed an acute right CRAO with macular oedema. There were no signs of uveitis or retinal vasculitis. Management consisted of intravenous acetazolamide (500 mg), ocular massage, and anterior chamber paracentesis. He was subsequently sent home with aspirin treatment, and referred to his family doctor for routine risk factors assessment.

The following morning, he returned to the eye casualty department with a left CRAO, which was treated in the same way. Vision was 6/60 in the right eye and hand movements in the left. Systemic inquiry revealed a 2 month history of general malaise, arthralgia, and myalgia. General examination revealed evidence of vasculitic rash (Fig 1) affecting the right elbow and nailfold infarcts (Fig 2). He was admitted for further investigation. His erythrocyte sedimentation rate in the first hour was 128 mm and C reactive protein (171 mg/l) were markedly raised. Liver enzymes were slightly abnormal (γGT, 165 IU/l, ALT, 85, alkaline phosphatase, 153). There were traces of blood and protein in the urine, with a normal creatinine clearance (86 mmol/l). He was commenced on oral prednisolone (60 mg/day) for presumed systemic vasculitis. Normal echocardiography and blood cultures excluded endocarditis. Ultrasound of his carotid arteries was normal. His vision gradually improved, and was recorded 6 days later as 6/12 in the right eye and 6/6 in the left. Patchy peripheral field defects remained.

Subsequently, his serum was positive to cryoplastic antineutrophil cytoplasmic antibody (cANCA) at a titre of 1:160, confirming the diagnosis of WG. Monthly pulses of intravenous cyclophosphamide were employed, with gradual tapering of oral prednisolone dose. There was also an incidental finding of homozygous AAT deficiency (Z allele) from serum protein electrophoresis.

Comment
WG is a rare disease with a reported annual incidence of 8.5 per million population in the UK, of which up to 16% present initially with ocular disease. CRAO in patients with WG is rare, with only several reported cases since 1960. Ocular and adnexal involvement is the result of parenchymal necrosis, small vessel vasculitis, and granulomatous inflammation. The orbit may be the site of primary inflammation and become secondarily involved from disease of the paranasal sinuses and nasopharynx. Mechanisms by which visual loss occurs in WG include vascular occlusion, macular oedema, inflammatory destruction of retina, optic nerve, or corneoscleral tissue.

Systemically, the most commonly affected tissues are the lungs and kidneys. The current therapy for WG is cyclophosphamide (daily or pulse) and corticosteroids. Multicentre trials are in progress to assess the optimum treatment duration for induction, and subsequent maintenance of remission.

This is the first reported case of bilateral CRAO in WG and AAT deficiency. AAT is normally present in serum and inhibits uncontrolled enzymatic destruction of connective tissue during inflammation. Deficiency of AAT results in progressive emphysema and liver injury. Research suggests an increased incidence of WG in patients with AAT deficiency. The incidence of AAT deficiency has been reported as being significantly increased in patients with anterior uveitis. We are not aware of any reports implicating AAT deficiency in CRAO, although it is possible that this is contributing to the ongoing inflammatory process underlying his vasculitis.

Complication of acneprucin in a patient with Behcet’s disease

We would like to bring to your attention a complication resulting from acneprucin in a patient with Behcet’s disease.

Case report
A 35 year old white man, who fulfilled the International Study Group criteria for Behçet’s disease, attended the Behçet’s clinic at the Birmingham and Midland Eye Centre. He complained of four red areas on his left arm (Fig 1). Two days earlier he had undergone acupuncture for what he described as “tennis elbow.” These red areas corresponded to where the acupuncture needles had been inserted. Examination revealed these areas to be pustules (inset) that were characteristic of Behçet’s disease, although only a small proportion of these cases are the result of central retinal artery occlusion (CRAO). We describe an unusual case of a patient with bilateral CRAO as a presenting feature of WG and α1 antitrypsin (AAT) deficiency.

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References

Figure 1 Photograph of right elbow showing vasculitic rash.

Figure 2 Retrorillumination photograph of right and left index fingers showing nailfold infarcts.
A positive pathergy test is an important diagnostic sign of Behçet’s disease. Its prevalence varies by geographic region, being less common in patients from Northern Europe. Nevertheless, patients with Behçet’s disease should be made aware of this potential complication if they intend to undergo acupuncture.

**Comment**

Fibrous histiocytomas of the corneoscleral limbus are rare tumours. We present a case report and a review of the clinical and histopathological findings from cases presented in the literature.

**Case report**

A 19 year old white male presented with a 6 month history of a painless growth on the inferior corneoscleral limbus of the left eye (Fig 1A and B). There were no other ocular symptoms. He had no medical history of note. His vision was 6/5 unaided in the right eye, and 6/6 unaided in the left eye. The growth was approximately 5 mm in diameter, vascularised, and yellowish in appearance. There were no other ocular abnormalities. The lesion was excised by conjunctival excision and silicone puncture.

Histopathological examination of the lesion showed a stromal hypercellular nodule consisting of spindle cells, small lymphocytes, multinucleated giant cells, and histiocytes. Some histiocytes had foamy cytoplasm. Many capillary sized blood vessels were present in areas, the spindle cells were arranged in a storiform pattern. A fine collagenous meshwork extended throughout the lesion. Immunohistochemistry confirmed the cellular composition of the lesion (Fig 1G, H) as rich in inflammatory cells, including foam cells and siderophages. No atypical nuclei or mitotic figures are present. Although some authors regard these tumours as reactive proliferations of fibroblasts, others do not accept this view because the lesions tend not to regress spontaneously. Recurrence is rare, with less than 5% of cutaneous benign fibrous histiocytomas recurring after local excision.

In contrast, malignant fibrous histiocytomas of the corneoscleral limbus characteristically appear in later life, between the ages of 50–70 years, with an equal distribution of males to females. They are highly aggressive tumours, and have been reported to have a local recurrence rate of 100% if a limited excision is performed. Recurrence can occur within a few months of excision. There are seven reported cases of corneoscleral malignant fibrous histiocytoma. Two of the cases had an enucleation and two cases underwent orbital exenteration.

**Figure 1** Fibrous histiocytoma arising from the corneoscleral limbus, showing the tumour extending into the cornea. It is moderately vascular and has a similar appearance to a dermoid. (B) Fibrous histiocytoma showing the inferior margin of the tumour. (C) Haematoxylin and eosin (H&E) stain, original magnification x20. (D) H&E stain, original magnification x100. (E) H&E stain, original magnification x400. (F) H&E stain, original magnification x400. (G) Immunoperoxidase CD68. Original magnification x200. (H) Immunoperoxidase CD45. Original magnification x200.
Follow up of these patients ranged from 18 months to 5 years and all were free from recurrence. The other three patients had a local excision. One patient developed local scleral recurrence 2 months later. (It is not evident from the literature if the borders of excision were clear of tumour.) He was found to have a metastasis in the liver 4 months later and after a parotidectomy and radical neck dissection, the patient developed multiple pulmonary metastases and died within 1 year. One of the patients was lost to follow up. The third patient who had a local excision went on to develop two recurrences, which subsequently needed excision and cryotherapy; however, after 1 year he was free of any recurrence. Malignant fibrous histiocytomas have a broad range of histological appearances; storiform-platelike, myxoid, giant cell, and inflammatory. The storiform-platelike type is the most common. The cells are predominantly plump pleomorphic spindle-shaped with occasional large, ovoid histioyte-like cells. Modest amounts of inflammatory cells, such as lymphocytes and plasma cells may be present. The differential diagnosis of a malignant fibros histiocytoma includes pleomorphic carcinoma, malignant melanoma, and other sarcomas.

