LETTERS TO THE EDITOR

Retinopathy associated with pancreatitis in a child with maple syrup urine disease

EDITOR,—Retinopathy associated with pancreatitis is an uncommon condition first described in 1975. To date, fewer than 50 cases have been reported, all involving adults. We report a case of pancreatitis with retinopathy in a young child with maple syrup urine disease, a rare metabolic disorder.

CASE REPORT
A 7 year old Indian female presented with maple syrup urine disease (MSUD) diagnosed in infancy. She has been maintained on a special diet since then, enjoying normal development. However, during December 1995 she was admitted to the hospital for management of acute gastroenteritis and dehydration. Laboratory studies included serum amylase and lipase which were within normal limits. She received hyperalimentation with glucose and intralipids through a femoral catheter. On her third hospital day she was noted to have mental status changes which improved with hydration. However, on her sixth hospital day her mental status deteriorated again. Her abdomen became diffusely tender. Laboratory studies revealed serum amylase 453 U/l (normal limits 40–128), calcium 8.2 mg/dl, phosphorus 2.2 mg/dl, albumin 2.6 g/dl, total protein 5.1 g/dl, and a white cell count of 12.4 x 10^9/l. The serum was noted to be lipaemic despite her receiving only intravenous dextrose in 1/2 normal saline solutions. All other routine laboratory values were within normal limits. Arterial blood gases were within normal limits. Amino acid levels of soleucine, histidine, and valine were within normal limits while leucine was 12.9 mg/dl (normal limits 1.0–5.2). During a neurological examination white lesions were noted in both fundi. In the next 48 hours, the patient’s general status improved. Her serum amylase levels declined while lipase levels increased to reach a peak of 2485 U/l (normal limits 23–208).

An ophthalmic evaluation 2 days after the initial elevation in serum amylase revealed visual acuity of 20/50 in both eyes (Snellen E) both at distance and at near. No afferent pupillary defect was recorded. Ocular versions were intact. Slit lamp biomicroscopy was unremarkable. Dilated fundus examination revealed the presence of bilateral white fluffy lesions and areas of haemorrhage (Fig 1). Findings were concentrated in the posterior poles only. Fluorescein angiography showed minimal capillary non-perfusion (Fig 2).

The patient slowly improved spontaneously. On follow up examination 1 month later, visual acuity was 20/25 in both eyes with no correction. Fundus examination was completely normal, with full resolution both of cotton wool spots and of haemorrhages.

COMMENT
Maple syrup urine disease is caused by a defect in branched chain ketoacid (BCKA) dehydrogenase. Occular complications of untreated disease or late diagnosis include optic atrophy, nystagmus, ophthalmoalgia, strabismus, and cortical blindness.

Pancreatitis is a known complication of branched chain organic acidemias (BCOA).

In a series of 108 paediatric patients with BCOA, nine cases of pancreatitis were found during a 10 year period. One of these cases was in a patient with MSUD. 1

Retinopathy as a rare complication of acute pancreatitis was first described in 1975. 2 It has been reported to occur either before 3 or after 4 the manifestation of acute pancreatitis. 5–6 All reported cases to date have been in young adults (ages 25–40 years), often associated with a history of ethanol misuse. None of the previously reported cases had associated systemic metabolic diseases.

The aetiology of retinopathy of pancreatitis is debatable. An older theory, supported by some experimental data, 7 holds that fat emboli found in many organs in cases of acute pancreatitis 8–9 cause ischaemic retinal infarcts. Retinal fat emboli have also been found in Purcher’s retinopathy, 10 a condition remarkably similar to pancreatitis associated retinopathy.

Another theory proposes embolisation of retinal vessels by complement induced fibrin clots and leucocaggregates as the causative mechanism. 11 However, experimental studies have failed to reproduce various features of the clinical picture of retinopathy of pancreatitis. 12, 13 The presence of lypaemia in our patient argues for the lipid embolism theory. Hypo-volaemia and hypoxia did not occur and are thus unlikely to have been involved in the pathogenesis of the retinal picture. Although our patient maintained almost normal levels of leucine, isoleucine, and valine throughout the above episode, it is unclear whether the presence of MSUD predisposed her to the development of retinopathy of pancreatitis.

