

# A CASE OF PEDIATRIC STURGE-WEBER SYNDROME

The steps that saved the vision of a 3-year-old girl.

BY JAYASREE VENUGOPAL, MS, MBBS

*Sturge-Weber syndrome is a congenital, nonfamilial disorder that can affect the eye in the form of conjunctival, episcleral, and choroidal hemangiomas. Surgical intervention is often needed, but it can be associated with sight-threatening intraoperative complications. Being prepared for these and taking suitable intraoperative precautions can help make the surgery easier and more predictable. This excellent teaching video by Dr. Venugopal demonstrates surgical measures that can be taken in such cases.*

—Soosan Jacob, MS, FRCS, DNB



Sturge-Weber syndrome (encephalotrigeminal angiomas) is a phakomatosis characterized by hamartomas involving the skin, the eyes, and the brain.<sup>1</sup> Glaucoma is reported in 30% to 70% of patients. Among them, 60% of cases are congenital and respond poorly to medical management.<sup>2</sup> Glaucoma surgery in Sturge-Weber syndrome is asso-

ciated with increased rates of severe complications like suprachoroidal hemorrhage, massive choroidal effusion, and serous retinal detachment.<sup>2</sup> This risk occurs secondary to the rapid movement of fluid from the intravascular space to the extravascular space because of the elevated venous pressure and sudden hypotony during surgery.

## CASE PRESENTATION

A 3-year-old girl presented with left-sided port wine stain and with buphthalmos, a cup-to-disc ratio of 0.8, an IOP of 30 mm Hg, and a visual acuity of 20/25 OS. The eye's corneal diameter was 14 mm (right eye was 11 mm), and the axial length measured 24.57 mm (right eye was 22.03 mm). A dilated fundus examination and B-scan did not reveal choroidal involvement. A neurology consultation and imaging did not suggest any neurological involvement.

I started the patient on medical management, but I could not achieve IOP control. Hence, I planned for trabeculectomy with mitomycin C and a collagen matrix implant under general anesthesia.

## WATCH IT NOW



## SURGICAL PLAN

There were several objectives during the surgery. I had to avoid sudden decompression of the globe to prevent rapid movement of fluid to the extravascular space, and I also had to avoid fluctuation of the IOP and the anterior chamber volume. I needed to create access to the suprachoroidal space for drainage in the event of suprachoroidal hemorrhage/effusion during or after surgery<sup>3</sup> and to reposition the flap as soon as possible after removing the corneoscleral block and performing a peripheral iridectomy.

My initial surgical steps were similar to those of a routine trabeculectomy with limbus-based conjunctival dissection, thorough cautery application, mitomycin C application (0.2 mg/mL for 1 minute), and scleral flap dissection. I made a radial sclerotomy in the inferotemporal quadrant 5 mm from the limbus, dissected the conjunctiva and Tenon capsule in the sclerotomy site, and applied cautery. The sclera was incised partially with a number 11 Bard-Parker knife. I made additional passes within the track of initial incision, while taking care not to cut the underlying choroid. The appearance of the dark color of the choroid between the lips of the scleral incision indicated that I had reached the suprachoroidal space.

I made a controlled paracentesis with a 26-gauge needle to prevent sudden decompression during subsequent

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**“ Glaucoma surgery in Sturge-Weber syndrome is associated with increased rates of severe complications.”**

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steps in surgery. To preserve chamber volume and prevent fluctuations in IOP, I introduced an anterior chamber maintainer. The next precautionary measure was to preplace two scleral flap sutures prior to removing the corneoscleral block so I could close the scleral flap immediately in the event of choroidal effusion or hemorrhage. I removed the corneoscleral block and performed a peripheral iridectomy. I then tied the preplaced sutures and added two more, while taking special care to tie them tightly.

After I sutured the scleral flap, I removed the anterior chamber maintainer and introduced air into the anterior chamber to preserve its depth. I kept a collagen matrix implant over the scleral flap, and I approximated the Tenon capsule and conjunctiva in separate layers. At the sclerotomy site, I left the underlying scleral incision unsutured for possible drainage in the postoperative period. I sutured the overlying conjunctiva with an 8-0 Vicryl suture (Ethicon).

## **POSTOPERATIVE RESULTS**

Postoperatively, the child's left eye had visual acuity of 20/25, an IOP of 8 mm Hg, a diffuse bleb, a well-formed anterior chamber, and no choroidal effusion. ■

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