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## Consensus Statement for the Management and Treatment of Sturge-Weber Syndrome: Neurology, Neuroimaging, and Ophthalmology Recommendations

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### Abstract

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**Background:** Sturge-Weber Syndrome (SWS) is a sporadic, neurocutaneous syndrome involving the skin, brain, and eyes. Given the variability of clinical outcome and the lack of prospective studies, consensus recommendations for management and treatment have not reached a conclusion.

**Objective:** This consensus statement aims to consolidate the current literature with expert opinion to make recommendations that will guide treatment and referral, particularly regarding the neurological and ophthalmological features of the disease as well as neuroimaging workup.

**Methods:** Thirteen national peer-recognized experts in neurology, radiology, and ophthalmology with experience treating SWS patients were assembled. Key topics and questions were formulated for each group and included: (a) risk stratification; (b) indications for referral; and (c) optimum treatment strategies. An extensive PubMed search was performed of English language articles published in 2008–2018, as well as recent studies identified by the expert panel. Clinical practice guidelines were recommended.

**Conclusions:** Any child with a high-risk facial port-wine birthmark (PWB) should be referred to a pediatric neurologist and pediatric ophthalmologist for a baseline neurological evaluation and ophthalmic evaluation, respectively, with periodic follow-up. In newborns and infants with a high-risk PWB and no history of seizures or neurological symptoms, routine screening for brain involvement is not recommended, but brain imaging can be performed in select cases. Routine follow-up neuroimaging is not recommended in children with SWS and stable neurocognitive symptoms. The treatment of ophthalmologic complications, such as glaucoma, differs based on the age and clinical presentation of the patient. These recommendations will help facilitate coordinated care for patients with SWS and may improve patient outcomes.

## Keywords

Consensus statement; Sturge-Weber Syndrome; Port-wine birthmark; seizures; glaucoma

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## Introduction

There is a critical need for a consensus statement regarding the management of Sturge-Weber Syndrome (SWS). In a prior manuscript, we provided a discussion of dermatologic management and treatment<sup>1</sup>. In the current manuscript, we aim to provide clinical practice guidelines to guide care of patients with SWS, while focusing on the neurological and ophthalmological features of the disease. The goals of this consensus are to 1) review the literature to provide an approach to risk stratification and evaluation of SWS patients; 2) offer guidance on what neurologic, neuroimaging, and ophthalmologic workup should consist of for patients with suspected or newly diagnosed SWS and indications for referral to an ophthalmologist or neurologist; and 3) assess the current treatment options for brain and eye manifestations with considerations for age and severity of disease.

## Methods

Eight national experts in neurology and radiology, and five national experts in ophthalmology, were consulted to form a consensus statement on the management and treatment of SWS, as part of a larger consensus statement<sup>1</sup>. Three key topics were

established: (1) evaluation of port-wine birthmark (PWB) and risk stratification; (2) consultation and referral; and (3) optimum treatment strategies. Questions were formulated for each key topic and an extensive literature review was performed using PubMed for English-language papers published between 2008 and 2018. Articles before 2008 or after 2018 were added by the expert panel based on importance. Search terms included “Sturge-Weber syndrome” and terms associated with each key topic: “clinical presentation”, “pathogenesis”, “risk prediction”, “port-wine birthmark or port-wine stain”, “diagnostic workup”, “triage”, “management”, “treatment”, “laser therapy”, “light-based therapy or treatment”, “photodynamic therapy”, “infantile hemangioma”, and “nevus simplex.” 112 total manuscripts were identified for consideration, of which 84 of those were relevant to neurology, radiology, and ophthalmology, and 47 manuscripts were directly referenced by the expert panel. Publications associated with each key topic were identified and distributed to each group, who then developed clinical practice guidelines (Tables 1–3). These guidelines were consolidated into key points that were then presented to all groups for electronic discussion and modification before achieving final consensus.

## Neuroradiology

### Neurological Manifestations and Risk Stratification

Key Point 1: Any child with a high-risk facial port wine birthmark should be referred to a pediatric neurologist for a baseline neurological evaluation and have periodic follow-up.

SWS is associated with neurological abnormalities, including seizures, stroke-like episodes, headaches, and developmental delays<sup>2,3</sup>. Seizures occur in approximately 75–80% of SWS patients.<sup>3</sup> A recent study of 277 SWS patients with brain involvement demonstrated epilepsy in 81.6% of those patients<sup>4</sup>. The age of onset is variable, but is usually within the first year of life; however, cases of seizure onset in adulthood have also been reported<sup>5,6</sup>. Recognition and management of seizures is essential, because early onset, frequent seizures can adversely affect cognitive and neurodevelopmental outcomes<sup>7,8</sup>.

