**Supplementary Table S1.** The following clinical summaries supplement the information given in Table 1. Abbreviations: HM hand motions vision; OD right eye; OS left eye; RD retinal detachment; VA visual acuity.

<table>
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<th>Patient</th>
<th>Clinical Summary</th>
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<td><strong>Patient 1</strong></td>
<td>was initially seen with a small left peripheral choroidal effusion and VA of 6/9 OD, 6/18 OS. There was a past history of chronic obstructive airways disease and mild ischaemic heart disease. Fundus examination showed a mottled, ‘leopard spot’ appearance. The effusions failed to respond to a course of oral corticosteroids. Over one year the left eye developed an extensive choroidal effusion with a subtotal RD, shifting fluid, and macular involvement (Figure 1). Surgery involved 4 partial-thickness sclerectomies of 5 by 7 mm, with a central 2 by 3 mm full-thickness sclerostomy in three of these. A fourth full-thickness sclerostomy was abandoned because of haemorrhage. Five weeks after surgery the effusions had resolved, with minimal subretinal fluid. Final vision was 6/36. The right eye developed smaller peripheral effusions over 4 months but did not require surgery. Vision was maintained at 6/9 in this eye.</td>
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<td><strong>Patient 2</strong></td>
<td>presented with bilateral, 360 degree, peripheral choroidal effusions, and pigmentary retinal changes suggesting previous chronic right RD. Vision was 6/9 OD, 6/6 OS with mild right macular oedema. The patient was being treated for systemic hypertension. Fluorescein angiography showed mildly slowed choroidal filling above the right macular but no other abnormality. There was no improvement following a trial of oral corticosteroids. The patient subsequently developed a right pigment epithelial detachment and choroidal neovascularisation involving the macular, but</td>
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vision remained at 6/9. Two years after initial presentation there were persisting choroidal effusions (Figure 2) with a left inferior RD. The patient underwent two partial-thickness and two full-thickness sclerectomies in the left eye. There was a persisting inferior RD but the effusions were reduced and VA improved from 6/36 to 6/24. Subsequently VA dropped to count fingers OD and three months after the initial operation the patient underwent four full-thickness sclerostomies in the right eye. These were repeated in three quadrant 5 months later, with reopening of the old sclerostomy in the fourth quadrant. The effusion and detachment had resolved completely within 3 months. Five months after the last operation the retinas remained flat, with ‘leopard spot’ mottling of the retinal pigment epithelium. Final vision was 6/12 OD, 6/18 OS.

**Patient 3** was a high hypermetope with choroidal elevation, detected as an incidental finding 5 months before right eye surgery, with subsequent involvement of the fellow eye, occurring 1 month before surgery in that eye. Examination showed bilateral annular choroidal elevation with inferior RDs and shifting fluid. Following surgery to the right eye there was a reduction in the choroidal effusion and RD but complete resolution took 5 months. After two operations on left eye there was reduced choroidal and subretinal fluid, but at final follow-up resolution was still incomplete. Vision at that stage was 6/18 right, 6/18 left.

**Patient 4** was found to have an asymptomatic, left, inferior, bullous RD detected during routine review for primary open-angle glaucoma. Despite macular detachment, presenting VA was 6/24. Past history included a non-ischaemic branch retinal vein occlusion. The time from detection of choroidal effusion to surgery was 4 months.
Surgery resulted partial reduction in the effusions, but the VA at last follow up remained only 6/60. The fellow eye was uninvolved.

**Patient 5** presented with an inferior bullous RD, choroidal effusion, and 6/9 VA. Rheumatoid factor was weakly positive, antinuclear antibody positive but other tests including C-reactive protein and erithrocyte sedimentation rate were normal. There were no ocular or systemic symptoms of vasculitic disease. The patient was treated initially with non-steroidal anti-inflammatory drugs and then systemic steroids without response, and was subsequently diagnosed as having idiopathic UES. At the time of surgery there had been an effusion for 500 days, with a total RD and a mottled appearing retina. Tortuous veins were noted with a nipped appearance as they exited the sclera. Following two inferior sclerectomies a superior effusion developed and this required a second operation with two superior sclerectomies. VA one month after this last operation was 1/60, but at most recent follow-up was only HMs with persisting subretinal fluid.

