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Glaucoma Management in Sturge–Weber Syndrome Using the Delphi Process

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Objective: We sought to identify areas of consensus and nonconsensus in the ophthalmic screening, diagnosis, and management of Sturge–Weber syndrome (SWS).

Design: Modified Delphi methodology.

Participants: North American glaucoma experts with prior experience managing glaucoma in patients with Sturge–Weber syndrome.

Methods: A modified Delphi process was used as a systematic and structured communication technique, consisting of 2 rounds of electronic questionnaires to a wider group, followed by an in-person meeting of selected experts. Questions that did not reach agreement were reformulated in each round, with the aim of reaching consensus. The University of Rochester Research Subject Review Board noted that this study was exempt from IRB approval. This study adhered to the Declaration of Helsinki.

Main Outcome Measures: Consensus was defined as agreement among at least 85% of participants for the electronic questionnaires or a minimum of 70% during the in-person meeting.

Results: Among other recommendations, the panel concluded that all patients with a facial port wine birthmark should be assessed for glaucoma within a month of birth, with shorter screening intervals thereafter in patients with bilateral eyelid involvement, choroidal hemangioma, and signs of elevated episcleral venous pressure. There was no consensus on timing of follow-up visits. In children aged $\geq$4 years with signs of glaucoma, medical intervention is the initial consideration. In children aged $<$4 years, the primary intervention is likely to be surgical, with some form of goniosurgery as the first-choice procedure.

Conclusions: Consensus for screening, diagnosis, and management in patients with SWS is designed to hopefully improve clinical practice and patient outcomes. Questions where consensus was not reached may highlight variations in practice, conflicting evidence, or areas that might benefit from further research and investigation.

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Sturge–Weber syndrome (SWS), also known as encephalotrigeminal angiomatosis, is a neurocutaneous syndrome characterized by vascular malformations of the face, choroid, and leptomeninges. It is the third most common neurocutaneous syndrome after neurofibromatosis and tuberous sclerosis, estimated to affect 1 in 20 000 to 50 000 people.[1] Presence of a facial cutaneous vascular malformation (port wine birthmark [PWB]), historically described as being distributed along the ophthalmic division of the trigeminal nerve, increases the risk of brain and eye involvement. Glaucoma is the most common ophthalmic complication, reported in up to 70% of patients, and may present from birth to adulthood.[2] The primary suspected pathogenic mechanisms of SWS glaucoma include goniodysgenesis/trabeculodysgenesis or increased episcleral venous pressure (EVP), thought to be responsible for early-onset glaucoma and later-onset glaucoma, respectively.[2–6]

Glaucoma is a risk factor for vision loss in patients with SWS. Management can be challenging, and there are no published consensus guidelines for management. It is often poorly responsive to medical treatment, and the surgical success rate can be limited by complications, including suprachoroidal effusion, expulsive hemorrhage, and retinal detachment.

We used a methodical research tool, the Delphi technique, to establish consensus-based recommendations to improve clinical practice.[7] The Delphi methodology has previously been employed successfully to collect data, analyze responses, and communicate and present a consensus from ophthalmic experts on controversial and complex clinical issues.[8–10] Through a series of

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questionnaires and standardized discussions, experts narrow down responses to complex clinical questions to reach mutual agreement. The Delphi method is modified by including in-person group discussions. We employed the modified Delphi method to achieve consensus in the management of glaucoma in patients with SWS.

Methods

A modified Delphi process was used as a systematic and structured communication technique, consisting of 2 rounds of electronic questionnaires followed by a face-to-face meeting.[11] A.V.L. served as facilitator and sent an email invitation to complete an initial questionnaire to ophthalmic specialist networks (Childhood Glaucoma Research Network and American Association for Pediatric Ophthalmology and Strabismus). Responses were screened to identify individuals who worked with this patient population frequently and determine their availability to respond to electronic questionnaires and participate in face-to-face meetings. The minimal experience criteria were determined by the working group, comprising North American experts in pediatric glaucoma who were interested and available, and are coauthors herein. Some of the participating authors also care for adults or are adult glaucoma fellowship graduates (T.C., B.E., D.W., L.B., S.F., and B.W.). Respondents were required to answer "yes" to at least 2 of the following screening questions to qualify for the study and were then automatically given access to the full questionnaire: (1) do you see at least 10 children with a PWB on or around the eyelids per year? (2) do you see at least 9 children with glaucoma associated with a PWB on or around the eyelids per year? (3) do you operate on at least 6 children with glaucoma associated with a PWB on or around the eyelids per year? and (4) do you operate on at least 21 children with glaucoma of any type per year?