Risk of SWS with brain involvement is greater in patients with hemifacial, forehead, and median locations of their PWB, since those locations involve skin derived from the frontonasal placode, which shares common progenitor cells with the brain<sup>9</sup>. Any child with a high-risk facial PWB should be referred to a pediatric neurologist for a baseline evaluation and have periodic follow-up. An electroencephalography (EEG) to assess abnormal brain activity and identify patients at risk for future neurologic events, although not diagnostic, may be useful in patients with neurologically asymptomatic PWBs<sup>10</sup>. The pediatric neurologist may then make recommendations regarding imaging and management of neurological complications. In 2018, De la Torre et al. provided a discussion of management and treatment options for neurological complications, along with specific clinical guidelines for neurology<sup>11</sup>. These recommendations remain valid. This review will focus on additional clinical guidelines, specifically related to neuroimaging.

## Neuroimaging

Key Point 1A: In newborns and infants with a high-risk PWB and no history of seizures or neurological symptoms, routine screening for brain involvement is not recommended.

Brain involvement may be detected by imaging in infants with a high-risk PWB even before the onset of neurological symptoms. However, negative neuroimaging in a normally developing asymptomatic infant with a facial PWB does not exclude SWS brain involvement. Such false negative findings have been reported in 3–23% of the cases in retrospective studies<sup>12–14</sup>. Negative MRI may provide false reassurance. False positive MRI in this early disease stage has been reported anecdotally, although its likelihood is low. Even if the MRI is true positive, neurologically asymptomatic children are unlikely to undergo immediate therapeutic intervention (exceptions in Key Point 1B), although it can increase vigilance for subclinical seizures. Nevertheless, future research may elucidate a better understanding regarding the utility of screening in this age group.

Key Point 1B: In newborns and infants with a high-risk PWB and no history of seizures or neurological symptoms, brain imaging can be considered for more subtle symptoms and extensive PWB. Indirect signs of SWS brain involvement and susceptibility-weighted imaging can optimize the yield of MRI in this group.

A screening brain MRI may be considered in select children with suspected SWS, for example, when presymptomatic treatment is contemplated. A retrospective study suggested the potential benefit of presymptomatic antiepileptic treatment combined with aspirin<sup>15</sup>, but no prospective studies have been completed. Some children with a particularly high risk for seizures may benefit from presymptomatic treatment, for example when bilateral SWS is suspected (e.g., extensive bilateral PWBs). Treatment should be preceded by MRI to establish presence and extent of early brain involvement. In a retrospective review, MRI features suggestive of SWS were detected in 43%–73% of individuals with high-risk PWB<sup>14</sup>. In a review of 32 children with high-risk PWB (hemifacial, median, and forehead), screening MRI had a sensitivity of 25%, specificity of 100%, positive predictive value of 100%, and negative predictive value of 77% for the detection of SWS brain involvement<sup>14</sup>.

Concerns regarding MRI in infants include the potential long-term cognitive effects of anesthesia and repeated gadolinium contrast administration with systemic deposition in tissues, including brain and bone marrow. The optimal MRI approach for presymptomatic screening has not been established; however, to minimize the risks of clinical MRI in presymptomatic children, a fast, non-sedated, non-contrast screening MRI can be considered, preferably including axial T1, T2 [or fluid-attenuated inversion recovery (FLAIR)], susceptibility-weighted imaging (SWI), with or without diffusion-weighted imaging (DWI) on a 3T scanner<sup>11</sup>. These images may detect early venous vascular abnormalities or accelerated myelination and atrophy. SWI can be particularly sensitive to detect early deep medullary veins during the first few months of life, even before contrast enhancement is detectable [Figure 1]<sup>12,16</sup>. Current research aims at development and validation of ultra-fast, non-contrast MRI acquisitions to provide accurate and safe presymptomatic imaging without sedation risks.

**Key Point 2A:** In children with suspected SWS, the first post-symptomatic imaging should be an optimized pre- and post-contrast MRI sequence, focusing on the detection of both vascular and parenchymal abnormalities associated with SWS.

