

DR. LINDA'S VASCULAR BIRTHMARK FACT SHEET

10 FACTS ABOUT STURGE-WEBER SYNDROME



- 1. Glaucoma is the most common serious eye problem of SWS, with a reported incidence of 30–70%. It is defined as an increase in intraocular pressure that causes pathological changes in the optic disk and typical defects in the field of vision.
- 2. A condition where the involved eye can have a darker colored iris (Heterochromia) may occur in those with SWS.
- 3. Various experts report that nearly 50% of all infants diagnosed with a port wine stain on the eyelid will be at risk for glaucoma. Involvement of the upper eyelid has the highest association with SWS.
- 4. Recent studies have pointed to SWS as the result of a genetic mutation associated with the nucleotide transition in gene GNAQ on chromosome 9q21.
- 5. Glaucoma associated with SWS usually affects only one eye, but in rare cases can affect both eyes.
- 6. Individuals with upper and/or lower eyelid involvement, but no glaucoma, are still at risk for late-onset glaucoma. It can occur in late childhood or adulthood.
- 7. With SWS, 60% of the glaucoma is diagnosed in early infancy (when the eye is susceptible to stretching effects of increased intraocular pressure—glaucoma). These infants can have enlarged corneal diameters and myopia (actual eye enlargement called Buphthalmos). For the other 40%, glaucoma begins later in childhood or early adulthood. When it is late onset, there is usually no eye enlargement.
- 8. Most doctors agree that the management of glaucoma associated with SWS is difficult. Lifelong anti-glaucoma drugs with a potential for systemic side effects are frequently needed and, often, multiple surgical procedures are needed.
- 9. Treatment of SWS should always involve an ophthalmologic physician who is a glaucoma expert and is familiar with this syndrome.
- 10. Eye exams for those with SWS should be conducted under anesthesia and performed at least every six months.



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