LETTERS TO THE EDITOR

Unilateral congenital mydriasis

EDITOR,—We read with interest the report by Richardson and Schulenburg¹ describing a patient with congenital mydriasis, and would like to contribute such another case to the literature.

In June 1992, a 4-year-old boy who had suffered photophobia in the left eye since infancy was referred to our hospital. His medical and family histories were unremarkable. Visual acuity was 20/20 right eye and 20/25 left eye. Ocular motility was normal. Both pupils were round, but the pupillary diameters were 3.5 mm right eye and 7.5 mm left eye in a lighted room. The right pupil reacted briskly to light and accommodation, whereas the left pupil reacted to neither stimulus as confirmed by infrared pupillometry. The other ophthalmic, neurological, and general examinations were all normal. Additional tests, including a head computed tomography scan, were normal. On pharmacological testing with 1% pilocarpine, the right pupillary diameter decreased from 3.5 to 2.5 mm, but the left diameter did not change. With 1% cyclopentolate the right pupillary diameter increased from 3.5 to 6.5 mm, and the left diameter increased from 7.5 to 8.5 mm. The refractive values increased from +1.25 to +2.5 D in the right eye, and from +1.75 to +3.00 D in the left; but, after pretreatment with a topical prostaglandin inhibitor (indomethacin), the left pupillary diameter did not increase owing to cyclopentolate. Infrared video transillumination in the sphincter zone showed no distinctive difference between the eyes.

A young boy was found to have an isolated unilateral fixed dilated pupil which was most probably congenital. All such patients reported¹ have been female and were affected bilaterally. A male patient with Waardenburg syndrome and a congenital unilateral mydriasis has been reported.²

The affected pupil in our patient did not react to light, accommodation, or a miotic. The ciliary muscle, however, functioned normally because of normal refraction change owing to the cycloplegic. This is evidence that the oculomotor nerve was intact. The affected pupil did react to cyclopentolate to some extent, and mydriasis was prevented by the prostaglandin inhibitor. Observation of the iris sphincter zone by infrared video did not reveal a distinct transparency. These facts indicate that the sphincter iridis exists morphologically but that its function is reduced. Its pathogenesis should be confirmed pathologically.

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Spontaneous hyphaema associated with anterior uveitis

EDITOR,—D S Fong and M B Raizman¹ presented five cases of spontaneous hyphaema in a variety of uveitides. The authors are correct in asserting that such hyphaemas are an uncommon accompaniment to anterior uveitis, and discuss the possible pathogenesis of such bleeding.

While hyphaema has occurred as a sequel to severe anterior uveitis of various causes, there also appears to be a particular association with Fuchs' heterochromic uveitis (FHU), a subject which was not touched on by the authors. Amsler² was the first to draw attention to the apparent intraocular vascular abnormality that led to hyphaema on paracentesis in Fuchs' disease, and this subtle but virtually universal sign, seen at the commencement of cataract surgery in FHU, now bears his name. Such bleeding is usually seen in the anterior chamber angle, but in some cases may occur from multiple sites on the anterior iris surface.

It is clear that not only a sudden decrease of intraocular pressure (as during paracentesis) but other forms of ocular trauma can induce hyphaemas in FHU. Such events have been reported after use of the Honan balloon, gonioscopy, applanation tonometry, and even after mydriasis.³ It is merely an extension of these phenomena that leads to the observation of 'spontaneous' hyphaema in some patients. Whether such events are truly spontaneous cannot be proved. On the contrary, it seems likely that in some cases the fragile angle vasculature in this disease is unable to maintain its integrity during what would normally be considered physiological changes in intraocular pressure during minor trauma (such as eye rubbing or wiping).

