STURGE-WEBER SYNDROME

Background

Sturge-Weber syndrome (SWS) is a rare vascular disorder affecting the skin, eyes, and brain. It is characterized by a facial birthmark called a port-wine stain and blood vessel growth abnormalities. The severity of symptoms is highly variable.

A facial birthmark or port-wine stain is the most visible feature present at birth. It presents on the forehead and upper eyelid, usually on one side of the face. The port-wine stain is caused by an enlargement of capillaries just beneath the surface of the skin and can vary in color from light pink to deep purple.

When present, eye abnormalities typically occur on the same side of the head as the port-wine stain. Abnormal blood vessels in various parts of the eye can cause visual impairment, vision loss or glaucoma. Some individuals may have eyes that appear enlarged or bulging because of the intense pressure.

The abnormal formation of blood vessels on the surface of the brain (leptomeningeal angioma) can cause a wide variety of neurological symptoms. Seizures, beginning in infancy or early childhood, are common. Cognitive impairment may be more severe in individuals with severe seizures or refractory epilepsy. Other neurological complications may include muscle weakness or paralysis on one side of the body (usually opposite of the port wine stain) and headaches. Abnormalities in the walls of arteries may also increase the risk of a stroke or a stroke-like episode. Emotional and behavioral conditions, especially depression, are common.

Prognosis of SWS is extremely variable. Some individuals are severely impaired due to neurological dysfunction, visual changes, and mobility needs. Others are more mildly affected with well-controlled seizures, intact vision, mild motor impairment, and normal intelligence with perhaps attention problems or milder learning disabilities.

There is no cure for SWS. Treatment is focused on managing the specific health issues and the related symptoms. Laser treatment of the port-wine stain and medication(s) to help control seizure activity and intraocular pressure are common options.

Top Takeaways for School Considerations

Sturge-Weber syndrome (SWS) is characterized by facial birthmark called a port-wine stain, neurologic complications, and visual impairment.

Blood vessel abnormalities on the surface of the brain can cause problems such as seizures, strokes, weakness, vision loss, and a range of intellectual challenges.

Staff should be educated about signs and symptoms to recognize a seizure and procedures to notify trained staff.

Side effects of anti-seizure medication can cause fatigue, inattention, and restlessness that may further interrupt school performance.

Individuals with SWS have varying levels of cognitive function. The greater the extent of blood vessel abnormality across the brain and the more severe the seizure activity is associated with intellectual disability.

Annual monitoring for glaucoma is recommended. Eye pressure can be extremely painful and may cause behavioral issues in individuals who are nonverbal. Some children may also have a visual field deficit.

Kennedy Krieger Institute's Specialized Health Needs Interagency Collaboration

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

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Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of risk for injury, risk for disturbed sensory perception and alteration in coping
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Student-specific triggers, avoidance, or intervention strategies
- Use of specialized equipment, adaptive equipment, and orthotics
- Activity, positioning, transferring (consider precautions and/or restrictions)
- Consider emergency care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Has the school staff been trained to implement the student-specific emergency plan?
2. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
3. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
4. Can rest breaks, safe spaces, or reduced stimulation times be built into the student's schedule?
5. Does the student require activity precautions to prevent injury?
6. Is the physical school environment safely accessible for the student's mobility needs (e.g., entry and exit, ramps, location of classes, access to elevator, doorways)?
7. Does the classroom environment support the student's needs and/or equipment (e.g., desk/seating options, maneuverability space, electrical outlets, flash pass for bathroom or

Resources

Kennedy Krieger Institute: Hunter Nelson Sturge-Weber Syndrome Center
kennedykrieger.org

Sturge Weber Foundation
sturge-weber.org/

The Vascular Birthmarks Foundation
birthmark.org/

Scan QR code or visit
KennedyKrieger.org/HealthInformation for more information.

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