

# Sturge-Weber Syndrome: A Review

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**Sturge-Weber syndrome is a rare disorder that occurs with a frequency of approximately 1 per 50,000. The disease is characterized by an intracranial vascular anomaly, leptomeningeal angiomas, most often involving the occipital and posterior parietal lobes. Facial cutaneous vascular malformations, seizures, and glaucoma are among the most common symptoms and signs. Stasis results in ischemia underlying the leptomeningeal angiomas, leading to calcification and laminar cortical necrosis. The clinical course is highly variable and some children experience intractable seizures, mental retardation, and recurrent strokelike episodes. In this review, we describe the syndrome's characteristic features, clinical course, and optimal management. © 2004 by Elsevier Inc. All rights reserved.**

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## Introduction

Sturge-Weber syndrome (SWS) is a rare, sporadic neurocutaneous syndrome affecting the cephalic venous microvasculature. An estimated frequency of 1 per 50,000 live births have SWS, although experts believe many more people have the disorder but have not yet been identified [1]. The hallmark intracranial vascular anomaly is leptomeningeal angiomas, most often involving the occipital and posterior parietal lobes, but it can affect other cortical regions and both cerebral hemispheres. An ipsilateral facial cutaneous vascular malformation usually affects the upper face in a distribution consistent with the ophthalmic division of the trigeminal nerve. Other clinical findings associated with SWS are seizures, glaucoma, headache, transient strokelike neurologic deficits, and behavioral problems. Hemiparesis, hemiatrophy, and hemianopia may occur contralateral to the cortical abnormality.

## Clinical Presentation and Diagnosis

A child with Sturge-Weber syndrome typically presents at birth with a facial cutaneous vascular malformation, a port-wine nevus, usually affecting the upper face ipsilateral to the angiomas. However, it is important to observe that most children with a facial cutaneous vascular malformation do not have SWS. When the cutaneous malformation is unilateral or bilateral and includes the ophthalmic division of the trigeminal nerve, the likelihood of SWS increases. The overall risk of SWS associated with any kind of facial cutaneous vascular malformation is approximately 8%. Children with the involvement of the eyelids are at elevated risk for eye and brain disease [2]. Rarely, some children with SWS lack a facial cutaneous vascular malformation but have the neurologic or ophthalmic components. The intracranial leptomeningeal angiomas is a key diagnostic feature in SWS.

When a child is born with a facial cutaneous vascular malformation covering a portion of the upper or the lower eye lids, imaging should be performed to screen for intracranial leptomeningeal angiomas. Leptomeningeal angiomas may not be apparent early on in infancy, but longitudinal studies have not yet been undertaken to define the optimal age of screening with magnetic resonance imaging. Imaging studies can indicate the degree and amount of cerebral calcification, atrophy, neuronal loss, and gliosis [3]. Computed cranial tomography provides adequate evaluation of brain calcifications [4]. However, calcifications may be absent or minimal in neonates and infants. Therefore magnetic resonance imaging with contrast is the preferred imaging modality for evaluation of the leptomeningeal angiomas. Metabolic imaging studies with single-photon emission computed tomography, positron emission tomography, or magnetic resonance spectroscopy may also assist in characterizing the extent of brain disease in SWS.

Seventy-five to 90% of children with SWS develop partial seizures by 3 years of age [5]. Of the longitudinal studies published, none demonstrate that early onset of

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seizures indicates poor prognosis. In fact, retrospective studies do not support the widely held belief that seizure frequency early in life in patients who have SWS is a prognostic indicator. However, some patients develop intractable epilepsy, permanent weakness, hemiatrophy, and visual field cuts, glaucoma, and mental retardation [6,7]. The hemiparesis and hemiatrophy are thought to arise from chronic cerebral hypoxia. Other findings common to patients with SWS are vascular headache (40-60%), developmental delay and mental retardation (50-75%), glaucoma (30-70%), hemianopsia (40-45%), and hemiparesis (25-60%) [8].

Diagnosis of SWS is made on the basis of the presence or absence of ophthalmologic or neurologic disease. The disease course, however, is variable and the patient must be continually monitored for complications.

