Sturge-Weber Syndrome

Sturge-Weber syndrome is also called "encephalotrigeminal angiomatosis". It is a congenital disorder. It causes a vascular birthmark and neurological abnormalities.

SYMPTOMS

Symptoms vary widely among patients.

- They may include eye and internal organ irregularities.
- The most apparent symptom is a facial birthmark or port wine stain. This is present at birth. It usually involves at least one upper eyelid and the forehead. The stain ranges in color from light pink to deep purple. It is caused by an overabundance of capillaries just beneath the surface of the affected skin.
- Neurological symptoms include excessive blood vessel growth on the surface of the brain (angiomas). These angiomas are typically located on the posterior or occipital region of the brain. They do cause seizures. The seizures often start before one year of age. They may worsen with age. The convulsions usually appear on the side of the body opposite the port wine stain. They vary in severity. A weakening or loss of use of the side of the body opposite the port wine stain (hemiparesis) may also develop.
- Developmental delay of motor and cognitive skills may occur.
- Glaucoma is increased pressure within the eye. It may be present at birth or develop later.
- Buphthalmos is enlargement of the coatings of the eye. It may also occur in the eye that is affected by the port wine stain.
- Sturge-Weber syndrome rarely affects other body organs.

TREATMENT

Treatment for this syndrome is symptomatic. Laser treatment is available to lighten and/or remove port wine stains. Anticonvulsant medications may be used to control seizures. Surgery and/or eyedrops may be prescribed to control glaucoma.

SWS related seizures can sometimes place patients in potentially life threatening situations. But the disease itself is not fatal.

RESEARCH BEING DONE

The NINDS supports a broad program of research to better understand congenital and seizure disorders such as Sturge-Weber syndrome.

Research is continually being done to learn more about this problem.

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