The routine use of computed tomography (CT) scans in children with new-onset SWS-related seizures or new neurological symptoms is not recommended due to low yield and the potential risks of radiation exposure. If a CT scan is done, an MRI with and without contrast administration should follow to establish the diagnosis of SWS and provide detailed insight on brain vascular and parenchymal abnormalities. A recent paper authored by a panel of experts recommends the use of pre- and post-contrast MRI, acquired under sedation (if needed) and including high-resolution volumetric sequences to optimize detection of subtle parenchymal changes and potential cortical malformations<sup>11</sup>. The post-contrast MRI acquisition should include T1-weighted and FLAIR images<sup>17</sup>, and the protocol should include SWI<sup>18</sup>. SWI can depict small caliber venous abnormalities, such as deep medullary veins and is highly sensitive to visualize calcifications that are often not present or subtle in early disease but can be progressive<sup>18–20</sup>. DWI can be useful to detect ischemic parenchymal changes, although these could be transient if MRI is done immediately after acutely developed clinical symptoms (see Key point 3A). The use of other, advanced MRI sequences is optional, their clinical yield remains to be established, and their potential benefit has to be weighed against the potential risks of prolonged sedation.

**Key Point 2B:** Routine follow-up neuroimaging is not recommended in children with established SWS and stable neurocognitive symptoms. In case of progressive neurocognitive symptoms during follow-up, multisequence brain MRI comparable to previous imaging should be performed for an accurate comparison.

If the child's cognitive development is steady, seizures are controlled, and there are no progressive neurological symptoms, routine follow-up neuroimaging is not necessary. On the other hand, delayed or declining cognitive function, worsening seizures, or development of new neurological symptoms (hemiparesis, visual field defect, etc.) can prompt a follow-up MRI to evaluate if there is progression in structural brain abnormalities. Follow-up MRI should include optimized multisequence acquisitions so interval changes can be assessed accurately. While the extent of leptomeningeal enhancement is usually stable, underlying brain parenchymal abnormalities such as atrophy and calcifications often show interval progression, especially during the first few post-symptomatic years<sup>20</sup>. Progressive atrophic changes have also been reported in adults with SWS, sometimes associated with late-onset new clinical symptoms, such as migraine attacks<sup>21</sup>. Recent studies demonstrated that post-symptomatic expansion of deep medullary veins can occasionally be detected during early disease by SWI<sup>22,23</sup>. Although data are preliminary, it is likely that expansion of these deep veins may be beneficial and represent an effective compensatory mechanism to offset the detrimental effects of impaired venous drainage.

**Key Point 2C:** Neuroimaging should be obtained in adults with PWB with or without glaucoma who have not had prior imaging; however, follow-up neuroimaging is not recommended in adults with established SWS and stable neurocognitive symptoms. Pre- and post-contrast brain MRI is recommended in case of new-onset or progressive symptoms.

Reports focusing on neuroimaging in adults with SWS are scarce. A recent review identified 31 reported cases where SWS was diagnosed in adulthood<sup>24</sup>, but the incidence of such a late manifestation is not known. Imaging in these cases is typically prompted by new-onset symptoms and, in rare cases, a first-ever neurological manifestation of SWS, such as seizure(s). SWS may be missed in cases with no facial PWB, such as the case of a 55-year old patient with a history of episodes of transient weakness in the right extremities<sup>25</sup>. Post-contrast FLAIR demonstrated fronto-parietal leptomeningeal capillary malformations, and SWI showed enlarged deep medullary veins and calcifications. Similar cases with adult-onset seizures along with the new diagnosis of SWS by neuroimaging have been reported<sup>26,27</sup>. Despite the scarce data, most of the SWS pathology reported in pediatric populations is present in adult patients. Routine imaging, however, is not justified in adults with previously documented SWS brain involvement, unless new symptoms emerge. In adult patients with new-onset symptoms or progression of previous symptoms, pre- and post-contrast MRI including SWI and DWI should be the choice of imaging. At this point there are no data to support that the optimal MRI sequences for adults should be different from those recommended in pediatric SWS patients.

Key Point 3A: SWS patients rarely hemorrhage and there is little evidence for acute ischemia. The available data do not show bleeding or clear strokes in SWS patients who present with acute neurological symptoms; however, prolonged new-onset or acutely/subacutely deteriorating and non-resolving neurological ‘stroke-like’ episodes justify repeat neuroimaging.