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Immediate postoperative objective refraction as an indication to final refraction in phacoemulsification surgery

EDITOR—The goal of cataract surgery is the rapid attainment of good visual acuity, ideally unaired for the intended focal distance and the refractive change on accurate intraocular lens (IOL) power calculation. Refractive prediction errors, if substantial, can be problematic for the patient. If the problem is one of symmetrical ametropia, spectacle correction is usually successful. Contact lens fitting may be necessary for those patients with anisometropia in whom spectacle wear would induce significant and intolerable anisokoria. IOL exchange as a secondary procedure may have to be considered in cases refractory to these measures.

In our study, we aimed to ascertain whether objective refraction at the end of surgery was feasible and if so, to determine how this refraction related to the final refraction at 4–7 weeks postoperatively with a view to suggesting guidelines for immediate exchange of implant in cases of gross refractive prediction errors.

CASE REPORT

Consecutive patients undergoing phacoemulsification cataract surgery after continuous curvilinear capsulorhexis with capsular bag implantation of Chiron C10UB injectable IOLs were refracted at the end of surgery on the table objectively with a streak retinoscope, and at 4–7 weeks postoperatively with a view to calculating the sphere and cylindric power and assessing the ametropia to be expected at 6 months postoperatively. Twenty-six eyes of 26 patients were included.

Data were analysed using statistical software (Minitab computer).

Figure 1 Scattergram of refractive change from immediate postoperative refraction to final refraction.

We were unable to refract two patients owing to the presence of corneal epithelial haze.

Figure 1 shows a few values clustering around the neutral—that is, “no change” line, and a single significant myopic change but the predominant feature is the shift towards hypermetropia. The single significant myopic change occurred in a patient who had topical anasthesia.

The mean change in refraction was 1.11 D hypermetropia with a standard deviation of plus or minus 0.94 D. The range was from 1.63 D myopic change to 2.75 D hypermetropic change. The 95% confidence interval was 0.73 D, 1.48 D. Paired two tailed Student’s t test performed on the immediate postoperative and final refraction results gave a p value of <0.001.

COMMENT

Hovding et al reported that about a third of the 188 patients in whom a PC-IOL was implanted after capsular cataract extraction ended up with more than plus or minus 1.0 D deviation from the predicted postoperative refraction. In about a tenth deviations of more than plus or minus 2.0 D from the calculated value were found.

The earliest reported IOL exchange was performed on the first postoperative day. In other studies the average interval between the primary surgery and the implant exchange ranged from 3.5 years to over 5 years.

We found that following phacoemulsification surgery, the preservation of corneal clarity together with the minimally induced surgical astigmatism allowed us to perform immediate postoperative objective refractions satisfactorily in most cases.

There was a statistically significant hypermetropic shift. We think that the most likely explanation lies in the reduced effectiveness of the IOL as it settles back in the postoperative period. After phacoemulsification cataract surgery with continuous curvilinear capsulorhexis, the depth of anterior chamber has been found to increase gradually, the anterior capsular size to narrow, and the refraction to tend towards hypermetropia.

The IOL used in our study was a Chiron C10UB—a plate haptic lens with no angulation and a predicted final anterior chamber depth of 5.59 mm.

Various factors which can affect the accuracy of immediate postoperative refraction should be considered.

We would advise a fundal examination be performed to exclude any posterior pole lesions hitherto undetected.

In conclusion, objective refraction immediately following phacoemulsification surgery is feasible; if a gross refractive prediction error is found, immediate implant exchange may be considered, taking into account the mean hypermetropic shift of about 1 dioptre from immediate postoperative objective refraction to the final refraction in the postoperative period in cases where the Chiron C10UB lens has been implanted.
or performed any valsalva manoeuvre. She denied sudden loss of the central vision in her left eye and felt unwell. She then left the club. She presented at the casualty department 2 days later. On examination her right visual acuity was 6/5 and left visual acuity was 1/60. She had no relative afferent pupillary defect. The anterior segments were normal and intraocular pressure was 15 mm Hg in the right eye and 17 mm Hg in the left eye. The posterior lens implant restored the left visual acuity to 6/60 after 6 months.

Intravenous fluorescein angiogram showed no retinal leak. Six months following latanoprost administration was striking.