For those limbal fibrous histiocytomas with a benign histopathological appearance, the management should be local surgical excision. Malignant fibrous histiocytomas need to be managed cautiously, preferably by wide local excision and cryotherapy at the earliest opportunity. If necessary, enucleation should be considered to fully excise a limbal malignant fibrous histiocytoma.

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References

Haemophilus influenzae corneal ulcer associated with atopic keratoconjunctivitis and herpes simplex keratitis

Haemophilus influenzae is a rare cause of corneal ulceration, usually associated with previous corneal damage. To ensure appropriate treatment, the organism must first be identified by cultures in enriched media. We describe a case of keratitis caused by H influenzae associated with two risk factors—herpetic keratitis and atopic keratoconjunctivitis.

Case report
A 43 year old Hispanic male presented with 1 week of redness, blurred vision, pain, burning, and itching in the left eye. Past ocular history was remarkable for bilateral epithelial and interstitial herpetic keratitis and atopic keratoconjunctivitis with a previous shield ulcer in the left cornea. The patient was not using any medications at the onset of the current episode. He had a history of anhidrotic ectodermal dysplasia and atopic dermatitis since birth. On examination, uncorrected visual acuity was 20/100 in the left eye. The skin of the eyelids was wrinkled, thickened, and hyperpigmented bilaterally with madarosis of the lashes. The conjunctiva had a mild papillary reaction on the right and a severe reaction on the left, with giant papillae in the superior tarsal area. In the left eye, corneal sensation was markedly decreased and a central corneal epithelial defect of 1.0 × 0.5 mm was present with an underlying anterior stromal infiltrate measuring 2.5 × 1.5 mm (Fig 1). Scant keratic precipitates and a small hypopyon were also present.

Scrapings of the corneal ulcer were taken and inoculated on blood and chocolate agar as well as in thiglycollate broth. Viral cultures and polymerase chain reaction (PCR) were also performed to rule out the possibility of a recurrent herpes simplex infection. They were subsequently negative. The patient was started on oloxicin and fortified cefazolin (50 mg/ml) drops every 1 hour. He was also started on oral aciclovir 800 mg five times a day. On the fourth day, corneal cultures were positive with a heavy growth of H influenzae sensitive to oloxicin (Fig 2). On the sixth day of treatment, uncorrected visual acuity improved to 20/30, the stromal infiltrate decreased to 1 × 1 mm, with a small epithelial defect, and there was no hypopyon. A suprastellar injection of dexamethasone was given and topical prednisolone acetate 1% three times a day was added. The papillae regressed and the corneal lesion healed completely, leaving a central scar with a deep stromal vessel.

Figure 1 Haemophilus influenzae corneal ulcer.

Comment
H influenzae is a tiny Gram negative cocccobacillus that is an uncommon cause of corneal ulceration. In most series it accounts for less than 3% of all corneal ulcers. It is a common cause of acute bacterial conjunctivitis, especially in children. Unlike Staphylococcus aureus, Streptococcus pneumoniae, and other bacterial causes of conjunctivitis, H influenzae seldom produces corneal ulceration. This is in marked contrast with H egyptus conjunctivitis where peripheral ulcers, infiltrates, and phtyctenules are commonly seen as complications of conjunctivitis. For H influenzae to infect the cornea, there must be an epithelial defect. Most of the reported cases of H influenzae keratitis have been associated with specific risk factors such as contact lens wear, application of cyanoacrylate glue, or systemic debilitating diseases. In this case, the patient had two significant risk factors—severe atopic keratoconjunctivitis and herpetic keratitis. Despite the negative results of the viral cultures and HSV-DNA PCR, we cannot rule out the possibility that a viral aetiology played a part in this case. Also, it seems likely that complications from the patient’s atopic disease delayed the healing of the ulcer.

H influenzae is a fastidious organism that needs media enriched with nicotinamide adenine dinucleotide (NAD), such as chocolate agar for growth. It will not grow in blood agar, unless there are also colonies of Staphylococcus aureus, which provide NAD. In that situation, H influenzae will then grow as satellite colonies around the Staphylococcus aureus. This case illustrates the importance of using chocolate agar as well as blood agar to make an aetiological diagnosis.

In conclusion, H influenzae is a rare cause of corneal ulceration, which can occur in patients with previous corneal damage from...
Blinking and operating: cognition versus vision

The difference in the refractive indices between the air and the tear film results in the tear film having the greatest optical power of any part of the eye. Eyelid blinks are important for maintenance of the tear film. Between blinks there is progressive thinning of the tear film, which becomes non-uniform on the ocular surface and may break up. This produces an irregular air-tear interface, with a reduction in image quality. The longer the period between blinks the greater the effect on the tear film and reduction in vision. Restoration of the tear film occurs immediately following a blink so that the ability to maintain a regular tear film is dependent on the blink rate. A reduction in the blink rate such as, for example, a pause between blinks of 15 seconds, has been associated with a change in the shape of the profile of the corneal tear film and up to a 6% reduction in visual acuity. More importantly, however, a reduction in blink rate leads to a reduction in contrast visual acuity.

The ability to distinguish between different layers during surgery, such as the posterior capsule of the lens and anterior vitreous face, or peeling a layer of the retina is difficult in vitreoretinal surgery, is dependent on the surgeon having and maintaining good contrast visual acuity. Blink rates and blink amplitude vary according to vision related behaviour. A reduction in the blink rate occurs with tasks of increasing visual difficulty. For example, visual tasks requiring concentration, such as video display terminal use, result in a decrease in average blink rate from 18.43 mm to 3.6 blinks/min.

In order to determine whether the blink rate of ophthalmic surgeons alters during intraocular surgery, the blink rate patterns of nine ophthalmic surgeons were recorded. Two observers recorded the blink rate during casual conversation and when the surgeons were using the operating microscope. None of the surgeons were aware that their blink rates were being recorded, which was done by two medical students during their ophthalmic attachment—that is, their presence in the theatre was accepted as part of their ophthalmic training. The blink rate for each surgeon was recorded in each condition between four and 10 times. The mean blink rate for each surgeon during casual conversation and while operating are presented in Table 1. There was a significant reduction in the average blink rate between both conditions (16.69/min and 4.75/min, p = 0.0001 [t test]), on average a third and a half fold decrease occurred while operating. It was also noted that the onset of conversations such as the request for an instrument or demonstration of an intraocular structure was associated with the onset of a blink response.