Liesegang⁴ reported that four of a cohort of 59 (6.8%) patients had episodes of spontaneous bleeding, and in a cohort of 151 patients with FHU in Manchester, we have observed this complication in six patients (4%). In two of the latter instances this was associated with frank rubeosis. In the remaining four cases no gonioscopic vessel abnormality was seen. Typically such patients present with sudden blurred vision with or without eye ache. In one instance acute glaucoma accompanies each episode.⁵ Signs of hyphaema may be subtle—a dusting of cells on corneal endothelium, and frank hyphaema visible only by gonioscopy. It would be interesting to speculate on the possible pathogenetic association between recurrent microscopic hyphaema and the development of glaucoma in some patients with FHU. One could also speculate that, as episodes of blurred vision and eye ache are not uncommonly reported retrospectively by FHU patients, that episodes of spontaneous microscopic hyphaema may be more common than we have observed.

Though Fong and Raizman identify a variety of uveitides as a cause of spontaneous hyphaema, and discuss common causes, it appears that FHU has a particular predilection for this phenomenon, for reasons unassociated with either rubeosis or iris hyperaemia. It certainly accounts for the majority of instances seen in the uveitis clinic at this hospital.

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Reply

EDITOR,—We would like to thank Nicholas Jones for reminding us that hyphaema can be seen in Fuchs' heterochromic uveitis. As he pointed out, hyphaema in FHU can be seen following a variety of conditions, such as minor trauma. However, it often is seen in the absence of inflammation.¹ In these patients, the mechanism of hyphaema is likely leakage from abnormal vessels. As discussed in our article,² Perry *et al*³ has reported histopathological confirmation of rubeosis in Fuchs' heterochromic iridocyclitis.

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Central serous retinopathy in systemic lupus erythematosus: a manifestation of the disease or of its treatment?

EDITOR,—In the paper by Eckstein *et al*¹ two patients with systemic lupus erythematosus (SLE) who developed typical central serous retinopathy (CSR) are described. As a result, CSR is reported as an unusual manifestation of SLE.

SLE is an autoimmune disease for the treatment of which glucocorticoids are usually employed. The treatment of patient 1 is not described, but patient 2 was treated with prednisolone, azathioprine, and cyclosporin A. The doses of steroids were increased a year before the development of CSR.

We recently reported the occurrence of CSR with unexpected frequency (5%) in a large consecutive series of patients with endogenous Cushing syndrome which is caused by prolonged exposure to endogenous cortisol.² CSR has also been associated with other conditions characterised by endogenous hypercortisolism (pregnancy, stress). Only few case reports describe CSR as a complication of systemic corticotherapy. However, some additional reports associate CSR with diseases treated with glucocorticoids (such as, Crohn's disease, ulcerative colitis). Based on the above observations we suspected a participation of glucocorticoids in the pathogenesis of CSR.

We are suggesting that glucocorticoid treatment alone or in conjunction with SLE may have played a role in the development of CSR in

patient 2 reported by Eckstein *et al.* This could also be the case for patient 1 if, as we presume, she had received corticotherapy. This represents an alternative explanation for the occurrence of CSR in patients with SLE.

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Reply

EDITOR,—I would like to thank E A Bouzas and G Mastorakos for their interest in our paper. In the September issue of the *Archives of Ophthalmology* they described three out of 60 patients with Cushing's syndrome who developed a central serous retinopathy during the course of their illness and quoted other circumstantial evidence that steroids may have a role in the aetiology of central serous retinopathy. However, in view of the vast number of patients who are on steroids for one reason or another this risk must be extremely low and a proper epidemiological study is required. Our paper pointed out that central serous retinopathy is in fact rare in systemic lupus erythematosus (although large numbers of patients are treated with steroids) and, furthermore, our patients were unusual in that although they developed the typical angiographic features of central serous retinopathy they both failed to get visual improvement with resolution of the retinopathy. In our view, the association between central serous retinopathy and steroid therapy is an interesting point which remains unproved.

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Pterygium excision with conjunctival autografting

EDITOR,—The excellent article by Allan *et al*¹ on pterygium excision with conjunctival autografting drew attention to the relative slowness of the procedure. They advocated spreading the free graft out on the cornea and transferring it to the donor site without lifting it clear at any point. However, once the donor conjunctiva is completely excised, it often develops a tendency to roll up into a ball. This has to be unravelled on the cornea to ensure that the donor conjunctiva is correctly oriented before moving it to the donor site.