Consensus was defined as 85% agreement to an answer, and borderline consensus as >75%. Questions that did not achieve consensus were reformulated, and the resultant new questions were then circulated to those who completed the first-round questionnaire. Questions that did not reach consensus during the 2 electronic rounds were again reformulated and discussed in the face-to-face meeting held at the University of Rochester Medical Center.

At the face-to-face meeting, each question for which consensus was yet to be achieved, along with relevant lead-in questions on which consensus had been achieved, was read aloud by a nonparticipating moderator (K.A.). The meeting was facilitated by a nonvoting working group participant (A.V.L.), who did not participate in the panel discussion other than to reflect the opinions expressed to help reconstruct the questions to achieve consensus where possible. Each member of the working group was required to comment on each question for a maximum of 2 minutes in random order as determined by a random name generator coded on Google Sheets. There was then a period of silence while each participant voted, in writing, maintaining confidentiality and using anonymous voting sheets, which were immediately folded to obscure their response, and collected by the support staff for collation. Results of the vote were immediately reported back to the group after analysis by support staff. If the agreement on any vote was <70%, the question was reformulated by group discussion in the same manner, followed by an open group discussion where needed. The participants anonymously voted again on the reformulated question, and if consensus or near consensus was not obtained after this second round, we defined the result as nonconsensus (Appendix 5, available at www.ophthalmologyglaucoma.org.). The University of Rochester Research Subject Review Board noted that this study was exempt from IRB approval. This study adhered to the Declaration of Helsinki.

Results

The initial questionnaire was emailed to the directories of the Childhood Glaucoma Research Network and American Association for Pediatric Ophthalmology and Strabismus with memberships of 205 and 1702, respectively. We received 171 responses total, of which 35 respondents met the necessary criteria and completed the entire survey. Eight of the 35 respondents did not manage patients surgically and thus did not complete the questions on surgical management. The second-round questionnaire was offered to the 27 individuals who had fully responded to the initial questionnaire and was completed by 26. Members of the working team, except the moderator (A.V.L.), also filled out the first and second questionnaires. Seven members of the working team attended the final face-to-face meeting to discuss 15 questions for which consensus had not been reached in the second-round questionnaire. Consensus or near consensus was achieved for 10 of 51 questions (19.6%) in the first round, 20 of 37 questions (54%) in the second round, and 15 of 15 questions (100%) in the third round. All questions can be found in the Appendix as Appendix 1: first-round questionnaire, Appendix 2: second-round questionnaire, Appendix 3: third-round questionnaire, and Appendix 4: summary of consensus statements (available at www.ophthalmologyglaucoma.org.).

Screening and Diagnosis

Patients with a facial PWB have a markedly greater risk of developing glaucoma than the general population and should be evaluated for ocular hypertension or glaucoma, as well as other SWS-related ocular comorbidities, within 1 month of birthmark detection if the birthmark involves 1 or both eyelids. Patients with a facial PWB not involving the lids may still carry a higher risk of developing glaucoma than the general population, and thus, selected patients should be similarly evaluated. Factors associated with a higher risk of glaucoma include eyelid involvement, presence of choroidal hemangioma, and signs of increased EVP (e.g., blood in Schlemm canal and episcleral vein prominence). There was no agreement on the frequency of repeat ophthalmic evaluations in patients with a facial PWB, but there was consensus that patients with these glaucoma risk factors should be monitored more closely for any development of ocular hypertension or glaucoma. There was a lack of agreement on referral to a glaucoma subspecialist for these evaluations, or whether there was an age at which the evaluation frequency could be reduced or even ceased.

