

Clinical Course

1 High-risk capillary malformation

An 8-month-old girl with an extensive congenital right-sided capillary malformation involving the upper face, trunk, and extremities presented for pulsed-dye laser therapy.

2 Early severe seizures

Her course was complicated by focal seizures progressing to status epilepticus, requiring hospitalization and transfer to Boston Children's Hospital.

3 MRI consistent with SWS

Brain MRI showed right-sided leptomeningeal enhancement, prominent subdural vessels, and asymmetric enlargement of the choroid plexus.

4 Surgical seizure control

Given severe focal epilepsy, she underwent right posterior quadrant disconnection. No recurrent seizures were reported at follow-up.

Clinical Images



References

- Mologousis MA, Bitar RA, Pinto AL, Rispoli JM, Liang MG. Early magnetic resonance imaging as a screen for Sturge-Weber syndrome-related seizures in infants with upper-facial capillary malformations. *Pediatr Dermatol*. 2026;43(1):56-63. doi:10.1111/pdc.7
- Valery CB, Iannotti I, Kossoff EH, et al. Retrospective analysis of presymptomatic treatment in Sturge-Weber syndrome. *Ann Child Neurol*. 2024;2(1):60-72. doi:10.1002/cns3.20058
- Sugano H, Nakanishi H, Nakajima M, et al. Posterior quadrant disconnection surgery for Sturge-Weber syndrome. *Epilepsia*. 2014;55(5):683-90. doi:10.1111/epi.12547

Discussion

1 Early recognition may change management

Upper-facial capillary malformations are an important dermatologic clue to Sturge-Weber syndrome. In a recent cohort of 33 infants with forehead or temple capillary malformations, 14 of 20 infants with an abnormal early MRI later developed seizures, while none of the 13 infants with a normal early MRI did.¹ These findings suggest that early, high-quality MRI may help identify infants at increased neurologic risk and support earlier referral, counseling, and surveillance.

2 Presymptomatic treatment is an emerging strategy

Early diagnosis may be increasingly actionable. In a retrospective study of 92 children with SWS, patients who received presymptomatic treatment were more likely to remain seizure-free at age 2 years than untreated patients (47% vs 12%).² Treatment regimens included low-dose aspirin with antiseizure medications, including levetiracetam and oxcarbazepine. These data are encouraging, but prospective trials are still needed before a standardized preventive regimen can be recommended.

3 Surgery may provide seizure control

Seizures in SWS can become medically refractory and contribute to neurologic morbidity. For selected patients with focal posterior disease, posterior quadrant disconnection can isolate epileptogenic temporal, parietal, and occipital cortex while preserving uninvolved frontal motor networks. In one series of patients with SWS, 8 of 10 were seizure-free 2 years after surgery.³ Our patient similarly had no recurrent seizures reported following right posterior quadrant disconnection.

DERMATOLOGY TAKEAWAY

Dermatologists may be the first clinicians to recognize a high-risk upper-facial capillary malformation. Early recognition can prompt neurology and ophthalmology referral, discussion of MRI screening, seizure-risk counseling, and multidisciplinary management before severe neurologic complications occur.