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Transconjunctival XEN45 implantation for secondary open-angle glaucoma management in a pediatric patient with WAGR syndrome

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ABSTRACT

Purpose: To report a case of XEN45 gel stent implantation in a pediatric patient with WAGR syndrome as a successful surgical intervention in the management of multifactorial secondary open-angle glaucoma.

Observations: A 6-year-old female with a history of WAGR syndrome, bilateral congenital aniridia, pseudophakia OD and glaucoma OD, was referred for a XEN45 gel stent OD. IOP was persistently elevated at 24 mm Hg despite two glaucoma medications. Implantation of the XEN45 gel stent was performed using a transconjunctival ab externo approach. There were no significant intra-or-postoperative adverse events associated with the stent. The patient achieved good IOP-lowering control without glaucoma medications across the 18-month follow-up period.

Conclusions: A XEN45 stent through a transconjunctival ab externo approach may be an effective surgical intervention in pediatric patients with secondary open-angle glaucoma associated with aniridia and aphakia.

1. Introduction

An example of glaucoma associated with non-acquired ocular anomalies is aniridia, a rare congenital eye disease that can be associated with Wilms' tumor, aniridia, genitourinary anomalies, and mental retardation (WAGR) syndrome caused by heterozygous contiguous gene deletions of PAX6 and Wt1 genes.[1] Aniridia affects the iris and other intraocular structures.[1] Although the term "aniridia" suggests the absence of an iris, presentations may range from severely hypoplastic iris tissue to a normal appearing iris.[2,3] Glaucoma secondary to aniridia has thought to occur due to the absence or abnormality of Schlemm's canal and/or trabecular meshwork dysgenesis, synechiae or ridge tissue formation between the iris stump and angle wall, or from anterior rotation of the iris stump.[4] Furthermore, as cataracts are a common complication of aniridia[4], glaucoma following cataract surgery (GFCS) can also develop secondary to lensectomy or intraocular lens implantation. Glaucoma treatment in both aniridia and GFCS can be challenging in pediatric patients and may require multiple medical and surgical interventions.

An emerging treatment modality for lowering intraocular pressure (IOP) in glaucoma patients is microinvasive glaucoma surgery (MIGS). These surgeries typically involve minimal scleral or conjunctival dissection, have good safety profiles, and have well-demonstrated IOP-lowering ability.[5] One MIGS device is the XEN45 gel stent (Allergan, Irvine, CA), a collagen implant typically placed using an ab interno approach into the anterior chamber that tunnels through the sclera to drain aqueous humor into the subconjunctival space.[6] Alternatively, some surgeons have begun using an ab externo approach where the stent is placed from the outside of the eye and may be done with or without conjunctival dissection.[7]

In this report, we describe the implantation of a XEN45 gel stent using a transconjunctival ab externo approach in a pediatric patient with WAGR syndrome and secondary open-angle glaucoma (SOAG) associated with congenital aniridia and aphakia.

2. Case report

A 6-year-old female was referred for surgical glaucoma management of the right eye (OD) as a treatment modality for refractory glaucoma secondary to congenital aniridia and aphakia in 2021. The patient was diagnosed with WAGR syndrome in 2018 with no relevant family history, signs of systemic disease, or other health concerns. Relevant ocular history included congenital bilateral aniridia, nuclear cataract OD, SOAG OD, amblyopia OD, exotropia OD, nystagmus in both eyes (OU),

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limbal stem cell deficiency secondary to aniridia OD, limbal keratitis OU and corneal scarring OD. Nuclear cataract lensectomy OD was performed in 2015 and a posterior chamber intraocular lens OD was implanted in 2018 due to inability to tolerate aphakic contact lenses and to prevent further limbal cell loss and limbal keratitis. Eye patching of the left eye (OS) for amblyopia was initiated in 2018. EUA scheduled in March 2019 confirmed a diagnosis of glaucoma OD. Initial medical management included combination brinzolamide 1% and timolol 0.5% OD twice daily (bid), bimatroprost 0.01% OD nightly (qhs), 1 mg/ml sodium hyaluronate drops OU bid, and moisturizing ointment OU qhs. In February 2020, IOP was 24 mm Hg OD and 14 mm Hg OS which remained elevated six months later (24 mm Hg OD and 17 mm Hg OS) (Tonopen). With persistently elevated IOP and signs of optic nerve damage despite medical management, the risks and benefits of various surgical interventions were discussed with the family, and a XEN45 gel stent implant was scheduled OD.