Consensus opinion was that in the absence of a qualified pediatric ophthalmologist, evaluation for the risk factors or development of ocular hypertension or glaucoma in these patients could be performed by an adult ophthalmologist, whether this be a comprehensive general ophthalmologist or a glaucoma subspecialist. A consensus could not be reached on the role of an optometrist in this specific scenario. There was also no consensus on the utility of home tonometry for glaucoma screening. Regardless of age, all patients with a facial PWB should be evaluated by an eye doctor. There was no agreement on which

other services should routinely be included in the child care team, although it was agreed that in some settings, trusted collaborating optometrists could assist with these evaluations. For the other systemic manifestations of SWS with PWB, either the child's pediatrician or their ophthalmologist would ensure appropriate evaluation and specialist referrals as appropriate. An important example is the risk of seizures due to brain/leptomeningeal involvement, for which magnetic resonance imaging of the brain with contrast may be indicated. A specialist dermatologist's input is also valuable for early management of the PWB itself. Genetic testing is not usually indicated.

Increased intraocular pressure (IOP), axial length, and corneal diameter, as well as cup-to-disc asymmetry, are all key factors for glaucoma diagnosis. Cup-to-disc ratio in and of itself did not reach consensus as a key diagnostic factor. A sedated eye examination or examination under anesthesia should be performed in patients of all ages with a facial PWB when a complete glaucoma assessment cannot be completed while awake. Gonioscopy and formal visual field testing should be performed to the best extent possible, appropriate for the child's ability and tolerance, regardless of presence or absence of glaucoma signs. After the age of 8 years, automated perimetry should be the visual field test of choice, where possible. Other testing modalities such as OCT of the retinal nerve fiber layer, ultrasound biomicroscopy, and diurnal IOP measurements were considered not fundamentally necessary for adequate evaluation, regardless of whether glaucoma signs are present, though these may be helpful in selected cases.

Medical/Surgical Management

Consensus was that medical IOP-lowering therapy is the first-line approach in patients with PWB and signs of glaucoma, especially in children aged $\geq$4 years. In children aged $<$4 years, medication is typically temporizing because most children eventually require surgical intervention.

For surgical interventions in children aged $<$4 years, goniosurgery was voted as the preferred choice, although this was not unanimous among the face-to-face participants. One member emphasized that the indication for surgery differs in children aged 2 to 3 months from those aged 6 months to 2 years, citing poor efficacy of goniosurgery in older children. Despite relatively lower complication risks of goniosurgery, this member strongly recommended trabeculectomy as the primary surgical intervention in children aged approximately $>$6 months.

When discussing preferred surgical treatment in patients aged 4 to 18 years with facial PWB and glaucoma, it was generally agreed that cyclodestructive surgery as a first procedure is less effective, although consensus could not be achieved for other procedures. It was agreed that goniosurgery is not the preferred first-choice procedure in this age range. Risk of choroidal detachment was cited as a reason to avoid goniosurgeries and trabeculectomy by those who did not prefer them. Members who preferred goniosurgery over trabeculectomy cited more complication risks with trabeculectomy. Those who preferred trabeculectomy cited low success rates of goniosurgery, and that modern trabeculectomy techniques may mitigate many complication risks. Consensus could not be achieved regarding the role of glaucoma drainage devices (GDDs).

Discussion

Sturge–Weber syndrome carries a lifetime risk of glaucoma in up to 70%[2] of affected individuals. Although much has been published regarding the ophthalmic manifestations of SWS, and the outcomes and complications of various surgical modalities, there are no consensus-based guidelines specifically addressing the issues unique to glaucoma associated with SWS.[4–6,12,13] Although Sabeti et al[14] published a consensus statement on SWS management and treatment, its scope was somewhat limited given the complexity of the disease and range of different practices and surgical preferences found among those who take care of these children. Our study addresses these gaps by offering more comprehensive and detailed consensus guidelines. Consensus development aims to integrate a deep understanding of any research relevant to the area with the collective clinical experience of a large enough group of participants to generate clinically appropriate recommendations. Consensus-based clinical practice guidelines are the preferred resource for both patients and clinicians in specific clinical scenarios.[15,16] The Delphi method was the first approach to consensus guideline development and has previously been used successfully to synthesize clinical decision-making and understanding in several areas of ophthalmology.[8–10]

