What is Sturge Weber Syndrome?

Sturge Weber Syndrome (SWS) is a rare, nongenetic condition resulting from abnormal development of blood vessels of the skin, eyes and brain. The spectrum of clinical manifestations includes so-called port wine stains, usually affecting one side of the face; glaucoma due to increased pressure within the eye; seizures; stroke-like events; migraine headaches; and focal neurological impairments. The condition poses a number of specific health risks due to its progressive, multi-organ involvement.

Most patients develop seizures and progressive neurological deterioration with hemiparesis, visual impairment and cognitive decline that may start in infancy or childhood. Early diagnosis of brain and eye involvement in infants with a port wine stain is critical to offering effective treatment and, potentially, modifying the severity of the disorder.

Sturge Weber Syndrome Program at Cleveland Clinic

The Epilepsy Center provides coordinated, interdisciplinary care for early diagnosis and treatment of patients with clinical symptoms of Sturge Weber Syndrome. Our cutting-edge, longitudinal care aims to reduce complications that impact long-term functional outcome, and includes aggressive medical and surgical management of seizures. Physicians in our SWS Program are also committed to research and education to advance our understanding and treatment of patients with Sturge Weber Syndrome.

The SWS Program offers:

- **a well-coordinated team approach** by expert physicians in the fields of epilepsy, neurology, neurosurgery, neuropsychiatry, ophthalmology, dermatology and developmental medicine, who deliver comprehensive, multidisciplinary medical and surgical care
- **a world-class reputation** for diagnosing and treating SWS and related conditions, including one of the largest programs in the world for pediatric and adult epilepsy evaluation, epilepsy surgery and other treatment options
- **a focus on families**, with specialists who address the needs of adult and pediatric patients of all ages
- **focused, coordinated care** to help patients access multiple services through one channel without undue delays

SWS Epilepsy Treatment

Epileptic seizures starting in the first year of life are usually the first and most common symptom of brain involvement in patients with SWS. Early, aggressive treatment of the epilepsy is essential to improving the
cognitive outcome as well as the quality of life, as shown in studies from our center and others. If medical treatment fails, evaluation for possible epilepsy surgery should be considered early in every patient with SWS.

**SWS Program Team**

The challenges of epilepsy surgery in a patient with Sturge Weber Syndrome are complex but surmountable in the hands of experienced epilepsy specialists and surgeons working as a team. At Cleveland Clinic Epilepsy Center, we have established a multidisciplinary team of dedicated pediatric and adult epilepsy specialists, neurosurgeons, neuropsychologists, neuroradiologists, functional neuroimaging experts, cognitive and behavioral experts, health psychologists and bioethicists to administer epilepsy care to SWS patients. The team meets weekly to discuss the best individualized plan of care for patients with complex epilepsy.

The collaboration of neurologists and neurovascular and pediatric stroke specialists is essential to address stroke-like events in SWS patients and to evaluate other vascular malformations affecting organs and extremities, which may need surgical or radiologic vascular intervention.

**Health Risks of SWS**

- **Epilepsy**, if poorly controlled, can have devastating consequences on the patient’s and family’s quality of life. In SWS, seizures tend to cluster over a number of days. An individualized treatment plan should start with first aid in the home setting, then progress to decision making on the next step of care at the emergency room and transfer to a pediatric facility with expert care for seizure treatment. Such a plan should be put in place for patients with the port wine stain and brain involvement. Ideally, patients should be seen in infancy, prior to the onset of seizures, to educate and instruct the family and establish a local network of care. A subsequent stepwise approach to epilepsy management should include early surgical evaluation when medication fails to control seizures and neurological function and development are at risk.
- **Skin** can be treated early with dermatologic laser therapy and surgery.
- **Eye pressure increase or glaucoma** leading to impaired vision is common, requiring early and serial assessment for medical and surgical management.
- **Migraine headaches** can be treated by pediatric headache experts in the pediatric headache clinic.
- **Airway and breathing problems** require the attention of pediatric ear, nose and throat specialists.

**Research**

Many questions about SWS are unanswered: What are the causes and are they preventable? Can we prevent seizures and stroke-like events? How do we best treat them medically and surgically?

Physicians in the Cleveland Clinic SWS Program are at the leading edge of interdisciplinary SWS treatment, education and research, which focuses on outcomes and finding biomarkers in the blood that may predict vascular risk underlying seizures and stroke-like events. By investigating these issues, we hope to improve the lives of patients and families.

**Referring Patients to the SWS Program**

If you have questions regarding a patient or our SWS Program, call Ingrid Tuxhorn, MD, at 216.444.8827 or toll free, 1.800.223.2273, ext. 48827, or visit clevelandclinic.org/epilepsy.