

Glaucoma in Aniridia: Management and Prognosis

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Purpose:

To present this relatively rare secondary congenital glaucoma and the efficacy of surgical treatment in controlling the IOP.

Patients and Methods:

Twelve eyes of 6 patients with aniridia and secondary congenital glaucoma (SCG) were included. Cataract was present in 6 eyes. Goniotomy was done as a primary procedure in 6 eyes with clear corneas. In eyes with hazy corneas, trabeculotomy was done in 4 eyes and subscleral trabeculectomy (SST) in 2 eyes. A successful control period was defined as the time from surgery until IOP exceeded 18 mm Hg.

Results:

Reduction of IOP by 34% was achieved (Preoperative mean of 26.7 mm Hg \pm 4.8 SD to postoperative mean of 17.7 mm Hg \pm 4.0 SD). Mean time for IOP control after primary procedure was 36.7 months \pm 21.4SD. Progressive rise in IOP was recorded in 6 eyes after lensectomy. Nine eyes (75%) necessitated a secondary procedure to achieve IOP control (SST with or without MMC). Choroidal detachment occurred in 3 eyes after filtration surgery and resolved spontaneously.

Conclusion:

Goniotomy and trabeculotomy are relatively effective and safe procedures in controlling glaucoma and should be considered as initial surgical procedure in pediatric glaucoma with aniridia. Filtration surgery proved to be effective as a secondary procedure to bypass the angle anomaly. Individual variation, and duration of the disease are important factors for the prognosis of glaucoma.

Introduction

Aniridia is a rare panocular disorder typically characterized by iris hypoplasia and a high risk of secondary glaucoma. This condition may show autosomal dominant transmission or can be sporadic. Other Clinical manifestations of aniridia may include cataract, and nystagmus. Decreased vision can occur secondary to corneal opacities, cataract, foveal and optic nerve hypoplasia, and glaucoma.

Aniridic glaucoma may occur at birth, or later. Its development has been attributed to congenital angle anomaly as well as progressive changes in the angle. (1)

The most common course of glaucomatous angle progression includes increased confluence of irregular attachments from the iris stroma onto the angle wall. These attachments migrate forward to obscure the scleral spur and posterior trabecular meshwork. (2)

Once glaucoma develops; medical therapy may prove inadequate (2). Surgical therapy also has uncertain prognosis and includes goniotomy, trabeculotomy, filtering procedures and cyclo-destructive procedures (3).

In this study we are reporting the efficacy of goniotomy , trabeculotomy and filtering surgery in controlling aniridic patients with secondary congenital glaucoma occurring in the first 3 years of life, with special attention to the pathological angle progression, and the impact of anterior segment surgery on prognosis of the glaucoma.

Patients & Methods

Twelve eyes of 6 patients with aniridia who had SCG at time of presentation were included in our study. Mean age at presentation ranged from 4 to 30 months.

Complete ophthalmic examination was performed in pediatric cases, started at office to assess the ability of fixation, presence or absence of nystagmus, and completed thoroughly under general anesthesia (EUA) (halothane +nitrous oxide +oxygen).

In glaucoma sheet we reported corneal diameter measurement, corneal clarity, limbal landmarks and presence of pannus (stem cell deficiency). Crystalline lens was inspected for cataractous changes or subluxation Fig (1).



Fig (1): Aniridia, mild cataract & SCG (evident by large corneal diameter) in 2 year old patient.

Measurements of IOP using Kowa hand held applanation tonometer were recorded as well. In eyes with clear media, evaluation of the optic disc for hypoplastic changes or glaucomatous cupping, as well as the central retina was done by indirect ophthalmoscope or by Posner 4-mirrored gonioscens. Fig (2)

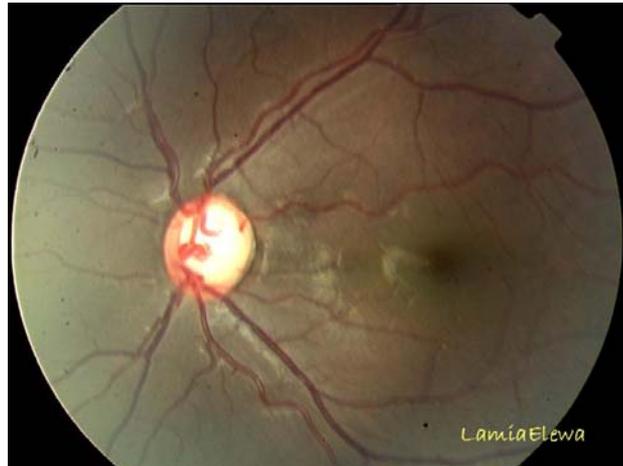


Fig (2): Glaucomatous cupping for same patient in fig 1.

Direct gonioscopy using Thorpe Gonioprism was done to assess the angle structures, iris stump, and ciliary body processes for hypoplasia.

