orphanesthesia

Anesthesia recommendations for

Sturge-Weber Syndrome

Disease name: Sturge-Weber Syndrome

ICD 10: Q85.8

ORPHAcode: 3205

Synonyms: Angiomatosis Aculoorbital-Thalamic Syndrome, Dmitri Disease, Encephalofacial Angiomatosis, Encephalofacial Hemangiomatosis, Encephalofacial Hemangiomatosis Syndrome, Encephalotrigeminal angiomatosis, Fourth phakomatosis, Leptomeningeal angiomatosis, Meningeal capillary angiomatosis, Meningo-oculo-facial Angiomatosis, Sturge-Kalischer-Weber Syndrome, Sturge-Weber Syndrome, Sturge-Weber-Krabbe Syndrome, SWS, SWS type 1 – facial and leptomeningeal angiomas, SWS type 2 – facial angioma alone – no CNS involvement, SWS type 3 – Isolated leptomeningeal angiomas.

Disease summary: The syndrome was first described by Rudolf Schirmer in 1860; in 1879, William Allen Sturge reported a child with seizures, facial angioma and glaucoma, and in 1922 Frederick Parkes Weber described the characteristic cerebral calcifications; it was later named Sturge-Weber syndrome (SWS) in 1935 by Hilding Bergstrand. SWS is one of a rare phakomatosis or neurocutaneous syndromes consisting of abnormal capillary malformations that can involve the face, eyes and leptomeninges of the brain [1-8]. It is most commonly associated with a somatic mosaic mutation in the GNAQ gene, thought to be enriched in endothelial cells leading to abnormal vasculogenesis and blood-brain barrier permeability [9-12].

The facial capillary angiomas (Port Wine Birthmarks) (PWB) are centered along the distribution of the trigeminal nerve and if present in an infant or child presenting with seizures, a diagnosis of SWS should be considered. There is a positive correlation between the size of facial PWB, the degree of SWS brain involvement and severity of neurological disability [12]. It should be noted that SWS may be present in a patient without any PWB and that not all patients with PWB have SWS.

Central neuraxial imaging may reveal characteristic angiomas along with calcification of the leptomeninges ipsilateral to the facial nevus. These may lead to atrophy of the cerebral cortex along with variable neurological and cognitive impairment.

Characteristically the main ocular manifestations include glaucoma, varicosities of the retinal vessels, hemangiomas of the choroid and retinal detachment. Optic neuropathy and buphthalmus secondary to the raised intraocular pressure (IOP) can occur in untreated cases of raised IOP. These may lead to varied visual field defects and even blindness.

Clinical features include seizures, which may be generalized or focal in origin most often occurring contralateral to the facial nevus. Developmental delay and cognitive impairment, along with headache, stroke-like events, hemiparesis and hemicerebral atrophy may be present. These result from ischemic and destructive effects of cerebral angiomas and

associated seizures. Facial angiomas may vary in color from light pink and flat to dark purple and raised. Cardiac lesions which may be associated with SWS include septal defects, valvular anomalies, transposition of the great vessels, aortic coarctation and rarely deep arteriovenous malformations.

The mainstay of treatment centers around seizure control. Seizures may worsen associated cortical hypoperfusion, potentially further impairing neurological function and development. IOP reduction in glaucoma can be achieved with medical treatments including carbonic anhydrase inhibitors or beta-blockers and if unsuccessful, surgery. PWB can be treated with pulsed laser therapy to varying degrees of results. Sirolimus (an mTOR inhibitor) has shown promise in small studies for both cutaneous and extracutaneous features in SWS, however, further research is still necessary [13,14]. Finally, the use of cannabidiol in patients with SWS and refractory epilepsy has been reported but also requires further research [14].

Diagnosis may be incorrect; if uncertainty exists, the diagnosis should be re-evaluated.

Every patient is unique; individual circumstances must always guide clinical care.

Medicine is in progress; new clinical knowledge may not be yet reflected in this recommendation.

Recommendations are not rules or laws; they provide a framework to support clinical decision-making. Although this recommendation has passed a structured review process, it does not meet the formal criteria of a guideline.

