



The Sturge-Weber Foundation

The stronger the wind, the tougher the trees

Sturge-Weber Syndrome & Brain Involvement

What does a brain with SWS look like/how is it different from a "normal" brain: The surface of the brain is where the angioma is located, however, because there may be a diversion of blood through the angioma which can "Steal" blood from the underlying brain, the tissue affected may undergo atrophy. This is because the tissue is chronically suffering from lack of adequate oxygenated blood supply.

What happens to the brain during a seizure: The seizure is an episode of altered brain function corresponding to an alteration in the electrical activity of the tissue. If the entire brain is involved the patient has a generalized seizure with complete loss of consciousness. If the activity is in only one portion of the brain, the patient may have some preservation of consciousness. Chronic hypoxia may increase the irritability of the brain and lead to seizures.

Why is it important for an SWS patient not to have a seizure that lasts longer than 5 minutes: Prolonged seizures may result in injury to normal or functioning neurons. For this reason we like not to have prolonged seizures and 5 minutes an arbitrary but reasonable time limit.

Why is SWS considered "progressive" and what does that mean: The effects of chronic hypoxia are somewhat cumulative over time. There is quite a bit of variability and for many children this is not progressive but for some it clearly is an ongoing process and the dynamics of cerebral blood flow in the region of the brain affected may change over time as venous pressure, arterial pressure, perfusion pressure etc. change.

What is calcification, how does it occur: The calcification is called dystrophic calcification and is the natural consequence of chronic hypoxia. It will occur in any situation which causes the tissue concerned to get insufficient oxygen over an extended period of time.

What is atrophy, how does it occur: Atrophy is another effect of the chronic hypoxia and will also occur in the situation where there is not enough oxygen delivered to the tissue. It frequently occurs with Calcification but not always. Atrophy can also occur after a more acute injury to brain tissue such as after a stroke or traumatic injury.

What other things happen to the brain with SWS: The Angioma, if present and causing a change in Oxygen delivery from early on, may result in malformations of the brain because of the lack of oxygen to the developing tissues.

What about blood flow and oxygen: Blood is the only major way of delivering oxygen to the tissues. Without adequate blood flow you cannot deliver adequate oxygen to the tissue.

How does aspirin therapy help/if it does: We don't know if it does help but Aspirin may help to prevent the development of clots in the vessels in those children who are prone to the "stroke like events" frequently talked about in SWS

Why does the person with SWS get more headaches/if they do: There is an increased likeli-

hood of headaches in SWS. About twice the general population risk. Several possible mechanisms including the changes in blood flow and the change in oxygenation both of which could cause headache and could trigger migraine.

What about strokes: There is no solid data to indication how often this occurs and some of us believe the apparant strokes are mostly the after effects of seizures.

Is there any way to tell how severe the case will be/what the progression will be:
No reliable way to tell except to see what has gone on up to now and guess at what is next.

Why do some individuals with SWS have mental retardation or delays: The angioma may affect the development of the brain, the seizures may affect the development and function of the brain, the medications may affect function of the brain and the size of the lesion may not leave enough normal brain to function normally.

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