COMMENT
Latanoprost and cystoid macular oedema in a pseudophake

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Latanoprost and cystoid macular oedema in a pseudophake

Editor—Latanoprost (0.005%) is a prostaglandin F2α analogue licensed for primary open angle glaucoma. We describe the occurrence of cystoid macular oedema in a pseudophakic patient following its use.

CASE REPORT
A 50 year old pseudophakic myope (−4.50 D) was prescribed latanoprost once daily, because of raised intraocular pressure despite treatment with twice daily betaxolol (1/2%) drops. The right eye was blind from retinal detachment and the left eye had undergone three retinal detachment repairs including one vitrectomy. He had required betaxolol twice daily in the left eye to control the intraocular pressure since then. In January 1994, a left phacoemulsification cataract extraction with posterior lens implant restored the left visual acuity to 6/60.

In March 1997, latanoprost was prescribed to control the rising intraocular pressure, 7 weeks after which the vision dropped to 6/9, the patient describing a central scotoma. Cystoid macular oedema was evident on examination with a left disc haemorrhage and a superonasal, peripapillary subretinal haemorrhage without evidence of other vascular abnormalities (Fig 1). Fundus fluorescein angiography (FFA) showed a petaloid appearance typical of cystoid macular oedema (Fig 2). The latanoprost was stopped, and within 2 weeks the symptoms of visual blurring and the cystoid macular oedema had improved. The patient continued on betaxolol to the left eye but inadequate intraocular pressure control made further filtering surgery necessary.

COMMENT
This eye had undergone retinal detachment repairs including scleral buckling, cryotherapy, vitrectomy, and then phacoemulsification and lens implantation. There was a 3 year gap before latanoprost was prescribed. These procedures may each be associated with cystoid macular oedema and selective use of pars plana vitrectomy used in its treatment, but all procedures occurred over 2 years previously with 6/6 vision in the intervening period. While we cannot exclude pre-existing subclinical, angiographically positive cystoid macular oedema, the timing of onset and relief of clinical symptoms and signs with latanoprost administration was striking.

Hoyng et al1 administered latanoprost (0.006%) twice daily for a month to a uncompli cated pseudophakic patients and fluorescein angiography showed no retinal leak. Six aphakic cynomolgous monkeys given seven times the usual daily dose for 6 months failed to develop cystoid macular oedema. However, intravitreal injections of PGF2α to pigmented rabbits showed a small but statistically significant leak by vitreous fluorophotometry. Animal work needs to be interpreted circumspectly, particularly regarding the rabbit which shows an atypical response to inflammation.3 These experimental results suggest that aphakic or pseudophakic pri mates do not normally develop cystoid macular oedema in response to prostaglandins.

Miyake and colleagues’ work on rabbits and baboons confirmed that aphakia and pseudophakia may be associated with impaired removal of prostaglandins by the ciliary processes (Bito’s pump), with resulting accumulation in the eye, since prostaglandins are not broken down intraocularly.4 Possibly Bito’s pump was impaired in our patient, further raising prostaglandin levels and increasing the concentration gradient.

This patient had already had a peripheral anterior vitrectomy during the second left retinal detachment repair, and in such a case it probably would facilitate diffusion of prostaglandins posteriorly.

Furthermore, there were extensive and repeated vitreoretinal procedures performed before administration of the medication and this was a complex case of posterior chamber pseudophakia. This case confirms the
accessibility

Visual acuity was right eye 6/12, left eye 6/12 taking the oral contraceptive pill. Snellen obscurations of vision. She had not been

A 20 year old obese white female presented to

Factor V Leiden mutation in association

with idiopathic intracranial hypertension

Editor,—Idiopathic intracranial hypertension has an association with prothrombotic condi-
tions. The recently described thrombophilic defect of activated protein C (APC) resist-
ance, caused by the factor V Leiden mutation, has been postulated to be a factor in some cases of our knowledge. In this report, we describe the first two cases reported of the factor V Leiden mutation identified in association with idiopathic intracranial hypertension. Both patients had been previously well and therefore should have a past history to suggest a clotting disorder.