In eyes with opaque media, ultrasound was used to assess the posterior segment, to detect glaucomatous cupping.

Diagnosis of SCG was based on; IOP elevation above 18 mmHg with optic disc changes suggestive of glaucoma with or without increased corneal diameter.

Genetic and pediatric consultation was done in all cases.

Surgical Procedures

In 6 aniridic eyes with SCG and clear corneas; Goniotomy operation was done. After adjustment of the surgical microscope with 45°-60° tilt, the Thorpe gonioprism was applied to the cornea to confirm optimal angle view. A 23-gauge needle attached to a syringe of hyaluronic acid was introduced into the anterior chamber (AC). Hyaluronic acid was injected to maintain a deep AC to avoid any trauma to the vulnerable crystalline lens.

The tip of the needle was approached to the angle. We started to engage gently any abnormal iris tissue extending anterior over the Trabecular meshwork (TM) by the tip of the needle, whileas injecting viscoelastic to dissect these fragile tissues. Then the tip of the needle was directed to the TM in its middle, and incision was created superficially to avoid injury to the underlying structure. The needle was then removed and the AC was irrigated. 80° to 120° of the angle was incised at each setting.

In eyes with hazy corneas; Trabeculotomy operation was attempted. A 2/3 thickness rectangular scleral flap (4x4 mm) was dissected forward until clear cornea. Identification of scleral spur was attempted and a 2 mm (*scratch*) radial linear incision was made across the corneoscleral transition zone. Then gradually deepened until the external wall of schlemm's canal was incised. The trabeculotomes (one after the other) were then introduced in the canal, and rotated with little force into the AC. Then closure of the scleral flap, and conjunctiva.

In eyes where Shlemm's canal identification was not possible, we shifted to (SST). Pre-placed scleral sutures were done. Paracentesis was done and Viscoelastic was injected into the AC to guard against sudden change in pressure gradient and maintain deep AC to avoid any trauma to the vulnerable lens. Two radial incisions were made in the deep limbal tissue; approximately 1.5-2 mm apart centered under scleral flap. The block was retracted and excised with vannus scissors. The pre-placement scleral sutures were tied. Water tight closure of the conjunctiva was done. The AC was gently irrigated to remove the viscoelastic.

Surgical success was defined as; IOP maintained at or less than 18mmHg without medication, and stabilization or decrease in optic disc cupping.

RESULTS

Family history was positive in 2 patients where as the other 4 cases showed sporadic inheritance.

Case #	Age at presentation (months)	Corneal diameter (mm)	Preop. IOP (mmHg)	Lens	Primary Procedure	Follow up (months)	Time At which 1ry procedure failed	2nd Procedure
1-	10	13	20	Clear	1 gonio	84	48m	SST
	10	13	22	Clear	1 gonio	84	48m	SST
2-	18	10(hazy)	32	Cataract	SST	30	12m	SST(M)
	18	10(hazy)	30	Cataract	SST	30	12m	SST(M)
3-	18	9(hazy)	28	Cataract(dense)	Trabeculotomy	30	15m	SST
	18	10(hazy)	36	Cataract(dense)	Trabeculotomy	30	15m	SST
4-	30	10	22	Cataract	2 gonio	52	12m	SST
	30	10	26	Cataract	2 gonio	52	24m	SST
5	20	14	22	Clear	1 gonio		-	-
	20	14	24	Clear	1 gonio		-	-
6	4	15	30	Mild	Trabeculotomy	62	60m	SST
	4	14	28	Mild	Trabeculotomy	62	-	-

Patients' clinical characteristics shown above in table 1.

Two patients (with positive family history) were referred from the genetic research unit, and the other 4 patients were referred by their pediatricians, either because of the white opacity (cataract) or the abnormal sized corneas. All cases presented bilaterally.

Their age ranged from 4 to 30 months (mean = 16.7 ± 8.5). SCG was present in all 12 eyes, mean preoperative IOP was 26.7 ± 4.8 mmHg (ranged from 20: 36mmHg).

Corneal diameter measurements ranged from 9mm to 15 mm (mean = 11.8 ± 2.2). Moderate corneal haze was present in 4 eyes (33%). Corneal pannus was evident in 6 eyes (50%) and was dense enough to hinder angle visualization in 2 eyes. Epithelial corneal erosions were present in 4 eyes.

Cataract was present in 8 eyes (67%). Glaucomatous optic disc changes was evident in 8 eyes. Gonioscopic examination was possible in 6 eyes; in which the iris stump, hypoplasia of the ciliary processes were evident .

Primary Surgical Procedures included; goniotomy in 6 eyes with clear cornea, trabeculotomy in 4 eyes, and SST in 2 eyes. No antimetabolites were used in any primary procedure. Lensectomy was done in 6 eyes with significant cataract.

