American Journal of Medicine and Medical Sciences 2026, 16(4): 1701-1704
DOI: 10.5923/j.ajmms.20261604.32

Care Tactics for Children with Primary Congenital Glaucoma with Sturge-Weber Syndrome

Turakulova Dilfuza Mukhitdinovna[1,*], Nazirova Zulfiya Rustamovna[2], Karabaeva Iroda Murodzhonovna[3]

[1]Candidate of Medical Sciences, Department of Ophthalmology, Pediatric Ophthalmology, Associate Professor, Tashkent State Medical University, Uzbekistan
[2]Doctor of Medical Sciences, Department of Ophthalmology, Pediatric Ophthalmology, Associate Professor, Tashkent State Medical University, Uzbekistan
[3]Master's Student, Department of Ophthalmology, Pediatric Ophthalmology, Tashkent State Medical University, Uzbekistan

Abstract The article presents a retrospective and prospective analysis of the examination and treatment outcomes of 21 children with primary congenital glaucoma associated with Sturge–Weber syndrome. Clinical and instrumental features of the disease course are analyzed, including vascular abnormalities of the anterior chamber angle and episcleral vessels and their impact on intraocular pressure. The effectiveness of a combined surgical approach aimed at simultaneous influence on the main aqueous humor outflow pathways, along with preoperative hemostatic preparation, is evaluated. The results demonstrate that the proposed surgical technique provides effective intraocular pressure control in most patients; however, it is associated with a high risk of prolonged postoperative hypotony and choroidal complications. The study concludes that management of glaucoma in children with Sturge–Weber syndrome requires an individualized surgical strategy and careful postoperative monitoring.

Keywords Sturge–Weber syndrome, Port-wine stain, Etamsylate, Maklakov tonometry, iCare 100 tonometry

1. Relevance

Sturge-Weber syndrome (encephalotrigeminal angiomatosis) is a rare congenital neurocutaneous disorder often associated with glaucoma in children. [1,5]

Sturge-Weber syndrome is a rare congenital disorder, occurring in approximately 1 in 20,000–50,000 live births. Glaucoma is observed in a significant proportion of patients with this syndrome—according to the literature, in 30–70% of cases. [3,6,7]

Glaucoma in this syndrome is characterized by early onset, a progressive course, and a high probability of irreversible vision loss already in childhood. According to clinical observations, glaucoma in Sturge-Weber syndrome is detected in a significant proportion of patients and is often resistant to standard treatment methods. [4,8,10]

A characteristic feature of glaucoma associated with Sturge-Weber syndrome is its complex pathogenesis, associated with developmental anomalies of the anterior chamber angle, increased episcleral venous pressure, and vascular malformations. This results in the low effectiveness of conservative therapy and a high complication rate after surgical interventions. Furthermore, in young children, diagnosis and monitoring of intraocular pressure are difficult, leading to late detection of the disease and a worse prognosis. [9,11,12]

Despite the existence of various medical and surgical approaches to the treatment of glaucoma associated with Sturge-Weber syndrome, there are still no uniform standards for the care of these patients, and treatment outcomes remain variable. The high risk of progression of glaucomatous optic neuropathy, the need for repeated surgeries, and long-term follow-up emphasize the social and medical significance of this problem. [13,14,15]

In this regard, an analysis of current glaucoma treatment methods in children with Sturge-Weber syndrome is relevant and necessary. This will allow us to evaluate the effectiveness of various therapeutic strategies, determine optimal patient management tactics, and improve functional outcomes, helping to preserve visual function and improve children's quality of life. [2]

The aim of the study is to examine current approaches to the treatment of glaucoma in children with Sturge-Weber syndrome, evaluate their effectiveness, and determine optimal management tactics.

