Ocular manifestations of Sturge–Weber syndrome

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Purpose of review
Sturge–Weber syndrome (SWS) is a rare, congenital disease which frequently involves the eye. It is important that ophthalmologists recognize this syndrome and are aware of its range of ocular manifestations. The aim of this article is to present our understanding of the pathogenesis and clinical manifestations of this syndrome and provide updated information on the treatment of SWS glaucoma and choroidal hemangioma.

Recent findings
SWS glaucoma usually fails medical management. Surgical options include angle procedures, filtering procedures, device placement, and combination procedures. Combination procedures have become popular in this population due to the single procedure failure rate of angle surgery and the complications associated with device implantation. Choroidal hemangioma is best treated by photodynamic therapy.

Summary
Lifelong monitoring for ocular complications related to SWS is essential. There is a need for consensus guidelines on care and surveillance of patients with SWS to provide the best care for these patients.

Keywords
choroidal hemangioma, encephalotrigeminal angiomatosis, glaucoma, port wine birthmark/stain, Sturge–Weber syndrome

INTRODUCTION

Sturge–Weber syndrome (SWS) is a rare, congenital neuro-oculocutaneous disorder. Without population-based evidence, estimates of incidence range from one in 20 000 to 50 000 live births [1]. Syndrome manifestations are varied and include unilateral port-wine birthmark (PWB), intracranial leptomeningeal angioma, hemianopia, glaucoma, choroidal hemangioma, hemiatrophy, hemiparesis, progressive seizures, and cognitive impairment [1–5].

Current literature provides insight about a few of these clinical findings. For example, prior to recent academic analysis of the distribution of the PWB, it was largely accepted that the PWB followed the distribution of the trigeminal nerve, particularly that of the ophthalmic branch. However, current literature clarifies that facial capillary malformations follow embryonic craniofacial patterns and that hemifacial, bilateral, and frontonasal capillary malformations are associated with higher risk of syndrome severity [6,7].

There have also been recent improvements in our understanding of behavioral disorders associated with SWS. The first systematically collected data on the topic shows that social communication disorders affect about half of patients with SWS while autism spectrum and sleep disorders each affect approximately one quarter of these patients. Even children that do not carry one of these diagnoses often require accommodations in school [8▪▪].

In addition, recent literature has emphasized the correlation between SWS and endocrine disorders. Within a registry of 1653 patients with SWS, authors found an estimated prevalence of growth hormone (GH) deficiency in patients with SWS to be 18 times that of the general population. This finding suggests the importance of considering GH deficiency as the cause of impaired growth velocity in children with SWS [9]. Case series have similarly demonstrated an increased rate of central hypothyroidism in patients with SWS. Further investigation is needed to determine whether anticonvulsant use as the cause of central hypothyroidism in this population is a significant confounding variable [10▪].

Although the symptoms discussed above usually present at birth, SWS is not inherited. Sporadic,

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KEY POINTS

- Lifelong monitoring for ocular complications related to SWS is essential.
- SWS glaucoma is complex and often requires a multidimensional approach to maintain effective management.
- Current literature suggests that photodynamic therapy has been successful in treating SWS choroidal hemangioma.
- Large-scale studies are needed to form consensus guidelines on multidisciplinary care and surveillance for patients with SWS.

somatic mutation of GNAQ, the genetic coding region for a protein called Gaq, leads to the development of SWS. Disruption of the normal coding sequence leads to cell proliferation and inhibition of apoptosis. Depending on when in development the mutation occurs, an altered copy of GNAQ can lead to SWS or simply sporadic PWB [11]. In severe forms, this mutation can induce impaired development of cell precursors in the neural crest during the first trimester of pregnancy and lead to characteristic central nervous system, dermatological, and ocular changes [12]. SWS is so variable that there are case reports of patients with an absence of facial angioma but with classic neuroimaging findings of parieto-occipital atrophy with heavy gyriform calcifications and angiomatous malformations in the parieto-occipital cortical and subcortical areas [13,14].