Translations may not always reflect the most recent updates of the English version.

Find more information on the disease, its centers of reference and patient organizations on Orphanet: www.orpha.net

A	**AIRWAY / ANESTHETIC TECHNIQUE**	Facial asymmetry, large tongues and lips along with friable airway hemangiomas may complicate mask ventilation and laryngoscopy, with a risk of bleeding if traumatized or if the sympathetic response is not adequately obtunded. Neuraxial anesthesia is not contraindicated; however, spinal angiomas (e.g., in overlap syndromes such as Klippel-Trenaunay) should be considered, and neuraxial imaging may be useful to exclude spinal involvement or elevated ICP.
B	**BLOOD PRODUCTS (COAGULATION)**	Aspirin use is increasingly common in patients with SWS and may affect perioperative hemostasis. Significant blood loss can occur during excision or resection of hemangiomas. Depending on the planned procedure, a type and screen should be performed, with crossmatching for surgeries at higher risk of bleeding. TXA should be considered in procedures with anticipated blood loss >500 mL, if not contraindicated.
C	**CIRCULATION**	Cardiac anomalies in SWS are uncommon but may include septal defects, valvular abnormalities, transposition of the great vessels, aortic coarctation, and arteriovenous malformations. Blood pressure should be maintained within the patient's baseline range, and sympathetic stimulation should be adequately blunted, as hypertensive surges may precipitate rupture of oral or intracerebral hemangiomas.
D	**DRUGS**	Seizure prevention with anticonvulsants is the mainstay of treatment in SWS. These agents may alter hepatic enzyme activity, affecting the metabolism of neuromuscular blocking and anesthetic agents. Conversely, some anesthetics may induce hepatic enzymes and reduce serum anticonvulsant levels perioperatively. Therapeutic drug monitoring should be performed to ensure adequate anticonvulsant levels. Premedication may be beneficial in reducing preoperative stress and attenuating hemodynamic responses that could affect hemangiomas.
E	**EQUIPMENT**	THRIVE (transnasal humidified rapid insufflation ventilatory exchange) and difficult airway equipment should be available. Cell salvage and a rapid infuser should be available for potential major hemorrhage.

Typical surgery and procedures

Neurological: Hemispherectomy, callosotomy, focal resections or surgical ablation of affected cerebral hemisphere for seizure control.
Ophthalmic: Ocular surgery, examination under anesthesia or glaucoma surgery such as trabulectomy, trabulotomy or insertion of drainage devices.
Dermatological and cosmetic surgery: Laser therapy for facial nevus formation, orthognathic surgery, soft tissue reduction, dental procedures, oral surgery for the removal of friable hemangiomas and cesarean sections have all been described [15-24].

Type of anesthesia

Patients with SWS tolerate anesthesia well [25,26]. Initial management should include appropriate investigations for associated anomalies, as the disease may present in varying ways and degrees, from isolated cutaneous manifestation to widespread systemic and airway involvement.

Regional anesthesia: Poor patient compliance and the presence of existing neuro-deficits may preclude the use of central neuraxial blockade. Neuraxial anesthesia is not specifically contraindicated unless other contraindications exist [16]. It is also important to note that there is the theoretical risk of spinal cord angiomas which may be present in possible overlap syndromes such as Klippel-Trenaunay syndrome and if these are suspected, neuroimaging may be appropriate.

General anesthesia: From an airway perspective, facial asymmetry, large tongues and lips as well as airway angiomas may make mask ventilation and laryngoscopy difficult [27]. Friable airway hemangiomas may engorge during stressful times and bleed easily if traumatized potentially obscuring the laryngoscopic view further [17,28]. Elevations in intracranial pressure (ICP) can occur not only because of laryngoscopy, but also during various events throughout the perioperative period, from induction to emergence. These increases in ICP may lead to rupture of angiomas as the abnormal blood vessels involved are often thin-walled and can exhibit impaired autoregulation. For this reason, it is important to maintain perioperative blood pressure within the patient's preoperative normal limits. It should also be noted that if large cerebral AV shunts are present, prolonged inhalational induction time due to the associated increased cardiac output may occur.

