Original Article

Outcome of Glaucoma Management in Sturge-Weber Syndrome: Case Series

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Abstract

Objective: To report the clinical findings and outcome of treatment for developmental glaucoma from Sturge-Weber syndrome in four consecutive children managed at a child eye health tertiary facility in Ibadan, Nigeria. **Methods:** A retrospective case series of four children with Sturge-Weber syndrome (SWS) and developmental glaucoma who underwent surgical treatment for glaucoma over a 3-year period. Information on patients' sociodemographic data, clinical features, and management are described. **Results:** Four male children, aged between 7 weeks and 11 years, with unilateral developmental glaucoma and varying extent of port-wine stain were included in the study. The primary surgery in three patients was trabeculectomy, while the fourth patient received a glaucoma drainage device. Shallow anterior chamber, choroidal effusion, and cataract were complications of glaucoma surgery observed. Secondary surgeries performed include anterior chamber reformation, suturing of leaking peritubular scleral channel, sclerotomy and fluid drainage, bleb needling, cataract surgery, and scleral buckle surgery. At the 1-year follow-up visit, one eye had complete success, one eye had qualified success, while the other two had treatment failure. **Conclusion:** The outcome of management of glaucoma associated with Sturge-Weber syndrome is modest. Though rare, SWS can cause significant ocular and systemic morbidity. Complications of therapy may be encountered, and multiple surgical interventions may be required for optimal management.

Introduction

Sturge-Weber syndrome (SWS) is a rare, sporadic, and congenital neuro-oculocutaneous disorder, which was first described about a century ago.[1,2] The initial reports described SWS occurring in children as a triad of extensive facial, scalp, and truncal capillary malformation (port-wine stain, PWS); contralateral focal seizure disorder; and ipsilateral intraocular vascular malformation associated with glaucoma.[1] There are, however, significant expansions of the original descriptions available in literature currently.[3] A somatic activating mutation leading to nucleotide transition in *GNAQ* on chromosome 9q21 is responsible for this congenital syndrome.[4] A multidisciplinary review has identified a critical gap in the management of the disease process must be the need of the hour for better understanding of its pathological processes.[5] There are few case reports from the African continent in South Africa,[6] Morocco,[7] Conakry,[8] and Senegal.[9] A population-based study reported an incidence of 0.19/100,000/year, and there is no known sex or racial predilection.[10]

A clinical classification of SWS, the Roach Scale classification, categorizes SWS as Type I, Type II, and Type III.[11] The most common ocular manifestations in the literature are glaucoma and choroidal hemangioma.[12] Other manifestations include visual field defects, retinal occlusion, ocular melanocytosis, and iris mammilations.[13] Glaucoma occurs in about 30–50% of patients with SWS,[2] and is found in Type I and Type II SWS.[14]

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The pathogenesis of glaucoma in SWS is complex as several theories have been proposed.[15] Mechanical abnormalities of the anterior chamber angle formation and elevated episcleral venous pressure from the abnormal vasculature are two common pathogenetic mechanisms implicated in glaucoma.[16] Angle closure glaucoma has also been associated with SWS with a differing pathogenetic mechanism.[17] Choroidal hemangioma may cause refractive error, foveal distortion, cystoid macula edema, retinoschisis, neovascular glaucoma, or retinal detachment.[12] In addition, patients on topiramate for the control of seizures may also be at the risk of developing glaucoma due to ciliochoroidal effusion.[18]

The management of glaucoma in SWS is at best challenging.[12,13,15] Treatment varies based on age and clinical presentation.[19] Currently, intraocular pressure (IOP) reduction is the only established strategy for retarding progressive optic nerve damage from glaucomatous optic neuropathy. Medical therapy as well as surgical options including angle surgeries, filtering procedures, drainage device placement, cyclo-destructive procedures, and combination procedures have been described.[12]

This study reports the clinical findings and outcome of treatment for developmental glaucoma from Sturge-Weber in four consecutive children managed at a child eye health tertiary hospital in Ibadan, Nigeria.