The reduction in blink rates observed in this study—that is, 16.69 to 4.75, are similar to that found by Patel et al for visual tasks such as video display terminal use. A reduction in blink rate to 4.75 translates to a reduction in contrast acuity of approximately 10% with four surgeons having an expected reduction of more than 60%. Such a reduction in contrast acuity is likely to have an effect on the ability to differentiate between different intraocular surgical layers. Blinking occurs between visual fixations and may be timed so as not to interfere with significant visual input. Blinking is low when information memory is operating, and cognitive processes might also create an abnormal tear film. Patients with a longer blink rate may have an increase in their blink rate. It is likely that video display unit screens be kept below eye level. During ophthalmic surgery the eye pieces are roughly perpendicular to the face, so that it might be reasonable to lower the height of the eye pieces to reduce the width of the palpebral aperture. Frequent instilation of artificial tears during surgery is not practicable and might also create an abnormal tear film. Blinking might also be suspended during parts of the operation or investigation where good contrast acuity is not essential and where disruption of the cognitive processes is likely to have a minimal effect.

| Table 1 Average blink rates [No/min] for ophthalmic surgeons during periods of casual conversation and while operating using the microscope |
|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|
| Surgeon         | 1               | 2               | 3               | 4               | 5               | 6               | 7               | 8               | 9               | Mean (SD)       |
| Casual          | 17.29           | 17.75           | 7.75            | 17.60           | 27.44           | 24.67           | 11.50           | 13.86           | 12.33           | 16.69           |
| Operating       | 9.71            | 9.00            | 0.29            | 7.40            | 1.68            | 8.67            | 1.59            | 3.86            | 0.54            | 4.75            |
| p Value         | 0.000002        | 0.0002          | 0.0002          | 0.000001        | 0.0001          | 0.0003          | 0.0004          | 0.0002          | 0.0003          |                 |

References


Blinking and operating: cognition versus vision

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Should we vaccinate for glaucoma surgery? Trabeculectomy is the most common non-laser surgical procedure performed for treatment of all forms of glaucoma. It involves the fashioning of a fistula from the anterior chamber of the eye to the subconjunctival space. This allows for extra drainage of aqueous humour to the subconjunctival space. This produces a localised elevation of the conjunctiva in the area of the trabeculectomy called a ‘filtering bleb’. Antimetabolites may be used intraoperatively and perioperatively to increase the success of glaucoma filtering surgery by their action on wound healing. 5-Fluorouracil or mitomycin C is administered to the scleral flap during the procedure. Post-operatively; subconjunctival injections of
5-fluorouracil may be given. This is known as an augmented trabeculectomy.

Infective endophthalmitis is a recognised complication of glaucoma filtering surgery. It may occur in the early postoperative period or it may happen years after surgery. Another eye may be involved by a precursor to endophthalmitis has been described as blebitis. Blebitis is an infection of the trabeculectomy bleb without vitreous involvement.

Clinical features of blebitis include pain, photophobia, conjunctival discharge, and severe conjunctival injection centred on an opeansal filtering bleb. A Siedel test may be positive (this indicates aqueous leakage from the bleb). There may be an anterior chamber reaction. There is no vitritis.

As many as one per 100 patients/year may develop infection of the bleb. Factors associated with increased risk of bleb related endophthalmitis include increased axial length, thin leaky bleb, conjunctivitis, upper respiratory tract infection, hibernar occurrence, and vitreous wicks. With the increase of antimitabolites in glaucoma surgery, the incidence of the thin walled cystic blebs seems to be increasing. These blebs are more prone to leakage. Some studies concluded that the incidence of bleb related endophthalmitis is higher when antimitabolites are used. This is more common with inferior limbal trabeculectomy. However, some studies show equal incidence in augmented trabeculectomy and trabeculectomy without antimitabolite augmentation.

There are few data available for the incidence of blebitis. In most reported cases conjunctival swabs were performed for culturing and sensitivity, but organisms causing endophthalmitis may only be present transiently on the ocular surface. In many reports, ocular surface cultures came back positive for Staphylococcus epidermidis and Staphylococcus aureus, which may both be found on healthy normal eyes.

The microbiology of bleb associated endophthalmitis is different from other causes of endophthalmitis. Clinicians should not extrapolate the results of the Endophthalmitis Vitrectomy study to the post-filteration surgery endophthalmitis that give the differing pathogenesis and unique spectrum of organisms. The most common organisms are Streptococci species. The second most common is Haemophillus influenza type b at over 23%. Of the streptococci, Staphylococcus aureus may account for approximately 12%. Between them H influenza type b and S pneumoniae probably cause more than 35% of blebitis and bleb associated endophthalmitis. The treatment of endophthalmitis is expensive. It usually involves admission of the patient and frequent use of expensive drops as well as surgical intervention. Inpatient treatment for blebitis has been priced at US$892 (approximately £540) per 24 hours. This can work out to more than £3000 for a 10 day stay in hospital. Furthermore, the cost of follow up visits and the morbidity that is involved need to be taken into account. Frequently these patients have pre-existing visual compromise and an episode of endophthalmitis may result in a rapidly acquired or blind registration, an individual disaster with wider social implications.

H influenza type b vaccine is licensed for use in healthy children. It is not licensed for use in those patients considered to be at risk for invasive H influenza type b disease such as sickle cell disease and those receiving treatment for malignancy. After the age of 13 months the vaccine is effective after a single dose. This vaccine has already shown benefit in ophtalmology by the dramatic decrease in the incidence of orbital cellulitis in immunised children. It consists of a capsular polysaccharide of H influenza type b conjugated to a protein carrier. Side effects of the vaccine include fever, headache, malaise, irritability, loss of appetite, vomiting, diarrhea, rash, urticaria, convulsions, erythema multiforme, and transient cyanosis of the lower limbs. Its cost to the NHS is as low as £8.83 for a single dose of 0.5 ml.

Pneumococcal vaccine is available. It is a polyvalent pneumococcal polysaccharide from each of 23 capsular types of S pneumoniae. The vaccine is recommended from the age of 2 for people with the following conditions: homozygous sickle cell disease, asplenia or dysfunc tion of the spleen, chronic renal disease, nephrotic syndrome, immunodeficiency, immuno suppression, chronic heart disease, chronic lung disease, chronic liver disease, and diabetes mellitus. It is effective after a single dose if the strains of S pneumoniae prevalent in the community are reflected in the poly saccharides contained in the vaccine. Its cost to the NHS is as low as £0.51 ml.