We achieved consensus that patients with facial PWBs are at an increased risk of glaucoma and should undergo ophthalmic evaluation. For patients with involvement of 1 or both eyelids, we recommend an expedited referral within 1 month of PWB recognition to ensure early detection of glaucoma and timely intervention if necessary. Although not part of the consensus panel's discussion, it follows that there is even greater urgency to be evaluated by a childhood glaucoma specialist should there be any suspicion of glaucoma in a child with a facial PWB. It has been well established that facial PWB involving the eyelids increases risk of glaucoma.[17–19] Many studies have used traditional dermatome boundaries based on the trigeminal nerve divisions (V1, V2, and V3) to explore this, whereas others have used alternative anatomic distributions such as upper versus lower lid involvement, in an effort to understand this risk. Several studies reported that PWB involving the V1 (ophthalmic branch of the trigeminal nerve) region was significantly associated with glaucoma, although their defined V1 region varied in including the lower eyelid.[17,20,21] Some also found V2 involvement to be a risk factor.[18,22] Many studies report that ipsilateral combined upper and lower lid involvement, as well as bilateral eyelid PWBs, are associated with elevated risk.[19,23–25] More recently, Waelchli et al[26] demonstrated that PWB distribution patterns can better be explained by the distribution of the embryologic vasculature of the face rather than that of the trigeminal nerve, which may explain some of the inconsistencies between studies.[27]

Because of the risk of preventable vision loss, current guidelines align with our recommendations to refer to a pediatric ophthalmologist to ensure an appropriate baseline eye examination is performed in all children with PWBs, which will enable timely detection of concurrent or subsequent IOP elevation or other signs of glaucoma. Periodic evaluations continue to be required thereafter, with more frequent intervals for those at higher risk of SWS-related ophthalmic complications. The baseline eye examination should include visual acuity measurement, tonometry, slit-lamp examination, and a dilated eye examination, which should be assessed under sedation if adequate measurements cannot be obtained in office.[14,28] In the absence of ophthalmologists, we generally propose referral to a provider contingent on their ability to accurately assess for glaucoma. Other factors that increase the risk of glaucoma include choroidal hemangioma, iris heterochromia, or signs of raised EVP.[3,25,29] Monocular glaucoma, typically ipsilateral to the PWB, is more common in infants with SWS.[30] Glaucoma in SWS has a bimodal presentation pattern, with most patients presenting in infancy (<3 years) or after 4 years in the first and second decades.[31] Glaucoma pathogenesis can vary depending on the age of presentation, with glaucoma in older patients usually attributed to elevated EVP and glaucoma of infancy typically caused by goniodysgenesis.[31]

The treatment of glaucoma in patients with SWS varies depending on factors such as patient age, underlying mechanism of glaucoma, clinical presentation, and the surgeon's expertise and preference. Goniosurgery is preferred to address goniodysgenesis and has been reported as an effective treatment for early-onset glaucoma (onset <4 years), often caused by goniodysgenesis, with outcomes improved by incremental angle procedures or adjunctive medical therapy.[32–34] One member felt that goniosurgery should be reserved for children aged <6 months. Goniosurgery is recognized for a superior safety profile compared with trabeculectomy, which is associated with higher complication risks. For example, Iwach et al[32] reported a 40% frequency of choroidal effusions after trabeculectomy in patients with early-onset glaucoma. Goniosurgery may provide temporary IOP control, especially when combined with medications, with the anticipation of further surgery in due course.[32,35] Yeung et al[35] specifically recommend trabeculectomy as the preferred initial surgical approach over goniosurgery in children aged >1 year. Goniosurgery effectively addresses glaucoma caused by goniodysgenesis; however, it is believed to be poor in addressing glaucoma due to elevated EVP, which is more commonly observed in older patients. In such cases, the main resistance to aqueous outflow is located distal to Schlemm canal, so bypassing or removing trabecular outflow resistance does not address the persistently elevated pressure in the distal venous system (Gedde, 2021 #3991). Some argue that a combined trabeculotomy/trabeculectomy may offer better outcomes by targeting goniodysgenesis through trabeculotomy, whereas trabeculectomy creates an alternative outflow pathway, bypassing the episcleral venous system.[36,37] Combined trabeculectomy/trabeculotomy procedures may be an area of future investigation.