Methods

This study was a retrospective case series of four children with Sturge-Weber syndrome and developmental glaucoma who underwent surgical treatment for glaucoma at the Pediatric Ophthalmology and Strabismus Clinic, University College Hospital (UCH), Ibadan, from December 1, 2018 to December 31, 2020.

Information collected included demographic data, presenting complaints, care obtained before presentation at the eye clinic, perinatal history, ocular examination findings at presentation including visual acuity, presence/absence of buphthalmos, strabismus, corneal diameters, pupillary reaction, optic disc findings, and intraocular pressure, type of surgery performed, complications, and follow-up findings.

Initial evaluation was performed in the eye clinic under sedation for the younger children using chloralhydrate at a dose of 50–100 mg/kg, while the older child was not sedated. Intraocular pressures were measured using Perkins handheld tonometer for the younger children and the Goldman applanation tonometer for the older child. Anterior segment examination was performed using a slit lamp biomicroscope (a handheld type was used where necessary). Fundal examinations were performed using the binocular indirect ophthalmoscope in the infants and via 90D fundus lenses in the older child. Axial length and pachymetry were measured using the ultrasound applanation technique. Patients were referred to the pediatrician for systemic evaluation, which comprised a physical examination and investigations.

Corneal diameters were measured with the calipers intraoperatively. Trabeculectomy was performed by a pediatric ophthalmologist, while the glaucoma drainage device (Fine Tube MT®) surgery was performed by a glaucoma specialist. For trabeculectomy, the surgery was performed under general anesthesia in aseptic conditions. After routine cleaning and draping, a fornix-based conjunctival flap was raised. A fornix-based conjunctival flap was raised following conjunctiva peritomy. A rectangular scleral flap 3 mm (horizontal) by 2 mm (vertical) was then dissected, and cellulose surgical sponges saturated with antimetabolite (5-Fluorouracil (50 mg/ml) in the two infants or Mitomycin (0.4 mg/ml) in the older child) were placed in the subconjunctival area and underneath the scleral flap for 3 minutes; after which, they were removed, and the surgical site was thoroughly irrigated with 30 ml of normal saline. A rectangular deep sclerostomy and a peripheral iridectomy were performed after paracentesis. The scleral flap was closed with two 10-0 nylon releasable sutures at the edges of the flap. The conjunctiva was closed with 10-0 nylon sutures ensuring a watertight bleb at the end of surgery. Subconjunctival injection of antibiotic and steroid combination was given.

The glaucoma drainage device surgery was performed using a membrane-tube (MT)-type glaucoma shunt device (Fine Tube MT®). The glaucoma drainage device surgery was performed using a membrane-tube (MT)-type glaucoma shunt device (Fine Tube MT®) under general anesthesia in aseptic conditions. After routine cleaning and draping, a fornix-based conjunctival flap was raised. The membrane reservoir of the drainage device was then inserted underneath the Tenon's capsule, and the wings of the membrane were inserted under the superior and lateral recti muscles. The membrane was sutured to sclera using 9-0 nylon. A partial thickness scleral tunnel of ~1 mm length and ~1 mm width was constructed between the proximal edge of the membrane reservoir and the corneal limbus at about 11 0'clock, and a track was created within sclera to the forniceal wound. This was for the passage of the tube tip via the lumen of 23-guage needle through the scleral tunnel and into the anterior chamber. The scleral track, conjunctiva, and Tenon's capsule were closed with 10-0 nylon sutures.

For this study, complete success was defined as IOP $\leq$ 21 mmHg without medication and qualified success as IOP $\leq$ 21 mmHg with anti-glaucoma medication as at1-year follow-up.

Results

Four patients were managed within the period under study; all were male children, and their ages at presentation ranged from 7 weeks to 11 years. They all had unilateral developmental glaucoma in the left eye, and all the patients had varying extent of PWS. The details of the clinical profile of the patients are presented in Table 1.