There have been no reports of epidemics of blebitis. If it was contagious, there would have been epidemics or clustering in our glaucoma clinics. We can find no evidence of case to case transmission. In fact, all reported cases and series appear sporadic. The association with upper respiratory tract infections and hibernar occurrence are strongly suggestive of respiratory infection with consequent spread to the predisposed eye. We are uncertain whether this is systemic or droplet spread. However, the need for topical antibody protection is negated by the presence of systemic antibodies against the specific bacteria. We believe that by minimising the possibility of systemic infections with these agents we diminish the likelihood of blebitis.

It is possible that these vaccines could be given to patients who are destined to have trabeculectomy. The cost for both vaccines would be less than £20.00.

Two hundred and fifty vaccinations could be paid for by the price of a single episode of bleb associated endophthalmitis. Assuming a long term infection rate of 2%, these vaccines could possibly prevent two cases of bleb associated endophthalmitis, representing a saving of £5000 to the NHS.

Apart from the cost, vaccination has the potential to prevent significant ocular morbidity. At the very least, these vaccines should be considered in high risk patients undergoing augmented trabeculectomy. We plan to conduct a prospective study of the effect of these vaccinations upon the incidence of blebitis and bleb related endophthalmitis.

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The wide field multifocal electroretinogram reveals retinal dysfunction in early retinitis pigmentosa

The diagnosis of retinitis pigmentosa (RP) is based on the presence of characteristic retinal pigment changes, visual field defects, and reduction in amplitude or loss of rod scotopic standard electroretinogram (ERG) responses, with a possible history of night blindness and a positive family history of RP. Multifocal electrophysiolog (mfERG), is a new technique that constructs a topograph ic map reflecting retinal function. Reports have suggested that the spatial resolution of mfERG is sufficient to detect focal changes in retinal function as RP progresses.

We describe a case of early RP in which the amplitude and implicit times of the patients’ standard ERG rod and maximal responses were normal in the right eye and equivocal in the left eye. However, the peripheral retinal mfERG amplitude and implicit times were reduced and delayed. These abnormalities were obtained using a custom built wide field mfERG, which facilitates assessment of a 90 degree retinal field.

Case report
A 29 year old woman was referred to the eye clinic by her optometrist. Abnormal retinal pigmentation was found in both fundi on routine examination. She had no visual problems and was otherwise systemically well. Her 51 year old mother is known to have RP. On examination, she had 0.50 dioptres of hypermetropia in both eyes. Her corrected logMAR visual acuities were 0.025 in the left eye and 0.025 in the right eye. Her colour vision, anterior segment examination, and intraocular pressures were normal. Fundus examination revealed semicircular arcs of intraretinal “bone spicule” pigmentation in the inferior mid-periphery of each retina. Her optic discs appeared normal and there was no evidence of attenuation of the retinal vasculature.
A Humphrey 120 point threshold related perimetry test was performed and the patient maintained fixation throughout the test. There was an arc of absolute visual field defect in the superior field, symmetrical in both eyes, 40 degrees from fixation, which was more extensive superotemporally than supranasally.

A Ganzfeld ERG test was performed in accordance with international standards. The implicit times and amplitude of the scotopic rod, photopic cone and flicker responses of the patient were normal. The scotopic maximal b-wave amplitude was reduced by 14% in the left eye and was normal in the right eye (Fig. 1).

A wide field mfERG was performed, using a technique previously described. The amplitudes of the central and peripheral mfERG responses were grouped and averaged (Fig 2A) and compared with similar responses from normative data (Fig 2B). The average amplitudes of the central mfERG response was 75 nV in the right and 101 nV in the left (normal range 74–122 nV) (Fig 2A). The average peripheral retinal mfERG responses were 29 nV in the right eye and 45 nV in the left eye (normal range 61–108 nV). The normal range is derived from a group of 40 controls, aged 20–40 years. In addition, the mfERG responses were reduced in areas that had normal visual field sensitivities.

**Comment**

Retinitis pigmentosa in its early stages of evolution is characterised by rod dominated photoreceptor dysfunction. Although mfERG is a photopic response, thought to predominantly reflect cone function, the nature of mfERG stimulation (that is, stimulation frequencies from 5 Hz to 75 Hz), indicates that this composite response may contain contributions from rods, in addition to cones and post-receptor cells.

The global nature of the Ganzfeld ERG requires approximately 30% of the retina to be dysfunctional before abnormalities can be detected. In this case report, the standard ERG did not help to confirm the diagnosis of RP. However, the spatial resolution of the peripheral wide field mfERG indicated peripheral retinal dysfunction, suggestive of RP. We conclude that wide field mfERG may have advantages over Ganzfeld ERG in the electrophysiological diagnosis of some forms of early RP. Further evaluation of wide field mfERG in the diagnosis and follow up of early RP is indicated.

**Day 1 review following cataract surgery: are we seeing the precise details?**

The Royal College of Ophthalmologists published cataract surgery guidelines in February 2001. This document includes protocols relating to postoperative visits suggesting that there are no additional risks to patients who are not reviewed on the first postoperative day. This is a change in recommendation from previous college guidelines in 1995 suggesting a review within 48 hours.

There may follow a growing impetus for ophthalmologists to dispense with the first day review, given the reduced demand on clinician time and the corresponding accrual of staffing and financial resource benefits. While we applaud the dissemination of practice guidelines, they constitute “merely tools, not precise details” to aid clinical decision making. They may have inherent limitations in particular circumstances and may require evaluation for effective application in clinical settings.

Four studies were quoted by the guideline authors, three of which advocated the omission of day after review and one of which was equivocal, suggesting that it was unsafe to abandon this practice unless raised intraocular pressures (IOP) were controlled. The numbers of patients included ranged from 100 to 387. The results of these studies are shown in Table 1.

**Comment**

In our view, deriving meaningful conclusions that may underpin clinical practice are difficult, owing to the varying methodological approaches.
approaches used in these studies. In Tufail’s study,1 extracapsular cataract extraction was the predominant surgical technique used. Cohen et al2 excluded more than 50% of patients with complicated ocular histories or complicated surgery and Whitefield et al3 had similar extensive exclusion criteria, although the number excluded was not mentioned.

We would draw attention to a recently published study by McKellar and Elder,4 which to our knowledge is one of the largest cohort studies, aside from national cataract surveys, reporting on first and seventh day complications of cataract surgery. Of 1000 patients, the study found that on the first postoperative day, complications were observed in 10% of eyes, of which 88% was raised IOP. Unlike most of the previous studies, all patients with available records were eligible, including those with preoperative risk factors and those with surgical complications. These figures align more closely with our “gold standard” of the National Cataract Surgery Survey than the previous mentioned studies. The events most frequently occurring within 48 hours after surgery in the national survey were corneal oedema (9.5%), raised IOP (7.9%), and uveitis (5.6%). Overall, 23.3% of patients had early postoperative complications ranging from minor to sight threatening conditions. The survey also found that several risk indicators were associated with poorer visual outcomes and complications related to cataract surgery: age, ocular co-morbidity (glaucoma, diabetic retinopathy, amblyopia, and previous ocular surgery), diabetes mellitus, stroke, type of surgical procedure, and grade of surgeon.

