



# BranchingOut

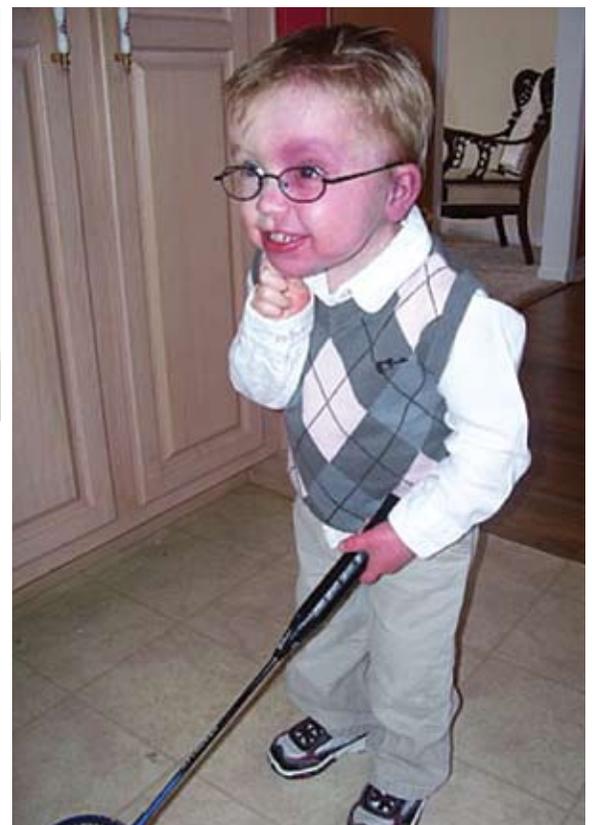
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# Glaucoma Risk in Children with SWS

By Robert Tomaino

When most people hear the term glaucoma, they probably associate it with the elderly – something involving the eyes of their grandfather or grandmother. But for some parents whose children have Sturge-Weber syndrome (SWS), glaucoma can occur at a very young age or even at birth.

**Sam and Mary Dalton** of Prince Edward Island, Canada, are well aware that glaucoma is not a disease of the elderly. Their youngest child, **Gabriel**, was born in 2005 and diagnosed with Sturge-Weber syndrome which came with accompanying concerns about glaucoma. Just two months later, Gabriel underwent surgery to relieve pressure from his left eye. Gabriel, now 29 months old, needs several different eye drops to help manage the disease and still faces the possibility of additional surgery in the future.



*With most diseases, we try to treat with drugs and save surgery as an adjunct to the drug therapy, said Dr. Chang. However, with pediatric glaucoma the opposite is true. Children often receive surgery first with drug therapy serving as the adjunct therapy. Unfortunately for children the drug options are limited due in part to the difficulty getting drugs tested and approved for children.*



Glaucoma is commonly thought to be one disease, but is actually a group of disorders characterized by optic nerve damage due to increased pressure within the eye. The reason for the increased pressure is varied.

In SWS, there are two peak times when glaucoma may occur – either at or shortly after birth or in older children between the ages of 5 and 9. According to **Peter Chang, M.D., Assistant Professor at the Baylor College of Medicine and the Cullen Eye Institute in Houston, Texas**, the cause of glaucoma in these two age groups is different.

In infants or newborns, glaucoma occurs as a result of poor development or blockage of the eye's natural drainage system. Fluid builds up in the eye, increasing the pressure within the eye. In older children, glaucoma is a result of increased blood pressure in certain blood vessels of the eye. The high pressure in the vessels leads to increased pressure within the eye, preventing fluid from draining out of the eye.

Although the two causes of glaucoma are different, there are instances when they both occur in one child. Children whose glaucoma is due to abnormalities of the eye's natural drainage system are at risk of developing

glaucoma as a result of high pressure of blood vessels when they grow older.

These different forms of glaucoma require different approaches to treatment. Gabriel Dalton underwent a procedure called a **trabeculectomy**, which creates a new passage that allows fluid to drain from the eye, reducing the pressure. Though Gabriel's surgery was successful, the pressure in his eye is still high and he must receive eye drops both in the morning and at night. According to Dr. Chang this is not uncommon.

For older children, a completely different surgery is required which involves the creation of a new drainage system for the eye. In this surgery, doctors

use a silicone implant to bypass the area of high pressure thereby allowing fluid to drain from the eye. As with younger children, these children also often need eye drops and other measures to help manage the glaucoma.