Table 1: Clinical features at presentation

Patient initials	BB	AE	OB	DM
Age/sex	11 years/M	10 month/M	10 weeks/M	7 weeks/M
Complaints	enlarged LE from birth; poor vision a year prior to presentation	cloudy cornea LE from birth; tearing, redness, photophobia, progressive enlargement LE	redness and tearing LE shortly after birth, enlargement of the LE, reddish discoloration of the left side of the face	reddish discoloration on the face, Whitish discoloration of the LE from birth, Enlargement LE
Visual Acuity	RE – 6/6 LE – Hand motion	strong aversion to occlusion of RE	RE – C, S, M LE – C, S, M	RE – C, S, M LE – C, S, M
Cornea diameter VCD by HCD (mm)	LE – 13.5 by 14.5	LE – 13.5 by14.0	LE – 13.5 by 13.5	LE – 13.0 by 14.0
Anterior segment features	Exotropia, buphthalmos, PWS, RAPD, PSC,	Exotropia, buphthalmos, PWS, hazy cornea, Haab's striae, AL RE: 20.8 mm, LE: 24.6 mm. CCT RE: 567 μm LE: 583 μm	PWS, hazy cornea, deep anterior chamber,	PWS, hemi-facial hypertrophy, Hazy cornea, deep anterior chamber,
Fundus	CDR 0.95 × 0.95	CDR 0.8 × 0.7	poor view of the posterior pole	VCDR 0.5
IOP	RE – 14 mmHg LE – 23 mmHg	RE – 12 mmHg LE – 16 mmHg	RE – 10 mmHg LE – 30 mmHg	RE – 10 mmHg LE – 12 mmHg

AL, axial length; CCT, central corneal thickness; CDR, cup: disc ratio; C, S, M, central, steady and maintained fixation; HCD, horizontal corneal diameter; IOP, intraocular pressure; LE, left eye; M, male; PSC, posterior subcapsular cataract; PWS, port-wine stain; RAPD, relative afferent pupillary defect; RE, right eye; VCD, vertical corneal diameter; VCDR, vertical cup: disc ratio.

All eyes had enlarged corneal diameters. Glaucomatous cupping was present in two patients at presentation. One of the patients (AE) had a history of birth asphyxia and seizures.

Systemic Features of Sturge-Weber in the Patients

The port-wine stain affected varying degrees of the facial skin. In all the patients, there was an extensive port-wine stain covering V1, V2, and V3 dermatomes on the left side of the face. In addition to this, the V2 and V3 dermatomes were also affected on the right side of the face in two patients.

A seizure disorder was present in two of our patients (50%). Cranio-orbital computed tomography (CT) scan was performed in two patients. CT scan finding was normal in one of them. The other patient had features of cerebral atrophy and calcifications. The specific radiologic findings were that of a smaller cerebral hemisphere on the left side with multiple cortical/subcortical hyper densities on the left fronto-temporo-parietal lobes, enhancement of the meninges of the left cerebral hemispheres, and generalized atrophy suggestive of intracranial calcifications.

Treatment

The primary surgery in three of the patients was trabeculectomy, while the fourth patient had a glaucoma drainage device (Fine Tube MT®) surgery. Three (75%) of the patients had additional surgeries performed and were described subsequently.

The control of IOP control was inadequate in three (75%) eyes, and they had additional medical treatment with antiglaucoma medications following the primary surgical intervention. At the 1-year follow-up period, one eye had complete success (BB), and one eye had qualified success (OB). Visual outcome was not satisfactory as 3 eyes in the series had amblyopia. (Table 2).