2. Materials and Methods

A retrospective and prospective study was conducted

* Corresponding author:
munojat1medical@gmail.com (Turakulova Dilfuza Mukhitdinovna)
Received: Mar. 4, 2026; Accepted: Mar. 28, 2026; Published: Apr. 8, 2026
Published online at http://journal.sapub.org/ajmms

involving 21 children diagnosed with Sturge–Weber syndrome and associated primary congenital glaucoma. The study was carried out at the Department of Ophthalmology, Tashkent State Medical University, during the period from 2018 to 2025.

The sample included 12 boys (55%) and 9 girls (45%). According to age distribution, 16 patients (76%) were infants, 4 (19%) were young children, and 1 (5%) was of preschool age.

Inclusion criteria: confirmed diagnosis of Sturge–Weber syndrome with glaucoma.

Exclusion criteria: absence of complete clinical data or follow-up.

All patients underwent comprehensive ophthalmological and general clinical examination, including: visometry; biomicroscopy; ophthalmoscopy; gonioscopy; echobiometry; Maklakov tonometry; iCare 100 tonometry; tonography. Magnetic resonance imaging (MRI) of the brain was performed to assess vascular abnormalities.

Tonometry was performed under general anesthesia 3–5 minutes after induction. Premedication included diphenhydramine 1% and atropine sulfate 0.1%. During induction, a combination of 0.5% sibazon, 40% sodium oxybutyrate (GHB), and 0.005% fentanyl was used.

All children underwent antiglaucoma surgery developed in our department using a combined method that involves simultaneous interventions on the outflow pathways in three directions: Burian sinusotrabeculotomy into the scleral sinus, cyclodialysis-cycloretraction with an autoscleral pedicle into the suprachoroidal space, and basal iridectomy with sclerectomy under the scleral flap into the episcleral venous system (Patent for Invention "Method for Surgical Treatment of Congenital Glaucoma" No. IAP 04890 dated May 12, 2014).

Ethical considerations. The study was conducted in accordance with the principles of the Declaration of Helsinki. Participation was voluntary. Written informed consent was obtained from the parents or legal guardians of all patients. Patient confidentiality and anonymity were strictly maintained.

Statistical analysis was performed using standard statistical methods. Quantitative data are presented as mean ± standard deviation.

3. Results

The study included 21 patients (12 boys and 9 girls). The majority were infants (76%).

All patients had a congenital capillary malformation of the facial skin (nevus flammeus), localized predominantly to one side of the face. The lesion appears as a flat, pink-red spot with indistinct borders, without signs of infiltration or elevation.

The coloration is relatively uniform, with a tendency to increase in intensity in the central areas.

The lesion extends into the innervation zone of the first (ophthalmic) and partially the second (maxillary) branches of the trigeminal nerve, including the periorbital area and buccal surface. The skin in the affected area is smooth, without scaling or ulceration.

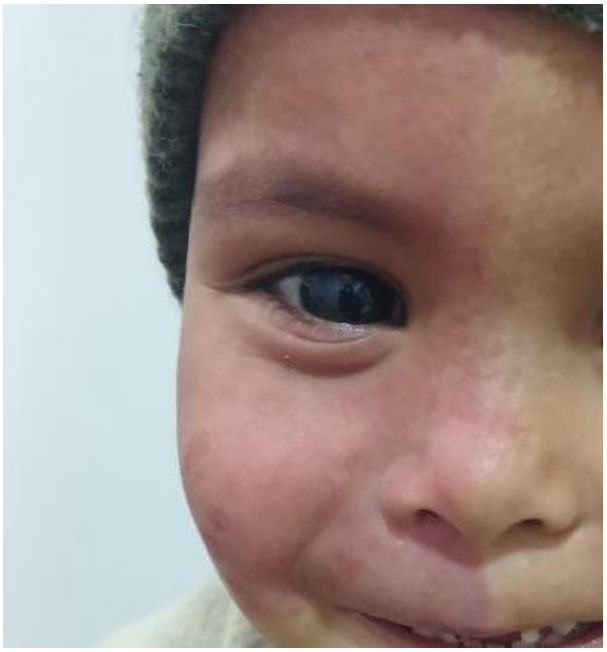

Figure 1. Congenital capillary malformation of the facial skin (nevus flammeus), localized predominantly in the area of one half of the face

The described vascular malformation corresponds to the classic cutaneous manifestation of Sturge-Weber syndrome, associated with the possible presence of leptomeningeal angiomatous malformation.

