

# PostScript

## LETTERS

### Long term survival of patient with invasive aspergillosis involving orbit, paranasal sinus, and central nervous system

*Aspergillus* infections of the paranasal sinuses are classified as invasive or non-invasive.<sup>1</sup> The vast majority are non-invasive with a good prognosis; however, the invasive type behaves as a malignant neoplasm with bone destruction, orbital and intracranial extensions, and a high mortality rate.<sup>2-4</sup>

We present one patient with invasive paranasal aspergillosis that extended into the orbit and cranial cavities, and who has survived for 9 years and 1 month.

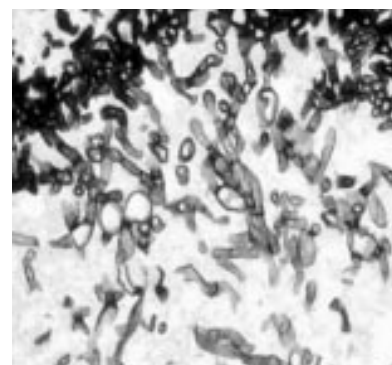
#### Case report

Along with headaches and periorbital pain beginning in July 1993, a 64 year old woman noticed a decrease vision in her right eye and visited us on 7 September 1993. She had poorly controlled diabetes mellitus. Her corrected visual acuity was 20/30 right eye and 20/20 left eye, and her critical flicker fusion frequency was 25 Hz right eye and 32 Hz left eye. A central scotoma with a relative afferent pupillary defect was present in the right eye. The right optic disc was slightly swollen.

A computed tomographic scan disclosed a mass in the right posterior ethmoid and sphenoid sinuses with destruction of the lateral wall of the sphenoidal sinus and the enlargement of the superior orbital fissure. The mass extended to the right orbital apex and the right cavernous sinus. Magnetic resonance imaging showed an enhanced mass, isointense on T<sub>1</sub> weighted and isointense and partially hypointense on T<sub>2</sub> weighted images (Fig 1).

Endoscopic right sphenoidectomy was performed on 18 September, and *Aspergillus* was histopathologically identified in a small mass isolated from the sphenoid sinus mucosa (Fig 2). Intravenous fluconazole (400-800 mg/day), oral flucitocine (8000 mg/day), and daily transnasal sinus irrigation with amphotericin B were started on 22 September. However, she lost all vision in her right eye on 4 October.

She was transferred to the department of neurological surgery on 13 October. Antifungal therapy was continued with the addition of oral itraconazole (200 mg/day). Carotid angiography demonstrated that stenosis of the right ophthalmic artery. A frontal craniotomy was performed on 1 November, and a fibrous granulomatous lesion was found in the right posterior ethmoid sinus, sphenoid sinus, cavernous sinus, and orbit that extended to the right optic nerve. The orbit was necrotic and the sphenoid sinus was severely eroded. Cultures and histopathological examinations were negative for *Aspergillus*.



**Figure 2** Histopathological section of mass obtained from the right sphenoid exhibiting *Aspergillus* with broad, separate, and branching hyphae (Grocott-Gomori methenamine-silver stain, x200).

The patient underwent debridement and removal of the frontal bone on 24 December because of the development of an epidural abscess. In February 1994, the abscess had extended from the right cavernous sinus to the temporal lobe and bacterial cultures were positive for *Pseudomonas aeruginosa*. The abscess subsided but still persisted following antibiotics and antifungal therapy.

On 13 September 1994, orbital exenteration, intracranial exenteration, and skin plasty were performed. Her diabetes mellitus is fairly well controlled by insulin injection and she has been free of aspergillosis for 9 years and 1 month after the initial presentation.

#### Comment

To the best of our knowledge, survival for more than 9 years of a patient with invasive paranasal aspergillosis has not been reported. The longest previous case of invasive paranasal aspergillosis is for 8 years and 2 months.<sup>1</sup>

The prognosis of this disease depends on the location and the duration of the infection, and patient's immunological status.<sup>3,4</sup> Sphenoidal aspergillosis is aggressive nature because of the close relation to the skull base.<sup>3</sup> Her poorly controlled diabetes mellitus could be a risk factor for the onset of *Aspergillus* infection; however, at present it is fairly controlled, presumably contributing to this long survival.

The sphenoid sinus mucosa was positive for *Aspergillus* but it was not found at the time of the intracranial surgery probably because of the extensive antifungal therapy before surgery and/or the low viability of the mycelium in a fungus ball.<sup>5</sup>

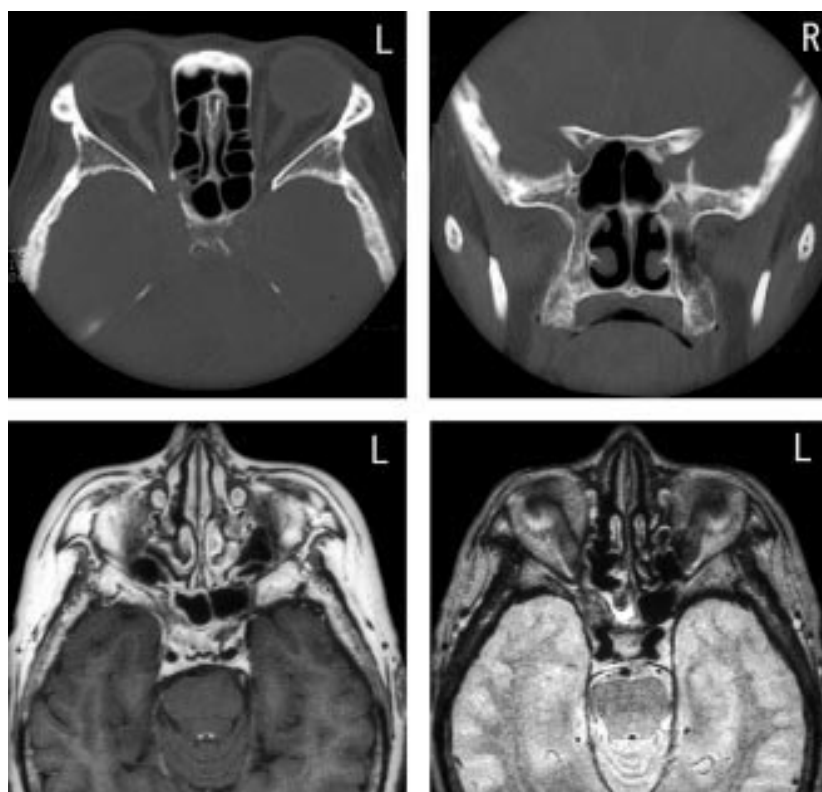
The long term survival of this patient with orbital-paranasal aspergillosis despite intracranial extension is attributed to early diagnosis, optimal antifungal therapy, complete surgical debridement, and the improvement in the patient's systemic condition.

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**Figure 1** (Top left) A computed tomographic scan shows a mass in the right ethmoid and sphenoid sinuses extending to the right orbital apex and the right cavernous sinus. (Top right) Coronal section shows bony destruction of the sphenoidal lateral wall. (Bottom left) Magnetic resonance imaging reveals the enhanced mass involving the right cavernous sinus and orbital apex on T<sub>1</sub> weighted with gadolinium contrast images. (Bottom right) Magnetic resonance imaging demonstrates isointense and partially hypointense on T<sub>2</sub> weighted images.

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## The effect of topical glaucoma medications evaluated by perimetry

The emphasis of treatment for glaucoma has been the reduction of intraocular pressure (IOP) to a safer level, which, in turn, theoretically will prevent further visual loss. It has been assumed that lowering IOP by medical means has no adverse effect, which could negate the beneficial effect of IOP reduction. However, several reports have raised the possibility that some may adversely affect visual function.<sup>1,2</sup> In several studies betaxolol and timolol were compared with respect to their effect on IOP reduction and perimetric findings.<sup>3-5</sup> Although timolol lowered IOP more effectively, betaxolol was more effective in preserving the visual field.

These findings suggest that IOP reduction is not the only parameter that demands attention. In this study, we attempted to evaluate various topical antiglaucoma medications in a normal population in terms of their short term effect on visual function; only minimal effects on IOP reduction were expected since this study population did not have glaucoma.

**Methods**

Five prospective, randomised, masked studies of levobunolol, dipivefrin, apraclonidine, betaxolol, and dorzolamide, respectively, were conducted over 5 years. In each study, 20 normal volunteers had baseline testing, including measurement of visual acuity (VA), IOP, visual field (VF) with the Humphrey computerised perimeter (HCP) program 24-2, and pupil size. One eye was randomly assigned to treatment and given a test dose of either a glaucoma medication or a placebo. VF testing was repeated in 1 hour. The same eye was later given the opposite drop (the drug if a placebo was used, a placebo if the drug was used) and a third set of visual field studies was done. Paired *t* test and signed rank test were used in the statistical analysis.

All subjects had normal ocular examinations, with best corrected visual acuity of 20/30 or better. The mean baseline IOP was 13.3 mm Hg right eye and 13.5 mm Hg left eye (range 10-18 mm Hg). When levobunolol was administered, there was a mean deviation of -2.9 dB compared with -1.9 dB for the

**Table 1** Effects of topical glaucoma medications on automated perimetry

	MD (dB)	C/W placebo
Dipivefrin	-0.86	-1.05
Apraclonidine	-0.36	-0.44
Levobunolol	-2.90	-1.90*
Betaxolol	-1.23	-0.79
Dorzolamide	+0.16	+0.12

\* *p*<0.05.

placebo (*p*<0.05) (Table 1). There were no significant differences in terms of other global values (short term fluctuation, pattern standard deviation, and corrected pattern standard deviation), visual acuity, and pupil size.

Thus, in a normal population, a single dose of several topical glaucoma medications had little or no effect on perimetry. The possible mild suppression of retinal sensitivity with levobunolol is of unknown clinical significance.