### Neuropathologic Deterioration

The leptomeningeal angiomas usually involves the occipital and parietal lobes, but can affect the entire cerebral hemisphere. In gross section, the leptomeninges appear thickened and discolored by the leptomeningeal angiomas. Enlargement of the choroid plexus is common. Calcifications are observed in meningeal arteries and in cortical and subcortical veins underlying the leptomeningeal angiomas. Laminar cortical necrosis can accompany calcifications, suggesting ischemic damage secondary to venous stasis in leptomeninges and in the cerebral vascular bed. With continued progression, neuronal loss and gliosis can occur. Some of the pathologic findings result from the primary leptomeningeal vascular malformation. Obstruction occurring within the vascular malformation can cause stasis, decreased venous return, hypoxia, and decreased neuronal metabolism [9,10]. In addition, the absence of normal leptomeningeal vessels may hinder neuroglial oxygenation, especially during seizures, when there is increased oxygen demand. Ischemia is associated with severe physiologic changes, including abnormal drainage into the deep cortical veins and hypertrophy of the choroid plexus, increased capillary permeability, hypoxia in the adjacent tissue, alterations in pH, calcium deposition, cerebral atrophy, and disruption of the blood-brain barrier [11-13].

Pathologic deterioration occurs in some patients with SWS; however, disease progression varies widely. Saltatory neurologic decline and strokelike episodes can be attributed to recurrent thrombotic deterioration, in which venous stasis in the leptomeningeal malformation causes repeated thrombosis, resulting in progressive, recurrent infarction that underlies loss of neurologic function [14]. Although cases of subarachnoid hemorrhage in SWS have been reported in one study, it is rare and not well documented [15].

In SWS, abnormalities of the skin, leptomeninges, eye, and cortex can be traced to malformation of an embryonic vascular plexus within the cephalic mesenchyme between

the epidermis (neuroectoderm) and the telencephalic vesicle [16]. It is presumed that at approximately 5 to 8 weeks of gestation interference with the development of vascular drainage of these areas subsequently affects the face, eye, leptomeninges, and brain. Low-flow angiomas involving the leptomeninges best describes the typical imaging findings associated with SWS [5]. The angiomas is accompanied by poor superficial cortical venous drainage, and enlarged regional transmedullary veins develop as alternate pathways for venous drainage [9,10,17,18]. The ipsilateral choroid plexus may become engorged [19-22]. Microcirculatory stasis promotes chronic hypoxia of both the cortex and the underlying white matter [18,20,23,24] and hinders neuroglial oxygenation, particularly at times of increased demand, such as when seizures occur. The resulting hypoxia is associated with several physiologic changes: disruption of the blood-brain barrier, increased capillary permeability, and alterations in pH [11-13]. Ultimately, there is tissue loss and dystrophic calcification. The key radiologic features, therefore, are vascular abnormalities, atrophy, and cortical calcifications [23-25]. The actual leptomeningeal angiomas is usually an anatomically static lesion, but documentation of the progressive nature of SWS is clear [26,27]. With increasing hypoxia, disease progression and neurologic deterioration may occur.

### Neurologic Complications

Children with SWS suffer from a variety of neurologic abnormalities, including epilepsy, mental retardation, and attention-deficit hyperactivity disorder, migraine, and strokelike episodes. Seventy-five to 90% of children with SWS have epilepsy. Focal seizures are initially observed in most children who have SWS. Fever and infection often precipitate seizure onset. If noncontrasted computed tomography obtained in the emergency room setting after seizure activity is reported as normal or reveals focal calcification ipsilateral to a cutaneous angioma, more complete cerebral imaging is warranted. Most seizures are focal, because the lesion responsible for the epilepsy in SWS is focal. Seizures are likely caused by hypoxia and microcirculatory stasis. Children with radiographic findings of intracranial angiomas usually develop seizures by the age of 3 years. Approximately half these children have frank mental retardation [28], whereas others display learning disabilities, attention disorders, or behavioral disturbances.