Distribution of eyes by disease stage showed that the initial stage was not observed, the advanced stage was observed in 7 eyes (32.9%), the advanced stage was observed in 11 eyes (55.2%), and the terminal stage was observed in 3 eyes (11.9%), respectively.

Preoperative examination data were as follows: the mean preoperative IOP was 32.75 ± 1.1 mmHg with a range (24–40 mmHg): at the advanced stage of the disease, the IOP was 29.5 ± 1.2 mmHg, and at the advanced stage, 36.8 ± 1.5 mmHg. and at the terminal stage, 40.2 ± 1.33 mmHg.

The anteroposterior axis (APA) of the eyeball, according to echobiometry data, was increased in 100% of cases: by 1–2 mm above the age norm (mean 21.5 ± 0.3 mm) in 7 eyes at the advanced stage, by 2–4 mm (24.8 ± 0.5 mm) in 11 eyes at the advanced stage, and by 5 mm or more (29.2 ± 0.5 mm) in 3 eyes at the terminal stage.

The average preoperative corneal diameter was 12.5 ± 0.76 mm (range 12–15 mm). During the study, preoperative corneal edema was recorded in 7 eyes (36.6%) and was superficial, manifesting as epithelial edema. In the remaining cases, the cornea remained clear—14 eyes (63.7%).

Gonioscopy revealed a predominantly wide-open anterior chamber angle in all patients with Sturge-Weber syndrome. The structures of the anterior chamber angle—the trabecular meshwork, scleral spur, and iris root—are visualized, but significant vascular changes are noted.

A characteristic feature is abnormal vascularization of the anterior chamber angle, represented by multiple dilated, tortuous blood vessels running along the trabecular meshwork and in the area of Schlemm's canal. The vessels are angiomatous in nature, often arranged in a reticular or radially oriented plexus.

The trabecular meshwork appears hyperemic, with vessels occasionally visible, without signs of true goniodysgenesis. Some patients exhibit thickening and increased pigmentation of the trabeculae, as well as a lack of normal differentiation of the angle structures. The severity of vascular changes correlates with increased episcleral venous pressure, leading to functional impairment of aqueous humor outflow despite an anatomically open anterior chamber angle.

Using ophthalmoscopy, optic nerve excavation was 0.3–0.4 in 7 eyes (33.3%) at the advanced stage. In the advanced stage, optic nerve excavation was 0.5–0.7 in 11 eyes (52.2%) and 0.7–0.8 in 3 eyes (14.2%), respectively.

All patients underwent surgery using our proposed method. In the preoperative preparation phase, all children were prescribed the hemostatic drug etamsylate at an age-appropriate dosage to prevent intraoperative intraocular hemorrhage.

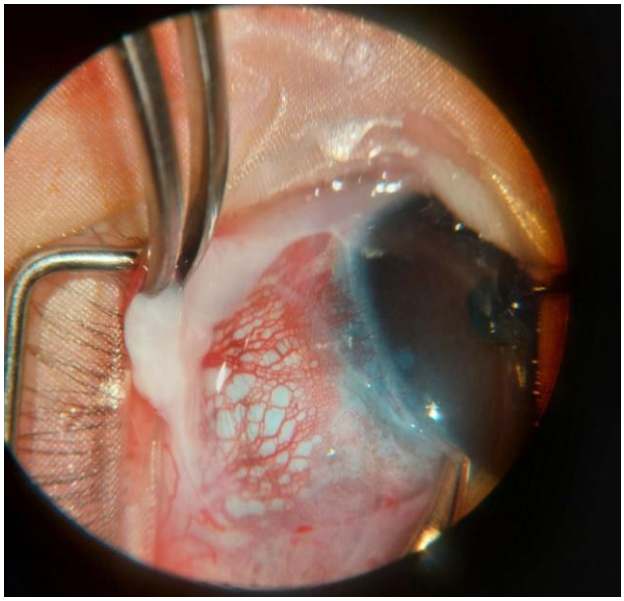

Figure 2. Intraoperative image of the scleral surface in a patient with Sturge-Weber syndrome

Intraoperative characteristics of the sclera and episcleral vessels.

