STURGE WEBER SYNDROME

What is it?

Sturge-Weber syndrome (SWS) is a rare genetic condition characterized by a congenital facial birthmark and neurological abnormalities. A facial birthmark or port-wine stain is the most apparent indication of SWS at birth. It appears on the forehead and upper eyelid of one side of the face. The port-wine stain can vary in color from light pink to deep purple and is caused by an overabundance of capillaries around the trigeminal nerve, just beneath the surface of the face.

SWS is also characterized by neurological concerns caused by the abnormal formation and growth of blood vessels within layers of tissue covering the brain and spinal cord. This abnormality can impair blood flow and lead to loss of brain tissue. Brain calcifications can also occur on the underlying tissue in the cerebral cortex of the brain; on the same side of the brain as the birthmark. Neurological symptoms most notably include seizures that begin in infancy and may worsen with age. The seizure activity usually happens on the side of the body opposite the birthmark.

What are the characteristics or complications?

Each case of Sturge-Weber Syndrome is unique and exhibits the characterizing findings to varying degrees. SWS affects the development of certain blood vessels, causing abnormalities in the brain, skin, and eyes. The three major features of SWS include a port-wine birthmark, a brain abnormality called a leptomeningeal angioma, and increased pressure in the eye. A person does not have to have all three features. Children with SWS can also be diagnosed with glaucoma because of the abnormal blood vessels in their eyes. Glaucoma has 2 peak periods; one in infancy and one in later young adulthood. People with SWS may also experience cognitive impairment and learning disabilities similar to ADHD. In addition to the distinct birthmark and neurological conditions, other characteristics and complications can include:

- Hemiparesis (opposite side of port wine stain)
- Internal organ irregularities
- Delayed mental and physical development
- Mood and behavior problems
- Vision field cuts
- Migraines, headaches
- Hormonal issues
- Cognitive and psychological impairment

What is the treatment?

There is no cure for SWS. Treatment is focused on the specific health issues and the related symptoms. A team of medical and developmental specialists are key to the best possible outcome. Those diagnosed with SWS will often be followed by numerous specialists which might include neurologists, ophthalmologists, dermatologists and neuropsychologists. The pediatrician will often manage the multidisciplinary approach. Some treatments might include: laser treatments for the port-wine stain, anti-seizure medications, medications to lower intraocular pressure or surgery for severe cases of glaucoma and PT/OT/SLP therapists to address cognitive and physical development.

Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

Supporting students with SWS in the school setting require educators and parents/guardians to work as a team. A well coordinated plan promotes success for the student in the educational setting. Some accommodations to consider for an 504/IEP could include:

- Early intervention assessment for young children
- PT/OT/SLP/vision evaluations
- Emotional support
- Education about seizure management for staff
- Offer preferential seating
- Know behavior signs for when a break is needed
- Offer designated rest area
- Copies of notes, whiteboard, presentations in proper font size
- Consider assistive technology
- Extended time for testing
- Accommodations if student is having surgery
- Revisiting work if student has seizures
- Extra time for assignments as needed
- Offer emotional support
- Staff education and training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including all affected systems and symptoms
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Current medication list for home and school
- Documentation/log of seizures including type of seizure, description of, typical length, characteristics, triggers, warning signs, how often seizures occur, and student's behavior following a seizure
- Orders for emergency medications, when to administer, dose, route
- Orders for hidden medical device like a vagus nerve stimulator including how to use/manage
- PT/OT/SLP/Vision services and hearing specialists assessment
- Bleeding precautions in event injury occurs at area of birthmark
- Plan for monitoring neurological changes, headaches, anxiety
- Orders for fluid intake goals for student to stay hydrated
- Provide rest area if needed
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

Kennedy Krieger Institute: Hunter Nelson Sturge-Weber Syndrome Center
https://www.kennedykrieger.org/patient-care/centers-and-programs/sturge-weber-syndrome-center

Sturge Weber Foundation
https://sturge-weber.org/

The Vascular Birthmarks Foundation
https://birthmark.org/

National Institute of Neurological Disorders and Stroke
https://www.ninds.nih.gov/Disorders/All-Disorders/Sturge-Weber-Syndrome-Information-Page