Seizures typically begin in early infancy and may occur in conjunction with other focal deficits, namely hemiparesis. Some reports suggest that mental retardation may be more common in children whose seizures begin before the age of 2 years or who have seizures that are not controlled with antiepileptic drugs [29]. However, it is important to underscore that longitudinal studies have yet to be conducted to define the true implications of early-onset seizures. Prior research has linked the extent of cerebral

angiomas with neurologic outcome. Although widespread vascular lesions may be associated with more severe seizures, there are no prospective studies that have examined correlates of seizure onset and mental outcome. Some children experience saltatory neurologic decline separate from overt seizure activity. Venous occlusions and hypoxia may account for this decremental neurologic deterioration. In summary, prior research findings do not clearly support the correlation between early-onset seizure and poor prognosis [26].

Clinically, most children reach age-appropriate developmental milestones in the first few months of life, but approximately half of all patients with SWS will not maintain this pattern [28-33]. The degree of developmental delay and mental retardation in SWS patients is dictated by the extent of neurologic involvement. Delays are much more common in patients with bilateral disease. Approximately 50-60% of patients with SWS will have developmental delay or mental retardation, or both. Anecdotally, patients who develop debilitating aspects of SWS often do so before grade school; hemiparesis, hemianopia, retardation, and epilepsy are often apparent but infrequently severely worsen before grade school.

Attention-deficit hyperactivity disorder is another comorbid condition associated with SWS. As with attention-deficit hyperactivity disorder patients in the general population, SWS patients need to be monitored for medication requirements to minimize impulsivity and inattention. Management of attention-deficit hyperactivity disorder can be achieved and may significantly improve daily functioning in some SWS patients.

Headaches affect 30-45% of patients with SWS. The temporal relationship between headaches, seizure clusters, and strokelike episodes is related to the pathogenesis of SWS [34]. An animal model of migraine demonstrates that dural stimulation causes distention of cranial vessels, stimulation of trigeminal afferents, and release of vasoactive peptides, resulting in vascular dilatation [35]. The leptomenigeal angioma in SWS patients may predispose them to neuronal hyperexcitability, causing changes in cortical perfusion and oxygenation consistent with theories on pathogenesis of migraine [36]. Patients with SWS are at increased risk of stroke because preexisting perfusion and metabolic defects subject them to prolonged phases of oligemia, a characteristic of migraine-induced stroke [37,38]. Vomiting, fever, and dehydration can create a hyperviscous state, placing the patient at further risk of thrombosis.

Transient focal deficits (strokelike episodes) are a unique feature in SWS, with the most common manifestation being transient episodes of hemiparesis or visual field defects not directly associated with epilepsy. These episodes may last hours to several days. Electroencephalography obtained during times of weakness indicates focal slowing but does not typically demonstrate epileptiform activity that could account for acute motor deficits. Importantly (and in contrast to Todd's palsy), strokelike

episodes precede the onset of seizures. Recurrent thrombosis is a hypothetical mechanism of strokelike episodes and neurologic deterioration in SWS. Prevention of thrombosis may delay or prevent neurologic deterioration. Aspirin and other antiplatelet drugs are thought to limit the number of recurrent strokelike episodes, thereby preventing further neurologic damage [39]. Prospective controlled trials are required to test relevant hypotheses on efficacy of antiplatelet therapies in SWS.

## Ocular Complications

Ocular complications arise primarily from vascular abnormalities of the conjunctiva, episclera, retina, and choroid. When the facial cutaneous vascular malformation involves the eyelid, vascular abnormalities of ocular circulation may occur. Glaucoma is the most common ophthalmic complication of SWS, occurring in 30-70% of patients [29,33]. Presentation of glaucoma is bimodal; 60% develop glaucoma in infancy when the eye is susceptible to increased intraocular pressure, 40% develop glaucoma in childhood or early adulthood. Early-onset glaucoma causes infants to develop enlarged corneal diameters and myopia. Late onset glaucoma prompts little to no eye enlargement [6,33].

Two mechanisms for development of glaucoma in SWS have been proposed [40]. In one scenario, anterior chamber angle anomalies, which are consistently observed in infantile glaucoma associated with SWS, increase resistance to outflow of aqueous fluid and consequently raise intraocular pressure. In contrast, patients with late-onset glaucoma usually have normal anterior chamber angles or only mild abnormalities. These patients, however, have clinical signs of raised episcleral venous pressure, a key factor in the second explanation for glaucoma development in SWS: elevation of intraocular pressure as a result of increased episcleral venous pressure caused by arteriovenous shunts within the episcleral hemangioma [41]. Patients with early-onset glaucoma treated surgically or medically may develop elevated intraocular pressure as conjunctival or episcleral hemangiomas become more evident. Table 1 summarizes the recommended treatments for associated complications in SWS.

