We have presented a case of type I Familial Amyloidotic Polyneuropathy. The underlying transthyretin mutation (TTR-glycine 54) has not been reported in any other family. The case illustrates the importance of taking a careful family history.

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Sir,

Secondary posterior chamber intraocular lens implantation in Fuchs' heterochromic uveitis

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In almost all patients with uveitis requiring cataract surgery, phakoemulsification with intraocular lens (IOL) implantation is the procedure of choice.^{1,2} The need for stringent peri-operative control of uveitis is well-documented.³ Some eyes however, will not tolerate IOL implantation and in some it is not possible accurately to predict postoperative reactions, including severe or persistent uveitis, cyclitic membrane formation and phthisis. Sometimes explantation of an IOL becomes necessary, which may be problematic.⁴ During the 1980s many surgeons, because of such potential problems, chose not to implant IOLs in eyes with uveitis on the grounds of safety.

Fuchs' heterochromic uveitis (FHU) is a peculiar uveitis which many have perceived to behave less aggressively after cataract surgery. This view is not universally shared,² and a higher inflammation rate has been reported from IOL implantation during extracapsular cataract extraction (ECCE)⁵ and in comparison with other forms of uveitis, in phakoemulsification surgery.² In the past some FHU eyes did not undergo primary IOL implantation but patients may subsequently request secondary IOL implantation, usually on the grounds of contact lens (CL) intolerance.

The outcomes and problems of secondary IOL implantation have been reported.^{6–8} However, there are no previous reports on the procedure in eyes with uveitis.

Case reports

The details of the four cases are shown in Table 1. All cases underwent primary surgery elsewhere and were referred to the Manchester Uveitis Clinic for secondary surgery. In all cases posterior capsule integrity was demonstrated preoperatively. A large-optic (7 mm) IOL was inserted under Healon® via a corneal section, into the ciliary sulcus. Two-hourly topical steroid was administered postoperatively, being tapered to zero according to recovery rate.

Case 1

A 43-year-old male bus driver with right FHU underwent ECCE but developed progressive contact

Table 1 Details of cases

Case number	1	2	3	4
Age, sex	43,♂	38,♀	23,♂	35,♂
Delay between ECCE and secondary IOL implantation	7 yrs	1 yr	15 yrs	11 yrs
Preoperative visual acuity	6/6	6/5	6/9	6/9
Amsler's sign	No	Yes	Yes	Yes
IOL type	HSM ^a	PMMA	PMMA	PMMA
Postoperative visual acuity	6/6	6/5	6/5	6/6
Maximum postoperative AC cell activity	±	±	±	+
Postoperative steroid discontinued	5 wks	5 wks	10 wks	12 wks
Postoperative complications	Nd-YAG capsulotomy	Nil	Nil	PS extension
Follow-up period	8.5 yrs	4 yrs	1.5 yrs	1.5 yrs
Inflammation at latest follow-up	Nil	Nil	Nil	Nil

^a HSM = heparin surface-modified.

lens (CL) intolerance and requested secondary IOL implantation. Surgery was uneventful. Three years later he underwent uneventful Nd-YAG posterior capsulotomy.

Case 2

A 38-year-old woman with FHU requested secondary IOL implantation despite satisfactory CL wear. Surgery was uneventful. No significant recurrence of postoperative inflammation occurred.

Case 3

A 23-year-old man with bilateral FHU underwent ECCE in the left eye. Subsequently he developed right secondary cataract and underwent uneventful phakoemulsification with IOL implantation, achieving 6/5 postoperatively. He then requested secondary IOL implantation in the left eye. Surgery and postoperative course were uneventful but 9 months postoperatively giant-cell deposition on the IOL reduced the visual acuity to 6/12, necessitating a 3-month course of topical steroid. The IOL surface was cleared, returning the visual acuity to 6/5.

Case 4

A 35-year-old man with right FHU underwent ECCE but became CL-intolerant after 11 years and developed peripheral corneal neovascularisation. There were peripheral anterior synechiae at 10 o'clock adjacent to a peripheral iridectomy and posterior synechiae (PS) to the capsule between 12 and 2 o'clock. At surgery the PS could not be safely divided but the IOL was nevertheless inserted transversely. There was IOL-iris touch. Postoperative uveitis was mild, but the PS

extended somewhat behind the IOL. Giant cell deposition did not occur and topical steroid was discontinued at 12 weeks.

Comment

In eyes with uveitis, the choices for secondary IOL implantation are limited. Angle- or iris-supported IOLs are inadvisable because of the risk of uveitis, glaucoma and hyphaema. The safety of trans-sclerally sutured IOLs in uveitic eyes is unproven, and so in this clinic, no aphake with active uveitis has been offered secondary IOL implantation unless the posterior capsule is intact, allowing sulcus implantation of a large-optic IOL.

There were no significant surgical problems in the four cases reported here. The integrity of the posterior capsule was confirmed in all preoperatively. Mild operative AC haemorrhage (Amsler's sign) is typical of FHU and occurred in three of four cases but was not problematic. In only one case (with preoperative synechiae) was iris-IOL touch a potential problem but no significant postoperative problems ensued. The visual outcomes were good in all cases.

Secondary implantation of a large-optic IOL into the ciliary sulcus has been successful in all four patients with FHU in whom this procedure has been performed at this centre. With appropriate patient selection, informed consent and preoperative control of inflammation, the procedure has been safe and effective for this form of uveitis and can be recommended as appropriate management of CL intolerance for the unilateral aphake with Fuchs' heterochromic uveitis.

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Sir,

Unilateral acute idiopathic maculopathy (UAIM) masquerading as Best's disease

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Acute onset of exudative maculopathy is a well-recognized feature of unilateral acute idiopathic maculopathy (UAIM).¹ We report a patient with unilateral acute idiopathic maculopathy (UAIM) characterized by predominantly macular involvement.

Case report

A 15-year-old, healthy female Caucasian patient was first noted to have a retinal abnormality during a routine visit to her optician. She was asymptomatic and had no significant ophthalmic or medical history or any history of medication. On a previous visit to her optician 6 months ago no fundal abnormality was detected. There was no history of amblyopia, squint or nyctalopia in the past. Her family history was unremarkable.

Figure 1 Fundus appearance at initial presentation.

Visual acuities were counting fingers at 1 metre in the right eye and 6/5, N5 in the left eye. Slit lamp biomicroscopy of the right eye revealed a normal vitreous cavity unaccompanied by signs of an active uveitis. The striking abnormality was the presence of a subfoveal grayish lesion at the level of the retinal pigment epithelium (RPE) measuring approximately 1/3 disc diameter in size surrounded by subretinal exudates associated with a fluffy white appearance. There was evidence of neurosensory retinal detachment overlying and surrounding the entire lesion as noted by the tenting up of the retina in the macular region (Figure 1).

Both optic discs were pink and healthy. Fluorescein angiography demonstrated hyper-fluorescence of a subfoveal lesion (Figure 2). The clinical picture was

Figure 2 Fluorescein angiogram confirming a hyperfluorescent lesion at the fovea with pigmentary macular disruption (white arrow) corresponding to the neurosensory retinal detachment involving the macula.