**Comment**

Flammer and Drance tested several drugs topically, all of which reduced retinal sensitivity on threshold perimetry in contrast with oral acetazolamide, which improved perimetry results in 75 patients with elevated IOP or in the very early stages of chronic open angle glaucoma.<sup>6</sup> Paterson found that acute IOP reduction increased retinal sensitivity.<sup>7</sup> Martin-Boglund and coworkers showed that the influence of topical glaucoma medications (epinephrine, timolol, betaxolol, pilocarpine) on the high pass resolution threshold was negligible in normal subjects.<sup>8</sup> Edgar and coworkers found no significant change with dipivefrin in normal subjects.<sup>9</sup> In our studies, only levobunolol caused a mild generalised decrease in visual function compared to controls on visual field testing of normal subjects. Short term fluctuation was not altered and there was no relative focal defect when levobunolol was compared with placebo. Since these were controlled trials, the placebo effect was not a factor. Vasoactivity, direct effect on neuronal elements, corneal epithelial alteration, and refractive change of current glaucoma drugs should be also considered. The mechanism by which various topical glaucoma medications affect retinal sensitivity remains unclear. The effect may be due to changes in blood flow to the posterior segment of eye. Further study is needed.

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## Delayed therapeutic success with endoscopic cyclophotocoagulation in treating refractory post-penetrating keratoplasty glaucoma

Endoscopic cyclophotocoagulation (ECP) was introduced as an alternative to trans-scleral cyclophotocoagulation for treating refractory glaucomas in order to minimise complications such as phthisis and hypotony by providing direct visualisation of the ciliary processes.

Glaucoma following penetrating keratoplasty, which has an incidence ranging from 10-52%, often proves refractory to medical treatment.<sup>1-3</sup> We introduce a case of refractory post-PKP glaucoma in order to demonstrate the efficacy of ECP in treating post-PKP glaucoma and to describe its potential delayed effect in achieving intraocular pressure control.

**Case report**

A 50 year old African-American man, who had undergone previous cataract surgery, anterior vitrectomy, and anterior chamber intraocular lens placement in his left eye in 1987, presented with pseudophakic bullous keratopathy and hand movement vision in his left eye. In April of 2000, the patient underwent a penetrating keratoplasty in which an 8 mm donor graft was placed in a 7.5 mm host site. Two weeks following the procedure, the patient developed elevated intraocular pressure in the 45-50 mm Hg range which was refractory to both medical therapy and discontinuation of topical steroids. An Ahmed valve was placed in June of 2000, yet his intraocular pressure eventually returned to the preoperative range despite the addition of four topical glaucoma medications (timolol 0.5%, brimonidine 0.2%, dorzolamide 2.0%, and latanoprost 0.005%).

In April of 2001, the patient underwent treatment with endoscopic cyclophotocoagulation via a limbal approach as described by



**Figure 1** Endoscopic view of the ciliary processes, in which the treated ciliary processes (left) appear white and contracted. The red aiming laser beam is directed at the anterior portion of the untreated ciliary processes.

Chen *et al.*<sup>4</sup> The patient received 300 degrees of treatment at settings ranging from 20–50 mW of energy with laser applied for 0.5–2 seconds until ciliary process whitening and contraction was observed (Fig 1).

During the first postoperative week, the patient was treated with topical polymyxin B/trimethoprim drops four times a day, and hyoscine (scopolamine) 0.25% drops three times a day. He was also treated with topical prednisolone acetate 1% four times per day, and was tapered off by the third postoperative week. Despite being restarted on all four of his glaucoma medications within 2 weeks of the procedure, the patient continued to have poorly controlled intraocular pressure in the 30–45 mm Hg range which persisted for more than 3 months. However, 14 weeks after the cyclodestructive procedure, the intraocular pressure suddenly began to decrease without any further surgical intervention. The patient's intraocular pressure has remained well controlled in the 10–15 mm Hg range for more than 1 year following ECP, and the total number of glaucoma drugs has been systematically reduced from four to two. Furthermore, the patient has not developed any signs of hypotony, phthisis, or graft failure.

### Comment

Previous studies have demonstrated significant pressure lowering within 2–4 weeks of endoscopic cyclophotocoagulation.<sup>4,5</sup> This represents the first reported case of late success with ECP, with intraocular pressure control achieved more than 3 months following ECP. Though both topical corticosteroids and cycloplegics may lead to a rise in intraocular pressure, the pressure remained elevated more than 2 months after discontinuing both types of medications.<sup>6,7</sup>

Reports vary regarding the number of degrees of treatment necessary to achieve effective results with endoscopic cyclophotocoagulation.<sup>4,5</sup> However, success is ultimately dependent on the extent of treatment along the anteroposterior axis of the ciliary processes as well as the size of the treatment zone.<sup>4</sup> The delayed response observed in this case likely represents incomplete treatment with late fibrotic changes in the ciliary processes, as signs of hypotony and phthisis remain absent.<sup>8</sup>

A high incidence of both acute (35–41%) and chronic (23–29%) graft failure has been associated with drainage tube implants in the treatment of post-PKP glaucoma.<sup>9,10</sup> In contrast, no cases of irreversible graft failure were observed in 16 post-PKP patients treated with ECP, with only a single patient (6%)

developing acute graft rejection.<sup>4</sup> We describe this case in order to demonstrate that the effects of ECP may be appreciated on the order of several months following treatment, and to illustrate, as shown previously, that ECP can often be used safely and effectively in treating refractory post-PKP glaucoma.

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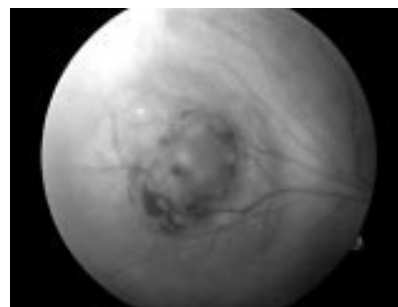
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### Simultaneous presentation of choroidal melanoma in mother and daughter

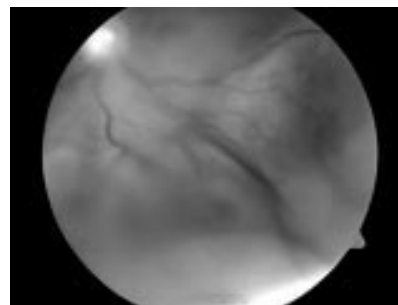
Despite being the most common primary intraocular malignancy, uveal melanoma is rare, with an incidence of only eight per million per year.<sup>1</sup> Familial cases account for only 0.6% of patients.<sup>2</sup> We report two members of the same family who were both independently found to have choroidal melanoma on the same day.

#### Case 1

A healthy 45 year old woman presented to her general practitioner with a 1 month history of photopsia and visual field defect. She was found to have a choroidal tumour in the right eye and referred to a general ophthalmologist, who she saw on 11 November 1999. The diagnosis of choroidal melanoma was confirmed and she was referred to our clinic, where she was seen on 22 November 1999. At our clinic the visual acuity was 6/9 with the right eye and 6/5 with the left eye. Both anterior segments and the left fundus were normal. The right fundus showed a superotemporal pigmented choroidal melanoma with a collar



**Figure 1** Patient 1. Fundus photograph of right eye. A choroidal melanoma was observed superotemporal to the macula.



**Figure 2** Patient 2. Fundus photograph of the right eye. A choroidal melanoma was observed inferonasal to the optic nerve.

stud configuration and extending within two disc diameters of the fovea (Fig 1). It measured 12.2 mm in diameter and was 5.1 mm thick. The patient was treated with proton beam radiotherapy.

#### Case 2

On 7 November 1999, the patient's 65 year old mother presented to her general practitioner in a different city with a 2 week history of blurred vision in the right eye. She was referred to her primary ophthalmologist who she saw on 11 November 1999, then to our clinic where she was given an appointment on 22 November 1999. Our assessment showed that the visual acuity was 6/36 with the right eye and 6/12+ with the left eye. Both anterior segments and the left fundus were normal. There was a pigmented choroidal tumour inferonasally extending from the disc to the ciliary body, measuring 19.2 mm in diameter and 6.0 mm in thickness (Fig 2). The patient was treated by enucleation.

Neither patient had a history of cutaneous melanoma or atypical naevi, nor could they recall any relevant family history of ocular or other disease. The mother's only sibling and three of four of the daughter's siblings have had a normal ocular examination elsewhere.

#### Comment

We report on the simultaneous presentation of mother and daughter each with uveal melanoma in the right eye with both individuals being seen by their ophthalmologist and by us on the same day.

Uveal melanoma is a rare disease, and instances of both parent and child being affected are even rarer. In one series of 4500 patients 17 kindreds were identified in which a first degree relative was also affected.<sup>3</sup> Singh *et al* reported on a single case of concurrent presentation with father and son presenting within 3 months of each other.<sup>4</sup> Our two

patients presented within only a few hours of each other. The chance of such simultaneous presentation must be extremely remote, but our report demonstrates that coincidence can occur in any disease.

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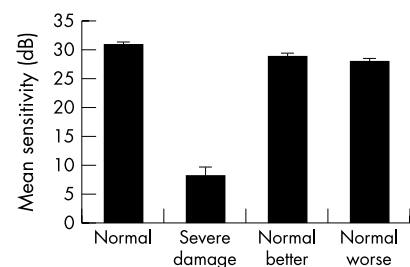
### Relative hypersensitivity in healthy eye by frequency doubling perimetry in patients with severely damaged contralateral eye

Frequency doubling technology (FDT, Humphrey-Zeiss and Welch Allyn, Dublin, CA, USA) has been developed to screen for glaucoma.<sup>1</sup> FDT could detect abnormalities in patients with full visual fields tested by Humphrey field analyser (HFA, Humphrey-Zeiss, Dublin, CA, USA) and nerve fibre layer defects<sup>2</sup> or normal visual fields and large disc cupping.<sup>3</sup> Inversely, FDT did not always detect visual field loss in patients with glaucomatous visual field defects determined by HFA (Fig 1), but it was rare.<sup>4,5</sup> That was because the ability of FDT to detect

glaucomatous visual field loss was limited or for some other reasons. In this study, we presumed that patients with severe damage in one eye had relative hypersensitivity in another eye.