## Psychosocial Aspects of SWS

Families of patients with mild impairments are often able to easily adjust to living with SWS [42]. However, for others, SWS is devastating, especially when children experience recurrent seizures, pervasive learning and behavioral problems, and disabling visual impairment. Also, classmates may taunt children with SWS because of apparent physical and social disabilities. Because the effects of SWS on individual patients and families are difficult to predict, support and education tailored to each family's specific needs are key [43]. The Sturge-Weber Foundation offers support for families dealing with this

**Table 1. Recommended treatments for associated complications in Sturge-Weber syndrome**

Therapy	Glaucoma	Partial Epilepsy <sup>†</sup>	Headache/Migraine <sup>‡</sup>	Strokelike Episodes	Neurobehavior
1st choice	Beta blocker drops	Carbamazepine	Ibuprofen	Aspirin 3–5 mg/kg/d <sup>§</sup>	Methylphenidate
2nd choice	Adrenergic drops or carbonic anhydrase inhibitor drops	Valproate Topiramate Phenobarbital Phenytoin	Abortive therapy with sumatriptan	Unknown	Clonidine
Other options	Trabeculectomy*	Epilepsy surgery	Preventive therapy with propranolol or nortriptyline	Unknown	Dextroamphetamine Risperidone

\* If drops fail, consider trabeculectomy. Surgery is associated with a high risk of complications because of elevated episcleral venous pressure from the hemangioma.

<sup>†</sup> Children with SWS may have generalized seizures or infantile spasms, but partial epilepsy (with or without secondary generalization) is most prevalent.

<sup>‡</sup> Approximately one third of children with SWS meet diagnostic criteria for migraine. Management in SWS is similar to management of common migraine not associated with SWS.

<sup>§</sup> Anecdotal evidence suggests that children with SWS receiving aspirin therapy (antiplatelet dose) have fewer strokelike episodes.

Abbreviation:

SWS = Sturge-Weber syndrome

disorder and up-to-date information regarding research progress ([www.sturge-weber.org](http://www.sturge-weber.org)).

### Radiologic Findings

Advances in neuroimaging techniques have afforded a more precise look at the pathology of SWS. Magnetic resonance imaging and computed tomography are the imaging modalities most widely used, although single-photon emission computed tomography, positron emission tomography, and magnetic resonance spectroscopy are also of value. Plain skull x-rays and angiography are less useful. Plain skull x-rays illustrate the classic “tram-line” or “tram-track” calcifications but are not helpful in the diagnosis of SWS early in life because calcifications may not appear until later. Angiography demonstrates an overall lack of superficial cortical veins, nonfilling of the dural sinuses, and tortuous course of veins toward the vein of Galen. Angiography also reveals evidence of venous stasis characteristic of SWS, although arterial flow is normal. Angiography, however, is no longer used to visualize the vascular anatomy because vascular anomalies can be observed more clearly and noninvasively with magnetic resonance imaging.

Magnetic resonance imaging is best for structural imaging and allows perspective on all aspects of SWS, except for calcifications, which are still best viewed using computed tomography (Fig 1) [5]. Complete evaluation of SWS includes spin-echo T<sub>1</sub>-weighted and T<sub>2</sub>-weighted images with administration of gadolinium contrast. T<sub>1</sub>-weighted images after enhancement are essential to defining the extent of the vascular malformation (Fig 2) [44,45]. Gradient recalled-echo images with a long echo

time are helpful in detecting calcifications. Magnetic resonance venography may be useful, but the flow within the malformation is generally too slow to be imaged using this technology. White matter abnormalities are common in patients with SWS and are thought to be caused by

