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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- 1 Richardson P, Schulenburg WE. Bilateral congenital mydriasis. *Br J Ophthalmol* 1992; 76: 632-3.
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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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- 1 Rutzen AR, Raizman MB. Fuchs' heterochromic iridocyclitis. In: Albert DM, Jakobiec FA, eds. *Principles and practice of ophthalmology*. Philadelphia: Saunders, 1994: 503-6.
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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