Twenty nine patients (23 men and six women) had one normal healthy eye and one severely damaged eye. Their ages ranged from 18–69 years (mean 51 years). Patients had been examined with threshold c-20 of FDT version 2.6 once. A normal healthy eye meant normal vision (20/20 or better), normal intraocular pressure (less than 21 mm Hg on two occasions), no inflammation, and normal fundus observed clinically. The damaged eye had poor vision (20/200 or worse) or severe visual field defects (mean deviation with FDT less than –6.0 dB). Damage was variously the result of optic nerve lesion by angle recession glaucoma, uveitic glaucoma, or tumour at the orbital apex. Primary open angle glaucoma, optic neuritis or tumour in the brain were excluded, since such conditions usually involve both eyes. As controls, 26 (20 men and six women) normal healthy volunteers were recruited for this study. Their ages ranged from 23 to 70 years (mean 51 years). Patients had been examined with threshold c-20 of FDT version 2.6 once. They had normal vision (20/20 or better), normal intraocular pressure (less than 21 mm Hg), and normal fundus findings. All subjects were examined with threshold c-20 of FDT version 2.6. Mean sensitivity was calculated from all 17 test areas. Mean sensitivity with one normal healthy eye and one severe damaged eye was compared to that in subjects with two normal healthy eyes with Mann-Whitney U test. All results of less than 20% of fixation loss, 20% of false positive, and 33% of false negative were adopted. The research followed institutional guidelines and the tenets of the World Medical Association Declaration of Helsinki. We obtained written informed consent from all patients before their entry in this study.

Eighty results of 55 patients were studied for analysis. One result in patients with two healthy eyes was excluded because of an unreliable result. Twenty nine eyes with severe damage in one eye had a significantly higher mean sensitivity (mean 30.8 (SE 0.47) dB) in one healthy eye than did both normal healthy



**Figure 2** In 29 patients with one normal healthy eye and one severely damaged eye, the mean sensitivity (30.8 (0.47) dB) in 29 healthy eyes was significantly higher than that in 26 patients with two normal eyes ( $p=0.0065$ , 28.8 (0.50) dB in 26 normal better eyes and  $p=0.0005$ , 27.9 (0.56) dB in 25 normal worse eyes).

eyes ( $p=0.0065$ , 28.8 (0.50) dB in 26 better eyes and  $p=0.0005$ , 27.9 (0.56) dB in 25 worse eyes) (Fig 2).

### Comment

It was interesting that there were 2 dB differences in sensitivity between patients with a normal healthy eye. The reason for eyes with severe damage in one eye having relative hypersensitivity was unclear. One possible explanation was because the pathway detected by FDT is thought to be a magnocellular pathway and a relatively less, complementary mechanism might work in magnocellular pathway.

In conclusion, patients with one severely damaged eye had relative hypersensitivity in one healthy eye. Estimation of such patients should be considered carefully.

### Acknowledgements

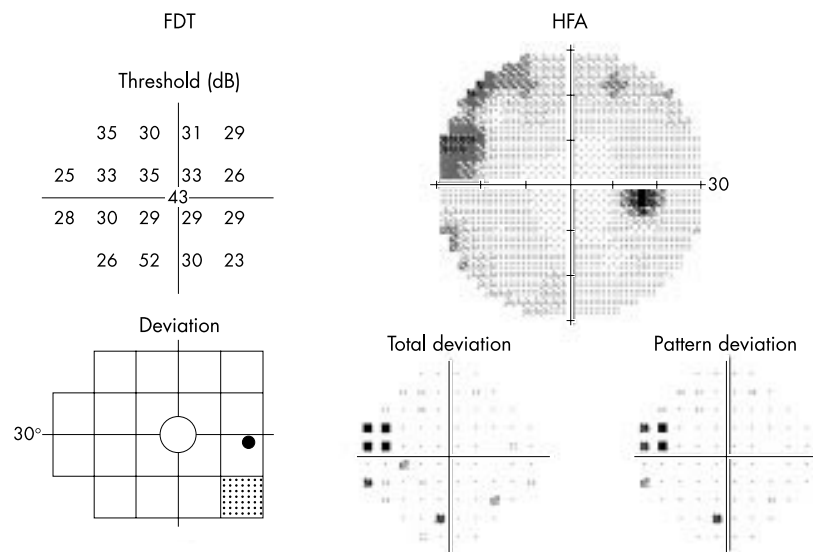
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**Figure 1** A 34 year old man had high intraocular pressure (29 mm Hg) and abnormal points in the right nasal field with Humphrey field analyser (HFA, right), but not with frequency doubling technology (FDT, left). His left eye was blind as a result of glaucoma.

## Solitary choroidal tuberculoma in a patient with chest wall tuberculosis

Tuberculosis has re-emerged as a serious public health problem in industrialised countries.<sup>1</sup> There are several explanations for the increased incidence but it is mainly due to an increase in immunocompromised hosts such as those who are older or with malignancies, those with AIDS, those who are immunosuppressed after transplantation, and the malnourished. However, a choroidal tuberculoma is rare except in cases with human immunodeficiency virus (HIV) infection.<sup>2,3</sup> We present a case of choroidal tuberculoma in an immunocompetent patient with an extrapulmonary tuberculoma in the chest wall, which is also rare.<sup>4,5</sup>

### Case report

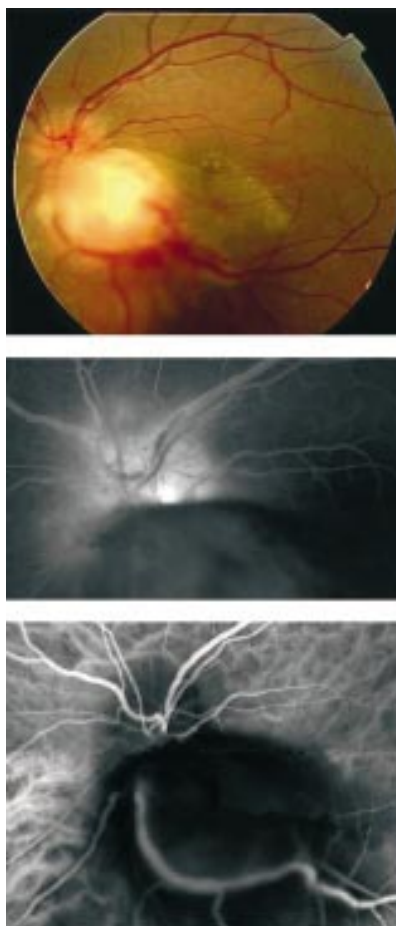
A 34 year old Filipina woman, who was in good health, complained of a pain in her right lateral chest. Computed tomography scan showed a well defined mass measuring 4 × 4 cm (Fig 1). Cultures of fluid aspirated from the mass showed acid fast bacteria. Polymerase chain reaction demonstrated *Mycobacterium tuberculosis* DNA in the aspirated fluid. Her sputum had never been positive for tuberculosis. Although the patient had no fever, cough, or anorexia, the mass was diagnosed as extrapulmonary tuberculoma with minimal pulmonary involvement. Anti-tuberculous treatment was started with isoniazid, rifampicin, pyrazinamide, and streptomycin after a 3 month presence of the cold abscess. Two weeks later, she complained of decreased vision in her left eye.

On initial examination, the best corrected visual acuities were 30/20 in the right eye and 80/200 in the left eye. Anterior segment examination was unremarkable, and no evidence of anterior or posterior inflammation was present.

Fundus examination showed an elevated yellow-white mass in the left eye that measured approximately 2 × 2 disc diameters just inferior and temporal to the optic disc (Fig 2A). The mass had a slightly irregular and fuzzy outline, and the disc had irregular margins and was reddish. There was a flat retinal detachment in the macula area, and fluorescein angiography (FA) demonstrated minimal early fluorescence with late moderate hyperfluorescence and peripapillary leakage in the lesion (Fig 2B). Indocyanine green angiography (IA) demonstrated persistent blockage of fluorescein (Fig 2C). Optic coherence tomography (OCT) showed a highly elevated mass associated with a serous retinal detachment. We excluded sarcoidosis, toxoplasmosis, or fungus infection by laboratory examinations;



**Figure 1** An enhanced chest computed tomography scan showing a capsulated mass (arrow) with destruction of the 12th cartilage (arrowhead).



**Figure 2** (A) Fundus photograph showing an elevated choroidal lesion just inferior and temporal to the optic disc. (B) Late fluorescein angiogram demonstrating minimal hyperfluorescence. (C) Indocyanine green angiography showing blockage of fluorescence.

the levels of angiotensin converting enzyme,  $\beta$ -D-glucan, titre of antibody for toxoplasmosis, and metastatic tumour from a breast cancer. HIV infection was also ruled out.

The visual acuity in the left eye decreased to 12/200 because of the retinal detachment, and vitreous cells and opacities, and retinal vasculitis were observed. Four to 6 weeks after beginning the anti-tuberculosis therapy, the mass became smaller and visual acuity improved. Although retinal folds were present in the macular lesion after 16 weeks of therapy, the best visual acuity was 20/20 (Fig 3). The cold abscess in the chest wall disappeared within 2 months, with drainage of the fluid and injection of streptomycin.

### Comment

There are only a few reported cases of solitary choroidal tuberculoma,<sup>1,3</sup> and it may present with or without active pulmonary tuberculosis. Ocular tuberculosis commonly presents in the choroid, and reaches the choroid by direct haematogenous spread from a primary infection.<sup>4</sup> The chest wall is also a rare site for tuberculosis,<sup>4</sup> and the co-occurrence of chest wall tuberculosis and choroidal tuberculoma has never been reported with or without HIV infection.<sup>1-5</sup> Rib tuberculosis was observed in 5% of all cases of bone and joint tuberculosis, and only in 0.1% of all hospital admission for



**Figure 3** A significantly smaller mass is demonstrated 16 weeks after the initiation of antituberculosis drugs.

tuberculosis.<sup>4</sup> It is usually secondary to haematogenous spread or, more rarely, due to direct extension of underlying pleural or pulmonary parenchymal disease.<sup>5</sup> In our patient, minimal pulmonary involvement was suspected in the apical lesion, not in chest wall mass lesion. Taken together, the tuberculomas in this patient may be caused by direct haematogenous dissemination. Anti-tuberculosis therapy was effective for both tuberculomas.

In conclusion, we report a rare case of choroidal tuberculoma with chest wall tuberculosis. With the re-emergence of tuberculosis, ophthalmologists should be aware that solitary choroidal tuberculoma as well as extrapulmonary tuberculosis can occur in immunocompetent individuals.