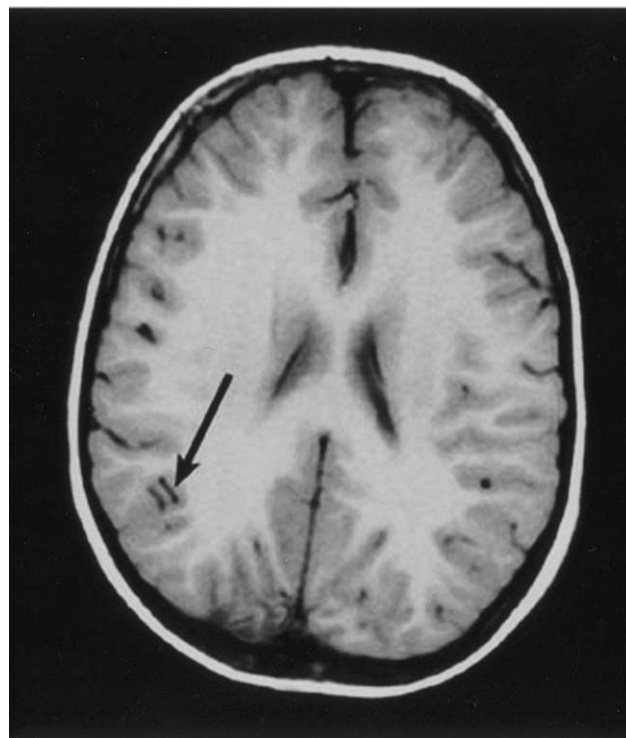
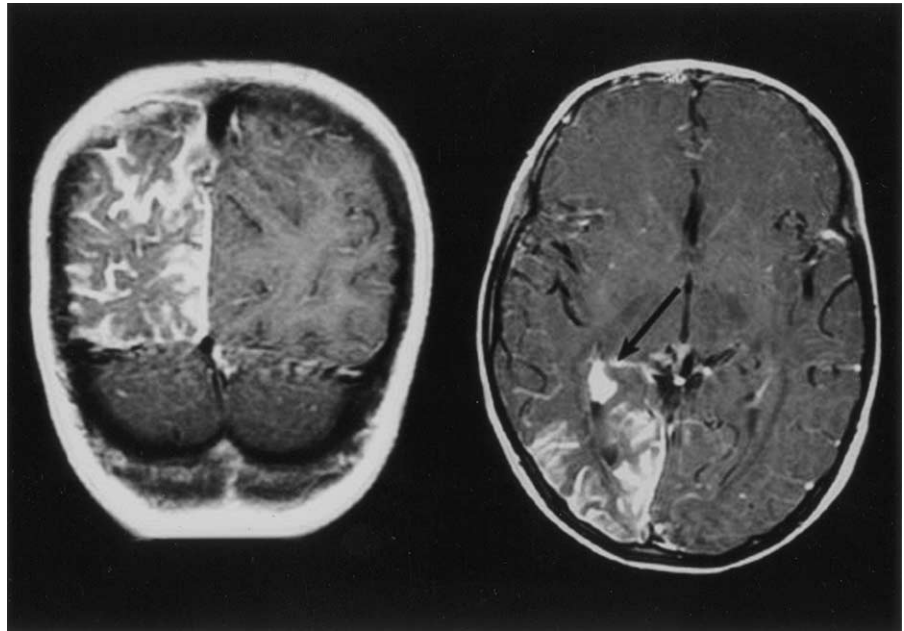


Figure 1. An axial computed tomographic image demonstrates abnormal calcification of the right occipital cortex (arrow) in a so-called gyriform pattern. The right hemisphere brain sulci are prominent, indicating brain atrophy.

Figure 2. Coronal and axial T<sub>1</sub>-weighted (TR = 516 ms, TE = 11 ms) gadolinium-enhanced postcontrast images demonstrate prominent leptomeningeal enhancement over the surface of the right occipital and parietal lobes. In the posterior horn of the right lateral ventricle, the choroid plexus is enlarged (arrow).



chronic ischemia and ischemia-related gliosis. T<sub>2</sub>-weighted magnetic resonance imaging demonstrates both hyperintense and hypointense white matter lesions.