In current clinical practice, children and adults with known SWS and new or progressive acute symptoms often undergo emergent imaging during an emergency room visit. However, the clinical value of this imaging in such a setting is questionable. One issue is that after prolonged or repeated seizures, status epilepticus or stroke-like episodes, MRI may show transient abnormalities<sup>28-31</sup> that could prompt follow-up MRI that shows normalization. In a recent retrospective study of 35 SWS patients, who presented to an emergency department with acute neurological symptoms, 89 urgent neuroimaging studies were reviewed, and none showed acute hemorrhagic or ischemic strokes<sup>32</sup>. The authors concluded that urgent imaging after breakthrough seizures does not result in a significant change of clinical management. Similarly, cerebral angiograms are often performed on SWS patients, have little value, and significant risk. A sudden, severe deterioration of the neurologic status, including prolonged loss of consciousness, has been reported in a few SWS cases, where acute imaging revealed acute thalamic hemorrhage<sup>33,34</sup>. It is therefore important to note that as SWS patients age, particularly with other health co-morbidities, they should be fully evaluated like any other patient for strokes and hemorrhages due to other causes and treated appropriately. The use of thrombolytics has not been investigated in SWS patients and needs to be carefully evaluated and could potentially be used if there is a strong clinical indication.

Key Point 4A: Presurgical evaluation in SWS can benefit from the use of advanced structural and functional imaging modalities. These studies and the subsequent surgery should be performed in specialized pediatric epilepsy centers that are experienced in processing and interpretation of advanced imaging and also in surgical techniques used in SWS (such as hemispherectomy).

Epilepsy surgery can be considered in patients (usually children) with SWS and drug-resistant seizures. The surgery is mostly reserved for those with unilateral involvement, with rare exceptions. The most common surgery type is hemispherectomy (structural or functional), while a posterior resection (preserving the frontal lobe including the motor cortex) is reserved for those with an intact frontal lobe. While multimodal MRI is essential to delineate the type, extent, and severity of vascular and brain parenchymal structural abnormalities in SWS, patients with drug-resistant seizures can benefit from additional, functional neuroimaging techniques during presurgical evaluation. Functional MRI of motor and language functions can be performed in non-sedated patients (such as older children) <sup>35</sup>. Motor mapping by fMRI can be useful in children where a posterior resection, preserving the primary motor cortex, is being considered. Language mapping by fMRI can evaluate the risk for post-surgical language deficit in those with left hemispheric surgery. The structural integrity of critical pathways involved in motor, language, and visual functions can be evaluated by diffusion tensor imaging tractography that can be performed in sedated subjects <sup>36,37</sup>. The data from such studies can predict the expected functional deficit from resecting brain tissue encompassing such pathways.

## Ophthalmology

### Ocular Manifestations and Risk Stratification

Key Point 1: Any child with a high-risk facial PWB should be referred to a pediatric ophthalmologist for a baseline eye evaluation and have periodic follow-up.

Approximately 50% of SWS patients show pathologic ocular changes, usually ipsilateral to the PWB, involving the eyelid, conjunctiva, episclera, anterior chamber, cornea, choroid, and retina <sup>38</sup>. Patients may present with anterior segment alterations such as cataract or glaucoma, and often with posterior segment alterations such as choroidal hemangiomas <sup>38</sup>. In darkly pigmented children in whom the PWB is difficult to discern, a conjunctival ‘blush’ may be the only sign of ocular involvement visible to the non-ophthalmologist. Patients with PWB overlying the eyelids or eye have a higher incidence of glaucoma during infancy and childhood <sup>39</sup>. A PWB in a high-risk distribution, defined as on the forehead from the midline to an imaginary line between the outer canthus of the eye and the top of the ear including the upper eyelids, was highly associated with the development of glaucoma in a cohort of 192 children with PWB <sup>40</sup>. Rarely, glaucoma may be bilateral, even in the presence of a unilateral PWB.

Because of the risk of preventable visual loss, every child diagnosed with SWS or with periocular vascular anomalies involving the eyelids should be referred to a pediatric ophthalmologist for examination. Should a pediatric neurologist be the first to diagnose a patient with SWS, this is an indication for referral to ophthalmology. The two most common causes of vision loss are glaucoma and amblyopia - both can be treated if detected early. Some children can have signs that prompt an ophthalmology referral such as enlarged corneal diameter in one eye, excessive tearing or rubbing, excessive light sensitivity, and/or cloudy appearance of the cornea, but a clinician should not wait for a child to have these signs to refer to a pediatric ophthalmologist. A baseline exam within the first few months of life will help to diagnose ocular involvement in SWS and will determine the timing of

treatment or follow-up. The baseline eye exam should include visual acuity measurement, intraocular pressure (IOP) check in the clinic and a full dilated eye exam. If the IOP cannot be measured in the office, and there is a high suspicion for glaucoma, arrangements may need to be made for sedated examination.