CASE REPORTS

Case 1

A 20 year old obese white female presented to the ophthalmic department with transient obsessions of vision. She had not been taking the oral contraceptive pill. Snellen visual acuity was right eye 6/12, left eye 6/12 and gross papilloedema with macular stars was noted. Computerised tomography and magnetic resonance angiography were normal and a lumbar puncture showed an opening pressure of 75 cm of water. CSF protein, glu-
cose, and cell count were normal as were U&Es, LFTs, TPTs, glucose, full blood count, and plasma viscosity. She was started on 250 mg warfarin daily and advised to lose weight. Over the next few days acuity reduced to right eye 6/24, left eye 3/6 with extensive field restriction. Urgent optic nerve fenestration of the left eye was under-
taken followed by the right 12 days later as acuity began to worsen in this eye also. Partial recovery occurred with Snellen visual acuities of right eye 6/60, left eye 6/36 but, unfortunately, the extensive field restriction remained. On thrombophilia screening, the APC resist-
ance ratio was reduced at 1.9 (normal 2.2–4.2) on two occasions and a heterozygous factor V Leiden mutation was identified. She was referred to the haematology department de-
sion, was not undertaken as she was thinking of planning a family.

Case 2

A 31 year old obese white female was referred by her general practitioner complaining of transient visual loss in her left eye for the pre-
vious 5 months and non-specific headaches for 2 years. She had stopped the oral contraceptive pill 12 months previously. Snellen visual acuity was right eye 6/5, left eye 6/5 and field analysis showed an enlarged blind spot in the left eye. Funduscopy revealed established papilloedema in the left eye and early papilloedema in the right. Urgent computer-
ised tomography was normal and a lumbar puncture had an opening pressure of 37 cm of water. CSF and blood investigations were unremarkable. Warfarin fields re-
mained stable on Diamox SR 250 mg twice daily and she has been able to lose weight from 15 stone. A thrombophilia screen showed a reduced APC resistance ratio of 1.9 on two occasions and a heterozygous factor V Leiden mutation was found. She was also referred to the haematology department for consider-
aition of anticoagulation and, after discussion, was begun on warfarin.

COMMENT

Prothrombotic abnormalities have recently been implicated in the pathogenesis of “be-

ign intracranial hypertension” (BIH). It has been suggested that CSF reabsorp-
tion due to damaged arachnoid villi, secondary to microthrombus formation, could be the cause of raised intracranial pressure. Arach-

noid villi dysfunction causing BIH has also been postulated in SLE and may be the result of venulitis, aseptic meningitis, or immune complex deposition and, in the antiphospholi-
pid syndrome, possibly due to micro-

occlusion. Such mechanisms affecting the optic nerves in addition could cause the poor outcome in case 1.

APC resistance is gaining greater recogni-
tion in the pathogenesis of ophthalmic disor-
s and has been implicated as a cause of central retinal vein occlusion in patients younger than 50 years. The factor V Leiden mutation may be heterozygous or ho-

mzygous and carries a lifelong increased thrombotic risk. The left and 50–100 fold respectively. As 5% of the population has the factor V Leiden mutation, it is felt that most individuals carrying the defect will never experience a thrombotic event unless they carry another genetic defect, such as a deficiency of protein C or S, or are exposed to a precipitating factor such as oral contra-

ceptives, pregnancy, or surgery. Currently the criteria for anticoagulation in idiopathic intracranial hypertension are unknown owing to the absence of controlled clinical trials. It is reasonable for haematologists to offer war-

farin as a short course or continuous in symptom-
atomatic patients, especially if the event is severe, and in those exposed to other risk factors who have a familial thrombotic tendency.

Without further studies addressing the association of idiopathic intracranial hyper-
tension and thrombophilia, routine testing for the factor V Leiden mutation and other thrombophilic factors cannot be recom-

mended in patients with idiopathic intra-
cranial hypertension outside a clinical trial basis. However, if the mutation is found, referral should be made to a haematologist for advice concerning anticoagulation, bearing in mind the increased bleeding risk and teratogenicity of warfarin. First degree blood relatives of the patient should be warned to avoid smoking and to seek medical advice before taking oral contraceptives or accepting elective surgery; the factor V Leiden mutation is of dominant inheritance.

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Do all patients with CMV retinitis require life long anti-CMV therapy?