Atrophy ipsilateral to the leptomeningeal angiomatosis is also common in SWS patients (Fig 3). Cerebral atrophy is accompanied by widening of the subarachnoid space and compensatory enlargement of the ipsilateral lateral

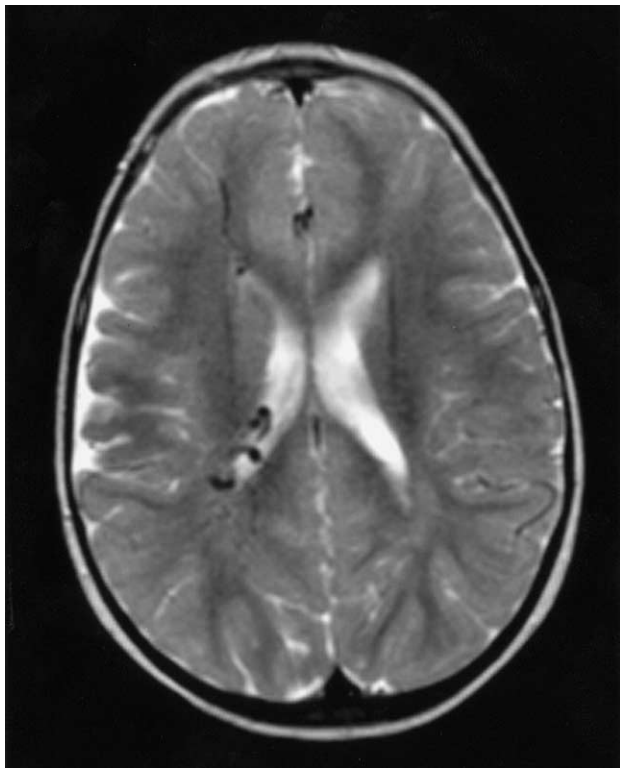


Figure 3. An axial fast spin echo T<sub>2</sub>-weighted (TR = 4000 ms, TE = 103 ms) image reveals lateral right frontal lobe atrophy and large centrally draining veins.

ventricle. Significant atrophy may result in hypertrophy of the cranium on the side of the vascular malformation [20,21,45].

Calcifications underlie the leptomeningeal angiomatosis and may manifest as a diffuse increased density suggestive of microcalcifications, or as gyriform calcification. Gadolinium-enhanced T<sub>1</sub>-weighted spin-echo magnetic resonance imaging is best for evaluation of the vascular malformation.

Advances in single-photon emission computed tomography demonstrate areas of hypoperfusion in patients with SWS [3,5,22]. Single-photon emission computed tomography measuring cerebral blood flow demonstrates hypoperfusion underlying the leptomeningeal angiomatosis and may detect latent angiomas not visible with other imaging studies. Single-photon emission computed tomography identifies areas of hypoperfusion before calcifications can be observed. Magnetic resonance imaging and single-photon emission computed tomography, used in conjunction, can reveal more remote areas of involvement [22].

Disturbances in glucose metabolism also can be visualized with 2-deoxy-2 <sup>18</sup>fluoro-D-glucose positron emission tomography or single-photon emission computed tomography. Perfusion abnormalities observed with single-photon emission computed tomography and positron emission tomography may precede clinical symptoms and the atrophy or calcification observed on computed tomography and magnetic resonance imaging [46]. Functional neuroimaging can reveal early abnormalities theoretically predictive of prognosis. Longitudinal studies are required to establish whether such imaging modalities can identify patients at increased risk for neurologic deterioration. Researchers are using positron emission tomography and single-photon emission computed tomography to evaluate the need for early resective surgery, which may alleviate

seizures and facilitate normal cognitive development in select patients.

Positron emission tomography scans have demonstrated that SWS patients with larger areas of mildly hypometabolic cortex paradoxically have more frequent seizures than those with severe asymmetric cortical hypometabolism [46]. This finding suggests that the extent and severity of the structural lesion does not directly indicate the severity of epilepsy, whereas the functional abnormalities beyond the structural lesion shown on magnetic resonance imaging are more prognostic. An association between higher cognitive function, shorter epilepsy duration, and a larger area of severely asymmetrical cortical metabolism has been demonstrated in a study performed by Lee et al [46]. Severely hypometabolic lesions that develop at younger ages seem to facilitate effective reorganizational processes, providing the patient with more normal cognitive abilities.