Complications

Shallow anterior chamber as a complication of filtration surgery was observed in two (50%) of the eyes operated in this series. Of these two, one of the eyes had trabeculectomy, while the other had glaucoma drainage device surgery. The eye that had trabeculectomy was managed conservatively, while the eye that had Fine Tube MT® implant underwent LE anterior chamber reformation and suturing of leaking peritubular scleral channel. Choroidal effusion was noted in two eyes; that is, in the eye that had Fine Tube MT® insertion and in one of the eyes that underwent trabeculectomy. Choroidal effusion was diagnosed after anterior segment examination that showed marked anterior chamber shallowing and subsequently large choroidal effusion seen via binocular indirect ophthalmoscope examination. The choroidal effusion following Fine Tube MT® insertion was managed conservatively, while sclerotomy and fluid drainage were performed for the other eye that had choroidal effusion. A retinal detachment was observed post sclerotomy, and the patient subsequently had scleral buckle and, thereafter, buckle readjustment for a re-detachment.

Further postoperative complication of cataract was observed in the eye that had Fine Tube MT® insertion, and a cataract surgery with primary posterior capsulotomy and anterior vitrectomy was performed. Bleb revision on the account of a thick fibrous tethering over the bleb was performed for a patient (DM), 7 months after the initial fine tube surgery, as well as for another patient (AE) after initial trabeculectomy.

Table 2: Treatment and treatment outcome

Patient initials	BB	AE	DM	OB
Prior medical treatment	+(timolol 0.5%)	-	+(timolol 0.5%)	-
Trabeculectomy	+	+	-	+
Antimetabolite	MMC	5FU		5FU
Glaucoma drainage devices	-	-	+	-
Additional surgical intervention	+	+	+	-
Suboptimal IOP	+	+	+	-
Additional medical treatment after surgical treatment	+	+	+	-
IOP at 1-year follow-up	17 mmHg	22 mmHg	35 mmHg	21 mmHg
Successful outcome				
VA at 1-year postop	+Hand Motion	–	-C, US, UM	+C, S, M
Complications				
Over filtration with flat AC	-	-	+	+
Choroidal effusion	+	-	+	-
Retinal detachment	+	-	-	-
Amblyopia	+	+	+	-
Hypotony	-	-	-	+

5FU, 5-fluorouracil; AC, anterior chamber; C, S, M, central, steady and maintained fixation; C, US, UM, central, unsteady and unmaintained fixation; IOP, intraocular pressure; MMC, mitomycin C; VA, visual acuity.

Clinical Photographs

Figures 1 and 2

Discussion

This case series represents, perhaps, the first series on glaucoma management in children with Sturge-Weber syndrome in Nigeria. Early presentation and diagnosis of glaucoma in children with SWS are necessary to optimize outcomes. Most of the children in the series presented during infancy although one child presented at pre-teen age. This is in keeping with literature review reporting onset of glaucoma in infancy in about 60%.[5] All our patients were male children. Male sex has been found to be a high-risk characteristic factor for development of glaucoma in patients with PWS.[20] Although majority of the patients in the case series presented in infancy, they already had significant glaucomatous changes in the eyes at presentation. Enlarged cornea diameters, absence of cornea clarity, buphthalmos, elevated IOP, and optic disc cupping were present in our patients. It was difficult to compare the presentation with other patients from other climes as there was sparse description of details of the clinical presentation in literature.

Figure 1: AE – 10-month-old male, PWS on the left involving V1, V2, V3 dermatomes, (white arrows), cornea opacity and buphthalmos