Editor,—It is becoming clear that some patients with cytomegalovirus (CMV) retinitis who respond to highly active antiretroviral therapy may be candidates for prophylactic treatment with ganciclovir or foscamet. When patients are treated with highly active antiretroviral therapy, the rate of reactivation of CMV retinitis is much lower than that found in patients not treated. This may be due to Ganciclovir or foscarabine being used in new patients who have not received previous treatment may reduce the development of CMV retinitis. Therefore, it is reasonable to consider continuing ganciclovir or foscarabine maintenance therapy for patients with CMV retinitis who respond to highly active antiretroviral therapy. However, the duration of maintenance therapy and the optimal dose of ganciclovir or foscarabine remains to be determined. In patients who are unable to tolerate ganciclovir or foscarabine, alternative treatments such as foscarnet or cidofovir may be considered. Combining multiple antiviral agents may be a useful strategy in the management of patients with CMV retinitis.
reactivations of their retinitis despite having no specific anti-CMV therapy. However, the factors underlying this improved immunity to CMV are not entirely clear. We report the case of a patient on highly active antiretroviral therapy (HAART) who has had no reactivation of CMV retinitis after 6 months without anti-CMV treatment. The unusual feature of this case is that the CD4 count has remained persistently low.

We discuss factors that may be relevant in this improved immune response to CMV infection and may be useful in isolating a group of patients on HAART who do not require lifelong maintenance with anti-CMV therapy.

CASE REPORT
A 33 year old man with a CD4 count of 27 cells $\times 10^3/l$ who had been diagnosed with AIDS in 1992 developed CMV retinitis in zones 1, 2, and 3 of the left eye in March 1996.

Treatment was commenced with intravenous ganciclovir $10$ mg/kg and maintenance therapy consisted of oral ganciclovir. After 3 months reactivation and progression of the retinitis necessitated a further induction course of intravenous ganciclovir with subsequent maintenance therapy with daily intravenous foscarnet.

Further reactivation led to the use of intravitreal ganciclovir and further re-induction course of intravenous foscarnet. As a result of difficulties with intravenous line sepsis intravenous cidofovir was commenced. In March 1997 the patient was started on HAART including protease inhibitor therapy but despite this the CD4 count remained low at $22$ cells $\times 10^3/l$.

The patient decided to discontinue anti-CMV therapy in April 1997 and has undergone frequent ophthalmological review since. He has received no anti-CMV therapy for 6 months and despite a continuing low CD4 count of 28 cells $\times 10^3/l$ and a HIV-1 viral load of 201 637 RNA copies/ml there has been no further reactivation of CMV retinitis.

COMMENT
Before the use of HAART, maintenance therapy with anti-CMV medication was required in all patients with CMV retinitis. Without therapy the average time to progression of the disease was 2–3 weeks.1 Several cases have been reported which describe lack of progression of CMV retinitis in patients treated with HAART and in each of these cases there was an associated rise in CD4 counts.2 There have also been reports of newly diagnosed CMV retinitis after commencement of HAART with improving CD4 counts,3 although considerable doubt has been shed on whether these were in fact new cases of CMV retinitis and not just reactivation of previously undetected disease.4 In this case the CD4 count remains persistently low and the HIV viral load high despite HAART.

This suggests that the immune response to CMV is not solely related to the CD4 count and that other factors are also involved in the recovery of immunity to CMV following treatment with HAART.

Bowen et al have demonstrated by quantitative and qualitative measurements of CMV DNA by polymerase chain reaction based assays that CMV viral load may be an indicator of patients at increased risk of reactivation of CMV retinitis.5 They suggest that this measurement can help with clinical management. It has also been shown that the detection of the early CMV antigen (p65 antigen) suggests elevated susceptibility to CMV infection.6

Transferring growth factor $\beta$ (TGF-$\beta$) inhibits the IL-2 induced proliferation of T lymphocytes, and expression of TGF-$\beta$ may be increased in CMV infection.7 Measurement of TGF-$\beta$ and other cytokines may also be useful in the monitoring of patients at risk of CMV disease.

By performing a controlled prospective trial involving measurement of CMV and HIV viral load, together with these immunological markers it may be possible to identify a subgroup of patients on HAART who do not require long term anti-CMV therapy.

However, early reports from Martin et al on the results of their study of the use of combined oral ganciclovir and intravitreal implant for treatment of CMV retinitis suggest that there is still a survival benefit from continued use of oral ganciclovir in the HAART environment.8 This once again raises the debate over whether CMV is just an opportunist taking advantage of immunosuppression or is it a cofactor acting in partnership with HIV.