In summary, up to 20 000 patients a year in the United Kingdom (10%) may have an untreated early postoperative complication such as corneal oedema or raised IOP if first day review was abandoned. And if McKellar’s study is representative, then 5% of patients would have raised pressure without any previous history or surgical complication and 0.9% of patients could have other potentially serious early complications. Nationally, that equates to almost 12 000 patients annually. It is worth noting that the American Academy of Ophthalmology in its white paper,5 concludes that there are enough significant early postoperative complications to warrant first day review. Are we sufficiently confident in our own practices to diverge?

Credit should be apportioned to the distinguished authors of the cataract surgery guidelines suggesting 24 hour follow up of patients who had undergone complicated surgery, had coexisting eye disease, or had large incision cataract surgery. We would like to reiterate the importance of explicit criteria as part of any review policy and suggest that clinical interpretation of individual circumstances is paramount.

To reconcile the need for an efficient, cost effective review protocol together with a necessity to give due consideration to the entirety of detrimental post-cataract complications, especially given the NHS resource constraints, is difficult. A pragmatic approach may be for clinicians to be discriminatingly aware of those patients most at risk of developing early complications and instituting review policies accordingly, together with an open door policy for patients who need or want reassurance on the first day following uncomplicated surgery. Furthermore, a multi-professional management approach involving the extended role of trained ophthalmic nurses in postoperative care may reduce demands on physician time.

At the moment, there is a paucity of good prospective literature on the subject and a need for future studies to address whether those identified complications would result in a change of management at the first postoperative day visit and whether patients would have a poorer outcome if the changes were not instituted.

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References

Table 1 Day 1 postoperative complications noted in clinical studies

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Case report
A 37 year old woman presented for an examination with hard contact lenses. Her best corrected visual acuity was 20/20 in both eyes. A diagnosis of neurofibromatosis type 1 was made on the basis of multiple cafe au lait spots, plexiform neurofibromas, and Sakurai-Lisch nodules in the iris in both eyes. The conventional ophthalmoscopic and biomicroscopic fundus examinations were unremarkable. A general medical examination showed no abnormalities except signs of neurofibromatosis type 1. SLO examination showed no abnormalities with a helium-neon laser (633 nm) and regions of multiple, bright patches with infrared imaging (780 nm) using the direct confocal mode (Fig 1) and dark patches with the indirect mode (Fig 2) at the corresponding regions in the posterior pole in both eyes. There were no scotomas in those regions using SLO micropеры.

Comment
In this case, we observed choroidal abnormalities in a patient with neurofibromatosis type 1. The conventional fundus examination, including biomicroscopic examination and fundus colour photography, did not show remarkable changes. However, the SLO examination showed regions of bright patches with infrared imaging using the direct confocal mode and dark patches using the indirect mode at both posterior poles. Infrared light penetrates the retina into the choroid more than visible light.5 Therefore, the bright patch regions seen with confocal infrared imaging and the absence of such regions under helium-neon light examination indicates that the patchy regions are of choroidal origin, as reported by Yasunari and colleagues.6 In addition, we observed dark patchy regions in the corresponding area using the indirect mode of infrared imaging (which also can obtain images of the deeper retinal layers non-invasively) instead of using indocyanine green fundus angiography. Yasunari and colleagues reported that choroidal abnormalities (100%) occurred more frequently than plexiform neurofibroma (29%) and Sakurai-Lisch nodules in the iris (76%).5

Figure 1 SLO image of the right eye using the infrared direct confocal mode.

Figure 2 SLO image of the right eye using the infrared indirect mode.
They also reported that bright patchy choroidal regions should be a new diagnostic criterion for neurofibromatosis type 1. The non-invasive SLO examination with confocal and indirect infrared imaging may be useful in the diagnosis of patients with neurofibromatosis type 1.

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References


Corneal melt and perforation secondary to floppy eyelid syndrome in the presence of rheumatoid arthritis

Floppy eyelid syndrome (FES) is an uncommon condition that it is often underdiagnosed or misdiagnosed owing to the somewhat trivial and non-specific symptoms with which it often presents. In association with the dry eye of rheumatoid arthritis it can, however, have devastating effects.

Case report

A 60 year old moderately obese man with well controlled rheumatoid arthritis (RA) presented to the eye clinic with recurrent red and gritty eyes. A diagnosis of dry eye syndrome with blepharitis was made. He was also found to have a mucocele of the left lacrimal sac. Lid hygiene and ocular lubricants yielded an initial beneficial response and he was discharged. He re-presented 5 years later with similar symptoms and reduced visual acuity (VA) of 6/36 in the left eye. A diagnosis of dry eye syndrome with secondary corneal epithelial changes was made. Topical lubricants failed to relieve the condition satisfactorily and he was therefore scheduled for punctal occlusion. However, upon admission for this 8 weeks later, he was found to have an asymptomatic perforation of the left cornea, with a VA of 6/60. The perforation was treated with glue and a bandage contact lens, topical antibiotic, steroids, and lubricants. Systemic immunosuppression was considered in view of the history of RA, but was withheld as a heavy growth of staphylococcus had been cultured from the cornea. The right eye demonstrated signs of dryness but was otherwise healthy with VA of 6/9.

Bilateral punctal occlusion was undertaken as planned, and in addition a left dacrocystorhinostomy (DCR) to eliminate the mucocele as a potential reservoir of infection. The eye, however, continued to slowly deteriorate, with persisting mucopurulent discharge, despite the DCR. Eventually uncontrolled endothelial damage was noted requiring evisceration.

He re-presented 8 months later with reduced VA of 6/60 in the right eye secondary to a corneal melt (Fig 1A). A chronic mucopurulent discharge had also developed in the right eye, but no lacrimal sac mucocele was identifiable. On this occasion, however, it was noted on examination that while everting the eyelids, all four lids exhibited excessive laxity (Fig 1B). This, together with a florid papillary tarsal conjunctival reaction and the chronic mucous discharge, led to a diagnosis of RA associated dry eye syndrome exacerbated by FES.

All four eyelids were immediately subjected to considerable shortening by pentagonal excision; the corneal melt was treated with a bandage contact lens, with topical antibiotic, steroids and lubricants. The response to surgery was dramatic with complete resolution of discharge and gradual spontaneous repair of the corneal melt (Fig 2). The VA eventually recovered to 6/9.