**Patient 6** presented with a bullous inferior RD with macular involvement, shifting subretinal fluid, and annular choroidal detachment. Fundus changes were present for 80 days before surgery. The sclera was noted to be thickened in the fellow eye on ultrasound examination but normal in the affected eye, however, at the time of surgery the sclera was both thickened and rigid. Inferonasal and inferotemporal quadrant sclerectomies were fashioned using a 6 by 7 mm superficial flap and 4 by 4 mm deep sclerectomy down to choroid. The superficial flap was closed with 7-0 Vicryl sutures. There was complete resolution of fluid within 44 days, and initial improvement in VA from HMs to 2/36. Later, macular pigmentary changes were associated with a final
Patient 7 was a high-hypermetrope who presented with a superior choroidal effusion, bullous inferior RD, and shifting subretinal fluid. The history suggested the macular was detached for one week but the duration of the effusion was not known. The ora serrata was clearly visible without indentation. Fluorescein angiography showed some vascular leakage but examination did not reveal any evidence of inflammation or any other cause of a serous RD. Like the involved eye, the fellow eye was also nanophthalmic, but appeared otherwise normal. Initial surgery involved four 3 by 4 mm partial thickness sclerectomies. This failed to treat the effusion or RD. A second operation six weeks later, involved two full-thickness sclerectomies. This produced a clinical improvement, but complete resolution of the RD and effusion took 11.5 months, with a final vision of 6/12.

Patient 8 presented with a 7 day history of visual loss OD. Examination revealed a bullous RD and mild to moderately elevated annular choroidal effusions. ‘Leopard spot’ fundal changes were noted, but shifting subretinal fluid was not demonstrated. Past history included right estropia with a squint operation at age 6 years, and associated right amblyopia. There was a systemic history of angina. A vitrectomy was undertaken to exclude rhegmatogenous RD, but this failed to demonstrate a retinal break, and thickened sclera was noted. A 90%-thickness 10 x10 mm sclerectomy was fashioned with internal drainage of subretinal fluid and injection of intraocular silicone oil tamponade. The silicone oil was removed 3 months later with complete resolution of the effusion and RD. Three and a half years later the left eye presented with choroidal effusions and serous RD. This eye therefore underwent two
sclerectomies without vitrectomy. This resulted in complete resolution of the effusion and RD. ‘Leopard spots’ were noted postoperatively in this eye. Final vision was 6/24 right, 6/9 left.

**Patient 9** presented with annular choroidal effusions such that the ora serrata was visible without indentation, 2 months after uncomplicated cataract surgery. VA was 6/24. There was a history of rheumatoid arthritis. The fellow eye was normal. Fluorescein angiography did not show any leakage. A RD with shifting subretinal fluid developed 11 months after the choroidal elevation and 8 months later the patient underwent 2 inferior quadrant sclerectomies with some resolution of fluid. Immediately after surgery the patient developed a rhegmatogenous RD due to a paravascular break in a fixed retinal fold. This was thought to be due to resolution of the effusion with subsequent tension in chronically elevated and shortened retina. The patient required vitrectomy and oil injection with two inferior sclerectomies and tight closure of the upper scleral windows. Light microscopy of the gluteraldehyde fixed specimen processed in the study laboratory showed characteristic features of uveal effusion syndrome, with disorganised collagen, interfibrillary deposits, and thickening. However, a specimen processed in the treating hospital by transmission electron microscopy did not find proteoglycan deposition between the collagen bundles. Numerous small calcific and lipoid bodies were scattered randomly through the collagen fibrils. Collagen diameter was noted to be 72 to 254 nanometers. Despite this apparent discrepancy the clinical appearances remained consistent with a diagnosis of uveal effusion syndrome and the patient continued to meet the entry criteria for this study. At last follow-up the patient had a persisting shallow inferior RD, choroidal effusions, vision of 2/60, and no plan to remove the silicone oil.
Patient 10 This 44 year old hypermetropic lady presented with a left, inferior, macular sparing retinal detachment associated with peripheral choroidal effusions. Past history included bilateral narrow angle glaucoma with peripheral iridectomies, and longterm treatment with timoptol and bimatoprost eyedrops in both eyes. After six months there was increasing subretinal fluid, that had extended to the inferior vascular arcade. In the year after presentation she underwent left deep sclerectomy in two sites, which resulted in resolution of fluid over a four month interval. She then represented with right photopsia and was found to have an inferior retinal detachment with a choroidal effusion. The macular was not involved. Four years after initial presentation she underwent right, deep sclerectomies in two sites, with a full resolution of fluid within 60 days. Surgery comprised 3 by 4 mm full thickness sclerectomy under a 4 by 5 mm scleral flap. Light microscopy of wax embedded material showed increased acidic mucopolysaccharides within slightly expanded interfibrillary spaces. Electron microscopy revealed thickened scleral fibrils that were noted to have a feathered appearance peripherally. Final visual acuity was 6/5\(^1\) right, 6/6\(^2\) left.