It is recommended that all children with suspected SWS undergo contrast-enhanced brain MRI to evaluate for this classic finding [7]. In the neonatal period MRI may not be sensitive. Therefore, it is best practice to confirm a SWS diagnosis after 1 year of age by identifying the characteristic abnormally enhancing leptomeningeal blood vessels within the parieto-occipital lobes [3,15▪].

Children with forehead PWB should also be evaluated by an ophthalmologist due to the range and severity of ocular manifestations associated with SWS. For this reason, the remainder of this article will delineate those ocular manifestations and focus on the treatment of the two eye conditions most common in SWS patients. These conditions are glaucoma and choroidal hemangioma, which occur in approximately 30–70% and 20–70% of SWS patients respectively [4,16,17].

GLAUCOMA

Childhood glaucoma is a costly disease that causes caregivers significant emotional and psychological burden [18]. Childhood glaucoma that is associated with systemic abnormality is estimated to comprise 12.9% of childhood glaucoma due to any cause [19▪]. Glaucoma associated with SWS is particularly burdensome as the ophthalmologist is clearly not the only specialist that these families need to see. Ophthalmologists need be confident in their understanding of this syndrome to decrease this load on families to the extent that is possible.

From the ophthalmologist's perspective, SWS glaucoma has unique complexities that make patient care challenging. There are two main proposed pathogenic mechanisms of this subsect of glaucoma: first, malformation of the anterior chamber angle and second, increased episcleral venous pressure from the ectatic vasculature of the PWB [4,20▪▪]. Angle malformation is likely responsible for early-onset glaucoma in SWS, whereas elevated episcleral venous pressure may play a greater role in later-onset cases. The congenital-type glaucoma accounts for about 60% of SWS glaucoma [20▪▪].

In addition to these early and later-onset subtypes, reports have been published of patients with SWS experiencing acute angle-closure attacks. Causes of such attacks include lens-induced pupillary block and posterior scleritis [21,22]. The author of a recent case report on acute primary angle-closure in SWS described her approach to treatment. With failure of medical management and knowledge that conventional laser iridotomy had a high risk of bleeding, the patient was successfully treated by lens extraction and intraocular lens implantation [23▪].

When managing chronic SWS glaucoma, it is best practice to attempt medical management prior to accepting the risks of surgery. Every child with PWB should have at least annual slit lamp exams with consideration for exam under anesthesia to measure corneal diameter and axial length for monitoring the development or progression of glaucoma. Ocular coherence tomography (OCT) can be used to surveil the optic nerve head. All patients with SWS should be monitored by intraocular pressure (IOP) and visual field testing when possible by age and cognitive ability [20▪▪]. If a patient would like to undergo cosmetic laser operation, there is no ophthalmological contraindication as there is no evidence to suggest that laser application has a clinically significant effect on IOP [24].

Small case series have shown topical glaucoma medications to be successful in controlling IOP and the progression of SWS glaucoma to varying degrees. Aqueous suppressants (beta blockers, adrenergic agonists, and carbonic anhydrase inhibitors) are the preferred medication for patients with congenital-type SWS glaucoma [20▪▪]. Brimonidine, an alpha

adrenergic agonist, is potentially toxic to the central nervous system and is therefore contraindicated in children less than 2 years of age [20▪▪]. Latanoprost, which lowers IOP by increasing uveoscleral outflow through remodeling of the extracellular matrix and decreasing resistance through the ciliary muscle, tends to work best in later-onset SWS glaucoma [4,25]. Despite the utility of beta blockers in topical formations and the role for oral propranolol in reducing hemangiomas, oral propranolol has not shown to be effective in lowering IOP as a single agent [26].