Comment

FES occurs most frequently in middle aged obese males, although it has been described in young, slim males, females, and one child. Typically, the upper tarsus is rubbery and the upper eyelids easily with gentle upward pressure. A florid papillary conjunctivitis and chronic mucus discharge are common. Severe corneal involvement is rare, with only four reported in the literature of ulceration in association with FES and only two cases of perforation. Although the exact pathophysiology of FES is uncertain, a sequence of events may lead to its development and to the secondary corneal changes has been proposed. Uncommon predisposing factors, possibly congenital, create a floppy upper tarsus. Whereas examination of post-excisional specimens has revealed normal tarsal collagen, elastin fibres are nearly absent. It is unclear whether this finding is causative or secondary. During sleep, a local pressure induced ischaemia may develop in the tarsus that, when relieved, results in a reperfusion injury which could injure tarsal elastin. In addition, there is a high incidence of obstructive sleep apnoea in FES patients and nocturnal dacyroadenitis could contribute to the local ischaemia and subsequent elastin damage.

Corneal involvement may occur through one or more mechanisms. Spontaneous nocturnal lid eversion resulting from pressure from the pillow on the upper lid may lead to repeated trauma of the corneal epithelium. Lash ptosis may contribute to this direct trauma. The cornea, however, may be damaged from a more subtle but important mechanism. Affected lid specimens demonstrate a marked polymorphonuclear infiltrate, which may be the sequelae of the reperfusion injury described above; this tarsal infiltrate and the associated papillary response may have direct toxic effects on corneal epithelium and stroma. It is perhaps intuitive that the corneal complications found in FES may be more severe when, as in our case, coexisting pathologies are present. Blepharitis and RA associated dry eye may both independently cause significant corneal pathology.

This case serves as a reminder that multiple pathologies may contribute to the clinical picture. If FES is not to be missed, ocular examination must include lid evisceration and inspection of the tarsus.

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References

**Ocular trauma with small framed spectacles**

Penetrating injuries are widely reported with spectacle related eye trauma, particularly in car accidents. The use of high grade plastics and secure frames have been shown to reduce the incidence of spectacle related eye trauma. Spectacle safety may be compromised in the trend for small frames and frameless spectacles and may place patients at risk of serious ocular injury.

We present the case of an aphakic patient who sustained a blunt injury following ocular compression by her spectacles. Her injuries could have been avoided if larger framed spectacles had been worn.

**Case report**

A 79 year old aphakic woman sustained a non-penetrating injury to her left globe by walking into a door. She noted a sharp pain and sudden loss of vision. The globe was compressed by her spectacles, which were smaller than her orbital rim. Her glasses were not damaged and there was minimal periocular soft tissue injury.

She was aphakic, following bilateral cataract extraction for congenital cataracts. The spectacle refraction was +9.00 with a short back vertex distance of 5 mm (Fig 1).

The pinhole acuity was 6/36, a quiet, deep anterior chamber was noted with no aqueous flare. The intraocular pressure returned to 14 mm Hg. The suprachoroidal haemorrhages resolved over 2 weeks and the visual acuity improved to 6/6.

**Comment**

The potential ocular damage from framed and frameless spectacles has been highlighted in a number of reports. These often result from minor road traffic accidents or inflation of airbags that damage the spectacles. The trauma is usually sufficient to break the lenses in the spectacles and the resultant globe laceration is the main cause of morbidity. The recent trends towards smaller framed spectacles has not been reported as a potential risk to the patient; however, in this case, with small framed spectacles (with a short back vertex distance), minor trauma was sufficient to cause serious eye injury. The patient’s previous larger framed spectacles would have prevented such an injury as the lenses would have been supported by the orbital margin and not the globe.

This case demonstrates the previously unconsidered risk of small framed spectacles in aphakic patients. The back vertex distance may be short, increasing the risk of blunt injury.

**References**


**Figure 1** Note the short back vertex distance in this aphakic correction.

**Figure 2** The choroidal rupture following blunt injury to the globe.
but approximately 1 year after the surgery there was gradual deterioration of vision to 6/24 in August 2000. At this stage the IOL was noted to be cloudy; there was also progression of her endothelial dystrophy. She underwent uneventful right penetrating keratoplasty with exchange of posterior chamber IOL in November 2000 and has a current right visual acuity of 6/12. The explanted opacified IOL is compared to a normal clear acrylic IOL in Figure 1B.

Comment
The safety and efficacy of AcrySof polyacrylic IOLs has been reported to be equal to or better than poly(methylmethacrylate) IOLs. The unexpected late opacification of the acrylic IOL (SC60B-OUV), implanted in the only "good" eye of both our patients, resulted in significant visual disability and clinical dilemma. This model of IOL was first produced in June 1997 by Medical Development Research (MDR, Inc). More than 60 000 of these lenses have been implanted worldwide, but only outside the United States. Reports of opacification of the IOL started coming through to the manufacturer in May 1999. (Summary of SC60B-OUV lens opacification investigation, personal communication from MDR, Inc, 20 July 2001.)

Several theories have been put forward to explain the late clouding of the IOL optic.1-4 Analysis of 23 explanted IOLs of the same model has shown that degeneration of the ultraviolet filtration material and calcium deposits within the optic biomaterial are responsible for the opacification of the IOL.5 Werner et al analysed nine explanted IOLs of the same model and demonstrated the presence of calcium phosphate salts in the deposits within the optics of the IOL.6 Investigations by the manufacturers identified four lots of polymer biomaterial formulated and prepared by Vista Optics (London) and used by MDR, Inc in the IOL manufacture, that correlated with opacification complaints (Summary of SC60B-OUV lens opacification investigation, personal communication from MDR, Inc, 20 July 2001).

Ninety two of the estimated 60 000 SC60B-OUV IOLs implanted were explanted and returned to the company. MDR, Inc ceased exporting SC60B-OUV lenses in June 2000 and claims that the opacification represents only 0.15% of total SC60B-OUV IOLs implanted. (Summary of SC60B-OUV lens opacification investigation, personal communication from MDR, Inc, 20 July 2001). However, this does not account for those patients who have not yet had their opaque IOLs identified or explanted. The lateness of the onset of opacification and resulting visual disability may mean that we are seeing only the tip of the iceberg.

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References

Treatment of superior limbal keratoconjunctivitis with a unilateral bandage contact lens

The typical patient with superior limbal keratoconjunctivitis (SLK) is a woman aged between 20 and 60 years of age with chronic red and itchy eyes. Although both eyes are usually affected, the condition maybe asymmetrical.1 After episodes of exacerbation and remission it usually resolves. The patient may also have abnormal thyroid function.2 SLK has been treated with silver nitrate or thermal cauterisation of the superior bulbar conjunctiva, pressure patching, and large diameter bandage contact lenses (BCL), topical trans-retinoic acid 0.1%, and recession or resection of the superior bulbar conjunctiva.3,4 Over 50% of patients with SLK are said to have keratoconjunctivitis sicca5 and recently upper punctal plugs have been used to treat SLK.

We report two cases in which a unilateral BCL wear ameliorated the symptoms of bilateral SLK and a possible explanation is discussed.

Case reports
Case 1
A 38 year old woman presented with a 3 month history of irritable photophobic eyes that were unresponsive to preserved lubricants. Her right eye was amblyopic. On systemic review she reported weight loss, heat intolerance, and insomnia.