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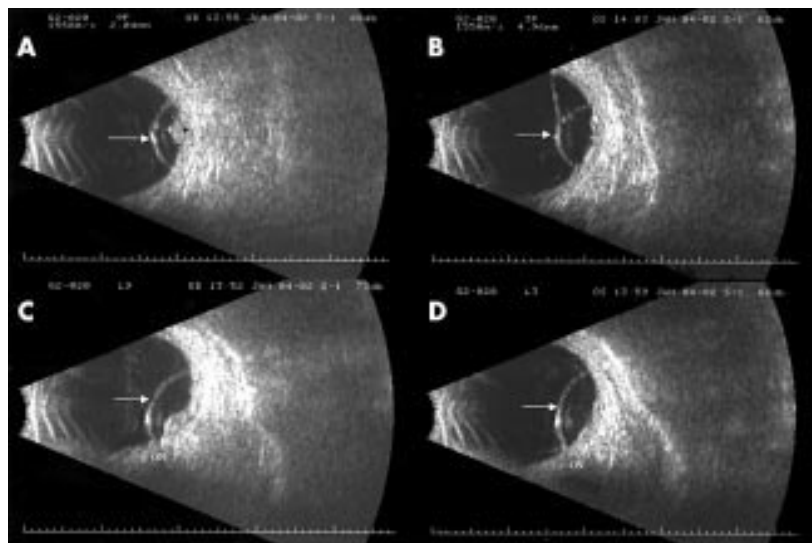
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## Giant neurosensory detachments associated with disciform lesions in neovascular age related macular degeneration

Age related macular degeneration (AMD) is the leading cause of blindness among the population over 65 years of age in Europe and North America.<sup>1,2</sup> Neovascular AMD, which is characterised by choroidal neovascularisation, often leads to severe central vision loss.<sup>3</sup> Choroidal neovascularisation may lead to



**Figure 1** (A, B) Transverse (cross sectional) B scans of both eyes showing bullous retinal detachments (arrows) overlying a localised scar (cross hairs). (C, D) Longitudinal (radial) B scans of both eyes showing bullous detachments (arrows) inserting into the optic disc.

development of fibrous tissue which replaces the normal retina and may be associated with serous or haemorrhagic detachment of the retinal pigment epithelium (RPE) and overlying retina.<sup>3,4</sup> We describe a case of neovascular AMD associated with large, bullous neurosensory detachments overlying bilateral macular disciform lesions.

#### Case report

A 69 year old white man presented to the Vitreoretinal Division at the Wilmer Ophthalmological Institute for evaluation of his macular degeneration. He was diagnosed with macular degeneration by an outside ophthalmologist in 1992. He reported slowly worsening vision in both eyes over many years. He denied any recent changes in his vision. Family history was significant for AMD affecting his father, sister, and brother. He denied history of ocular trauma, surgery, or laser.

On ophthalmological examination, the best corrected visual acuity was "hand movement at 4 feet" in the right eye and 4/200 in the left eye. There was no relative afferent pupillary defect. Extraocular movements were full in each eye. Intraocular pressures were 15 mm Hg in the right eye and 17 mm Hg in the left eye. Anterior segment examination was remarkable for moderate nuclear sclerotic and cortical cataractous changes in each eye. Extended ophthalmoscopy showed cup to disc ratios of 0.3 without evidence of optic nerve head oedema or pallor. The maculae showed disciform lesions in both eyes with overlying large and bullous neurosensory detachments. Shifting subretinal fluid was not identified. Given the extent of the neurosensory elevation, B scan echography was performed in order to quantify these lesions. B scan images showed bullous elevation of the retina in the posterior pole in each eye corresponding to the neurosensory detachments and the localised areas of scar tissue beneath the detachments (Fig 1). The maximum elevation of the neurosensory detachment measured 2.5 mm in the right eye and 5.0 mm in the left eye at the centre of the lesion. The retinal periphery was unremarkable in both eyes.

#### Comment

Previous studies have measured neurosensory detachments in AMD.<sup>5,6</sup> In a study of 16 eyes

with neurosensory detachments secondary to neovascular AMD, the authors found that the average maximal height of the lesions at baseline measured 272  $\mu\text{m}$  by confocal microscopy.<sup>5</sup> The measurements ranged from 146  $\mu\text{m}$  to 584  $\mu\text{m}$ . Using confocal techniques, Bartsch *et al* described a case with a neurosensory detachment secondary to AMD which measured 1300  $\mu\text{m}$ .<sup>6</sup> The use of ultrasound in ophthalmology has increased significantly over the past three decades to encompass a variety of indications.<sup>7</sup> Quantification of lesion dimensions is one aspect that has proved to be a significant tool for documenting findings noted on clinical examination. B scan echography provides two dimensional images to document the topographic features such as shape, location, and extent. Various probe positions (transverse and longitudinal) facilitate accurate delineation of the lateral and radial borders of intraocular lesions.<sup>7</sup> The case illustrated in this report clearly demonstrates that neurosensory detachments associated with neovascular AMD can be significantly larger than previously described. This may contribute to significant loss of central vision. Understanding the pathophysiological mechanisms which determine the maximal elevation of the neurosensory detachments in neovascular AMD may help in designing treatment strategies targeted towards preventing or restricting this process.

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### Successful photodynamic therapy for subretinal neovascularisation due to Sorsby's fundus dystrophy: 1 year follow up

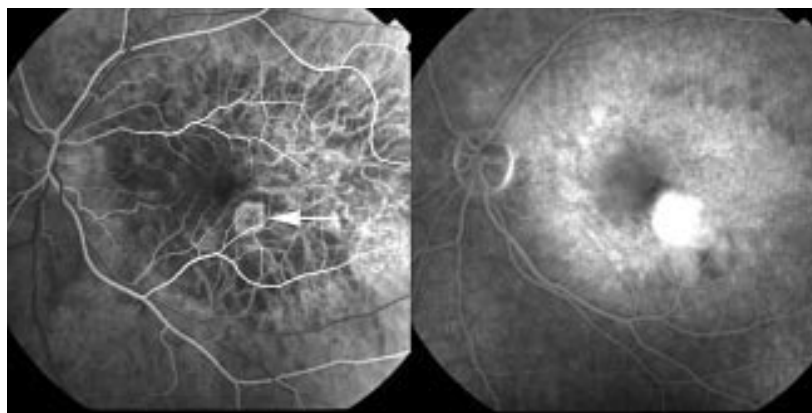
Sorsby's fundus dystrophy (SFD) is a rare but severe autosomal dominant disease. Clinically it is characterised by severe central visual loss, mainly due to submacular choroidal neovascularisation (CNV) during the fourth or fifth decade of life.<sup>1,2</sup> Blindness therefore occurs during the patient's most productive years of employment. We report a case of successful treatment of CNV in SFD with photodynamic therapy (PDT) and verteporfin.

#### Case report

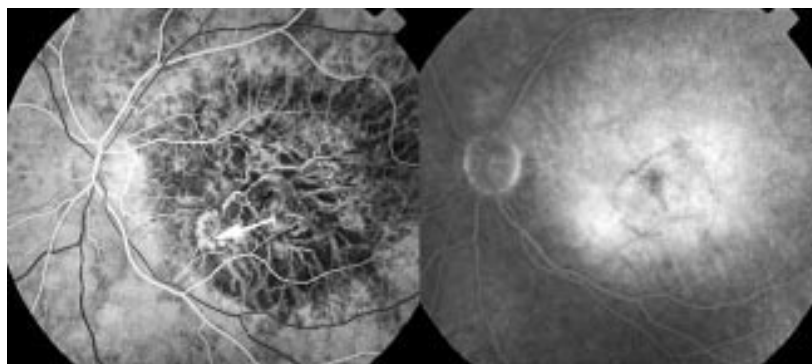
A 40 year old white man (occupation photographer) presented in 1999 with sudden blurring and distortion of vision in the right eye. Visual acuity was 6/6 in the right eye, and 6/4 in the left. Funduscopy and fundus fluorescein angiogram (FFA) demonstrated a large subfoveal CNV. This was deemed unsuitable for laser photocoagulation owing to its location and size. Subsequently, acuity in the right eye deteriorated to 3/60 with the formation of a disciform macular scar. Standard flash electroretinogram (ERG) was normal, while dark adapted ERG was abnormal. Family history revealed that his mother and maternal grandmother went "blind" in their 50s. The patient's cousin had also suffered from recent vision loss. A clinical diagnosis of SFD was made based on the patient's age, family history, and retinal appearance. This was confirmed by molecular genetic assessment. Restriction digest analysis (using *NsiI*) showed that both the patient and his affected cousin were heterozygous for the Ser181Cys mutation in the tissue inhibitor of metalloproteinases-3 (*TIMP3*) gene.<sup>3</sup>

In 2001 the patient reported visual disturbance in his left eye. Visual acuity in the left eye had decreased to 6/36. FFA revealed a left extrafoveal, predominantly classic, CNV (Fig 1). It was known that submacular CNV in SFD responded poorly to conventional argon laser treatment,<sup>4</sup> so we elected to undertake photodynamic therapy (PDT) with verteporfin.<sup>5</sup> The protocol used for treatment was as previously described.<sup>6</sup>

Further PDT treatments were applied to the left macula at 3, 6, and 12 months. These supplemental treatments were prompted by fresh leakage seen on FFA. At 1 year, a small subretinal scar was seen at the site of the original CNV and some leakage was noted at



**Figure 1** Early and late FFA of left eye before PDT showing leakage in a predominantly classic, extrafoveal, choroidal neovascular membrane (arrow).



**Figure 2** Early and late FFA of left eye at 1 year follow up showing staining only in the area of the original neovascularisation and leakage just at the inferonasal edge away from the fovea (arrow).

the inferonasal edge of this scar (Fig 2). Further PDT treatment is planned for this. Visual acuity in his left eye improved from 6/36 to 6/12 and this has been maintained for the 1 year of follow up.

