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CORRESPONDENCE

Sir,

Delayed vitreous haemorrhage after paediatric cataract surgery in Lowe syndrome

Delayed vitreous haemorrhage is an exceedingly rare complication of cataract surgery; in the paediatric population, it can result in significant visual morbidity. The case presented demonstrates that Lowe syndrome patients can have disorders of primary haemostasis, and may be predisposed to this complication.

Case report

A 4-week-old term male infant presented with absent red reflexes bilaterally. Examination revealed bilateral dense cataracts, with no view to the posterior segment. B-scan ultrasonography was unremarkable.

The patient underwent left lensectomy and anterior vitrectomy. Fundus examination immediately post-operatively revealed a small peripapillary haemorrhage, which remained unchanged at 1 day post-operatively.

Two weeks later, uncomplicated cataract surgery was performed on the right eye. Simultaneous examination under anaesthesia of the left eye revealed corneal haze, hyphaema, and a dense vitreous haemorrhage (VH). There was no retinal detachment (RD) on B-scan. A pars plana vitrectomy (PPV) was planned, but the patient was lost to follow-up. When the PPV was performed 3 weeks later, a funnel RD with significant proliferative vitreoretinopathy was noted. Densiron-fluid exchange was also performed. The patient was again lost to follow-up and returned 3 months later, with a total fibrotic RD (Figure 1). The family declined further surgery, and at last follow-up, the left eye was phthisical.

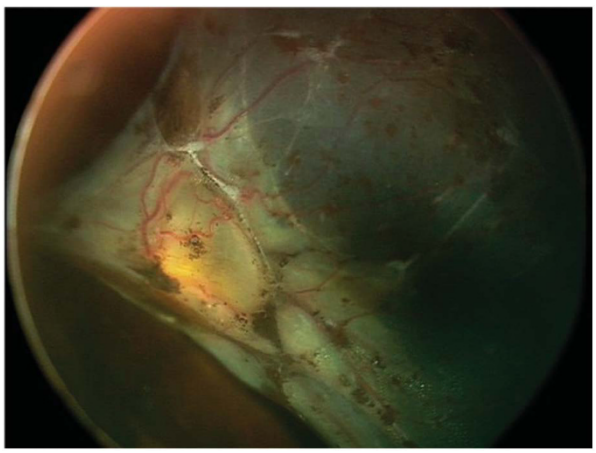

Figure 1 Post-operative and fundus examination of the left eye revealing a retinal detachment, and significant proliferative vitreoretinopathy.

Genetic testing revealed a c.940-11 G>A splice site mutation in the oculocerebrorenal gene (*OCRL-1*), confirming the diagnosis of Lowe syndrome. The patient subsequently developed the classic findings of renal tubulopathy, hypotonia, Rickets syndrome, and developmental delay. Testing revealed abnormal platelet function (Table 1).

Comment

Lowe syndrome is an X-linked recessive disorder, associated with mutations in the *OCRL-1* gene (locus Xq26).[1,2] Affected males develop bilateral cataracts requiring surgery.[3,4] Delayed VH is an exceedingly rare complication of paediatric cataract surgery.[5,6] There have been isolated reports of intraoperative hyphaema in Lowe syndrome patients undergoing cataract and goniosurgery.[7–9] To our knowledge, this is the first reported case of delayed post-operative VH after routine cataract surgery in a Lowe syndrome patient.

The *OCRL* gene regulates signalling pathways essential for normal platelet function.[10–12] OCRL mutations have been linked to systemic haemorrhagic events. Routine coagulation testing can be normal, but many patients have increased platelet closure times. The Platelet Function Analyser (PFA-100) test measures platelet adhesion and aggregation.[13,14] It can be elevated in primary haemostatic disorders, as was the case in our Lowe syndrome patient.

Surgeons need to be aware of systemic haemostatic disorders in Lowe syndrome patients undergoing cataract surgery. Routine haematological testing may be within normal limits. More specialised testing may be necessary to identify this increased bleeding risk.

Conflict of interest

The authors declare no conflict of interest.

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Table 1 Haematological testing revealing elevated PFA-100 closure times, and otherwise normal coagulation profile and complete blood count

PFA-100 closure time		*Coagulation profile*			*Complete blood count*	
EPI (seconds) (N = 60–115)	ADP (seconds) (N = 75–170)	INR (N = 0.90–1.10)	PT (seconds) (N = 12.71–15.1)	aPTT (seconds) (N = 31.6–43.5)	Haematocrit (%) (N = 34–40)	Platelets (10^9/l) (N = 140–450)
142	226	0.93	13.1	29.2	34%	338

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Sir,
Glaucoma and retained-triamcinolone in pediatric cataract surgery

Herein we describe a case report of a child operated for bilateral congenital cataract receiving identical treatment in both eyes, where glaucoma developed in the eye with long-term retained-triamcinolone, but not in the eye without it.

Case report
A 6-week-old infant presented to one of us (MEW) with bilateral cataract. Cataract surgery with limbal approach primary posterior capsulectomy and vitrectomy, and intraocular lens (IOL) implantation (30 D Rayner 570C, East Sussex, UK) were performed for both eyes. Two milligrams (0.05 ml) of intracameral preservative-free triamcinolone acetonide (Triesence, Alcon, Fort Worth, TX, USA) was used in both eyes. In addition to being an anti-inflammatory agent, triamcinolone acetonide helps to identify any residual vitreous strands in the anterior chamber,[1] an important advantage in younger children who routinely undergo planned vitrectomy.

Residual anterior chamber triamcinolone was noted in both eyes on the first postoperative day and at 1 week after surgery. However, at the 3-week postoperative visit, triamcinolone was not detected. Surgical removal of visual axis opacification (VAO) was required in both eyes (3 and 8 weeks postoperatively in left and right eyes, respectively). Triamcinolone was not used during the surgical removal of VAO.

Six months after surgery, the patient was noted to have an asymmetric myopic shift, greater in the left eye than in the right eye. As IOP measurement was not possible in the clinic, timolol eye drops were prescribed and examination under anesthesia (EUA) was scheduled. Axial elongation and myopic shift of refraction was documented in the left eye at the time of EUA (Table 1). Phospholine iodine 0.125%, b.i.d., was added as a topical drop for left eye. One month later (7 months post surgery), a 360-degree suture trabeculotomy *ab externo* was performed in left eye along with an IOL exchange (21 D Rayner 570C, East Sussex, UK). No residual triamcinolone was detected and no additional triamcinolone was used. An additional EUA was performed at 10 months after cataract surgery because the IOP could not be obtained in the office. Retained-triamcinolone was detected in the vitreous cavity of the left eye. None was found in the right eye. The left eye underwent a *pars plana* vitrectomy and removal of triamcinolone. Several large 'chunks' of triamcinolone were cut and aspirated without difficulty. IOP remained under control in both eyes until last follow-up at 3 years after cataract surgery.

Comment
In the case reported here, intracameral triamcinolone acetonide made its way into the vitreous cavity in the left eye and escaped detection for >10 months. In the many of our pediatric cataract and IOL surgeries, the triamcinolone acetonide remained in the anterior chamber and disappeared over a 3- to 14-day period. The lack of spread into the vitreous cavity in most cases is likely due to the capsular-fixated IOL that acts as a sufficient barrier to migration from the anterior chamber into the vitreous cavity. We do not