The iCare™ rebound tonometer<sup>†</sup>, introduced a decade ago, can measure IOP in many children without the need of an exam under anesthesia or even topical (eyedrop) anesthesia. A dilated eye exam can assess whether the child has a choroidal hemangioma, with its “tomato ketchup” fundus appearance, and assess the optic nerve for glaucomatous optic neuropathy [Figure 2]. The pediatric ophthalmologist may then make further recommendations regarding the treatment of possible amblyopia, glaucoma or retinal disease. The risk of not treating amblyopia, glaucoma or retinal disease is permanent visual impairment in the affected eye. In the case of glaucoma in a young child, this may also cause severe ocular pain and discomfort. Discussion of specific risks or benefits should occur between the treating physician and the appropriate subspecialized ophthalmologist as the spectrum of ocular disease can vary greatly and is tailored to the individual patient.

### Determination of the Optimum Treatment

Key Point 2: The treatment of glaucoma in patients with SWS differs based on the age and clinical presentation of the patient.

In SWS, the ocular issues that arise are thought to be due to the increased venous pressure from episcleral and choroidal hemangiomas that “back up” the normal drainage pathways for the eye<sup>41</sup>. The increased downstream venous pressure leads to increased intraocular pressure (IOP), and can also cause fluid buildup under the retina resulting in serous retinal detachments<sup>41,42</sup>. Reyes-Capo et al report that the glaucoma associated with SWS or PWB’s occurs in 30–50% of affected cases in children younger than 36 months, however other references cite a lifetime risk of glaucoma upwards of 70%<sup>41,42</sup>. Glaucoma, caused by high pressure or wide IOP fluctuations, damages the structures of the eye resulting in an irreversible optic neuropathy and subsequent progressive visual impairment. Thus, early detection and treatment is vital. The glaucoma associated with a PWB has a bimodal presentation, with some patients presenting in infancy (0–3 years) and others that present later in life<sup>41</sup>. Glaucoma may present early during infancy and childhood in about 60% of patients or later during childhood and adolescence in about 40% of cases, with age of onset reported up to 41 years<sup>5,38</sup>.

The glaucoma that presents in infancy is likely related to both the increased episcleral venous pressure as well as an anatomic developmental abnormality of the infant trabecular meshwork system<sup>41,42</sup>. In a child younger than 3 years old, glaucoma may present acutely with enlargement of the cornea and globe, clouding of the cornea due to edema, pain and light sensitivity. In some patients the onset can be more insidious with ipsilateral ocular enlargement the only clue that glaucoma is present. In young children, the disease is almost always treated surgically<sup>41,42</sup>, followed by medications and/or laser.

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<sup>†</sup> <https://www.icare-usa.com/>



The glaucoma presenting in older individuals likely occurs as a result of the increased episcleral venous pressure with otherwise normal anatomic development<sup>41</sup>. In these patients, the glaucoma may first be managed with medications, either topical or systemic. Surgery or laser treatment is often necessary if medications are not sufficient to control the disease. Surgical options need to be carefully considered as these eyes are more at risk for intraoperative and postoperative complications because of the increased pressure gradient across the choroidal vasculature, which can result in suprachoroidal hemorrhage or recalcitrant serous retinal detachments<sup>41,42</sup>.

### **Psychosocial outcomes and quality of life in patients with SWS**

Quality of life (QoL) is a construct that encompasses many components of well-being, including physical, functional, emotional, social, and family<sup>43</sup>. Living with diseases like SWS is more complex than clinical findings alone. The QoL of individuals with facial PWB is affected by the skin condition and the procedures used to treat the PWB. In addition, caregivers' QoL may be significantly impacted by the facial PWB. As research on QoL issues for SWS is limited, we provide an overview of the impact of any type of visible skin disease on QoL [Table 4]<sup>44,45</sup>. These are likely representative of many aspects of the disease experience for individuals with PWB.

In addition to the presence of a facial PWB, epilepsy has a significant negative impact on QoL, and these patients may experience stigmatization and social isolation<sup>46</sup>. In the United States, 13.1 percent of children with epilepsy have depression, and 23.3 percent have anxiety<sup>47</sup>. Intellectual disability is also common in SWS, noted in 60% of patients<sup>11</sup>. In addition to decreased QoL of the patient with intellectual handicaps, family members also experience more stress and face many challenges to support the patient's needs and transition to adulthood<sup>48</sup>. Therefore, physicians caring for these patients should inquire about QoL<sup>49</sup>. It is important to note that addressing QoL and psychosocial issues depends on a strong patient- and family-physician relationship. Maintaining a positive attitude and providing reasonable hope through all medical care can often lessen the emotional toll of body image issues due to visible skin disease<sup>50</sup>. For individuals suffering from psychosocial issues, social workers, psychologists, psychiatrists, and patient support/advocacy groups are often able to help patients and families who need community and support to reach their full potential<sup>51</sup>.