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Patient No 2
A 44 year old white man with AIDS had a CD4+ cell count of 25 cells ×10^3/l and no evidence of CMV retinitis on serial eye examinations. Combination antiretroviral therapy with saquinavir, zidovudine, and lamivudine was begun in March 1996 and 1 month later the patient developed a temporal scotoma in his left eye. Examination of the left eye showed mild anterior uveitis, vitritis, and a large area of retinal atrophy in the nasal midperiphery consistent with inactive CMV retinitis (Fig 2). The area of inactive retinitis has remained stable over the past 12 months despite the fact that he has received no anti-CMV therapy. A CD4+ T cell count in October 1996 was 111 cells ×10^3/l.

COMMENT
We describe two patients who presented with mild uveitis and an area of retinal atrophy consistent with inactive CMV retinitis despite never having received specific anti-CMV therapy. Both patients had prior CD4+ cell counts below 50 cells ×10^3/l, and each experienced an elevation to above 50 cells ×10^3/l in response to combination antiretroviral therapy concurrent with the onset of their symptoms. We hypothesise that these patients developed subclinical CMV retinitis in the setting of severely suppressed CD4+ T cell counts, but became symptomatic when HAART induced elevations of their CD4+ cell counts enhanced the immune response to CMV. This led to the development of a significant and symptomatic uveitis, a finding uncommon in patients with CMV retinitis before the use of HAART. These observations support the notion that HAART induced restoration in immune function can lead to spontaneous and sustained resolution of CMV retinitis. The fact that patient no 1 developed reactivation of CMV retinitis soon after his CD4+ T lymphocyte count fell below 50 cells ×10^3/l supports the initial diagnosis of CMV retinitis. Additionally, CD4+ cell counts may continue to provide valuable information regarding the risk of reactivation of opportunistic infections in patients receiving HAART. These findings are in accordance with recent publications addressing the relation between protease inhibitors and sustained inactivity of CMV retinitis. Additional studies are required to further delineate the role of HAART and CD4+ cell counts in the natural history of CMV retinitis.

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Figure 1 Matching the ages of donors and recipients. (A) Number of corneal transplants placed into recipients aged 0–5 in 1996, by age of donors. Total 15. (B) Utilisation of corneas from donors aged 0–5 in 1996, by age of recipient. Total 44 (22 not used, and one allocated to recipient of unknown age). Data from UKTSSA.
maintained in culture for 68 days (over 9 weeks), the final endothelial assessment, by vital staining and light microscopy, having confirmed its suitability for use. The right eye was operated on 1 week later using a cornea from a 25 year old donor as no infantile cornea was available.

At 2 months postoperatively at examination under anaesthesia both corneal grafts were clear (Fig 2c,d). Examination under anaesthesia was repeated 1 year later. Again both grafts were clear, and specular microscopy of the graft endothelium revealed a cell density of 2235 cells/mm² in the graft from the infantile donor, and 1410 cells/mm² in the graft from the older donor (Fig 2e,f). Retinoscopy showed high myopia in both eyes, but both fundi appeared normal.

COMMENT
In most eye banks organ cultured corneas that are not transplanted within 4–5 weeks of retrieval are discarded. This is based on the finding that the rate of decline of endothelial cell density increases after 35 days of culture in a single aliquot of culture medium.1 Pels et al reported that storage of corneas in culture for 3–7 weeks induced a mean cell loss of about 11% while preservation for 9–17 weeks (medium changed after 6 weeks), resulted in a mean cell loss of about 43%. The actual cell loss, however, varied significantly among examined corneas and therefore it has been suggested that the suitability of an individual cornea for transplantation should be based on the quality of the endothelium during final assessment rather than the length of storage.1 It has also been suggested that the increased cell loss after 40 days of culture might be caused by the depletion of nutrients and accumulation of waste products in the culture medium. A larger volume of medium, or the renewal of the medium, may postpone or prevent this process. However, with prolonged organ culture endothelial survival is not the only factor in determining the suitability of tissue for transplantation. If there is epithelial overgrowth onto the posterior corneal surface this can potentially lead to problems of implantation of epithelial cells into the anterior chamber.