#### Comment

SFD was first described by Sorsby in 1949.<sup>1</sup> Mildly affected patients suffer colour vision deficits and night blindness.<sup>2</sup> In such patients, mid-peripheral drusen are often seen. Histologically, a confluent, lipid containing layer is seen deposited within the inner layer of Bruch's membrane.<sup>7</sup> Consistently, in the fourth to fifth decade of life affected patients suffer sudden, severe vision loss due to CNV. A few experience more gradual vision loss due to macular atrophy. All patients invariably progress to vision loss sufficient for blind registration.<sup>8</sup>

Despite some evidence to suggest improvement in night blindness with vitamin A supplements<sup>9</sup> by far the most significant visual deficit in SFD relates to the complications of CNV. Effective treatment needs to be devised for this. Unlike age related macular degeneration, even if the CNV in SFD is juxtafoveal or extrafoveal, argon laser therapy is ineffective.<sup>4</sup> Also, CNV natural history in age related macular degeneration can result in a variable final visual acuity.<sup>10</sup> In SFD, however, visual prognosis after CNV, particularly when associated with the *TIMP3* Ser181Cys mutation, is always very poor.<sup>8</sup> It is particularly noteworthy therefore that 1 year after treatment, visual acuity has improved and has been maintained at 6/12 when acuity would

be expected to have declined to 6/60 or less. Significantly, this has allowed the patient to continue in his career as a photographer.

This is the first report suggesting that treatment may limit severe visual deficit in an SFD patient and for an extended period. Photodynamic therapy with verteporfin should therefore be considered in other SFD patients when they suffer CNV.

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### Association of HLA type and Mooren's Ulcer in Chinese in Taiwan

We read with interest the article by Taylor *et al.*,<sup>1</sup> suggesting a possible association between HLA-DR17(3) and/or DQ2 and susceptibility to Mooren's ulcer on the basis of cases collected globally, though none were Chinese. We have collected HLA data on cases of peripheral ulcerative keratopathy and investigated the genetic relation between Mooren's ulcer and HLA type in Chinese people.

In total, eight patients with non-infectious peripheral destructive corneal ulcer were treated in our referral clinic. Full systemic and ocular examinations were performed to diagnose Mooren's ulcer. A laboratory examination to rule out the possible rheumatological and infectious causes, included complete blood count with platelet count, serum complement fixation, circulating immune complexes, antinuclear antibodies, rheumatoid factor, anti-neutrophil cytoplasmic antibodies, erythrocyte sedimentation rate, C reactive protein, rapid plasma reagent/fluorescent treponemal antibody absorption test, antibodies of herpes simplex, herpes zoster, and *Toxoplasma*, hepatitis B and C tests, liver function tests, blood urea nitrogen and creatinine, fasting blood sugar, urinalysis, chest x ray, sinus x ray, and kidney, ureter, and bladder x ray (KUB) study. Complete ocular evaluations included slit lamp microscopy, conjunctival and corneal swabs for cultures of possible infective agents, and tear function tests such as Schirmer's test and tear break up time (TBUT). All of our patients were Chinese and two were given the diagnosis of Mooren's ulcer. Both patients had a normal other eye, and were otherwise healthy, except for previous hepatitis B infection, which is very common (up to 90% in those more than 40 years old) in Taiwan.

#### Case report

##### Patient 1

A 67 year old woman presented with a 3 week history of a painful, tearing and a photophobic right eye in June 2002. Slit lamp biomicroscopy revealed an inferior peripheral corneal ulcer and adjacent conjunctival injection of her right eye. This crescent shaped ulcer caused thinning to 30% of the corneal thickness, thereby weakening the central edge of the inferior peripheral cornea. In addition,

**Table 1** HLA class I and II types of two Mooren's ulcer patients

HLA typing	Patient 1	Patient 2	Antigen frequencies (%) of Chinese in Taiwan
HLA-A		A2	28.82–32.81
		A11	18.00–36.06
	A24		14.88–19.76
	A33		8.72–11.56
HLA-B		B46	8.55–17.23
	B54		1.61–4.32
	B61		2.71–3.77
		B75	1.64–5.40
		BW6*	
HLA-DR		DR4	11.69–15.41
		DR9	13.56–16.36
		DR16	1.31–5.44
		DR17(3)	<b>0.98–8.37</b>
		DR51†	
HLA-DQ		DR53‡	
	DQ2		<b>7.51–15.66</b>
	DQ4		5.03–9.88
		DQ5	<b>10.60–21.50</b>
	DQ9		4.31–13.06

\*BW6 associations: B46, B54, B61, B75; †DR 51 associations: DR15, DR16, DR1; ‡DR53 associations: DR4, DR7, DR9.

overlying epithelial defect was noted by fluorescence staining.

#### Patient 2

A 60 year old woman was referred for a painful, red right eye with incipient peripheral corneal perforation of 3 months' duration. She reported a history of extracapsular cataract extraction of her right eye 8 months before, in November 2001. On examination, there was marked thinning of the right superior cornea from 10 to 2:30 o'clock with pannus and an infiltrated leading edge. Within the marginal ulcer, around 90% of the areas was thinned to 10% of the corneal thickness. Rheumatological evaluation was normal. This ulcer perforated 4 days after admission and emergency repair with multilayered amniotic membrane covered with a conjunctival graft was performed smoothly. Afterwards the destruction of peripheral corneal stroma ceased to progress and the anterior chamber was reformed 3 days after surgery.

Blood samples of these patients were obtained and tested for HLA-A, B, C, DR, and DQ typing by the polymerase chain reaction (PCR). Specific sequence primer (PCR-SSP)<sup>2</sup> low resolution method. HLA-A, B, C, DR were tested using One Lambda (One Lambda Inc, Canoga Park, CA, USA) Micro SSP genetic HLA class I and II typing trays. HLA-DQ was tested by using Dynal all set typing trays (Dynal Biotech Ltd, Warral, UK). The HLA types of these two Mooren's ulcer patients are listed in Table 1. HLA phenotype frequency data of the Chinese population in Taiwan were obtained from recently published data.<sup>3</sup>

#### Comment

According to Craig's report, 10 of 12 Mooren's ulcer patients (83%) were HLA-DR17(3) and/or HLA-DQ2 positive. According to published population studies, the HLA-DR17(3) antigen frequencies are 4–19% in India, 10–20% in black South Africans, and 23% in white northern Europeans. The HLA-DQ2 antigen frequencies are 36–45% in India, 17–19% in black South Africans, and 33% in white northern Europeans.<sup>4</sup> These findings

suggest predisposition of HLA-DR17(3) and HLADQ2 might have some significant association with susceptibility to Mooren's ulcer.

The HLA-DR17(3) and DQ2 antigen frequencies for Chinese people are 1–8% and 7–15%, respectively. If we combine the data of our two female Chinese Mooren's ulcer patients with those of patients in Craig's study, we find that 11 of 14 (78.5%) patients with Mooren's ulcer are HLA-DR17(3) and DQ2 positive, which is still higher than in ethnically matched control populations. In Craig's article, 100% of non-white Mooren's ulcer patients are HLA-DR17(3) and DQ2 positive, but if our patients are included in this assessment, the frequency decreases to 90% of non-white patients.

Another interesting finding was the increased frequencies of HLA-DQ5. In the Mooren's ulcer group, HLA-DQ5 was found in 50% patients, whether or not our data and Craig's are considered as a whole. The HLA-DQ5 antigen frequencies are 21–25% in Indian people, 13–22% in black South Africans, 10–32% in white northern Europeans,<sup>4</sup> and 10–21% in Chinese. Therefore, our data support the possible linkage of HLA-DR17(3), HLA-DQ2 gene with Mooren's ulcer proposed by Craig's article, and suggest HLA-DQ5 might be another candidate gene of HLA associated with Mooren's ulcer.

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#### Corneal endothelial deposits secondary to rifabutin prophylaxis for *Mycobacterium avium* complex bacteraemia

We report a case of corneal endothelial deposits in a patient positive for human immunodeficiency virus (HIV) who had received rifabutin prophylaxis for *Mycobacterium avium* complex bacteraemia.

#### Case report

A 50 year old man was referred to the corneal clinic with bilateral scattered endothelial deposits. He was asymptomatic at the time of presentation.

His history indicated that he had been HIV positive since 1992 and had been commenced on treatment in 1995. Since then he had suffered from tuberculosis and pneumonia but there was no history of any eye problems.

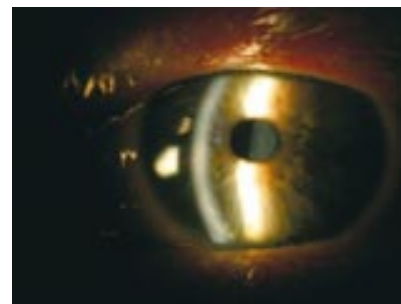
His systemic health was currently good and his CD4 count was 540 cells × 10<sup>6</sup>/L.

His ophthalmic history revealed loss of vision in the right eye in 1986 following an episode of herpes zoster in this eye.

On examination his right visual acuity was no perception of light and his left visual acuity was 6/9. Both eyes were white. Corneal examination revealed bilateral endothelial deposits, scattered through out the cornea, stellate in the middle but more confluent in the periphery (Fig 1).

There was no associated uveitis. The intraocular pressures were within normal limits. There were posterior synechiae and a white cataract in the right eye, which precluded any fundal view. The left eye had a clear lens and fundal examination was entirely normal.

A detailed history of his medications indicated that he had received rifabutin for 2 years but had been off this treatment for 18 months before his referral to the eye clinic.



**Figure 1** Example of bilateral endothelial deposits.

Serial photography over the past 9 months has not shown any change in the appearance of these deposits.

### Comment

Rifabutin is used to prevent *Mycobacterium avium* complex (MAC) disease in patients with HIV and CD4 counts of less than 100 cells  $\times 10^6/l$ .<sup>1</sup> Rifabutin causes inhibition of DNA dependent RNA polymerase in sensitive strains of *Escherichia coli* and *Bacillus subtilis*. However, its mode of action against *M avium* is unclear.<sup>2</sup>

It has been associated with uveitis, which may be difficult to differentiate from other causes of uveitis in patients with AIDS.<sup>3,4</sup>

Uveitis is unusual at the recommended oral dosage of 300 mg/day, but becomes common as the total daily dose approaches 1 g.<sup>5</sup>

Corneal endothelial deposits secondary to treatment with rifabutin have been reported in children positive for HIV.<sup>6</sup>

The deposits are usually bilateral and initially peripheral and stellate. Of interest is the fact that these deposits occur without any associated uveitis. They increase in number with continued administration of rifabutin but appear not to be sight threatening.