Invariably SWS glaucoma seems to become refractory to medical management and the patient is recommended to undergo surgery. The main surgical options include angle procedures, filtering procedures, device placement, cyclodestructive procedures, and combination procedures. Angle procedures, goniotomy, and trabeculotomy, are preferred initial surgical options due to their relatively low risk of adverse events [20▪▪]. Although early diagnosis and performing early trabeculotomy may improve success rates of angle procedures, the generally high single procedure failure rate has led physicians to investigate other procedures [27▪].

Filter procedures, particularly trabeculectomy, have recently been reported successful. A prospective, randomized study of 20 eyes compared trabeculectomy with mitomycin C, a chemotherapeutic agent, to trabeculectomy with Ologen implant, a biodegradable collagen implant designed to decrease subconjunctival scarring and bleb-related complications. The Ologen implant was found to be equally as effective as mitomycin C without the complications of thin polycystic bleb, blebitis, and shallow anterior chamber. This small study shows promise for the utility of the Ologen implant in patients with increased hemorrhagic risk as in patients with SWS [28▪▪].

In the trabeculotomy–trabeculectomy combination procedure, anterior chamber abnormalities are overcome by creating two outflow tracts for the drainage of aqueous humor. Trabeculotomy allows communication between the anterior chamber and Schlemm's canal while trabeculectomy establishes a fistula between the anterior chamber and the subconjunctival space. In this way the episcleral venous pressure can be overcome [29▪]. Two groups of investigators have recently had high success rates with this procedure. Complications of the procedures were uncommon but included exudative choroidal detachment, hypotony, and cataract [29▪,30▪].

Various drainage devices have been used in SWS patients. The Ahmed valve significantly reduced the need for antiglaucoma medications but was complicated by hyphema, hypotony, choroidal detachment, tube-cornea touch, and visually significant cataract [31▪,32]. The Molteno tube has been used less frequently than the Ahmed implant as it is reported as having limited success with relatively high (>50%) postoperative complication rates. Complications included choroidal effusion, cataract, and retinal detachment [33]. The Ex-Press shunt is the latest device to enter the literature of SWS glaucoma. It has been used alone, in conjunction with mitomycin C, and in combination with trabeculectomy. Complications with this device include overfiltration, decompression retinopathy, choroidal detachment, hyphema, and pupillary block. Across varying causes of childhood glaucoma the Ex-Press shunt with mitomycin C better controlled IOP, had better visual acuity scores, had fewer complications, and had fewer reoperations than trabeculectomy with mitomycin C [34–36].

Cryocoagulation is a safe alternative treatment option for SWS glaucoma [37,38]. Cyclocrycoagulation in 69 eyes was only complicated by conjunctival edema. Transscleral contact cyclophotocoagulation with diode laser is more effective in decreasing IOP than cyclocryocoagulation but can be complicated by uveitis, hypotony, and hyphema [38].

CHOROIDAL HEMANGIOMA

Choroidal hemangioma is more commonly observed in an eye with PWB involving the upper eyelid. The diffuse engorgement of preexisting as well as proliferating blood vessels can give the appearance of a 'tomato catsup' fundus [2]. Diagnostic imaging for choroidal hemangioma includes fundus fluorescein angiography, indocyanine green chorioangiography, B-scan ultrasound, OCT, and even MRI [39–41].

Management depends upon whether the choroidal hemangioma is diffuse or circumscribed. Diffuse choroidal hemangioma (DCH) may be subtle and often occupies more than half of the fundus without well defined borders. Circumscribed choroidal hemangioma (CCH) is usually observed as a subtle reddish mass in the posterior choroid. CCH must be distinguished from choroidal melanoma to ensure proper treatment [40].

Vision loss from DCH may result from refractive error, foveal distortion, or exudative retinal detachment. External beam radiotherapy, stereotactic radiotherapy, proton beam radiotherapy and plaque brachytherapy have been used with success. However, the most up to date literature endorses photodynamic therapy with verteporfin. Authors suggest avoiding overlapping spots during treatment to avoid the theoretical risk of fibrosis [40–43]. The procedure may lead to initial worsening of visual acuity and exudative detachment at 1 week that can

continue up to 1 month after treatment. By 1 month postprocedure there is usually gradual improvement. After 6 months the full effect of a photodynamic therapy (PDT) application may be assessed [40].