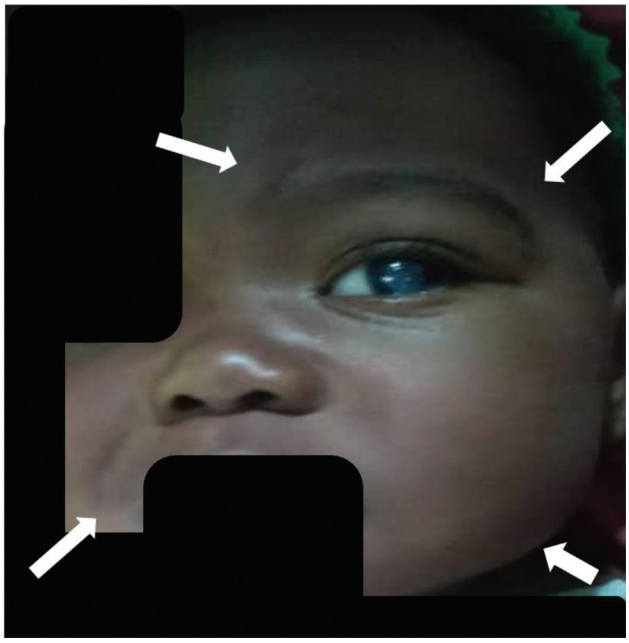

Figure 2: DM – Port-wine stain involving V1, V2, V3 dermatomes (white arrows), left hemifacial hypertrophy and cornea oedema

Two of our patients in this series were on monotherapy with Timolol at their initial presentation having been prescribed with the medication at the clinic where they initially presented before referral to us. Medical therapy can be used as first-line or as adjunct to other treatment options.[16] There are still controversies as to the adequacy of medical management and it is not sufficient for long-term glaucoma control.[15] This prompted the clinical decision to proceed to surgical therapy in all our patients. Awad *et al.*[21] reported success rates of about 30% with use of anti-glaucoma medications at a mean follow-up duration of 5 years.

Antimetabolite augmented trabeculectomy was the primary surgical therapy in three of the four patients in our series, and this is an important surgical option in the treatment of glaucoma associated with SWS. Outcomes of treatment were modest as at the 1-year follow-up period, one eye had complete success, one eye had qualified success, and despite adjunct medical therapy, the third eye still had treatment failure. The oldest among our patients was 11 years. The other patients were all under 1 year with their buphthalmic globes making handling of tissues difficult, and this may have affected outcomes. Better surgical outcomes following trabeculectomy were reported by Mohamed *et al.*[22] in Egypt. The study cohort consisted of 10 eyes of eight patients with a mean age of 3.75 years, a mean preoperative IOP of 29 mmHg, and mean corneal diameter of 12.6 mm. The mean 12-month postoperative IOP was 12.2 mmHg. All eyes achieved success, 80% had complete success, and 20% had qualified success. Ali *et al.*[23] also reported favorable outcomes with trabeculectomy in seven eyes of six patients with late-onset glaucoma with a mean age of 22.4 years and an age range of 7–38 years. After a long-term follow-up period ranging from 9 months to 9 years, IOP control was optimal in two eyes without any antiglaucoma medication, four eyes needed medication to achieve control, while one eye required three trabeculectomies and postoperative medication. In the study by Mohamed *et al.*,[22] the age at presentation ranged from 3 to 5 years, while majority of the patients in our study were infants. Age at presentation could be a proxy for severity such that the younger age in most of our patients could indicate worse severity of glaucoma. This difference in study cohorts could account for the difference in outcomes. Racial differences may also account for this difference as conjunctival scarring and fibrosis are more common in young children and black African pigmented eyes.

A glaucoma drainage device was inserted in the eye of the youngest patient in our series. Outcome was also poor in this patient with significant postoperative complications and subsequent multiple surgeries within a 1-year follow-up period. Over filtration, shallow AC, choroidal effusion, and treatment failure occurred following the Fine Tube® insertion for the patient. As at the 1-year follow-up visit, IOP was 35 mmHg even with anti-glaucoma medication. Efficacy of glaucoma drainage devices such as the Ahmed, Molteno, and Baerveldt used in SWS glaucoma has been reported. Slightly similar to our experience in the present case series, Amini *et al.*[24] concluded that implantation of a single-plate Molteno tube as primary surgery for childhood glaucoma resulting from SWS was associated with a modest success rate and a high rate of complications. On the contrary, Budenz *et al.*[25] reported that a two-staged Baerveldt glaucoma implant procedure combined with posterior sclerectomies was safe and effective for childhood glaucoma associated with SWS. Direct comparisons of these two studies (in which, non-valved glaucoma drainage devices were implanted), with our present study is difficult, because the Fine Tube® has a "valve" mechanism, and there were differences in the characteristics of the study populations. In our study, surgery was performed in a 7-week-old infant, but theirs mainly involved older children. The study by Amini *et al.*[24] included nine eyes in children at a mean age of 9.6 years who all had choroidal hemangioma. After a 6-month postoperative period, no eye achieved complete success in IOP control, and at the last follow-up visit, the mean number of postoperative glaucoma medications was two. In the case series by Budenz *et al.*[25] the mean age and preoperative IOP were 6.6 years and 24.8 mmHg, respectively. Their study cohort, however, included a 1.5-month-old child who had good IOP control and was not on any medications after 10 months of follow-up.