It is possible to simplify this technique and, in the process, to quicken the transfer and ensure the correct orientation of the graft every time. The limbal side of the conjunctival graft is freed first. Next, the nasal end of the incision is extended radially for the required distance. The area beneath the graft can now be freed by using blunt dissection, while elevating the free edge of the graft. Once the underside of the

Figure 1 The conjunctival autograft is gently immobilised using a small chalazion clamp. Once dissected free, the clamp is rotated to the donor site. The autograft is automatically in the correct position for suturing at the host site.

graft is free, a small chalazion clamp is introduced, with the solid blade beneath the graft. Light pressure is then applied (with no need to tighten the screw) thus immobilising the flap. Tension is then gently exerted on the flap, and the remaining sides of the trapezoid can be quickly completed (Fig 1). The clamp is then simply swung round to the prepared recipient site. The limbal edges of the autograft are next secured with a 10/0 nylon suture (the suture is passed and cut, but not tied, so allowing the surgeon to position both limbal sutures without releasing the clamp). The clamp is then gently removed and the sutures tied. The autograft is then secured at the nasal side of the donor site. Care must be exercised throughout in exerting only light pressure on the chalazion clamp to avoid crushing the autograft. However, once the technique is familiar, the autograft transfer is much quicker.

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Reply

EDITOR,—The variation in autograft transfer technique suggested by C McLean would certainly help to ensure the correct graft orientation; but the correct orientation can usually be maintained without difficulty by ensuring that the graft is not lifted clear of the ocular surface during excision.

Lifting the free graft clear of the ocular surface causes it to shrivel and twist dramatically. As the graft has no rigidity, the forces of surface tension acting through the fluid film coating the graft are not neutralised until the minimal volume is assumed and the graft has rolled up into a ball. So long as the graft is in contact with the eye, however, the fluid film coating the graft remains in continuity with that coating the ocular surface. Surface tension then acts to flatten the graft onto the eye. Spreading the graft out on the cornea immediately after excision, allows it to be slid into position at the excision site in the correct orientation.

Graft transfer with this technique is relatively quick. Overall, the procedure can be expedited somewhat using a continuous 10/0 nylon suture (running around three sides of the graft between the two principal points of

fixation at the limbus) to secure the graft. This suture can be removed at 3 weeks and inspires less postoperative inflammation than degradable sutures. Our clinical impression is that pterygium recurrences are more likely to occur in the context of continued postoperative inflammation.

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Causes of visual handicap in the Royal Blind School, Edinburgh, 1991-2

EDITOR,—We have performed a follow up study to that done in 1985 by Phillips *et al* at the Royal Blind School, Edinburgh¹ in order to detect trends in the causes of childhood blindness in the school. During the academic year 1991-2 all 93 children at the school were examined by one consultant ophthalmologist (BWF). The main cause of blindness was ocular in 46 children and neurological in 47 children. Thirty children were blind due to a recognised genetic syndrome. Five of these were autosomal dominant with a positive family history. The remaining cases were autosomal recessive or sporadic. Twenty seven had structural abnormalities of the eye or brain of unknown aetiology - 'developmental'. Sixteen children were blind due to retinopathy of prematurity (ROP) and a further three children were blind because of central nervous system complications of prematurity. Eight children had blindness associated with birth asphyxia and in eight blindness followed meningitis or encephalitis. Three cases were due to trauma.

While many of our findings are similar to those of Phillips *et al* some trends were evident. The number of congenital cataract cases has fallen from 12 to four, possibly because of improved early surgical management. The number of cases of ROP has risen from 11 to 16. This may be related to improved survival of very low birthweight infants.² It remains to be seen whether the advent of cryotherapy or laser treatment for acute ROP will reduce the incidence of blindness due to this cause.³

A recent population based survey in Ireland reported the causes of blindness in 172 children.⁴ The proportion of conditions associated with neurological handicap, such as cortical blindness and ROP, was higher in the Royal Blind School (26% v 16% and 17% v 11% respectively). Conditions causing less severe forms of visual handicap, such as some cases of optic nerve hypoplasia, were relatively under-represented in the Royal Blind School (3% v 22%). This may be because of the trend towards 'mainstream' education for less handicapped children.

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