CCH are often asymptomatic but can lead to visual loss by retinal degeneration over a subfoveal tumor, retinal detachment of the fovea, cystoid retinal edema leading to retinoschisis, neovascular glaucoma, or even total retinal detachment. Cautery, diathermy and cryotherapy are poor treatment options as these methods destroy adjacent tissues. Argon laser is not recommended due to its high recurrence rates. Laser photocoagulation, transpupillary thermotherapy (no utility in subfoveal lesions), ruthenium plaque therapy, external beam irradiation, proton-beam irradiation, and photodynamic therapy are all viable treatment options for CCH [40].

Although radiotherapy and proton beam therapy are effective treatments for both DCH and CCH, they are not widely available due to the need for specialized centers for procedure performance. In addition these methods can be complicated by radiation retinopathy, optic neuropathy, macular ischemia, and subretinal fibrosis. These complications are much more extensive than the possibility of choroidal atrophy from PDT [40].

The aim in treating any choroidal hemangioma is to shrink the portion of the tumor that is near or within the macula while allowing the resolution of subretinal fluid. PDT accomplishes these treatment goals in a relatively safe, same-day, office-based procedure that is familiar to most retina specialists [40]. Case reports have discussed the utility of adding anti-vascular endothelial growth factor (VEGF) agents as an adjunct to PDT due to the therapy's effect of inducing high VEGF levels [2,40,43]. More study with these agents is necessary to determine their utility in SWS patients.

Although PDT is often effective after one application, a case report of a 7-year-old male with SWS DCH needed five sessions over the course of 1 year before resolution of the choroidal hemangioma and subretinal fluid. The author draws attention to Chinese reports that echo his observation that Asian patients often require multiple PDT treatments [44].

It is important to note that some patients are unable to tolerate PDT. A case report of a 5-year-old patient with exudative retinal detachment secondary to choroidal hemangioma was not able to tolerate the procedure, and so the physician performed a temporizing partial thickness sclerectomy in each quadrant of the retina. When the retinal detachment relapsed 2 years later, the sclerectomies were revised and patient underwent successful PDT [45▪].

ADDITIONAL OCULAR FINDINGS

Although glaucoma and choroidal hemangioma are by far the most common ocular manifestations of SWS, we must be aware of additional ocular manifestations. For example, when analyzing visual field defects it is important to remember that not only glaucoma, but also SWS-related occipital lobe involvement could be the culprit [20▪▪].

A recent case report of a 12-year-old male with SWS highlights additional ocular findings. This patient with bilateral PWB was observed to have hyperpigmented iris with iris mamillations, glaucoma, and DCH. The authors call for thoughtful use of imaging upon encountering atypical presentations of SWS eye disease. They were able to individualize treatment of this patient using OCT assessment of the macula, choroid, and peripapillary retinal nerve fiber layer [46].

CONCLUSION

After dermatologic laser, the PWB may be less visible or not visible at all. Despite these excellent cosmetic results, the risk of glaucoma and other ocular complications remains unchanged. Patients need to be reminded of their continued need for ophthalmologic care. SWS is a lifetime disease and although these patients may remain asymptomatic throughout childhood, they can present with significant eye complications later in life. Ongoing monitoring is essential.

Due to the rarity of SWS and the even more profound rarity of SWS-related ocular conditions, much of the data presented here is from small studies and case reports. Better investigations would be possible with standardization of neuroimaging and tissue banking across health centers. With better data the need for consensus guidelines on multidisciplinary care and surveillance could be fulfilled [47▪▪]. Collaboration would ideally include SWS patients and their families to ensure the approach is a humanistic one [48].

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Conflicts of interest

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- ▪ of special interest
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