### **Conclusion**

This consensus statement reflects the current state of knowledge on the non-cutaneous manifestations of SWS and is meant to guide clinical decision-making. This document highlights the importance of consultation with other members of the SWS patients' care team, including neurologists, ophthalmologists, and dermatologists, to manage SWS collaboratively and ensure proper assessment and treatment of patients to improve patient outcomes. A natural history study currently underway through the Brain Vascular Malformation Consortium, part of the Rare Diseases Clinical Research Network (RDCRN) of NIH, will obtain parallel longitudinal clinical and imaging data that will add to our knowledge base and improve clinical decision-making for SWS in the future.

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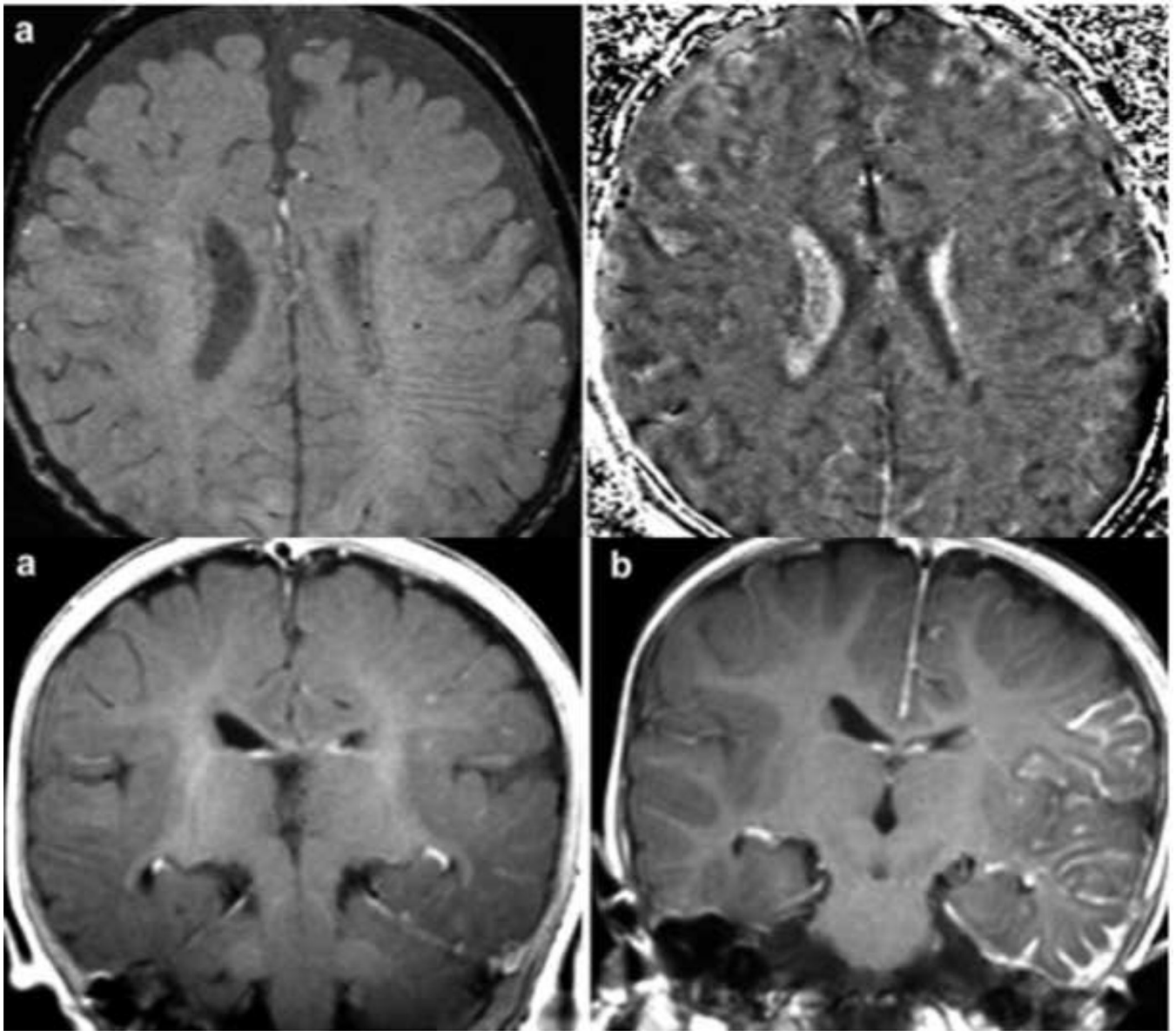
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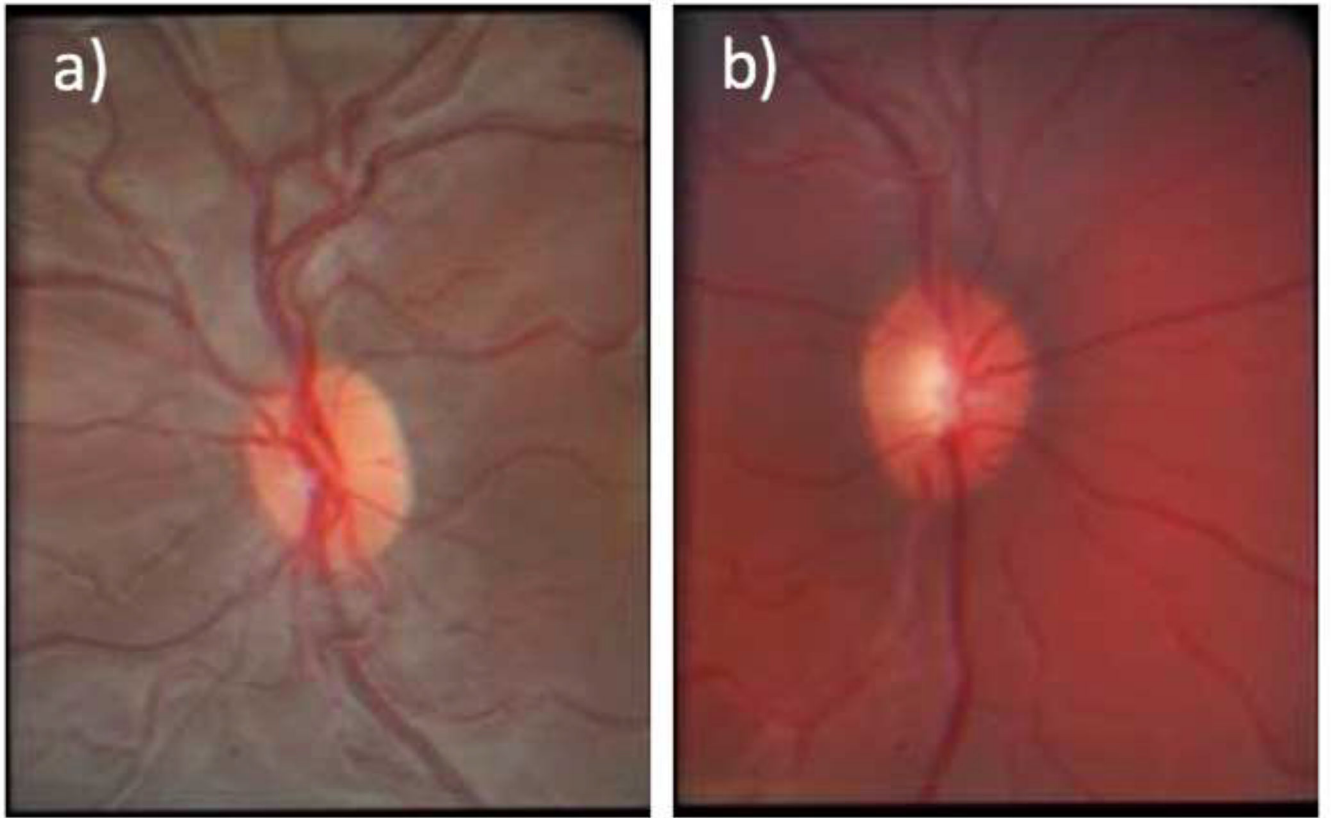
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**Figure 1.** Detection of enlarged deep medullary veins by SWI in the left hemisphere of a 4-month old girl before the first seizure. Post-contrast T1-weighted MRI showed leptomeningeal enhancement only 8 months later, after the first clinical seizure (adopted from Mentzel et al., 2005).



**Figure 2.**

a) Normal fundus photo of the left eye; b) Fundus photo of the right eye demonstrating choroidal hemangioma and increased cup to disc ratio, consistent with glaucoma.

