



STURGE-WEBER SYNDROME

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Sturge-Weber Syndrome (SWS) is a rare disorder that is present at birth. This syndrome affects the blood vessels of the central nervous system, eyes, and the skin. In SWS blood vessels of the brain and face grow more rapidly and dilate abnormally. This appears to be due to genetic mutations. As a result there is an increase in the number and size of blood vessels, called angiomas, which can alter the color of the skin. In SWS the skin of one side of the face is usually involved, though rarely it can be bilateral. The skin appears more red, purple or blue over the forehead and around an eye, but an entire side of the face can be affected. This color change is referred to as a birthmark or port-wine stain. Port-wine stains continue throughout life and can get larger, more elevated, and darker in color.

Just as the skin of the face is involved because of the vascular malformations, the vessels of the brain are also involved and these angiomas can grow within certain regions of the brain. Because of the vascular malformations pushing on the brain, complications that can occur include seizures, paralysis, developmental delays, and glaucoma.

Generally speaking birthmarks or vascular malformations can arise anywhere on the skin. In SWS these are usually isolated to the skin of the face. It is important to note that if SWS is suspected, a neurologist should be seen. The neurologist can request certain imaging studies of the head to determine the best course of treatment. Depending on the extent of the disease a neurologist may prescribe medications to limit seizure activity, and to improve quality of life. An ophthalmologist should be involved in the patient's care to monitor and treat problems with vision called glaucoma. Glaucoma is a loss of vision because of complications due to high blood flow to the eyes.

The port-wine stains can be tender, painful, congested with blood as to alter facial features, and even become infected. These port-wine stains should be evaluated by a dermatologist. Depending on location and how large or involved the vascular malformation is, it could cause obstruction of vision or cause problems with feeding. Treatment is usually not delayed to prevent negatively impacting vision or an infant's nutritional development. Treatment can be as limited as the use of a **laser** called a pulse dye laser (PDL) or be much more involved requiring surgery to treat the vascular malformations. With PDL a focused high energy beam of light is directed toward blood vessels in short bursts, the bursts of energy heat the blood vessels and cause them to shrink. As the blood vessels shrink, blood can no longer move through the malformed vessels in the skin, and the red discoloration clears. Multiple sessions may be necessary.

The prognosis of SWS is dependent on the severity of the disease. The type of seizure and when the seizures began, the extent of vision loss, and involvement of the facial features determine the severity and future of the patient.

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