

Challenges of Surgical Management of Glaucoma in Aniridia: A Case Series

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Introduction: Congenital Aniridia is a rare condition which is mainly associated with PAX6 gene mutations, though other genes such as FOXC1, PITX2 and CYP1B1 have also been implicated.¹ Glaucoma associated with Aniridia has been reported to be between 6% and 75% of patients² and though it usually occurs later in life, it can also be present at birth. Treatment of glaucoma is difficult and challenging³ due to the angle anomalies and the progressive angle changes that take place. Surgical intervention is the most commonly utilized mode of management to achieve good intraocular pressure (IOP) control.¹

Methods: Clinical records of patients who had surgical intervention for glaucoma with aniridia between May 2016 and June 2019 were reviewed. Relevant data was retrieved and analyzed.

Results: We report 4 patients with aniridia, 2 of which had associated bilateral glaucoma and the other 2 with unilateral glaucoma. All 6 eyes with aniridia and glaucoma had surgical intervention in

Table: Summary of the patient's surgeries and post-operative clinical state

Patients' initials	Age at diagnosis of glaucoma	IOP prior to Surgery; RE/LE (mmHg)	No of AGMs prior to surgery	Type of Surgery	IOP post-op day 1 (mmHg)	IOP 6m post-op (mmHg)	Other surgeries
I. H.	5 days	36/30	1	Biltrabs + 5FU	10/10	20/16	Bil GDD (fine tube)
S. M.	2 ½ yrs	24/18	2	Biltrabs + 5FU lens extraction of dislocated lens from LE, vitreous noted in LE	08/02 Total hyphema LE	13 RE LE phthisical	
K. J.	6 yrs	36/28	3	GDD (RE fine tube)	07 RE	10	RE temporal SICS + PCIOL 9m post op
					14 LE 3 AGMs	On 2 AGMs RE. 27 LE, on 4 AGMs	
O. A.	16 yrs	36 LE (only eye)	2	GDD (LE Ahmed tube)	15 LE	21 On 2 AGMs	

IOP= Intraocular pressure, RE= right eye, LE= left eye, AGM= anti-glaucoma medication, m= month, GDD= glaucoma drainage device, Bil= Bilateral, trab= trabeculectomy, 5FU = 5-Fluoro-uracil, SICS + PCIOL= Small incision cataract surgery with posterior chamber intraocular lens implantation

the period under review. Age range at presentation was 5 days – 16 years. Male to female ratio was 1:3. Four eyes had trabeculectomy surgery and two had glaucoma drainage device (GDD) surgery as primary procedure. Three eyes had more than one surgery. Of the three eyes that had more than one surgery, two eyes had GDD (fine tube) after trabeculectomy. One eye had other surgeries (capsulectomy for an encysted bleb and temporal small incision cataract surgery with posterior chamber intraocular lens). All patients had good IOP control (10 – 21mmHg) as at 6 months post-operatively. The table below gives a summary of the surgeries and post-operative clinical state of the patients.

Discussion: The treatment of glaucoma associated with aniridia can be challenging.³ Outcome from medical therapy and surgical therapy has not been satisfactory.⁴ All six eyes in our series were initially started on medical therapy prior to surgery (the definite treatment) but none had significant IOP reduction. Inconsistent success rates ranging from 0% to 83% has been reported with trabeculectomy.⁵ Of the four eyes that had trabeculectomy one eye had good outcome with IOP less than 21mmHg. In a series by Wiggins et al⁴, one of 15 trabeculectomies were successful. Grant and Walton⁶ reported failures in all their patients that had trabeculectomy. However, Nelson et al² reported successful outcome in 11 of 14 eyes. Glaucoma drainage device surgery appears to be effective in obtaining control in aniridic glaucoma. Reported success rates ranges from 66% to 100%³. The two eyes that had GDD as initial surgery had good outcome at 6 months. Two eyes had GDD surgery as secondary procedure with good control of IOP within the first 3 months. Wiggins et al⁴ reported success in 5 of 6 eyes following GDD and all the patients in their series had previous multiple ocular surgeries. Management of aniridia with glaucoma is difficult. Patients may require more than one glaucoma surgery to increase the chance of successful IOP control with or without anti-glaucoma medications.

Conclusion: Though intervention with GDD shows promise in the control of glaucoma in patients with aniridia the challenges of management can sometimes lead to unsatisfactory outcomes.

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