

UK Paediatric Glaucoma Society (UKPGS) Annual Meeting
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Approved CPD 6 points (Royal College of Ophthalmologists)

Abstracts

27 - Case report of monozygotic twins with Hallerman-Streiff syndrome presenting with microphthalmia, bilateral congenital cataracts, posterior synechiae and early-onset glaucoma

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Introduction: Hallerman-Streiff syndrome is characterised by bird-like faces with mandibular hypoplasia, abnormal dentition, proportionate dwarfism, hypotrichosis, microphthalmia, and congenital cataracts. Ocular manifestations, present in about 90% of cases, also include glaucoma, retinal detachment, uveitis, and iris synechiae.

Case Description: Monozygotic twins born at 32 weeks were found to have features consistent with Hallerman-Streiff syndrome, including microphthalmia and bilateral congenital cataracts. One infant had spontaneous resorption of both cataracts prior to age 4 months with early-onset of glaucoma and anterior uveitis. She underwent extraction of residual capsular material, however developed exudative retinal detachments, which intermittently progressed and resolved spontaneously. IOP in the right eye fluctuated, and the left eye maintained high pressures in the 40's mmHg despite medical therapy. Her sister conversely did not show spontaneous cataract resorption and underwent cataract extraction at 3 months. Around 5 months postop, she developed peripapillary exudates, which progressed to bilateral exudative detachments similar to her sibling. Her IOP ranged from 15 to 35 mmHg, with variable response to topical medications.

Discussion: Previous case reports suggest spontaneous resorption of the cataractous lens may be associated with ocular inflammation and glaucoma (Hopkins & Horan, 1970). Here we describe two cases from monozygotic twins, presenting with posterior synechiae suggestive of intraocular inflammation prior to cataract surgery. Both twins developed exudative retinal detachments and intermittent IOP elevation, which were difficult to control medically. We will discuss the challenges of how to surgically manage glaucoma in the microphthalmic eye with uveitis and exudative detachment in the setting of Hallerman-Streiff syndrome.