This case demonstrates that these endothelial deposits do not appear to resolve upon termination of rifabutin therapy in the short to medium term. A longer period of observation is required to determine if these deposits alter in the long term.

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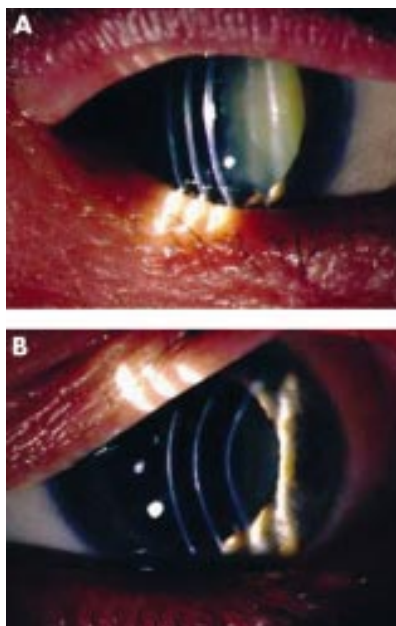
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### Corneal ectasia following deep lamellar keratoplasty

Keratoconus is a bilateral non-inflammatory corneal ectasia with an incidence of approximately one per 2000 in the general population.<sup>1</sup> Contact lenses are the most common treatment. When contact lenses fail, a surgical approach is necessary for visual rehabilitation. Penetrating keratoplasty has been the traditional and most common mode of treatment and has excellent results.<sup>2,3</sup> However, more recently, deep lamellar keratoplasty



**Figure 1** Anterior segment photographs comparing both eyes with deep lamellar keratoplasty, the left eye (B) shows marked apical thinning with subepithelial scarring and nuclear sclerotic cataract in the right eye (A).

(DLK) is gaining popularity as an alternative option for the surgical management of keratoconus.<sup>4</sup> It has obvious advantages in that endothelial rejection is rare and it is essentially an extraocular procedure.<sup>5</sup>

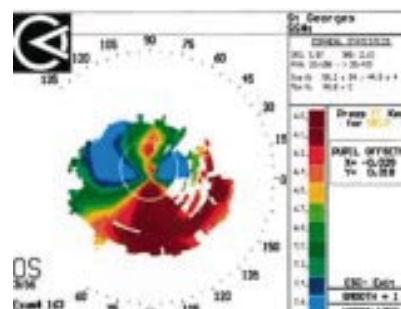
Recurrent keratoconus following penetrating keratoplasty is rare but has been described.<sup>6,7</sup> We report on the first case of recurrent ectasia following deep lamellar keratoplasty supported by clinical and histological evidence.

### Case report

A 38 year old chronic schizophrenic male was referred to the anterior segment clinic with advanced bilateral keratoconus. He had previously been treated with hard contact lenses. His condition had deteriorated over the years and he was now keen on surgical intervention. On examination, visual acuity was counting fingers in both eyes with no improvement with pinhole. Anterior segment examination revealed bilateral advanced cones with subepithelial scarring. Fundal examination was unremarkable.

The patient underwent uneventful right lamellar keratoplasty with lyophilised corneal tissue under general anaesthetic. The left eye had the same procedure with intraoperative Botox injection to the upper lid 1 year later. Again, the procedure was uneventful but postoperatively, he developed a persistent central epithelial defect which later became infected. There was no improvement in the patient's condition despite intensive antibiotics and steroids and the patient underwent an emergency left lamellar keratoplasty 1 month later. Twelve months postoperatively, corrected VA had improved to 6/60 right eye and 6/18 left eye. Topographical data and refraction were not done at this time and he was noted to have bilateral lens opacities.

Three years later the patient was reviewed with decreased vision in both eyes. On examination, unocular visual acuity was counting fingers and hand movements in the right and



**Figure 2** Corneal topography of the left eye showing inferior steepening of the vertical meridian following deep lamellar keratoplasty consistent with recurrence of ectasia.

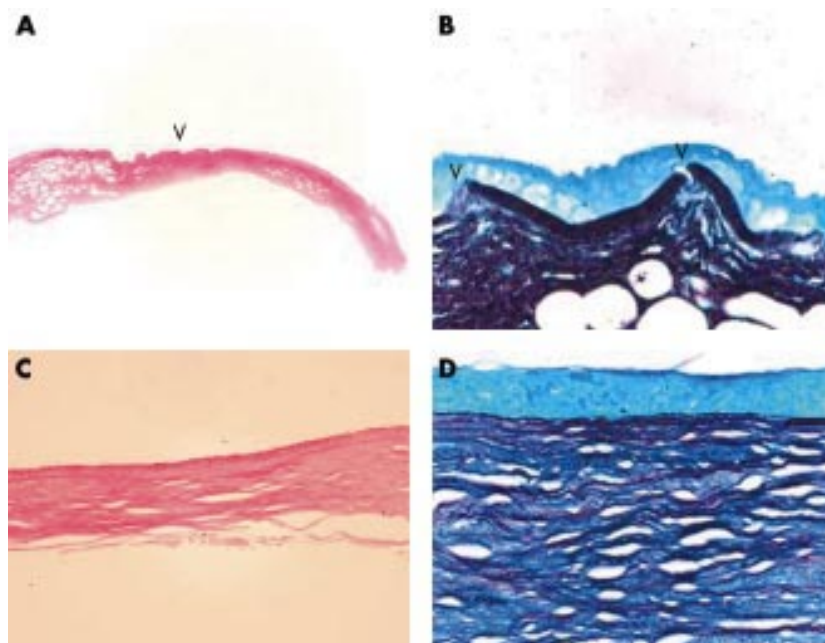
left eye, respectively. Anterior segment examination revealed bilateral central subepithelial corneal opacities more pronounced in the left eye with severe apical thinning (Fig 1). He had bilateral nuclear cataracts more marked in the right eye. Corneal topography demonstrated marked inferior steepening in the left eye consistent with keratoconus (Fig 2). A repeat DLK was performed on the left eye and histology from the second grafted corneal button showed degenerative thinning consistent with ectasia. The original host tissue in comparison revealed breaks in Bowman's membrane which is typical of keratoconus (Fig 3). At his last clinic visit 1 month later, having undergone bilateral cataract extraction and IOL implantation in the preceding year, his visual acuity was 6/9–2 right eye with +3.25/–8.00 $\times 140$  correction and 6/9 left eye with +4.00/–4.50 $\times 180$ .

### Comment

Lamellar keratoplasty (LK) has been an established procedure for corneal pathology for over a 100 years. Advances in surgical techniques such as deep lamellar anterior keratoplasty have expanded the application of lamellar surgery and have achieved visual results approaching those of penetrating keratoplasty while reducing the rate of rejection and improving the long term graft stability.<sup>4</sup> The procedure can be defined as the excision of superficial stromal layers. A number of techniques have been used for dissection of the stroma such as air, viscoelastic, and fluid injection.<sup>9–12</sup> The entire stroma can be completely excised so that only Descemet's membrane and endothelial cells remain.

Studies have shown that in deep lamellar keratoplasty, endothelial rejection reaction is rare with cell counts being maintained for a longer period.<sup>5</sup> This confers obvious advantages over penetrating keratoplasty in the treatment of keratoconus. However, it is still a relatively new procedure and is technically more challenging.

There have been a few cases in the literature of recurrent keratoconus following penetrating keratoplasty successfully treated with regrafting.<sup>6,7</sup> In all these cases, the pathogenesis of this complication was unclear. In our patient, ectasia recurred in the left eye 3 years after deep lamellar keratoplasty and this was confirmed both clinically and histologically. Donor factors include the possibility of ectatic disease which may have been missed or remained subclinical throughout the donor's life. New screening methods utilising the Orbscan are being explored looking at the topography of donor corneas that could



**Figure 3** Histopathology slides of the original host keratoconic cornea showing apical thinning and subepithelial scarring in the low magnification (A) High magnification with Jones stain shows disruption of Bowman's membrane (B). In comparison, the deep lamellar keratoplasty corneal button shows apical thinning (C) but no disruption of Bowman's membrane (D). Low power (haematoxylin and eosin stain) high power (Jones stain).

prevent potential problems with using ectatic corneas if routinely employed.<sup>13</sup>

Another consideration is that the cryo-lathed corneal lenticule used at the time of repeat surgery may have been inherently thin as the procedure was done as an emergency. Adequate preparation of the donor tissue is therefore necessary before surgery. The inflammatory pathways activated following the first DLK failure due to infection, in particular the metalloproteinase system (gelatinolytic activity of stromal collagenase (matrix metalloproteinase-1 (MMP-1)), may play an important part through thinning of the stromal tissue.<sup>14</sup>

These lenticules are devoid of keratocytes; invasion of the graft by host keratocytes, which may be metabolically prone to producing abnormal corneal architecture, may contribute to ectasia.

In summary, we have reported the first case of recurrent ectasia in a relatively new treatment option—deep lamellar keratoplasty for keratoconus. Protection of the lamellar graft from infection and inflammation is important in order to obtain the best visual potential. Preoperative and postoperative data such as refraction, topography, and pachymetry are vital in monitoring progression of these patients. As lamellar surgery becomes increasingly popular it is important to recognise such late complications which may require further surgical intervention in the future.