**Table 1.**

Key Points for Neurological Management and Treatment in SWS

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1. Any child with a high-risk facial port wine birthmark should be referred to a pediatric neurologist for a baseline neurological evaluation and have periodical follow-up.

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**Table 2.****Key Points for Neuroradiological Management and Treatment in SWS**

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1A. In newborns and infants with a high-risk PWB and no history of seizures or neurological symptoms, routine screening for brain involvement is not recommended.

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1B. In newborns and infants with a high-risk PWB and no history of seizures or neurological symptoms, brain imaging can be performed in select cases, e.g., in those where presymptomatic treatment is being considered. Indirect signs of SWS brain involvement and susceptibility-weighted imaging can optimize the yield of MRI in this group.

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2A. In children with suspected SWS, the first post-symptomatic imaging should be an optimized pre- and post-contrast MRI sequence, focusing on the detection of both vascular and parenchymal abnormalities associated with SWS.

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2B. Routine follow-up neuroimaging is not recommended in children with established SWS and stable neurocognitive symptoms. In case of progressive neuro-cognitive symptoms during follow-up, multisequence brain MRI comparable to previous imaging should be performed for an accurate comparison.

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2C. Neuroimaging should be obtained in adults with PWB with or without glaucoma who have not had prior imaging; however, follow-up neuroimaging is not recommended in adults with established SWS and stable neurocognitive symptoms. Pre- and post-contrast brain MRI is recommended in case of new-onset or progressive symptoms.

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3A. SWS patients rarely hemorrhage and there is little evidence for acute ischemia. The available data do not show bleeding or clear strokes in SWS patients who present with acute neurological symptoms; however, prolonged new-onset or acutely/subacutely deteriorating and non-resolving neurological 'stroke-like' episodes justify repeat neuroimaging.

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4A. Presurgical evaluation in SWS can benefit from the use of advanced structural and functional imaging modalities. These studies and the subsequent surgery should be performed in specialized pediatric epilepsy centers that are experienced in processing and interpretation of advanced imaging and also in surgical techniques used in SWS (such as hemispherectomy).

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**Table 3.**

**Key Points for Ophthalmological Management and Treatment in SWS**

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1. Any child with a high-risk facial port wine birthmark should be referred to a pediatric ophthalmologist for a baseline eye evaluation and have periodical follow-up.
  2. The treatment of glaucoma in patients with SWS differs based on the age and clinical presentation of the patient.
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**Table 4.**

## Psychosocial Outcomes and Quality of Life in Patients with SWS

<p><u>Impact of Visible Skin Disease on Patients</u> (Ablett and Thompson, 2016)</p>	<ul style="list-style-type: none"> <li>■ Feeling a sense of being different from others due to their appearance leading to isolation, especially if they are teased about their birthmark</li> <li>■ Avoiding intimate behaviors due to embarrassment about their appearance</li> <li>■ Need to educate others due to a lack of sympathy or underestimation of the impact of the skin condition on the individual's life by medical professionals</li> <li>■ Feeling a sense of powerlessness and separation as many individuals have been viewed as having "special needs" due to their skin condition</li> </ul>
<p><u>Impact of Visible Skin Disease on Parents and Caregivers</u> (Ablett and Thompson, 2016)</p>	<ul style="list-style-type: none"> <li>■ Treatments for skin disease are often time consuming, which takes away from spending time with spouses, other family members, and work</li> <li>■ Feeling psychological strain when strangers make comments about their child's skin condition and during painful treatments for the skin condition</li> <li>■ Feeling blamed for not preventing sequela of the skin disease</li> </ul>
<p><u>Negative Impact of Treatments for Visible Skin Disease on Patients</u> (Ablett and Thompson, 2016 and Bemmels et al., 2013)</p>	<ul style="list-style-type: none"> <li>■ Addiction to attaining a perfect result</li> <li>■ Missing school or work for treatments</li> <li>■ Adjusting to an evolving appearance</li> <li>■ Wondering when treatments will end</li> <li>■ Experiencing stigma related to undergoing surgery due to missing school or work and their changing appearance</li> <li>■ Strains on the parent (or caregiver) and child relationship as some resent their parent's suggestion that there was something about their appearance that needs to be changed</li> </ul>
<p><u>Positive Impact of Treatments for Visible Skin Disease on Patients</u> (Ablett and Thompson, 2016 and Bemmels et al., 2013)</p>	<ul style="list-style-type: none"> <li>■ Improved self-esteem and reduced stigmatization when the appearance is more "normal" after treatments</li> <li>■ Less staring, questioning, and teasing</li> </ul>