After conjunctival incision in a child with Sturge-Weber syndrome, pronounced changes in the episcleral vascular bed, characterized by diffuse angiomatous transformation, were detected. The episcleral vessels are sharply dilated, tortuous, and congested, forming dense vascular plexuses with a reticular pattern, spreading over a significant area of the sclera.

The vascular formations are located directly on the scleral surface, have an unclear architecture, and lack the usual segmental vascular pattern. Increased vascularization and congestive blood flow are noted, indicating a congenital vascular malformation of the episcleral layer. Thinning of the underlying sclera is visible, with localized translucency of the choroid. There are no signs of inflammatory infiltration. The severity of the angiomatosis causes increased tissue bleeding during surgical procedures.

These changes are consistent with episcleral angiomatous malformation, characteristic of Sturge-Weber syndrome, and are considered one of the pathogenetic factors for increased episcleral venous pressure, leading to the development of secondary glaucoma.

Complications were observed in the postoperative period. The most common complications in the early postoperative period (up to 7 days) included central choroidal obstruction, hyphema, increased intraocular pressure, shallow anterior chamber syndrome, and hypotony.

On the first postoperative day, hypotony (-) 0.5–1.0 and mild choroidal edema were diagnosed in 14 eyes (65.2%) with a clinical picture typical for this complication. These patients were prescribed atropine sulfate instillations at age-appropriate doses twice daily. Normal intraocular pressure was detected in 7 eyes (34.8%).

On the third postoperative day, the number of patients with hypotony increased to 18 eyes (85.7%). B-scans revealed significant choroidal edema, so conservative treatment was prescribed for these patients (atropine sulfate eye drops at age-appropriate doses twice daily, and 3% sodium caffeine benzoate instillations into the conjunctival sac 4–5 times daily). On the fifth day, severe hypotony greater than (-) 1.0 and CCO detachment were observed in 7 eyes (25.9%). These patients were prescribed instillations of 3% sodium caffeine benzoate into the conjunctival sac 4-5 times daily, 0.4% dexamethasone lymphotropic, 0.5 ml once daily, and a pressure bandage once daily. In 3 eyes (14.3%), intraocular pressure was within normal limits. On the seventh postoperative day, hypotony persisted in 4 eyes (19.3%), and CCO detachment was observed. It developed against the background of shallow anterior chamber syndrome and unsuccessful conservative treatment. These patients were recommended to continue conservative treatment in the hospital (dexamethasone instillation, 2 drops 6 times daily and continue dexamethasone lymphotropic). Following conservative treatment, IOP returned to normal, and the patients were discharged home under the care of a local ophthalmologist.

With continued conservative treatment, intraocular pressure returned to normal in two patients, while hypotension persisted in two others. All four patients were discharged under the care of a local ophthalmologist.

For the two patients with persistent hypotension, conservative treatment was continued, including dexamethasone, atropine, and caffeine instillations. No signs of choroidal detachment were detected, but the anterior chamber remained shallow, and intraocular pressure remained at -0.5.

4. Discussion

The aim of this study was to evaluate the effectiveness of

surgical management of glaucoma in children with Sturge–Weber syndrome. The results demonstrated that the combined surgical approach provides effective intraocular pressure control in most cases. However, a high incidence of postoperative hypotony and choroidal complications was observed. These findings are consistent with previous studies reporting high surgical complexity and complication rates in Sturge–Weber-associated glaucoma [6,7]. However, some authors report lower rates of postoperative hypotony, which may be related to differences in surgical techniques. The strengths of this study include a combined retrospective and prospective design and detailed clinical analysis. Limitations include a relatively small sample size and the absence of long-term follow-up in some patients.

