Aqueous misdirection, also known as ciliary block glaucoma or malignant glaucoma, is a rare type of secondary angle-closure glaucoma in which aqueous humor is abnormally directed posteriorly into and/or behind the vitreous humor. This posterior accumulation of aqueous humor causes anterior movement of the vitreous, lens, and iris, which ultimately leads to a flattening of the anterior chamber, iridocorneal angle closure, acute ocular hypertension, and glaucoma.

Though first described by von Graefe in 1869, the root cause of aqueous misdirection remains unclear. One hypothesis is that an abnormal seal between the ciliary body and the lens or anterior hyaloid directs aqueous humor posteriorly, while another hypothesis suggests a role for choroidal expansion and abnormal vitreous fluid conductivity. Aqueous misdirection most typically occurs following filtering surgeries for chronic angle-closure glaucoma, although it can also occur after other ocular surgeries or rarely spontaneously. The overall incidence of aqueous misdirection has been estimated to be 2% to 4% after glaucoma procedures. A history of hyperopia, microphthalmos, nanophthalmos, or chronic angle closure increases the risk of aqueous misdirection.

Patients with Down syndrome have a range of refractive errors, however, a subset of patients have hyperopic eyes and are theoretically at higher risk of aqueous misdirection. Two cases of spontaneous aqueous misdirection have been reported in association with keratoconus and corneal hydrops in patients with Down syndrome. We report the first case of aqueous misdirection in a patient with Down syndrome after a trabeculectomy procedure.

CASE REPORT

A 44-year-old male with Down syndrome was referred for the evaluation of narrow angles. On initial examination, his visual acuity was 20/20 in the right eye and count fingers in the left eye, and his intraocular pressures (IOPs) were 15 and 34 mm Hg in the right and left eyes, respectively. The patient was found to have shallow anterior chambers with completely closed angles on gonioscopy in both eyes. The cup-to-disc ratios were 0.4 and 0.9 in the right and left eyes, respectively. Because general anesthesia was required for this patient, both eyes were operated upon in a single session. A trabeculectomy with mitomycin C was performed in the left eye, and a surgical iridectomy was performed in the right eye. There were no intraoperative complications, and the immediate postoperative course was uncomplicated.

Three weeks after the trabeculectomy, the patient presented with a flat anterior chamber of the left eye with an IOP of 32 mm Hg. No evidence of suprachoroidal hemorrhage or choroidal effusions were seen on ophthalmoscopy or with ultrasound, and the patient was diagnosed with aqueous misdirection. Atropine and acetazolamide were started, resulting in lowering of the IOP to 12 mm Hg and deepening of the anterior chamber 2 days later. After atropine and acetazolamide were tapered and discontinued, the patient presented with recurrence of aqueous misdirection in the left eye with an elevated IOP and a shallow anterior chamber. The patient developed recurrent episodes of aqueous misdirection until atropine and acetazolamide were tapered at a very slow rate over a 5-year period and eventually discontinued without recurrence.

Six years after initial presentation, an Ahmed seton was placed with a concurrent pars plana vitrectomy and cataract surgery in the right eye. Aqueous misdirection was not encountered at any point in the right eye. At last follow-up, the vision was 20/80 in the right eye and counting fingers in the left eye with an elevated IOP and a shallow anterior chamber. The patient was found to have shallow anterior chambers and completely closed angles on gonioscopy in both eyes. The cup-to-disc ratios were 0.4 and 0.9 in the right and left eyes, respectively. Because general anesthesia was required for this patient, both eyes were operated upon in a single session. A trabeculectomy with mitomycin C was performed in the left eye, and a surgical iridectomy was performed in the right eye. There were no intraoperative complications, and the immediate postoperative course was uncomplicated.

Patient Consent

The research was approved by the University of Iowa’s Institutional Review Board (#202003353), which granted a waiver of informed consent for our retrospective data review of the medical chart.

DISCUSSION

Down syndrome is the most common chromosomal disorder with an estimated incidence of 1 in 700 births in the United States. Down syndrome has been associated with a wide range of abnormalities of the eyes, including refractive error, glaucoma, cataract, strabismus, nystagmus, nasolacrimal duct obstruction, Brushfield spots of the iris, and keratoconus. Two cases of spontaneous aqueous misdirection in patients with Down syndrome.
syndrome have been reported in association with keratoconus and corneal hydrops.\textsuperscript{2,20} Case-series studies identified glaucoma in 0% to 6.7% of patients with Down syndrome.\textsuperscript{12-16,18} However, the type of glaucoma was not specified. A substantial proportion of patients with Down syndrome have hyperopia (13% to 59%) or high hyperopia (0% to 6.3%),\textsuperscript{12-19} which suggests that angle closure may be a frequent mechanism of disease.

Aqueous misdirection should be suspected in patients with Down syndrome who have flat anterior chambers and elevated IOP, especially in the postoperative setting or with corneal hydrops. Medical management for aqueous misdirection with cycloplegics and aqueous suppressants was effective in our patient with Down syndrome. To maintain the anterior chamber and mitigate aqueous misdirection, cycloplegics may be used immediately after anterior chamber and suprachoroidal injection of aqueous shunt with vitrectomy in malignant glaucoma. To our knowledge, medical management was not required for our patient. A prophylactic vitrectomy and a pars plana seton should be considered when glaucoma surgery is indicated in patients with Down syndrome that may be at higher risk for aqueous misdirection, such as those with hyperopia, angle-closure glaucoma, and especially a history of aqueous misdirection in the fellow eye.\textsuperscript{20}

CONCLUSIONS

Hyperopia, narrow iridocorneal angles, and the associated risk for angle-closure glaucoma are relatively common in patients with Down syndrome. We present the first reported case of a patient with Down syndrome who developed aqueous misdirection after trabeculectomy. It is unclear whether patients with Down syndrome, like our patient, who are treated with a trabeculectomy are at increased risk for aqueous misdirection due primarily to underlying features of hyperopia and angle-closure glaucoma or due to intrinsic features of Down syndrome. Regardless of the cause, it may be prudent for ophthalmologists to consider the potential for aqueous misdirection when performing a trabeculectomy for patients with Down syndrome.

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