The response to these surgeries depends on the individual. Some children may not need any additional surgery, but may need medications. Other children may not need any medications to control their glaucoma. However, there are instances when children may need multiple surgeries in addition to using many different eye drops until a helpful combination is found. Other supportive measures may also be necessary, for example, Gabriel occasionally wears an eye patch over his unaffected eye as a way to strengthen his affected eye.

"Patching was very challenging at first," said Sam Dalton in an email interview. "It still has its moments but from being persistent, things have really paid off." Although doctors said Gabriel would not be able to see much more than 3 or 4 feet in front of him with the weakened eye,

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## Glaucoma Risk...

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Gabriel is able to see Sam and Mary from more than 20 feet away and wave back. He can “see his sippy cup from far off,” Sam added.

Even if a child shows no signs of glaucoma following surgery and does not need medication or other measures to help manage the disease, there is always the risk that glaucoma could recur.

“Surveillance is important even for successfully treated patients,” stresses Dr. Chang. Periodic eye examinations are required to ensure that an increase in eye pressure is caught early enough to treat before permanent eye damage occurs. Although there may be a few signs pointing to glaucoma – a puffy or swollen eye; sensitivity to sunlight; frequent squinting or closing of the affected eye – glaucoma often develops without any warning signs.

If the increased eye pressure damages the optic nerve (the nerve that transmits visual stimuli from the eye to the brain), the damage is permanent. However, Dr. Chang explained that some nerves affected by glaucoma that were thought to be “dead” and beyond saving are not. These nerves are actually in a sort of “coma.” Although they appear damaged, they can actually be brought back if the glaucoma is caught early enough.

Gabriel Dalton’s eye pressure remains high and to avoid optic nerve damage, the doctors feel that he will need surgery again later this year. Sam is ready. “We hope for the best but realize surgery may be the only option.”

Pediatric glaucoma research is ongoing always with the hope that better, safer procedures will be developed. This is especially important for children with SWS.

“People with SWS are at a higher risk for bleeding complications during and after surgery,” said Dr. Chang.

In the meantime, current surgical and drug options will continue to help children like Gabriel. Sam also cites the support from families with children with SWS and the Sturge-Weber Foundation (SWF) as being invaluable.

“We were told the very worst case situations with SWS,” said Sam. “We were devastated at first. It was the lowest point of my life, because we lost hope and didn’t believe our son would have any quality of life. However, after contacting the SWF we learned that things can be good.”

So far Gabriel hasn’t let SWS or glaucoma slow him down and although Gabriel will need more surgery, Sam, who

is hopeful for the future, counts his blessing as he shares his son’s story. Gabriel has a soon to be 6-year-old sister named Sophia and a 4-year-old big brother named Xavier.

“They are all the best of friends,” says Sam. “They chase one another and play games all over the house. Gabriel can keep up even when he has his eye patch on. So far he is beating glaucoma... a fight we hope he will win!”

*Rob Tomaino is a free lance writer with a special interest in SWS/PWS/KT.*

**Ian Hubling** is an SWF Partner from Ontario, Canada and has been the prime mover of the Pediatric Glaucoma and Cataract Family Association. [www.pgfa.org](http://www.pgfa.org). Ian and Linda’s son **Calvin** is now 15.

Ian suggests that if parents want to keep up with the latest in pediatric glaucoma issues, they can subscribe to an email notification list through the



*Calvin Hubling*

PGCFA. They will receive information on news articles of interest, updates on research, etc. Dr. Alex Levin of the Hospital for Sick Children in Toronto (also an SWF Consultant) reviews industry abstracts and studies and summarizes them. Dr. Levin’s summaries, as opposed to the original articles, are always parent-friendly.

SWF Partner and Board member **Jackie Brown** shares this bit of information:

“Our daughter who is now 8 has been patching since 6 months old. There is a web site, [readingglassesforless.com](http://readingglassesforless.com) that sells disposable eye patches with designs. The girls’ ORTOPAD eye patches are pink and light blue and have flowers and smiley faces on them. There is also a boy’s version. On their web site, look under professional optical supplies”.

Also try [Fresnelprism.com](http://Fresnelprism.com) for kids’ eye patches. They donated a supply of colorful patches that were available at last year’s Conference in Anaheim.