Slit lamp examination revealed bilateral superior conjunctival hyperaemia, superior punctate epithelial erosions, and four to five filamentary and micropannus on the superior cornea of each eye. Both superior tarsal conjunctivae had moderate papillary reactions. Schirmer’s test without anaesthesia was 14 mm on the right and 15 mm on the left at 5 minutes. Non-preserved lubricants every 1–2 hours and Lacrilube ointment at night were prescribed.

Thyroid function tests were normal 6 months later and the propylthiouracil was stopped. Thyroid function tests were normal 6 months later and the propylthiouracil was stopped.

Comment
The pathogenesis of SLK is unclear. It may be the result of mechanical irritation from increased pressure of the upper eyelid against the globe and/or increased motility of the upper bulbar conjunctiva from hypothroidism or ageing. Increased upper eyelid tightness may be the result of thyroid eye disease or chronic inflammation and, in addition, may

Case 2
A 54 year old woman was referred with a 3 year history of sore, gritty eyes, worse on the left. The tear break up time was <10 seconds and Schirmer test without anaesthesia was right 0 mm and left 1 mm after 3 minutes. There was some relief from lubricants, though occasionally the pain was so severe that she required oral analgesia.

On examination punctate epithelial erosions were found on the superior bulbar conjunctiva of both eyes, but were more marked on the left (Fig 1). A silicone hydrogel BCL (Pure Vision, Bausch and Lomb) was inserted into the left eye, which rapidly ameliorated the symptoms in both eyes. Lubricants were continued for the right eye.

Three months later she remained asymptomatic and no fluorescein staining was seen. The BCL was removed. Within a month her bilateral ocular discomfort returned and the left BCL was refitted with immediate symptomatic relief in both eyes. Two months later the BCL was lost and her bilateral ocular discomfort recurred within 2 days. Since then her BCL has been replaced every 3 months. Occasionally she has used lubricants for the right eye.

One year following her presentation she became tachycardiac and hyperthyroidism was diagnosed. Her endocrinologist commenced carbimazole and β-blockers. Her treatment was later changed to propylthiouracil as she suffered from carbimazole induced arthralgia. Thyroid function tests were normal 6 months later and the propylthiouracil was stopped.

Figure 1  Case 2, left eye. (A) The superior bulbar conjunctiva, at presentation, showing hyperaemia and rose Bengal staining. (B) After 3 months of bandage contact lens wear the superior conjunctival hyperaemia had resolved and the patient was asymptomatic.

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impair the normal turnover of bulbar conjunctival epithelial cells. This may be aggravated, in some patients, by blepharospasm, which increases the force on the globe.  

Therapeutic lenses can produce rapid symptomatic relief. They may be helpful in the treatment of SLK as they relax the tarsus and facilitate healing of punctate epithelial erosions by protecting the ocular surface from blinking as it has a keratitis and corneal vascularisation. Decrease bilateral reflex blinking. Measurable 1-2 hours after lens insertion that BCL wear reduces the tactile corneal surface sensation in patients, increase oxygen transfer and have extended wear silicone BCL, as used in our study, seems to be either undiagnosed forme fruste keratoconus, with a residual stromal thickness of over 500 μm, from the data of controlled studies and are of good merit, whereas others have less scientific merit in that they are long term follow up studies without controls. For ophthalmologists wishing to look into the subject of substantial information available regarding idiopathic keratectasia (iatrogenic keratoconus), with a little over 60 cases published. At this point, although some are likely to be due to over-ablation, for many cases such as this the exact aetiology remains unknown and is likely to be multifactorial, and one of these factors is residual corneal thickness. The fact that keratectasia can occur, after what would be considered minimal ablation, highlights the unpredictability of occurrence, but with over a million cases of LASIK or PRK occurring each year, the stimulus to identify contributing factors is significant.

Keratectasia after PTK
Takahashi and colleagues have elegantly described an interesting and rare complication of phototherapeutic keratectomy (PTK) in their recent report of an unusual case of keratectasia after PTK. The hypothesis that risk of ectasia is proportional to residual stromal base, or depth of ablation, fits with the assumed biomechanical aetiology of this recently reported complication of laser refractive surgery. The generally accepted empirical minimum thickness of 250-300 μm of corneal stroma, excluding flap thickness, remains speculative, as we do not understand the underlying pathophysiology. Indeed, although Holland et al highlighted the association of thin residual stromal thicknesses, post-PRK and LASIK, with keratectasia, they also described this complication after surface ablated hyperopic PRK ablation, where the centre was minimally ablated and residual stromal thickness was greater than 360 μm. The authors suggest, in the reported case, that band-shaped keratopathy (BSK) may have compressed the tensile strength of the cornea. This seems unlikely as this condition generally affects the superficial anterior cornea, and usually does not penetrate deeper than Bowman’s layer. Its suitability for treatment by PTK. However, further clinical detail which the authors have not provided might reveal underlying corneal pathology with secondary “rough” BSK rather than “smooth” BSK.

However, there are a number of reasons, other than simple biomechanical compromise, for keratectasia following PTK in this case: (1) forme fruste keratoconus—as no preoperative topography or surface asymmetry values are presented to enable the reader to rule this out; (2) clinical keratoconus, which seems less likely in respect of patient’s age and a preoperative cylindrical error of only −1.50 D; (3) idiopathic keratectasia, possibly secondary to widespread deregulated keratocyte apoptosis. The latter has been demonstrated after LASIK, with a considerable and longstanding decrease in keratocytes in the peri-ablation area. Also, Helena et al demonstrated apoptosis to a depth of at least 50 μm after all of the following procedures: epithelial scrapes, corneal scrape PRK, transepithelial PRK, and LASIK. Epithelial scrapes and LASIK demonstrated keratocyte apoptosis to depths of up to 75 μm and 100 μm, respectively. The authors have recently identified a keratocyte free zone 160 μm into the stroma following LASIK, and theoretically more widespread apoptosis as a response to excimer laser photorefractive surgery, may contribute to keratectasia.

While it is difficult to ascertain why keratectasia occurs, in this case with a residual stromal thickness of over 500 μm, from the data provided the most likely aetiologies would seem to be either undiagnosed forme fruste keratoconus or idiopathic keratectasia. Currently, recent reviews illustrate the dearth of substantive evidence in this area. However, the authors suggest, in the reported case, that band-shaped keratopathy (BSK) may have compressed the tensile strength of the cornea. This seems unlikely as this condition generally affects the superficial anterior cornea, and usually does not penetrate deeper than Bowman’s layer. Its suitability for treatment by PTK. However, further clinical detail which the authors have not provided might reveal underlying corneal pathology with secondary “rough” BSK rather than “smooth” BSK.
radiotherapy for ARM D this would be a good source of material and is well referenced. There are a few chapters on the clinical manifestations, diagnosis, and surgery of ARM D but there is no real mention of laser treatment. Many of these chapters are of limited scope and do not provide a comprehensive overview of the ophthalmic assessment and management of ARM D. Although not stated in the book it reads as if it is the proceedings of a clinical meeting. The chapters do not read in a coordinated way and essentially present the results of individual units describing their methods and results of radiotherapy. As such it is a useful source of information for the clinician with an interest in this topic but it is of limited value for ophthalmologists wishing to obtain a balanced view of current treatment of ARM D.

