

Case Report

Bilateral XEN Gel Stent Implantation in Juvenile-Onset Open-Angle Glaucoma

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Keywords

Juvenile-onset open-angle glaucoma · Minimally invasive glaucoma surgery · XEN Gel Stent · Glaucoma

Abstract

The purpose of the current case is to report the successful management of juvenile-onset open-angle glaucoma with bilateral XEN Gel Stent (Allegran Inc., Irvine, CA) implantation in a patient first diagnosed during pregnancy with co-existing dysautonomia. Treatment with the XEN Gel Stent provided dramatic reductions in intraocular pressure (IOP), and glaucoma medications sustained up to 23 months postoperatively. The success of this case may suggest that this minimally invasive procedure could be an effective treatment option for younger patients who require substantial reductions in IOP and glaucoma medication burden.

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Introduction

Juvenile-onset open-angle glaucoma (JOAG) is a rare subset of primary open-angle glaucoma (POAG) with an age at onset between 5 and 35 years [1]. It is often associated with a rapidly progressive disease that can be refractory to medical therapy. Surgical intervention is usually required to control intraocular pressure (IOP) and slow disease progression.

Minimally invasive, angle-based procedures are typically preferred in JOAG cases as they restore trabecular meshwork function without bypassing it via the creation of a filtering bleb [2]. They also avoid manipulation of the conjunctiva that could be needed for future surgical intervention. However, these procedures are generally less effective in reducing IOP compared to trabeculectomy and tube-shunt procedures [3].

The purpose of the current case report is to introduce another minimally invasive glaucoma surgery, the ab interno XEN Gel Stent, for the treatment of JOAG. While it requires bleb formation, an ab interno approach can provide excellent IOP reduction while sparing most of the patient's conjunctiva. To the best of our knowledge, there is only one existing report of the use of XEN Gel Stent in JOAG, with a focus on its management during pregnancy. The follow-up period of this report was limited to 2 months [4]. Herein, we report the 23-month follow-up of a non-pregnant JOAG patient who underwent successful XEN Gel Stent implantation in both eyes.

Case Presentation

A 48-year-old Caucasian woman presented to the Glaucoma Service at the Massachusetts Eye and Ear Infirmary in August 2018 for a routine 6-month IOP check for bilateral, mild-stage JOAG. The patient denied any pain or changes in vision. Her best-corrected visual acuity was 20/20 OD and 20/20 OS. IOP measurements were 19 mm Hg in the right eye and 38 mm Hg in the left eye. Gonioscopy showed grade 3 open angles. The anterior chamber and the lens were clear bilaterally, and the cup-to-disc ratios were 0.75 OD and 0.70 OS. Humphrey visual field analysis revealed a full field OD and scattered, non-specific defects OS. Optical coherence tomography showed an average retinal nerve fiber layer thickness of 65 μ m OD and 61 μ m OS.

The patient's glaucoma had been treated at an outside institution where elevated IOP was first noted at the age of 30 during her first pregnancy. Her maximum recorded IOP was 25 mm Hg OD and 47 mm Hg OS. She had previously undergone selective laser trabeculoplasty OU in 2007, and again OS in 2017. Her glaucoma was currently being treated with 4 topical medications OU: latanoprost q.h.s., timolol 0.5% b.i.d., brinzolamide t.i.d., and brimonidine t.i.d. This reliable patient endorsed excellent adherence to this complex medication regimen.

The patient had a positive family history of glaucoma, and she had received long-term oral prednisone for possible systemic lupus erythematosus in 2012, 6 years prior to presentation. She had not received any systemic steroids since that time. Genetic testing to rule out myocilin-associated JOAG was not performed. It is therefore possible that her glaucoma was attributable to multiple mechanisms, including both JOAG and steroid-induced glaucoma. Notably, the patient also had a current diagnosis of dysautonomia, a disorder of the autonomic nervous system that usually involves failure of the sympathetic or parasympathetic components of the autonomic nervous system [5]. She took many measures to manage a host of symptoms associated with the disorder, including variable heart rates and blood pressures. Her condition was being treated with pyridostigmine bromide, an orally active cholinesterase inhibitor.

When the patient's IOP was markedly elevated in August 2018, latanoprost was switched to latanoprostene bunod q.h.s. OU, netarsudil was added q.h.s. OU, and she was tentatively scheduled for a trabeculectomy with mitomycin-C OS. With these changes in the medication regimen, her IOP decreased to 17 mm Hg OD and 29 mm Hg OS prior to surgery. The trabeculectomy was cancelled in favor of a minimally invasive procedure according to patient preference and more in line with her age and glaucoma stage.