Necessary additional preoperative testing (beside standard care)

Patients with SWS should be evaluated for any associated airway, cardiac and CNS anomalies [29].

Airway anomalies and their investigation are described in the next section.

Cardiac anomalies are rare in SWS. Cardiac investigations such as an ECG and ECHO may be required to look for possible cardiomegaly and cardiac failure if clinical suspicion exists. These may occur secondary to the cardiac anomalies associated with SWS which include septal defects, valvular anomalies, transposition of the great vessels and aortic coarctation [27,29]. AV shunting from large arteriovenous angiomas in the skin, subcutaneous tissues, muscle and intracerebral hemangiomas may lead to a high output cardiac failure. An ECG may also help elucidate any cardiac conduction defects secondary to possible elevated levels of

anticonvulsants e.g. phenytoin. GNAQ polymorphisms have been linked with increased myocardial injury in female patients undergoing cardiac bypass [30].

Contrast CT and MRI of the central neuraxis may be used to demonstrate if present, any of the characteristic cerebral features of angiomas. These include enlarged choroid plexi, cortical atrophy underlying the angioma, calcification and abnormal draining of medullary and subependymal veins. Neuraxial imaging may also be useful to demonstrate any associated raised ICP or potential spinal leptomeningeal angiomas. While spinal leptomeningeal angiomas have not been reported in SWS to date, there is always the potential for overlap syndromes for example with Klippel-Trenaunay Syndrome so caution would be recommended prior to performing central neuraxial blockade [31].

Laboratory testing should include blood levels of anticonvulsants to ensure levels that are within the therapeutic range. A full blood count may help exclude any possible bone marrow depression secondary to anticonvulsant therapy. A platelet count is also important considering the increased frequency of thrombosis and/or bleeding. Patients with heart failure on diuretic or digoxin therapy should also have their electrolytes checked.

Particular preparation for airway management

In preparation for a potentially difficult airway THRIVE (transnasal humidified rapid insufflation ventilatory exchange) should be prepared along with a variety of face mask sizes, endotracheal tubes, laryngoscopy blades and handles. THRIVE has become an essential proactive strategy in anticipated difficult airway management [32]. It may be beneficial as it extends safe apnea time while allowing uninterrupted access to the oral cavity which is of particular benefit during anticipated difficult airway management [33]. If THRIVE is unavailable, consider other methods of preoxygenation such as standard nasal cannula at high flow rates.

The use of videolaryngoscopes is not only helpful in difficult airways but may help prevent injury to airway angiomas by allowing navigation around them [20]. Fiberoptic intubation may be considered the safer option in a potential difficult airway if the patient is compliant and has the advantage of being able to quantify any airway involvement below the level of the vocal cords [20]. There should also be access to a difficult airway trolley for alternative oxygenation strategies should the need arise.

Emotional stress with its potential hemodynamic effects can cause swelling of angiomas with increased risk of perioperative bleeding [34]. This can be partially avoided by ensuring a good preoperative rapport with the patient, and with adequate premedication.

Facial asymmetry may make head positioning, mask ventilation and intubation difficult. It is important to take time to adequately position the patient into the optimal position using pillows and/or blankets if required [29].

If examination or imaging is suggestive of airway angiomas, consultation with Ear, Nose and Throat specialists may be useful for further elucidation of lesions by upper airway endoscopy. These angiomas may involve the mucous membranes of the mouth including the lips, gingiva, tongue, palate, floor of mouth and may also be present as far down as the larynx, trachea and bronchi [25,27,28,35]. These may also be identified on any preoperative MRI, if airway structures are included in the imaging [20].

Particular preparation for transfusion or administration of blood products

Significant blood loss and difficulty with hemostasis can be encountered during excision or resection of any hemangiomas. Depending on the proposed surgery, a type and screen should be performed with a crossmatch required for any surgery at higher risk of bleeding. Tranexamic acid (TXA) should be considered in major surgeries with potential blood loss of more than 500 mL as it has been found to reduce bleeding by 25% with a low probability of thromboembolic events [36-38]. Evidence shows a dose-dependent increased risk of seizures with TXA administration particularly at doses exceeding 2 grams per day or 100 mg/kg [39]. Risk factors include those over 75 years, preoperative renal failure, high dose TXA (>100 mg/kg) and those undergoing cardiac bypass surgery and gastrointestinal hemorrhage where high doses of tranexamic acid are used [40,41]. The mechanism is thought to be via competitive antagonism at the inhibitory ligand gated glycine and GABA-A receptors in the brain [40,42].