XEN45 gel stent implantation (Fig. 1) was performed under general anesthetic in February of 2021 at Royal University Hospital in Saskatoon, Saskatchewan. Prior to surgery, visual acuity (VA) was 20/400 OD and 20/150 OS, and the cup-to-disk ratio was 0.6 OD and 0.1 OS. Eyes were EUA and IOP readings were 14 mm Hg OD and 20 mm Hg OS (Tonopen). Corneas were clear, anterior chambers were deep and formed, aniridia was present OU, and limbal corneal vascularization was prominent OD. Fundus examination was normal bilaterally. A 7-0 vicryl suture was inserted into the superior corneal limbus to bring the eye into infraduction. The sclera was marked approximately 2.5mm posterior to the limbus and 3mm posterior to the initial mark (Fig. 1A). A temporal paracentesis was created and viscoelastic was injected prior to the insertion of the stent. The XEN45 gel stent was inserted under the subconjunctival space (Fig. 1B) and injected approximately 2.5mm from the limbus (Fig. 1C). Bleb formation immediately followed (Fig. 1D). Once the stent was adequately positioned within the anterior chamber, incision sites were closed with 9-0 vicryl sutures and 0.4mg/ml mitomycin C was injected into the subconjunctival space. Combination tobramycin and dexamethasone drops were applied prior to patching and shielding.

Postoperatively, the patient was prescribed difluprednate and moxifloxacin drops for 2 weeks. All previous IOP-lowering medications were discontinued. On postoperative day 1, IOP was 10 mmHg OD. At two weeks follow-up, IOP remained stable at 10 mmHg OD and VA was 20/400 OD and 20/150 OS. IOP remained stable at both the 12-month (7 mmHg OD) and 18-month (7 mmHg OD) appointments. At 18-months, VA remained stable at 20/400-2 OD and 20/125-3 OS.

3. Discussion

Here we present a case of transconjunctival ab externo XEN45 stent implantation in a pediatric patient with WAGR syndrome and multifactorial SOAG. The XEN45 gel stent showed good safety and efficacy, with no significant intra-or-postoperative adverse events and good IOP control for 18 months postoperatively.

It is imperative to comment on the relatively low IOP recorded during the EUA prior to surgery compared to previous clinic measurements. Multiple factors can affect IOP pressure readings before surgery such as breath-holding, Valsalva maneuvers, corneal irregularities, and inhalational anesthetic agents.[8,9] As inhalational anesthetics can significantly impact IOP,[9] the IOP reading OD prior to surgery did not guide treatment. Rather, persistently elevated pressures in clinic with the presence of glaucomatous damage indicated necessity for surgery.

Medical therapy is generally used for the initial treatment of glaucoma regardless of etiology. However, given the complex nature of aniridic glaucoma, medical therapy often fails.[10] Trabeculectomy has long been considered the gold-standard for surgical treatment of aniridic glaucoma even though it is associated with a relatively high failure rate in congenital aniridic glaucoma.[11,12] Poor outcomes with trabeculotomy in GFCS have also been cited, especially in pediatric patients diagnosed under the age of one and who remain aphakic.[13] Alternatively, MIGS procedures have gradually become more common and may change the approach to complex pediatric glaucoma treatment.

There have been a handful of studies that evaluated the use of the XEN45 stent in cases of pediatric glaucoma though to our knowledge, there is no case that has described the use of the transconjunctival ab externo approach XEN45 stent implantation in pediatric multifactorial SOAG. Lindland et al., used the ab interno approach to XEN45 implantation in a 60-year-old male with congenital aniridic glaucoma and achieved good IOP control at two years alongside timolol monotherapy.[14] Additionally, Ruparelia et al., performed an open conjunctiva ab externo XEN45 implant on a 10-year-old boy with aphakic glaucoma secondary to cataract surgery at 8 weeks old.[15] His original XEN implant showed good IOP control for six months, however, conjunctival dehiscence and contraction occurred, resulting in stent removal. A replacement ab interno XEN was implanted and good IOP control was achieved for the following six months. Arad et al., described an XEN-augmented Baerveldt technique in 10 cases of refractory pediatric glaucoma where previous surgical interventions had failed. Short term results were promising with a median follow-up period of 13 months and IOP levels $<$20 mm Hg without adjunctive medication therapy in 6 of the 10 cases.[16]

A study performed by Smith et al., is one of few where a XEN was performed using the transconjunctival ab externo approach in a child.[17] The stent was used as a revision procedure after a failed trabeculectomy and ab interno XEN. They achieved good IOP lowering control for six months postoperatively. They accredited this to the greater control of distal stent placement with the ab externo approach, allowing the surgeon to confirm that the stent remains subconjunctival and not sub-tenons.