After consideration of the risks and benefits of various approaches, the surgeon and patient decided to proceed with XEN Gel Stent implantation OS in September 2018, in an ab interno fashion without opening the conjunctiva. Approximately 0.1 mL of mitomycin 0.4 mg/mL were injected subconjunctivally. On postoperative day 1, IOP measurements were 15 mm Hg OD and 02 mm Hg OS. The anterior chamber was deep in both eyes. The postoperative course consisted of 1 week of moxifloxacin therapy and atropine while the eye was hypotensive (IOP <5 mm Hg). Prednisolone acetate 1% was given 8, 6, and 4 times a day for 2 weeks each and was then tapered over the next 4 weeks. The patient remained on prednisolone acetate 1% once a day for an additional 2 weeks. Ketorolac was also tapered over 4 weeks.

While the left eye healed from surgery, IOP was maintained in the mid-teens in the right eye on the previously described topical regimen. In December 2018, the surgeon and the patient decided to proceed with XEN Gel Stent implantation in the right eye in an effort to decrease her glaucoma medication burden. The procedure was performed in a similar manner to above. On postoperative day 1, IOP measurements were 05 mm Hg OD and 11 mm Hg OS. The stent was slightly tented up in the conjunctiva, but it was successfully repositioned with a Q-tip. The postoperative course was similar to that of the left eye with the exception of ketorolac. After the initial tenting of the stent, ketorolac was avoided for fear of stent exposure, as significantly greater rates of conjunctival contraction have been observed in patients who received postoperative ketorolac compared to those who received steroid treatment after tube-shunt surgery [6].

At postoperative week 7.5, the patient presented to the clinic with an IOP of 19 mm Hg in the right eye off all glaucoma medications. Needling with subconjunctival injection of 5-fluorouracil (5-FU) was performed OD to reduce IOP and prevent scar tissue formation. The patient's IOP dropped to 14 mm Hg following the procedure. Over the next 16 months, IOP ranged from 05–19 mm Hg OD and 02–15 mm Hg OS off all glaucoma medications. Her best-corrected visual acuity remained 20/20 in each eye, and her visual fields remained stable and full in both eyes with scattered, non-specific defects OS. On optical coherence tomography, her average retinal nerve fiber layer thickness ranged from 58 to 64 OD and from 58 to 63 OS. At the last follow-up in July 2020, her IOP measurements were 13 mm Hg in each eye off all glaucoma medications. Images of the patient's filtering bleb in the right eye as of January 2020 are shown in Figure 1. The left eye had a similar appearance to the right.

Discussion

This case illustrates the successful management of JOAG with XEN Gel Stent implants. Both eyes achieved stable IOP control off all glaucoma medications up to 23 months postoperatively. At the most recent follow-up visit, IOP was reduced by 4 mm Hg (24%) OD and 16 mm Hg (55%) OS from preoperative measurements, and the number of IOP-lowering medications was reduced by 5 in each eye.

As mentioned previously, angle-based procedures such as goniotomy, ab interno trabeculectomy, or trabeculotomy are typically deployed as first-line treatment for JOAG. Specifically, Arora et al. [7] observed mean reductions of 10 mm Hg in IOP and 0.7 medications 12 months after ab interno trabeculectomy with the trabectome (NeoMedix Corp., Tustin, CA). Moreover, Yeung and Walton [2] observed a comparative 12.7 mm Hg reduction in IOP after goniotomy. IOP has also been shown to be reduced by 12.5–16 mm Hg following gonioscopy-assisted transluminal trabeculotomy, with reductions in medications ranging from 1.3–1.74 [8, 9]. More recently, Khouri et al. [10] suggested ab interno trabeculectomy with the Kahook

dual blade as an alternative to goniotomy, after achieving a >30% reduction in IOP 18 months postoperatively in a case report of a 14-year-old male with JOAG.

The substantial reductions in IOP and glaucoma medications observed in the current case align more closely with those reported following ab externo trabeculectomy in JOAG patients, which range from 16–22 mm Hg and 2.3–2.8 medications [11–13]. However, ab externo trabeculectomy is not recommended as a first-line treatment for JOAG despite the potential for greater IOP reduction [2, 7]. Rather, angle-based procedures are preferred as they spare the conjunctiva and avoid the potential of life-long bleb-related complications [2, 7, 8]. As bleb morphology is critical to ocular health, the potential for long-term bleb-related complications should be emphasized when discussing surgical options with these young patients.