5. Conclusions

The study demonstrated that combined surgical treatment is effective in controlling intraocular pressure in children with Sturge–Weber syndrome.

From a healthcare perspective, early diagnosis and individualized surgical strategy are essential to improve visual outcomes and reduce complications.

It is recommended to ensure careful postoperative monitoring and further research with larger patient groups.

ACKNOWLEDGEMENTS

The authors would like to thank the staff of the ophthalmology department for their assistance.

Financial Support and Sponsorship

None.

Author Contributions

T.D.M. – study design, supervision
N.Z.R. – data analysis, manuscript editing
K.I.M. – data collection, manuscript preparation

Conflict of Interest

The authors declare no conflict of interest.

REFERENCES

[1] Higueros E, Roe E, Granell E, Baselga E. Sturge-Weber Syndrome: A Review. Actas Dermosifiliogr. 2017 Jun; 108(5): 407-417. English, Spanish. doi: 10.1016/j.ad.2016.09.022. Epub 2017 Jan 23. PMID: 28126187.

[2] Yuen NSY, Wong IYH. Congenital Glaucoma from Sturge-Weber Syndrome: A Modified Surgical Approach. Korean J Ophthalmol. 2012; 26(6): 481–484. doi: https://doi.org/10.3341/kjo.2012.26.6.481.

[3] Management of Childhood Glaucoma in Sturge-Weber Syndrome: A Case Report. DOI: 10.18240/ijo.2018.04.23.

[4] Cibis GW, Tripathi RC, Tripathi BJ. Glaucoma in Sturge-Weber syndrome. Ophthalmology. 1984 Sep; 91(9): 1061-71. doi: 10.1016/s0161-6420(84)34194-x. PMID: 6493715.

[5] Hassina S., Krichene M.A., et al. Congenital Glaucoma in Sturge-Weber Syndrome. Scholars Journal of Medical Case Reports. 2024; 12(05): 889–891. DOI: 10.36347/sjmcr.2024.v12i05.081.

[6] Wen T., Wang L., Luo H., Tang L. Sturge-Weber syndrome secondary glaucoma: From pathogenesis to treatment. Eye and Vision. 2025; DOI: 10.1186/s40662-025-00432-6.

[7] Sadovnikova N.N., Brzheskiy V.V., Zertsalova M.A., Baranov A.Yu. Clinical presentation, diagnosis, and treatment of glaucoma associated with Sturge-Weber syndrome. Ophthalmology Reports. 2025; 18(2): 35-42. https://doi.org/10.17816/OV648669.

[8] Savranova T.N., Rozukulov V.U., Yusupov A.F., Asadov D.A., Saidzhonov S.S. Surgical treatment of glaucoma with filtration bleb activation in Sturge-Weber syndrome. 2022; 15(3): 128-135. https://doi.org/10.21516/2072-0076-2022-15-3-128-135.

[9] Comi A.M. Sturge-Weber syndrome. Handbook of Clinical Neurology. 2015; 132: 157-168. doi: 10.2147/TACG.S363685.

[10] Egorov E.A., Katargina L.A. Pediatric ophthalmology. - M.: GEOTAR-Media, 2019.

[11] Katargina L.A., Tarasova L.N. Congenital and early childhood glaucoma. - M.: Medicine, 2018.

[12] Fedorov S.N., Zakharov V.P. Surgical treatment of glaucoma in children. - M.: Eye Microsurgery, 2017.

[13] Malyugin B.E., Pashtaev N.P. Modern methods of surgical treatment of glaucoma. - M.: Ophthalmology, 2016.

[14] Katargina L.A. Glaucoma in children with Sturge-Weber syndrome // Bulletin of Ophthalmology. — 2020. — No. 4. — pp. 25–31.

[15] Budenz D.L., Gedde S.J. Glaucoma in Sturge-Weber syndrome. // Ophthalmology. — 2000. — Vol. 107. — P. 136–143.