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### Opacification of SC60B-OUV lens implant following routine phacoemulsification surgery: case report and EM study

In 1949, Sir Harold Ridley implanted the first artificial intraocular lens (IOL) to reduce refractive error following cataract extraction.<sup>1</sup> Numerous designs of IOL implants have followed and a variety of materials have been used in their manufacture, including poly(methyl methacrylate) (PMMA), silicone, acrylic, and hydrogel based materials. Important requirements of IOL implant material are to not excite an inflammatory response and the ability to remain transparent within the eye for an extended period of time. In recent years, there have been reports of opacification of IOL implants such as calcification on the optical surface of the Hydroview lens<sup>2</sup>; “glis-tenings” of fluid filled vacuoles in the optic of the AcrySof IOL<sup>3,4</sup>; and “snowflake” crystal-line opacification of three piece rigid PMMA lenses.<sup>5</sup>

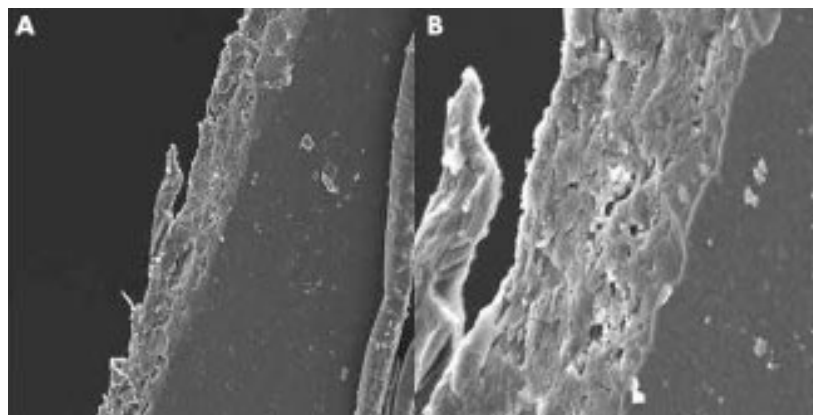
In particular, late postoperative opacification of a particular hydrophilic acrylic IOL, the SC60B-OUV, has been reported<sup>6</sup> and analysis of these explanted IOLs have shown the presence of granular deposits within the optic.<sup>7</sup> We report examination, using electron microscopy, of a similar explanted IOL removed following late postoperative opacification, which appears to have different surface morphology from those reported previously.

#### Case report

An 82 year old female patient with Fuchs' endothelial dystrophy underwent uneventful phacoemulsification and foldable lens implantation into the capsular bag of the left eye. Two weeks later, the best corrected visual acuity was 6/9. Fifteen months later, she underwent a similar procedure with a different foldable lens in the right eye leading to a visual outcome of 6/9. At that time, the left visual acuity had dropped to 6/18 and red reflex assessment of the dilated eye with a direct ophthalmoscope was very similar to that of a senile nuclear cataract. On slit lamp examination, the intraocular lens optic was found to have become uniformly cloudy (Fig 1). The patient was offered a lens exchange procedure and this was carried out 6 months later. Extensive capsular fibrosis and capsular dehiscence meant that the lens could not be explanted in one piece. The



**Figure 1** Slit lamp examination of opacified IOL.



**Figure 2** SEM examination of explanted IOL showing 5 µm thick degraded layer of outer surface. (A) ×2000, (B) ×7000.

haptics were left in situ and the optic was transected before explantation. Anterior vitrectomy and peripheral iridectomy were carried out and an anterior chamber implant was inserted. Five months postoperatively, her vision had recovered to 6/9.

The explanted lens underwent detailed examination at the School of Pharmacy and Biomolecular Sciences, University of Brighton. The surface and the interior portion of the explanted (test) lens were examined and compared to an identical unused SC60B-OUV (control) lens. Fourier Transformed infrared (FT-IR) spectroscopy was performed using a diamond attenuated reflectance unit on a Perkin-Elmer 1620 spectrophotometer under control pressure. For analysis by scanning electron microscopy (SEM), both the control and test lens were cut to produce cross sections to enable the visualisation of the interior and exterior surfaces. All sections were then sputter coated with palladium and photographed at ×20, ×2000, and ×7000 magnification using a Joel JSM 6310 scanning electron microscope.

#### Comment

The opaque lens was a 12.5 mm SC60B-OUV (manufactured and distributed by Medical Developmental Research Inc, USA). The lens is hydrophilic in nature and is a composite of poly(2-hydroxyethyl methacrylate) (HEMA) and PMMA with a polymerisable ultraviolet absorber. The source of the polymer was Vista Optics, UK.

The FT-IR spectra showed identical typical hydroxyl and carbonyl stretching adsorption bands for both test and control lenses. Examination of the control lens by SEM at ×20 and ×2000 magnification showed smooth, unblemished inner and outer surfaces. The inner and outer surfaces of the test lens did not appear different from the control lens when viewed at ×20 magnification. At ×2000 the outer (opaque) surface of the test lens was markedly different from the interior portion. A 5 µm thick section of the outer surface appeared to have degraded giving a sponge-like morphology (Fig 2).

The dimensions of this degraded portion only occurred within the visibly opaque section of the explanted lens and the degradation did not extend across the transected surface.

The FT-IR spectroscopy indicated that there were no significant differences in the surface chemistry of the original and explanted lens. From the SEM results it can be concluded that the opacity observed on the lens removed

from the patient, corresponds to a 5 µm thick interfacial layer. The structure observed is typical of high water component, swollen hydrogel systems such as poly(HEMA) polymerised in 80% water. Such damage may therefore have been caused by slow degradation of the polymer matrix or dissolution of unpolymerised monomer/oligomers and swelling of incompletely polymerised material in the core of the optic.

These findings are different to those previously reported by Werner *et al* who found granular deposits in a region beneath the anterior and posterior surfaces with intact surface structure. The time frame for the appearance of the opacification and clinical description appears to be equivalent in both studies. The reason for the difference in SEM findings is unclear. If our findings represented an earlier phase of the same degenerative process then one would expect some residual surface degeneration in their study. Conversely, if ours is a later phase then one might expect the presence of granular deposits in the substance of the optic.

Although our lens showed the same clinical appearance of postoperative opacification before explantation as other studies, the EM results suggest that our findings may represent a different degenerative process. The nine explanted lenses examined by Werner *et al* came from the same surgeon in Turkey, and the type of opacification may be due to a "batch" effect as well as a polymer effect. In conclusion our findings may represent a different degeneration in IOL structure to that previously described.

The manufacturer has withdrawn all SC60B-OUV IOLs made from materials obtained from Vista Optics, UK and these IOLs are now being manufactured by polymer from a new source (Benz Research, USA). Out of 12 patients who received this IOL at our institution, seven experienced significant clouding, three have corrected vision of 6/12 or better, and two have died. Of the seven patients with significant clouding, two have undergone exchange, two await exchange, two are considering exchange, and one declined exchange. Figures from the Medical Devices Agency (MDA) state that of 3200 lenses distributed in the United Kingdom, only 27 reports of clouding have been received. There may be under-reporting of cases and we encourage reporting of all cases to the MDA. Vigilance is clearly necessary with this IOL to ensure that the change in polymer manufacture has resolved the problem.

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#### Isolated foveal retinoschisis as a cause of visual loss in young females

Foveal or macular retinoschisis is an uncommon retinal disorder, usually seen in patients affected with generalised retinal disease such as X linked retinoschisis,<sup>1</sup> Goldmann-Favre syndrome,<sup>2</sup> and enhanced S-cone syndrome.<sup>3</sup> There have been a handful of previous reports of patients exhibiting foveal retinoschisis in whom there appeared to be limited concomitant peripheral retinal disease,<sup>4-6</sup> suggesting the existence of a distinct disorder. We report the clinical findings in four female patients presenting with a reduction in central acuity and exhibiting isolated bilateral foveal retinoschisis, and investigations including scanning laser ophthalmoscopy (SLO) autofluorescence imaging, optical coherence tomography (OCT), and electrophysiology.

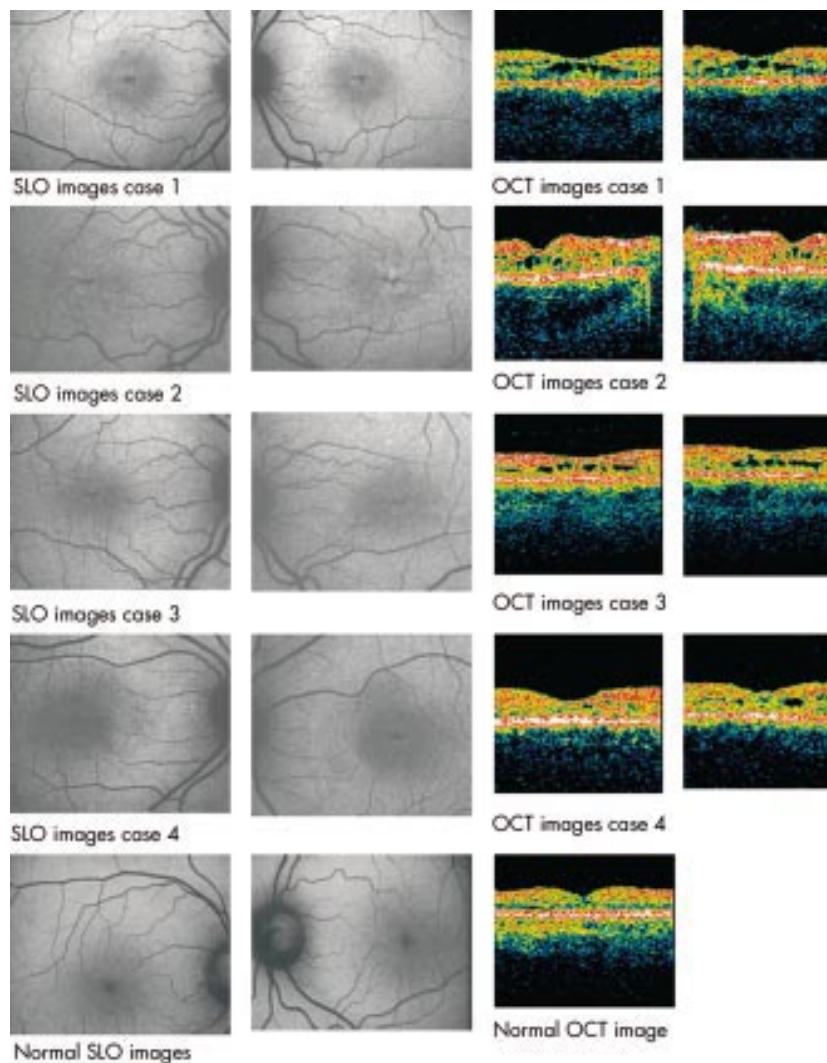
#### Case reports

##### Case 1

A 17 year old girl presented with bilateral reduction in central vision. With refraction (+0.50DS right, +0.25DS left) her visual acuity was 6/18. On examination the only abnormal finding was thickening of the neurosensory retina at the fovea with a radial pattern of striae bilaterally. There was no leakage suggestive of macular oedema on fluorescein angiography.