For patients with early onset glaucoma with associated angle abnormalities, conventional therapy via surgical intervention with either goniotomy or trabeculotomy is usually required. The outcomes in SWS are worse than observed with primary congenital glaucoma, and often require additional surgery with trabeculectomy or a glaucoma drainage device.[26] Cycloablation was also a viable option in the older child with poor preoperative vision, but it was not considered at the time of treatment.

Regarding complications of surgery, similar to this present case series, Mohamed *et al.*,[22] Amini *et al.*,[24] and Budenz *et al.*[25] reported no intraoperative complications. Contrastingly, Iwach *et al.*[26] reported intraoperative choroidal effusions in 24% of patients undergoing trabeculectomy especially among the early onset group. Shallow AC, blebitis, and thin polycystic blebs have been reported post trabeculectomy.[22] Pathogenetic mechanisms for increased risk of complications include choroidal hemangiomas, unopposed high choroidal vascular pressure, significant vascular fragility, use of antimetabolites, and transudation that occurs with IOP reduction.[26,27] Following glaucoma drainage device surgery, Amini *et al.*[24] and Budenz *et al.*[25] also reported postoperative choroidal effusion, which they attributed to higher preoperative IOP. Retinal detachment observed following trabeculectomy in our series was also reported after Molteno tube implantation by Amini *et al.*[24] and after trabeculectomy by Awad *et al.*[21]

Recent reviews of the long-term efficacy and safety of surgical interventions have been reported.[13,15,28] Combination therapy utilizing medical and surgical therapies seems to be beneficial in improving treatment outcomes. In this present series, patients were on combination therapy as medical therapy was instituted either before surgery or as an adjunct to surgery when optimal IOP levels were not achieved. Similar observation was made by Amini *et al.*[24] Outcomes were modest in this study indicating that management of glaucoma is SWS is often challenging, and treatment decisions ought to be individualized. Surgical success across the studies was variable and differed with the type of surgery as well as demographic characteristics of study cohorts. The aggressive course of glaucoma and the accelerated rate of fibrosis following surgical procedures in blacks could be factors responsible for our modest treatment outcomes. As our study cohort involved mostly infants, the excessive healing process in young patients and the stimulatory effect of abnormal episcleral vessels, which enhance scarring could be contributory. The average increase in postoperative IOP compared to preoperative IOP in our series was probably due to the postoperative complications and multiple surgeries that eventually resulted in fibrosis in our patients.

There are some limitations to our study. We were only able to describe our experience with four patients, which reflects the rarity of this syndrome. The retrospective nature of the study made uniformity of clinical documentation impossible. Furthermore, the follow-up duration was limited to one year after surgical intervention.

In conclusion, glaucoma management in children with Sturge-Weber syndrome in our setting is challenging, and the outcomes are modest. Though rare, it can cause significant ocular and systemic morbidity; hence, early diagnosis is important in its management. Multiple surgical interventions may be necessitated to achieve optimal intraocular pressure control and caregivers need to be adequately counselled. The complications encountered following our surgical approach were managed medically and surgically. Ophthalmologists managing such patients should anticipate these in the postoperative care.

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Conflicts of interest

None to declare.

Ethics approval

Obtained from the University of Ibadan/UCH Ethical Review Committee 20/0302.

Consent for publication

Yes.

Availability of data and material

Not applicable.

Code availability

Not applicable.

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