Particular preparation for anticoagulation

Aspirin use is increasingly common in patients with SWS, often recommended for the management of seizures and stroke-like episodes [43,44]. Recurrent thrombotic episodes within the abnormal angiomas may also require the use of antiplatelet agents. Antiplatelet therapy may impact perioperative hemostasis and should be considered when planning surgery.

Particular precautions for positioning, transportation and mobilization

Not reported.

Interactions of chronic disease and anesthesia medications

The mainstay of treatment in SWS involves the prevention of seizures with anticonvulsants. First line anticonvulsant medications include oxcarbazepine, carbamazepine and levetiracetam [45]. When ineffective, second line or adjuvant therapies that are utilized include lacosamide, lamotrigine, phenobarbital, cannabidiol, topiramate and clobazam, with benzodiazepines often being utilized for acute and prolonged seizures and clustered seizure management [45]. Other medicines commonly used by patients with SWS include ibuprofen, paracetamol and triptans for headache control. Importantly, carbamazepine and oxcarbazepine can be associated with thyroid insufficiency which may exacerbate hypothalamic pituitary dysfunction sometimes observed in patients with SWS [46]. These drugs also have the potential to affect enzyme metabolic pathways, which could subsequently affect the metabolism of muscle relaxants and anesthetic agents [15]. Conversely, some anesthetic agents may induce hepatic enzymes and reduce serum anticonvulsant levels to below the therapeutic range in the postoperative period.

Anesthetic procedure

Adequate premedication, in addition to anxiolysis, may facilitate seizure prophylaxis and placement of an intravenous line prior to induction of anesthesia in cognitively impaired children [29].

Airway equipment as described above should be prepared.

Induction, maintenance and emergence from anesthesia should be as smooth as possible as any light plane of anesthesia, straining, coughing or even laryngoscopy itself may increase IOP, ICP, or blood pressure theoretically risking angioma rupture and hemorrhage due to their abnormal autoregulation [25]. The rapid acting μ-opioid receptor agonist remifentanil may be particularly advantageous for both conscious sedation and as part of balanced general anesthesia in this population group [21]. Its pharmacodynamic profile makes it ideal in helping minimize the stress response to intubation, allowing for rapid titration during periods of increased surgical stimulation and allows for smooth extubation [47-49]. Processed EEG can help ensure adequate depth of anesthesia throughout the perioperative period [17].

Gentle laryngoscopy and tracheal intubation using a well lubricated endotracheal tube should be performed to minimize the hypertensive response and rise in ICP that can potentially lead to rupture of any cerebral angiomas [15,17,23,25,28]. Nasal intubation should only be performed once nasopharyngeal angioma involvement has been excluded.

Ketamine and succinylcholine, which may both cause transient increases of ICP and IOP should be avoided [24,29]. Anticholinergics should also be avoided in those with narrow angle glaucoma [15,20].

Endocarditis prophylaxis should be considered according to current guidelines [29].

Particular attention should be paid to hydration as dehydration may increase the risk of intravascular thrombosis.

Particular or additional monitoring

Not reported.

Possible complications

Complications can be grouped into airway, circulatory, CNS and metabolic complications.

Airway complications as discussed above can include problematic mask ventilation, hypocarbia, intubation difficulties or bleeding into the airway from angioma damage [29].

Perioperative sleep deprivation, hyperthermia, electrolyte disturbance, hypoxia, hypoglycemia, and hypotension may precipitate seizures and should be avoided [15,20]. Preparation should be made to treat seizures if they occur [50].

Vascular insufficiency might occur in organs such as the pituitary, thymus, lung, liver, spleen or pancreas where other angiomas may be present. This may lead to secondary metabolic and associated clinical disorders.