The older age of the JOAG patient described in the current case (48 years) provided a stronger basis for the use of a procedure that creates a filtering bleb and could also provide potentially greater reductions in IOP and medication use. With that said, Smith et al. [14] reported the successful management of congenital glaucoma in 3 young children, with stable control of IOP off all medications and favorable bleb morphology up to 2 years after XEN surgery.

To the best of our knowledge, there is only one existing report of the use of bilateral XEN Gel Stents in JOAG. Similar to the current case, the authors report substantial reductions in IOP from >40 mm Hg preoperatively to 14 mm Hg in both eyes 2 months postoperatively [4]. Unlike the current case, their procedure was performed without intraoperative mitomycin-C due to pregnancy. The patient subsequently required multiple 5-FU augmented needling procedures. In contrast, our JOAG patient required a single needling procedure with subconjunctival 5-FU in the right eye.

The need for both postoperative needling and 5-FU injections following XEN Gel Stent insertion is fairly well documented [15]. Specifically, in several prospective studies of XEN insertion with subconjunctival mitomycin-C needling rates have ranged from 30.7–43.2%, although it has been reported as low as 2.4% [16–21]. In the present case, needling with an injection of 5-FU successfully reduced the IOP in the right eye from 19 to 14 mm Hg. Evidence suggests that 5-FU augmented needling revisions are both a safe and effective option for bleb management, with the potential for substantial IOP reductions [22].

Conclusion

The current case introduces the XEN Gel Stent as a minimally invasive treatment option for JOAG patients. Bilateral XEN Gel Stent implantation reduced this patient's IOP by 24% OD and 55% OS, and reduced the patient's glaucoma medication burden by 100% OU. The XEN Gel Stent may potentially provide greater reductions in IOP and glaucoma medication burden compared to angle-based procedures that are currently recommended as first-line treatment for JOAG. The XEN Gel Stent may particularly benefit JOAG patients who – while still young in comparison to the average POAG patient – may be of an age at which topical and systemic therapy has proved insufficient, and substantial reductions in IOP and medication burden are needed.

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Statement of Ethics

The patient consented to the publication of this case and the images therein in writing. All research was conducted in accordance with the Declaration of Helsinki.

Disclosure Statement

The authors declare that they have no conflicts of interest to disclose.

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Author Contributions

D.S.-V. was the attending physician of the case. E.K. researched the case, and both authors contributed to the writing and revising process.

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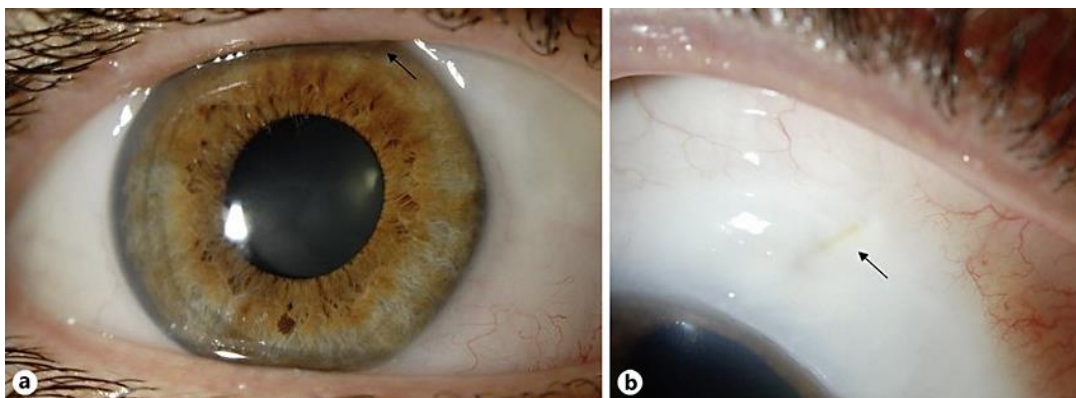


Fig. 1. Images of the filtering bleb in the right eye 16 months postoperatively. Arrows indicate the visible XEN Gel Stent. **a** View of the right eye head-on. **b** View of the right eye with the patient looking down.