##### Cases 2 and 3

Female dizygotic twins 19 years of age both reported a mild non-progressive reduction in



**Figure 1** A prototype Zeiss confocal scanning laser ophthalmoscope (SLO) recorded autofluorescence images using argon laser blue light and a broadband pass barrier filter with a short wavelength cut off at 521 nm. Single line bilaminar scans of the macular retina were performed using the OCT 2000 scanner (Zeiss Humphrey Instruments, San Leandro, CA, USA).

their central vision. Best corrected visual acuities ranged from 6/12 to 6/18. On examination the only abnormal finding was thickening of the neurosensory retina at the foveal retina in all four eyes. Fluorescein angiography showed no evidence of macular oedema.

#### Case 4

A 22 year old girl presented with difficulties for near vision over the previous 4 years. Best corrected visual acuity was 6/12 with each eye with a small hyperopic correction. The only abnormal findings on ophthalmoscopy were fine cystoid retinal changes at both foveae and small areas of retinal pigment epithelium (RPE) depigmentation at the maculae. Fluorescein angiography showed small areas of transmission defect with increased fluorescence within the foveal avascular zone of both eyes; there was no leakage suggestive of macular oedema.

In none of the three families was there any family history of eye disease. Moreover, the six parents were examined and all found to have normal acuities and retinal macula appearance. The results of SLO autofluorescence

imaging and OCT imaging are shown in Figure 1. In all cases, there was a radial hyperautofluorescence at the central macula, within an otherwise uniform normal retinal signal. The splitting of the neurosensory layer at the central macula was confirmed on OCT imaging. The ganzfeld ERGs were normal and the pattern electroretinogram (PERG) subnormal in amplitude in all four patients (Fig 2).

#### Comment

All four patients have localised central retinal disease as confirmed by electrophysiology, and are therefore distinct from those cases with generalised retinal disorders listed above, as well as the reported families with inherited macular oedema.<sup>7</sup> Similarly, they are unlikely to be manifesting homozygotes, or XO cases of X linked retinoschisis. The morphology of the central retina in each of the cases does seem to be identical to the findings in hemizygotes affected by X linked retinoschisis.<sup>8</sup>

Instead, these cases closely resemble clinically the patients described by Lewis *et al*<sup>9</sup> and Lorenz *et al*.<sup>5</sup> When viewed collectively it is likely that this disorder is autosomal recessive

in inheritance. It is of interest that all eight cases are female. This might represent a chance finding (this is unlikely:  $p = 0.016$ , considering the identical twins in the report by Lewis *et al*<sup>9</sup> as one case), a real underlying sex difference in the prevalence of this rare condition, or the under-reporting of similar male cases as a result of their assignment to a diagnosis of XLRs. There are a few other cases in the literature demonstrating a similar foveal appearance with minimal peripheral changes<sup>6,9,10</sup> which may be manifesting a different disorder. It is very difficult to predict the long term prognosis for our patients, as such cases are rare and longitudinal data are unavailable. Future genetic analysis, such as screening for novel mutations in NR2E3, the gene responsible for enhanced S-cone syndrome, may shed light on the aetiology of this rare disorder.

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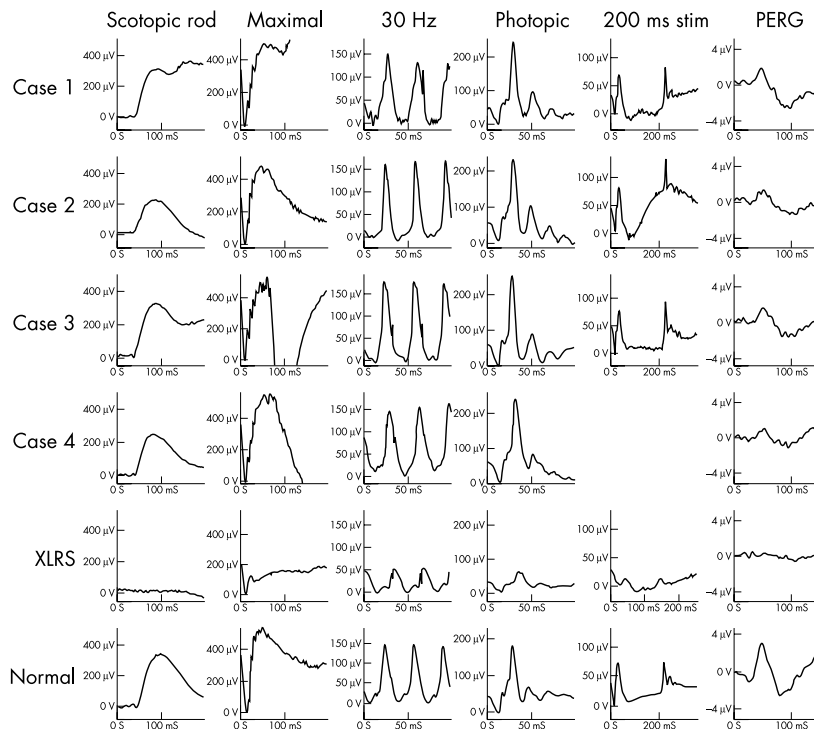
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**Figure 2** Full field electroretinogram (ERG) and pattern ERG were recorded using standardised methods according to International Society for Clinical Electrophysiology of Vision (ISCEV) standards. Long duration photopic stimulation was performed to separate ON and OFF photopic pathways.

## NOTICES

### Monitoring cataract surgical outcomes

The latest issue of *Community Eye Health* (No 44) discusses the monitoring of cataract surgical outcomes in the Third World. 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: [eyesource@ucl.ac.uk](mailto:eyesource@ucl.ac.uk); website: [www.jceh.co.uk](http://www.jceh.co.uk)). Annual subscription (4 issues) UK£25/US\$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: [eyesource@ucl.ac.uk](mailto:eyesource@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's website ([www.secondsight.org.uk](http://www.secondsight.org.uk)) or by contacting Dr Lucy Mathen ([lucymathen@yahoo.com](mailto:lucymathen@yahoo.com)).

### Specific Eye ConditionS (SPECS)

Specific 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 50 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 website [www.eyeconditions.org.uk](http://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](mailto:k@eyeconditions.org.uk); website: [www.eyeconditions.org.uk](http://www.eyeconditions.org.uk)).

### The British Retinitis Pigmentosa Society

The British Retinitis Pigmentosa Society (BRPS) was formed in 1975 to bring together people with retinitis pigmentosa and their families. The principle aims of BRPS are to raise funds to support the programme of medical research into an eventual cure for this hereditary disease, and through the BRPS welfare service, help members and their families cope with the everyday concerns caused by retinitis pigmentosa. Part of the welfare service is the telephone help line (+44 (0)1280 860 363), which is a useful resource

for any queries or worries relating to the problems retinitis pigmentosa can bring. This service is especially valuable for those recently diagnosed with retinitis pigmentosa, and all calls are taken in the strictest confidence. Many people with retinitis pigmentosa have found the Society helpful, providing encouragement, and support through the Help line, the welfare network and the BRPS branches throughout the UK (tel: +44 (0)1280 821 334; email: [lynda@brps.demon.co.uk](mailto:lynda@brps.demon.co.uk); website: [www.brps.demon.co.uk](http://www.brps.demon.co.uk)).

### Surgical Eye Expeditions International

Volunteer ophthalmologists in active surgical practice are needed to participate in short term, sight restoring eye surgery clinics around the world. Contact: Harry S Brown, Surgical Eye Expeditions International, 27 East De La Guerra, C-2, Santa Barbara, CA 93101-9858, USA (tel: +805 963 3303; fax: +805 965 3564; email: [hsbrown.md@cox.net](mailto:hsbrown.md@cox.net) or [seintl@seintl.org](mailto:seintl@seintl.org); website: [www.seintl.org](http://www.seintl.org)).

### MSc course in Community Eye Health

The International Centre for Eye Health is offering a full time MSc course in Community Eye Health from 29 September 2003 to 19 September 2004. The course is not clinical and is specifically for eye health professionals wanting to work in the field of community eye health. The course is designed in keeping with the aims, priorities, and strategies of Vision 2020—the Right to Sight. The course costs £3939 for home students and £14 110 for overseas students. Further information: The Registry, 50 Bedford Square, London WC1B 3DP, UK (tel: +44 (0)20 7927 2239; fax: +44 (0)20 7323 0638; email: [Adrienne.Burrough@lshtm.ac.uk](mailto:Adrienne.Burrough@lshtm.ac.uk); website: [www.lshtm.ac.uk](http://www.lshtm.ac.uk)).

### Institute of Ophthalmology: Professor Alan Bird's 65th Birthday Meeting

The Institute of Ophthalmology is holding a meeting to celebrate Professor Alan Bird's 65th Birthday on 10-11 July 2003, at The Beveridge Hall, Senate House, University of London, Malet Street, London. Session one on Retinal Dystrophies will be chaired by Professor Tony Moore and session two on The Ageing Macula will be chaired by Professor Steve Ryan. It is expected that CME credit will be awarded. Admission is free. Places for the meeting are limited and booking is essential. There will also be a dinner held on Thursday evening for guests and partners, the cost is £40 (US\$62). Further details: Miss Laura Short, Institute of Ophthalmology, 11-43 Bath Street, London EC1V 9EL, UK (register on the website: [www.ucl.ac.uk/foo](http://www.ucl.ac.uk/foo)).

### Glaucoma Society 24th Annual Meeting and Dinner

The Glaucoma Society 24th Annual Meeting and Dinner will take place on 20 November 2003, from 8:30 am to 5:00 pm at The Royal College of Physicians, London, UK. Further details: Ms Janet Flowers (email: [glausoc@ukeire.freeserve.co.uk](mailto:glausoc@ukeire.freeserve.co.uk)).



## Successful photodynamic therapy for subretinal neovascularisation due to Sorsby's fundus dystrophy: 1 year follow up

S C Wong, K C S Fong, N Lee, et al.

*Br J Ophthalmol* 2003 87: 796-797

doi: 10.1136/bjo.87.6.796

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Updated information and services can be found at:

<http://bjo.bmj.com/content/87/6/796.full.html>

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### Notes

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