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Clinical Ophthalmic Pathology.

This is an excellent, easy to read, well illustrated book. It is one of the first of its kind to bring pathology alive by describing diseases via pathogenesis as opposed to anatomy. For the trainee in ophthalmology, optometry, and visual science it, therefore, provides a more logical approach to the understanding of ocular diseases. As the book attempts to cover many subjects it sometimes does not do justice to all. It would have been better to have had the reading lists at the end of each chapter. However, as it stands the book is an excellent introduction to pathology complementing the clinical textbooks. If read together with clinical texts it certainly will broaden the knowledge base of all trainee ophthalmologists. As a result of its logical and simple approach I was left frustrated at times with the lack of background knowledge. However, going through pathologically based chapters including injury and repair, immunity, genetics, growth, degeneration, vascular disorders, and disorders of the nerve and muscle, I was left entertained, as a clinician, with a greater understanding of pathological processes.

The final chapter for the clinician in the laboratory I felt could have been expanded, delivering more detail, particularly, on the current molecular methods used in pathological practice today.
A Dick

NOTICES

Childhood blindness
The latest issue of Community Eye Health (No 40) discusses news in childhood blindness, with an editorial by Clare Gilbert, senior lecturer at the International Centre for Eye Health. For further information please contact: Journal of Community Eye Health, International Centre for Eye Health, Institute of Ophthalmology, 11–43 Bath Street, London EC1V 9EL, UK (tel: +44 (0)20 7608 6910; fax: +44 (0)20 7250 3207; email: eyeresource@ucl.ac.uk; website: www.jceh.co.uk). Annual subscription (4 issues) UK£23.50/$40. Free to workers in developing countries.

International Centre for Eye Health
The International Centre for Eye Health has published a new edition of the Standard List of Medicines, Equipment, Instruments and Optical Supplies (2001) for eye care services in developing countries. It is compiled by the Task Force of the International Agency for the Prevention of Blindness. Further details: Sue Stevens, International Centre for Eye Health, 11–43 Bath Street, London EC1V 9EL, UK (tel: +44 (0)20 7608 6910; email: eyeresource@ucl.ac.uk).

Second Sight
Second Sight, a UK based charity whose aims are to eliminate the backlog of cataract blind in India by the year 2020 and to establish strong links between Indian and British ophthalmologists, is regularly sending volunteer surgeons to India. Details can be found at the charity website (www.secondsight.org.uk) or by contacting Dr Lucy Mathen (lucymathen@yahoo.com).

Specific Eye Conditions (SPECS)
Specil Eye Conditions (SPECS) is a not for profit organisation which acts as an umbrella organisation for support groups of any conditions or syndrome with an integral eye disorder. SPECS represents over fifty different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. We also include groups who offer support of a more general nature to visually impaired and blind people. Support groups meet regularly in the Boardroom at Moorfields Eye Hospital to offer support to each other, share experiences and explore new ways of working together. The web site www.eyeconditions.org.uk acts as a portal giving direct access to support groups own sites. The SPECS web page is a valuable resource for professionals and may also be of interest to people with a visual impairment or who are blind. For further details about SPECS contact: Kay Parkinson, SPECS Development Officer (tel: +44 (0)1803 524238; email: k@eyeconditions.org.uk; www.eyeconditions.org.uk).

XXIXth International Congress of Ophthalmology
The XXIXth International Congress of Ophthalmology will be held on 21–25 April 2002 in Sydney, Australia. Further details: Congress Secretariat, C/- ICMAS Australia Pty Ltd, PO Box 2609, Sydney, NSW 2001, Australia (tel: +61 2 9241 1478; fax: +61 2 9251 3552; email: ophthal@icmasaust.com.au; website: www.oophthalmologyaust.com).

12th Meeting of the European Association for the Study of Diabetic Eye Complications (EASDEC)
The 12th meeting of the EASDEC will be held on 24–26 May 2002 in Udine, Italy. The deadline for abstracts is 15 February 2002. Three travel grants for young members (less than 35 years of age at the time of the meeting) are available. For information on the travel grants, please contact Pr CD Agardh, President of EASDEC, Malmö University Hospital, SE-205 02 Malmö, Sweden (tel +46 40 33 73 66; email: carl-david.agardh@endo.mas.lu.se). Further details: NORD EST CONGRESSI, Via Aquileia, 21–33100 Udine, Italy (tel: +39 0432 21399; fax: +39 0432 50687; email: nordest.congressi@ul.net.uno.it).

3rd Interdisciplinary Symposium on the Treatment of Autoimmune Disorders
The 3rd Interdisciplinary Symposium on the Treatment of Autoimmune Disorders will be held in Leipzig, Germany on the 6–8 June 2002. Topics to be covered include: basic aspects of autoimmune diseases, experimental therapeutic concepts, and clinical studies providing novel concepts or novel focus on established therapies. There will also be the presentation of the Nils-Illa-Richter Award (application deadline is April 2002, further details on the web site). Further details: Prof. Dr. med. Michael Sticherling, Department of Dermatology, University of Leipzig (email: stiem@medizin.uni-leipzig.de; website: www/autoimmun.org); Fördergesellschaft zur Therapie von Autoimmunerkrankungen e.V. (email: autoimmun.org@gmx.de).

International Society for Behçet’s Disease
The 10th International Congress on Behçet’s Disease will be held in Berlin 27–29 June 2002. Further details: Professor Ch Zouboulis (email: Zoubbere@zedat.fu-berlin.de).

Singapore National Eye Centre 5th International Meeting
The Singapore National Eye Centre 5th International Meeting will be held on 3–5 August 2002 in Singapore. Further details: Ms Amy Lim, Organising Secretariat, Singapore National Eye Centre, 11 Third Hospital Avenue, Singapore 168751 (tel: (65) 322 8374; fax: (65) 227 7290; email: Amy_Lim@snec.com.sg).

BEAVRS Meeting
The next BEAVRS meeting will be held in the Dalmahoy Hotel near Edinburgh on 31 October to 1 November 2002. Further details: Susan Campbell, Medical Secretary, Gartnavel General Hospital (email: susan.j.campbell.wg@northglasgow.scot.nhs.uk).

CORRECTION
The authors of the letter “Recurrent corneal ulceration as late complication of toxic keratitis”, appearing in the February issue of Br J Ophthalmol (2002;86:245–6), would like to add an author, SH Santander.