## Treatment

Patients with SWS require consistent and thorough monitoring for development of glaucoma, seizures, headache, and strokelike episodes. Medical and surgical management of glaucoma associated with SWS continues to be challenging (Table 1). Lifelong medical treatment coupled with frequent surgeries is standard. The goal is to control intraocular pressure to prevent optic nerve damage. Medications should be administered to decrease the production of aqueous fluid or promote the outflow of aqueous fluid. Beta-antagonist eye drops, adrenergic eye drops, and carbonic anhydrase inhibitors are the treatments of choice. Trabeculectomy and goniotomy are typical surgical options.

Laser therapy for facial cutaneous vascular malformations should begin soon after diagnosis for the best results. A vascular-specific pulsed dye laser can improve the appearance of the facial cutaneous vascular malformation, typically within 10 treatments. The location of the facial cutaneous vascular malformation predicts the response to laser therapy. Central forehead lesions respond best, whereas central facial lesions do not respond as well [47–51]. Many patients benefit psychologically from removal of the facial cutaneous vascular malformation [52]. Without laser therapy, the lesion grows and typically darkens, developing vascular ectasias that promote nodularity and superficial blebbing. This development may lead to overgrowth of the soft tissue and bone beneath the lesion. Hypertrophy and nodularity within the lesion develop in 65% of patients by the fifth decade [52].

Prevention of recurrent seizures may diminish the effects of hypometabolism and hypoxia; therefore, the goal is complete seizure control. Management principles for recurrent seizures associated with other conditions also apply to seizure prophylaxis in SWS. Children are initially placed on carbamazepine, with phenobarbital and phenytoin as second-line therapies (Table 1). If control is not

achieved, valproate or topiramate may be added to carbamazepine, with the ultimate goal of monotherapy seizure control with valproate or topiramate. Children who receive no relief from frequent, debilitating seizures are candidates for epilepsy surgery. Although there is no conclusive evidence that surgical management in infancy provides a better prognosis, delay of surgical treatment may result in further cognitive deterioration [53]. A retrospective clinicopathologic review of infants requiring epilepsy surgery indicated that in 7 of 8 patients, epilepsy was absent or significantly diminished postoperatively, supporting the benefits of early surgery [54]. Hemispherectomy is definitive surgery for recurrent seizures, yet substantial operative risk remains [39]. Most candidates for epilepsy surgery have significant developmental delay. Few data are available, but anecdotal experience suggests that surgical relief of catastrophic epilepsy may result in resumption of developmental progression. For each patient, the timing of surgery must be carefully considered after fully assessing the procedure's relative risks and benefits [55]. The retrospective study by Kossoff et al. concluded that hemispherectomy did not affect the outcome [56].

Transient focal deficits presenting with hemiparesis or visual field defects not directly linked to seizures should be monitored diligently. Prophylactic aspirin is recommended for the prevention of these episodes. Aspirin may delay the neurologic deterioration that often accompanies SWS. Anecdotal data suggest that aspirin therapy is safe and effective. However, no randomized, controlled clinical trials have tested its use in children with SWS. We recommend the antiplatelet dose of 3 to 5 mg/kg/day for children with recurrent strokelike episodes. Children prescribed aspirin should receive varicella immunization and yearly influenza immunizations because of the association with these infections and with Reye's syndrome [39].

Headaches can be debilitating in patients with SWS. The frequency and severity of headaches is higher in SWS than in the general population. Many children report a temporal relationship between their headaches and seizure activity. The leptomenigeal angioma may predispose children to neuronal hyperexcitability, which may account for the migraines. Children with SWS often respond to standard abortive and preventive migraine management to cope with headaches (Table 1). To the best of our knowledge, there are no reported serious adverse events from the use of triptans in SWS.

## Prognosis

The prognosis in SWS varies widely. Although patients with widespread hemispherical disease or bihemispheric disease are at greatest risk for neurologic complications, many function virtually normally. Clearly, a subgroup of patients with limited central nervous system involvement as defined by neuroimaging studies has a particularly malignant clinical course, with intractable epilepsy, head-

ache, strokelike episodes, and cognitive deterioration. A longitudinal study must be conducted to identify risk factors for neurologic deterioration.

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