Trabeculectomy and GDDs are typically recommended for glaucoma with onset >4 years, although the higher risk of postoperative complications compared with both goniosurgery and the same surgery in non-SWS eyes is an important consideration. Trabeculectomy and combined trabeculotomy-trabeculectomy can be effective in creating an alternative outflow pathway from the episcleral venous system.[35,38,39] Choroidal effusions occur in 17% of patients with SWS and late-onset glaucoma.[32] Some have proposed that the risk of subconjunctival tissue fibrosis and subsequent bleb failure can be mitigated by using adjunctive antimetabolites during surgery, such as mitomycin C. In patients with SWS, Senthilkumar et al[40] observed a 70% success rate at 2 years with GDD implantation in those with late-onset glaucoma, and Karaconji et al[41] reported an 80% success rate with Ahmed glaucoma valve implantation after failed primary trabeculectomy. Other studies have supported similar GDD efficacy; however, they did not differentiate between early-onset and late-onset glaucoma.[39,42]

Regardless of age, we recommend medical therapy as the first-line treatment modality for all patients with SWS and glaucoma, consistent with existing literature.[35] However, apart from agreeing that goniosurgery should not be considered the first-line surgical choice for children aged >4 years, there was no consensus on the preferred surgical approach in this group, and in children aged <4 years, the success of medical therapy is limited, with surgery often required. Factors dividing the panel included the lower efficacy of goniosurgery in this age group than younger children, and a higher complication rate experienced when performing incisional filtering surgeries in these eyes, than many other forms of childhood-onset glaucoma. The consensus regarding no recommendation for angle surgery as first-line surgical treatment choice in children aged >4 years is in alignment with the literature and the presumed mechanism of late-onset glaucoma in SWS.

A possible limitation of our study relates to our exploration of participants' preferences based on 2 broad age groups distinguished by age of glaucoma onset: before 4 years and after the age of 4 years. We made no distinction within the <4-year-old group between very early-onset neonatal and infantile glaucoma and glaucoma developing after 3 to 6 months old. This may be relevant as certain surgeries may have different outcomes in the very young; for example, in neonatal-onset primary congenital glaucoma, which is considered a similar form of goniodysgenesis to that of early SWS-associated glaucoma, goniotomy has a significantly poorer prognosis than when performed in slightly older children aged between 1 month and 1 year.[43] Although we sought to maintain consistent language and format across rounds, another limitation is that the survey was inherently subject to the wording and structure chosen by the study team, which may have introduced variability in participant interpretation.

Perhaps the greatest limitation of the Delphi process is that it reflects only the expert practice patterns of those

involved. The purpose of the large lead-in surveys administered in the months before the final panel meeting is to submit to this panel only those areas where consensus in the larger population of practitioners is absent. By choosing a panel with a large collective experience in the subject area, and adhering to the Delphi technique, the representative value of the process is greatly enhanced. Although the expert committee was self-selected, the Delphi process minimizes the risk of undue influence from any single individual. The lack of consensus on some issues may stem from variations in resource availability, differences in patient populations, specific areas of expertise, and unique personal experiences of the panel members. Any consensus opinion may not be applicable to every patient. Additionally, the Delphi process is hard to adapt to individual patients, and therefore, target pressures and surgical decision points were not handled as a multitude of individual factors lead to these decisions in any given patient.

Our study is intended to fill the gaps in glaucoma management guidelines for SWS by providing comprehensive and detailed consensus recommendations. We employed the Delphi technique to develop recommendations based on current knowledge and clinical practice. Key areas of focus included screening strategies, particularly concerning the location of PWBs, as well as management approaches, care team coordination, diagnostic testing, and medical/surgical interventions. Topics where consensus was not achieved highlight areas that may benefit from further research and discussion.

Footnotes and Disclosures

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Abbreviations and Acronyms:
EVP = episcleral venous pressure; **GDD** = glaucoma drainage devices; **IOP** = intraocular pressure; **PWB** = port wine birthmark; **SWS** = Sturge−Weber syndrome.

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