To the best of our knowledge, we report the first case of a transconjunctival ab externo approach to XEN45 stent implantation as the initial surgical therapy in pediatric SOAG associated with aniridia and aphakia. Given the demonstrated IOP-lowering efficacy and favorable safety profile, it may be reasonable to use the XEN45 prior to trabeculectomy, a more invasive alternative for glaucoma surgery. As suggested by Mendel et al., an advantage of the XEN45 for initial surgical therapy is that it does not preclude the ability to perform additional surgery later, which is often required during the lifetime of congenital glaucoma patients.[18]

4. Conclusions

The transconjunctival ab externo XEN45 implantation achieved good IOP-lowering efficacy with 18 months of follow-up in our pediatric patient with WAGR syndrome and uncontrolled SOAG associated with aniridia and aphakia. The procedure demonstrated a favorable safety profile and may be a reasonable option for initial surgical therapy in the pediatric glaucoma population. Additional studies with larger sample sizes and follow-up durations are warranted to further evaluate the use of the XEN45 stent in this population.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

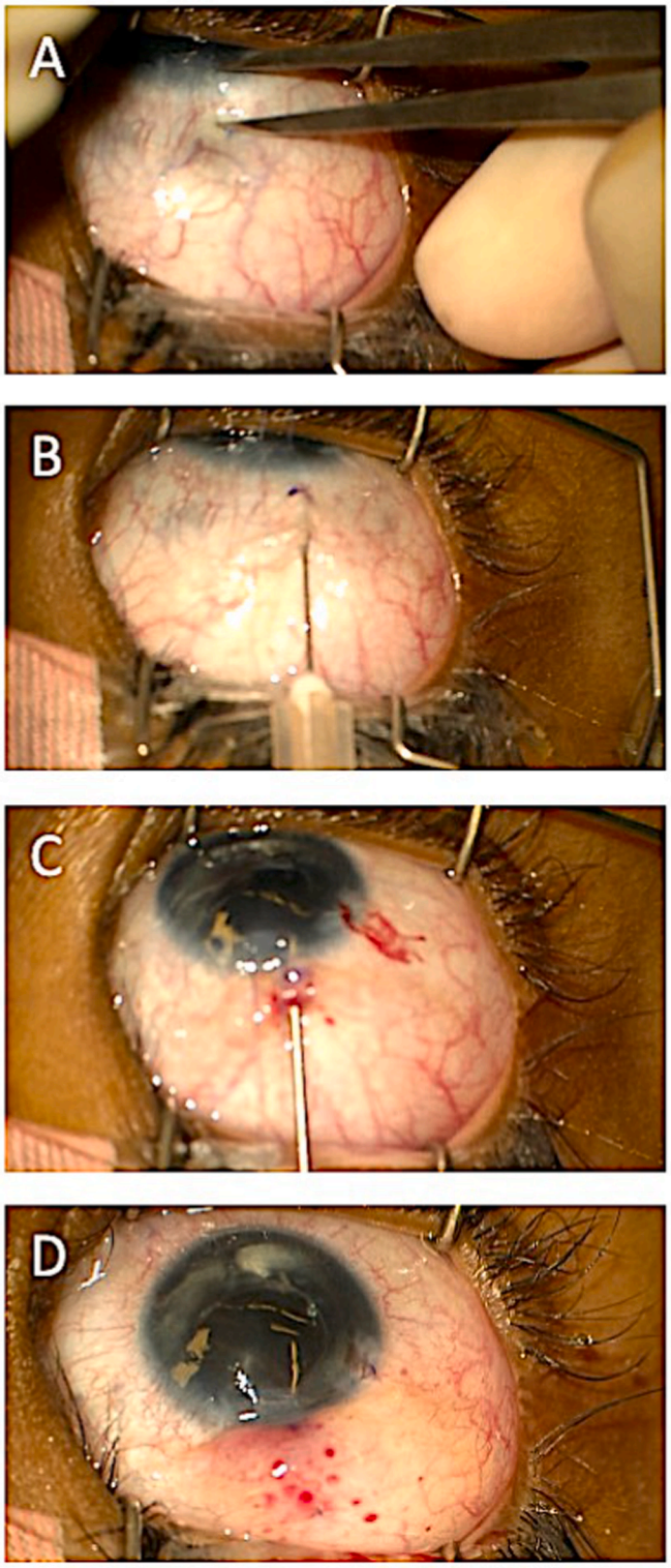

Fig. 1. Transconjunctival ab externo approach to XEN45 implantation demonstrating the initial 2.5mm marking of the conjunctiva (A), creation of a tunnel in the subconjunctival space (B), direction of the injector towards the sclera at the 2.5mm marking and insertion of the XEN45 stent into the anterior chamber (C), and bleb formation at the end of the case (D).

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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