There has been a report of corneas safely preserved in culture for 7 weeks before transplantation,2 and we have found that an infantile cornea preserved for 9 weeks in organ culture can be transplanted successfully. There is usually no need to extend the storage time of adult corneas, but it may be appropriate to extend the preservation time of some infantile corneas in order to maximise the chance of their most appropriate utilisation, and further research is needed to optimise storage methods and evaluate the cost/benefit of longer storage times.

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Figure 1 Fundus drawing showing spiculed retinal pigmentary changes and a choroidal melanoma with inferior hemiretinal serous detachment.

Figure 2 (a) Right eye, and (b) left eye of 3 month old infant with Peters’ anomaly preoperatively. (c) Right eye 8 weeks after penetrating keratoplasty with cornea from 20 year old donor. (d) Left eye 9 weeks after penetrating keratoplasty with cornea from 13 month old donor preserved in organ culture for 9 weeks. (e) Contact specular microscopy of right eye 1 year postoperatively with cell density of 1410 cells/mm². (f) Contact specular microscopy of left eye 1 year postoperatively with cell density of 2235 cells/mm².

Choroidal melanoma in a patient with retinitis pigmentosa and Usher’s syndrome

EDITOR,—Usher’s syndrome is an autosomal recessive condition characterised by retinitis pigmentosa and hearing loss. It is the most common cause of combined blindness and deafness in the USA. Usher’s syndrome has two well defined subtypes including type I and type II.3 Type I has more severe and early onset findings.

CASE REPORT
A 39 year old woman with retinitis pigmentosa and type I Usher’s syndrome developed blurred vision in the left eye over a 4 month period. The best corrected visual acuity was 20/50 in the right eye and 20/80 in the left eye. Anterior segment examination and intraocular pressures (16 mm Hg) were normal bilaterally. Ophthalmoscopic examination showed mild optic disc pallor, marked vascular attenuation, and retinal pigmentary changes in both eyes. Inferotemporally in the left eye, there was a 15 × 10 mm dome-shaped choroidal melanoma with a secondary retinal detachment affecting the inferior half of the fundus (Fig 1). There was no evidence of retinal invasion or seeding. A-scan and B-scan ultrasonography demonstrated a 5.0 mm thick choroidal mass with low to medium internal reflectivity, asymmetric reflectivity, having con-

satisfactory response to radiation. At 10 months follow-up, the subretinal fluid dried up completely but radiation papillopathy developed. The papillopathy resolved over 5 months, leaving more optic disc pallor. At 22 months follow-up, the patient underwent cataract surgery with posterior chamber lens implantation in both eyes because of advanced posterior subcapsular cataract. At 40 months follow-up, the vision was hand movements in the right eye and tumour thickness was 2.8 mm. There was no radiation retinopathy or metastatic disease.

COMMENT
Abnormalities involving chromosomes 1 (type II), 11, and 14 (type I) have been observed in patients with Usher’s syndrome. Reported chromosome alterations in uveal melanoma involve chromosomes 3, 6, 8, and 9; therefore, the simultaneous occurrence of these two conditions, as in our patient, is probably coincidental.

The development of retinal detachment in a patient with retinitis pigmentosa is rare because of adhesions between the retinal pigment and sensory epithelium. Only a few cases of retinitis pigmentosa with retinal detachment have been reported.’ Our patient illustrates a non-rhegmatogenous retinal detachment with retinitis pigmentosa.

Our patient developed radiation papillopathy with an optic disc dose less than 5000 cGy, which is unusual.4 The atrophy/gliosis of the optic disc in retinitis pigmentosa might increase the susceptibility of the disc to irradiation. On the other hand, she did not develop radiation retinopathy despite a high dose of radiation to the retina. Perhaps the atrophic retina in retinitis pigmentosa was unable to elicit a radiation response.

Choroidal melanoma can rarely lead to a pseudoretinitis pigmentosa picture secondary to retinal invasion and dispersion of melanoma cells on the retina.5 Retinal invasion was not present in our patient and retinal pigmentary changes were noted in both eyes, ruling out pseudoretinitis pigmentosa. Retinitis pigmentosa can be associated with several fundus tumours including giant drusen resembling astrocytoma and acquired vasoproliferative tumours.6 To our knowledge, this is the first report of choroidal melanoma in a patient with retinitis pigmentosa.

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