During a mean follow up period of 53 ± 19.7 months (range 30 to 84), all patients were monitored for IOP, optic disc cupping, gonioscopic findings, and visual function whenever possible.

Reduction of IOP by 34 % was achieved (preoperative mean of 26.7 ± 4.8 mmHg to postoperative mean of 17.7 ± 4.0 mmHg). There was progressive IOP rise in the 6 eyes which underwent lensectomy.

Our successful control period was defined as the time from surgery until IOP exceeded 18 mmHg. Mean time for IOP control after primary procedure was 36.7 months \pm 21.4 SD.

Nine eyes (75%) necessitated SST as *secondary procedure* to achieve IOP control. We used antimetabolites only in 2 eyes (MMC in a concentration 0.3mg/dl). Choroidal detachment occurred in 3 eyes after filtration surgery and resolved spontaneously.

IOP was controlled in all eyes after secondary procedures through a relatively short follow up period (mean of 8 months \pm 3.4 SD). Lens opacities progressed in 2 eyes (SST without MMC). In 2 eyes with relatively long time follow up (62 months), gradual rise in IOP was observed, and gonioscopic examination; showed anterior progression of peripheral iris tissue on to the TM (fig.3).



Fig (3): Gonioscopic view shows anterior progression of peripheral iris tissue on to the TM.

DISCUSSION

Surgical management of aniridic glaucoma is a difficult issue. Blake 1952 reviewed the experiences of 41 ophthalmologists who had operated on aniridic eyes with glaucoma; he concluded that there was no convincing opinion to the best surgical treatment (4)

The mechanism of aniridic glaucoma in infancy is almost associated with abnormalities of schlemm's canal or angle function (5). This was evident in our study by the bilateral presentation of the glaucoma in this age group (mean age at presentation= 16.7 ± 8.5 months) and was our reason to start by goniotomy or trabeculotomy to restore the physiological direction of aqueous outflow.

Goniotomy has been suggested early in the management of aniridic glaucoma, Barkan 1953, reported a case of successful goniotomy at 9 months of follow up. However, other authors with larger number of patients reported lower success rate (20% or less). (6),(7), (8)

In our study, we had a success rate of goniotomy 100% during a follow up period ranged from 4 to 12 months. This success rate decreased to 33% only by the 48 months of follow up. That was explained by the perfect response for the younger age group to goniotomy and the progressive nature of the disease with continuous opposition between the iris stump and the trabecular meshwork.

In 1999, Chen and Walton reported the results of prophylactic goniotomy, in 55 eyes of 33 patients who had aniridia without glaucoma. Forty nine eyes (89%) had IOP control during a follow up ranged from 8 months to 9 years without medication (9).

Trabeculotomy relies on the external approach (ab externo), which avoids the lens and zonules; and thus considered safer than goniotomy and also applicable to eyes with hazy corneas. In a report of 12 eyes by Adachi et al 83% obtained IOP control after first or second trabeculotomy, with a mean follow up of 9.5 years. The high success rate was because these eyes were in young patients (average age 4 years) that in general did not show progressive angle changes(8).

We did Trabeculotomy in 4 eyes (age range of 4 to 18 months) and we recorded a success rate of 75% during a follow up period ranged from 30 to 62 months. With longer IOP control evident in the younger patient (age 4 months at time of interference, and IOP control over 62 months).

Subscleral Trabeculectomy (SST) in aniridia is a little bit dangerous as lens subluxations was recorded in as many as 56% of eyes with aniridia and large corneas. This increase the risk of vitreous loss as well as inadvertent damage to the lens. (1).

Although Nelson et al (1) reported that 5 of 14 aniridic patients who were subjected to primary SST needed secondary surgical interference, Okada et al (10), believed that filtration surgery is effective in controlling the intraocular pressure of young aniridic patients with glaucoma, where they reviewed the chart of trabeculectomy procedures which were applied on 10 eyes of 6 aniridic patients with glaucoma. The mean IOP control period after the filtering surgery was 14.6 months.

Our results agree with Okada et al series; we did SST in 11 eyes and we had not experienced vitreous loss in any case. Only postoperative transient choroidal detachment occurred in 3 eyes. IOP was controlled in 8 eyes (73%) without medication and in 3 eyes with one medication, through a relatively short follow up period (mean of 8 months \pm 3.4 SD).

We had 8 eyes (67%) with cataract. Six eyes necessitated lensectomy, all of which unfortunately showed progressive increased IOP readings (aphakic glaucoma).

During our follow up period (mean 53 \pm 19.7 months) gonioscopic examinations showed patches of iridotrabecular attachments in all eyes with or without progressive rise in IOP.

To conclude; *Goniotomy and trabeculotomy are relatively effective and safe procedures and should be considered as initial surgical procedure in pediatric glaucoma with aniridia. Filtration surgery proved to be effective as secondary procedure to bypass the angle anomaly. Individual variation and duration of the disease are important factors for the prognosis of glaucoma.*

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