Hypoxia, hypercarbia, and hypertension which can have detrimental effects on IOP should be avoided in those with glaucoma.

Kossoff described outcomes for 32 patients following hemispherectomy. 47% experienced immediate postoperative complications which consisted of bleeding (4), infection (4), and re-operation was required in a further 3 patients because of seizures (1), hypertension (1) and

shunt (1). Perioperative deaths due to uncontrollable bleeding from diploic veins during burr hole creation, hyperkalemia from blood products, postoperative fluid shift and autonomic instability have been described after neurosurgical procedures [51].

Postoperative care

Seizure prophylaxis is of vital importance. As stated above perioperative sleep deprivation, electrolyte disturbance, hypoxia, hypocarbia, hypoglycemia, and hypotension should be avoided as abnormalities here may precipitate seizures. Any febrile episodes in children should be appropriately treated due to the potential for febrile-induced seizures [8]. It is important to ensure that oral diet along with regular anticonvulsant medication are resumed as soon as possible in the postoperative period. Patients in whom oral intake cannot be established, consultation with a neurologist may be required to adequately titrate nasogastric, rectal or even intravenous anticonvulsants. Hydration should be maintained.

Disease-related acute problems and effect on anesthesia and recovery

Seizures may be related to underlying cerebral pathology. Seizures may also be precipitated by any of the factors mentioned above. Potential for cerebral herniation in patients undergoing neuraxial anesthesia with concomitant raised ICP should also be considered.

Ambulatory anesthesia

Not reported.

Obstetrical anesthesia

Any parturient with port-wine nevus in the ophthalmic division of the trigeminal nerve should have a contrast MRI to rule out any cerebral angioma. This is especially important if the patient has a history of seizure disorders as there is the potential for angioma rupture during labor secondary to both the hypertensive pain response and Valsalva maneuvers during labor.

While regional anesthesia is safe in the majority of cases neuroimaging assessment will help clarify any possible concerns to central neuraxial blockade such as raised ICP or vertebral canal angioma. As stated above there should be awareness to the possibility of inadvertent dural puncture and potential cerebral herniation in those with raised ICP when performing epidural anesthesia. Excessive absorption or intravenous injection of local anesthetic may also provoke seizures [50]. It should be noted that pre-existing neurological deficits of the spinal cord or peripheral nerves is a relative contraindication to central neuraxial blockade.

Regional anesthesia is preferred to general anesthesia for cesarean section if time permits, ICP is normal, and the patient agrees [52].

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This recommendation was prepared in 2015 by:

Author(s)

Conan McCaul, Anesthesiologist, Department of Anesthesia, The Rotunda Hospital, Dublin, Ireland
cmccaul@rotunda.ie

Richard Katz, Anesthesiologist, Department of Anesthesia, The Rotunda Hospital, Dublin, Ireland

Disclosure: The author(s) has no financial or other competing interest to disclose. This recommendation was unfunded.

This recommendation was reviewed by:

Reviewer(s)

Anne Comi, Director of the Hunter Nelson Sturge-Weber Center at Kennedy Krieger Institute, Baltimore, USA
comi@kennedykrieger.org

Csaba Juhasz, Pediatrician, Wayne State University School of Medicine PET Center and Translational Imaging Laboratory Children's Hospital of Michigan, Detroit, USA
juhasz@pet.wayne.edu

Please note that this guideline has not been reviewed by an anesthesiologist but by two disease experts instead.

Disclosure: The reviewer(s) have no financial or other competing interest to disclose.

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Conan McCaul, Anesthesiologist, Department of Anesthesia, The Rotunda Hospital, Dublin, Ireland
cmccaul@rotunda.ie

Richard Katz, Anesthesiologist, Department of Anesthesia, The Rotunda Hospital, Dublin, Ireland

Reviewer

Beate Küppers, Anesthesiologist, University Hospital Pisa, Italy

Disclosure The reviewer has no financial or other competing interest to disclose.

Editorial Review

Christine Gaik, Anesthesiologist, Department of Anesthesiology and Intensive Care Medicine, University Hospital Giessen and Marburg, Campus Marburg and Philipps University of Marburg, Germany
gaikc@med.uni-marburg.de