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Sir,

Surgical repair of bilateral full thickness macular holes in a patient with blue sclera secondary to osteogenesis imperfecta

The management of full thickness macular holes (FTMH) in patients with osteogenesis imperfecta has not to our knowledge been reported. We describe a patient with blue sclera secondary to osteogenesis imperfecta, who underwent surgical repair of bilateral FTMH.

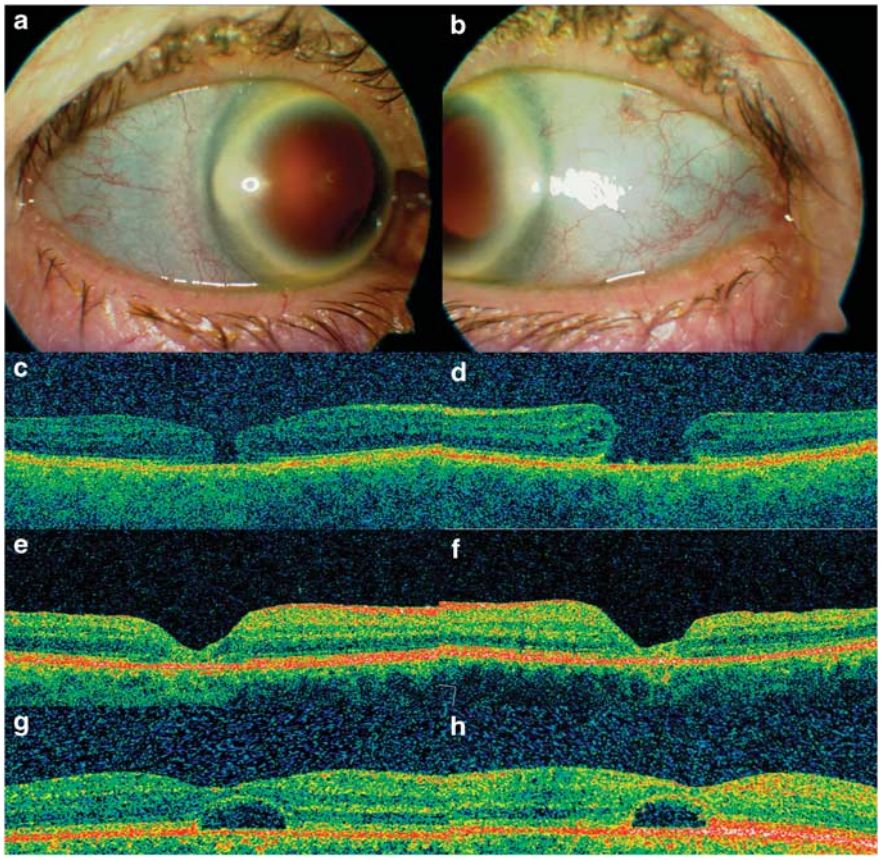

Figure 1 Photographs showing bilateral blue-grey scleral colouration OD (a) and OS (b). Pre-treatment OCT appearance of FTMH OD (c) and OS (d). Post-treatment OCT appearance showing closure of FTMH OD (e) and OS (f). Punched-out retinal defect on OCT in patient's son OD (g) and OS (h).

Case report

A 73-year-old lady was referred complaining of a 2-month history of reduced central vision in the right eye. Her left vision had been poor since childhood and she had not noticed a change. She was hypermetropic with spherical equivalents of +7.00 OD and +6.50 OS, and had blue sclera secondary to osteogenesis imperfecta type 1.

Examination revealed LogMAR acuities of 0.46 OD and 0.90 OS. Blue sclera was noted in both the eyes (Figures 1a and b) together with nuclear sclerotic cataract and bilateral FTMH (Figures 1c and d).

The amblyopic left eye had initial surgery in order to assess possible technical difficulties associated with the blue sclera and high hypermetropia. The left eye underwent 20 g combined phaco-vitrectomy, inner limiting membrane peel, 20% C_2F_6 gas tamponade and she postured face down for 5 days. Following successful closure of the left macular hole (Figure 1f) she underwent a similar procedure to the right eye but with 23 g instrumentation leading to successful closure of the right macula hole (Figure 1e). Postoperative corrected LogMAR acuities were 0.16 OD and 0.94 OS.

The patient's 46 year-old son, who had been registered blind since childhood was also examined. He did not have blue sclera but had a circular foveal abnormality bilaterally with punched-out retinal defects on OCT that have been described in cone dystrophy and Stargardt disease (Figures 1g and h).

Comment

Osteogenesis imperfecta is an inherited connective tissue disorder resulting from deficiency of type 1 collagen. The main ocular manifestation is blue sclerathat has been shown to be associated with reduced scleral rigidity[1] and it has been proposed that this may lead to increased susceptibility to retinal detachment[2] and retinal haemorrhage.[3] Our literature search did not reveal any reported association with FTMH.

Vitrectomy surgery in patients with blue sclera secondary to osteogenesis imperfecta has been described in the treatment of retinal detachment[4] and vitreous opacities,[5] the latter employing 25 g sutureless vitrectomy. We describe the treatment of FTMHs in such a patient, employing both 20 and 23 g vitrectomy. Ports were sutured in both the 20 and 23 g procedures as the surgeon (PRS) felt than the lack of scleral rigidity and thinness did not allow self-sealing of scleral tunnels. No operative complications were encountered.

Conflict of interest

The authors declare no conflict of interest.

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Sir,

Paediatric adjustable strabismus surgery

We commend Nihalani and Hunter[1] on an excellent review of the adjustable suture strabismus surgery literature, especially with regard to paediatric squints. As the authors mention, the benefits of adjustable sutures have been demonstrated in both adults and children, although there is a general reluctance in attempting this technique in children owing to the perception that it would not be tolerated by the patient.

Between 2007 and 2010, our experience of adjustable suture use in a paediatric population aged between 8–15 years (mean 12.3 years old) reflects the benefits of this technique. Our current practice involves a hangback technique with a 6/0 vicryl adjustable, slip-knot tie, followed by sub-Tenon's levobupivacaine (5 mg/ml). Within 2–3 h of recovery from general anaesthesia, topical tetracaine hydrochloride 1% is instilled and a prism cover test is performed. The suture is adjusted as required and the conjunctiva closed with 8-0 vicryl in the treatment room. Ninety-five percent (20/21) of our paediatric patients were able to tolerate this technique, with only one requiring a second general anaesthetic. The vast majority (90%) had a resultant angle <10 prism dioptres (Figure 1).

While we agree that a certain amount of experience is required with adjustable sutures for this population, we feel the most important factor is suitable patient selection and thorough preoperative counselling of both the patient and the parents. We find that with suitable scrutiny it is possible to identify those who will tolerate the adjustment, such that the surgeon should not feel intimidated by the prospect of adjustment in this group of patients.