Sturge-Weber Syndrome

WHAT IS STURGE-WEBER SYNDROME?

Sturge-Weber syndrome is characterized by a reddish discoloration of the skin on one side of the face (port wine stain) and malformation of blood vessels of the brain [See figure 1].

Fig. 1: Sturge-Weber Syndrome is characterized by a reddish discoloration of the skin on one side of the face.

DOES EVERY CHILD WITH A PORT WINE STAIN OF THE SKIN HAVE STURGE-WEBER SYNDROME?

No. The reddish discoloration of the face may be an isolated finding. Sturge-Weber syndrome is diagnosed when the brain is involved as well.

IN WHAT WAY IS THE BRAIN INVOLVED?

Abnormal brain vessels may cause a variety of symptoms. Seizure activity, neurological deficits, developmental delay, and weakness are some of the more common findings. Many children, however, have normal intelligence and development.

WHAT TESTS ARE DONE TO LOOK FOR BRAIN INVOLVEMENT?
Neuro-imaging studies (CT scan / MRI), may be utilized to evaluate for abnormal vascularity of the brain.

**WHAT CAUSES STURGE-WEBER SYNDROME?**

Sturge-Weber syndrome is not a genetically inherited disease. The abnormalities associated with Sturge-Weber syndrome develop very early in pregnancy - probably between the second and third month of pregnancy.

**WHEN IS STURGE-WEBER DIAGNOSED?**

The port wine stain is usually noted shortly after birth. Further testing may reveal involvement of the brain (Sturge-Weber syndrome).

**WHY IS THE SKIN OF THE FACE SO RED?**

Redness (port-wine stain) of the skin is caused by numerous, dilated, small blood vessels. The forehead and upper eyelid are often involved while the lower eyelid and cheek are occasionally involved.

**IS THERE ANY TREATMENT FOR THE SKIN?**

The skin appearance can be improved in some situations by laser treatment.

**WHAT EYE PROBLEMS CAN OCCUR WITH STURGE-WEBER SYNDROME?**

The surface of the eye occasionally has large, dilated vessels which cause a pink or red eye appearance. These blood vessel abnormalities can occur inside the eye as well, leading to an elevated pressure within the eye. If this elevated pressure begins to cause damage to the optic nerve, this is condition is referred to as “glaucoma.”

Glaucoma is the most serious eye problem associated with Sturge-Weber. About 50% of children with Sturge-Weber develop glaucoma between infancy and later childhood. Symptoms of glaucoma may include a large and/or cloudy eye, light sensitivity, and excessive tearing.

If the optic nerve or visual areas of the brain are involved, a defect in the peripheral vision (“visual field”) may result. Children with Sturge-Weber should have regularly scheduled examinations by an ophthalmologist to monitor for eye problems.
HOW IS GLAUCOMA TREATED?
Drops may be used to help control the pressure, but surgery may also be indicated.

WHERE CAN I LEARN MORE ABOUT STURGE-WEBER SYNDROME?

- Sturge-Weber Foundation

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