

## INSIGHT INTO CHILDREN'S GLAUCOMA AND CATARACTS

### The Red Reflex

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The red reflex test is an examination technique performed by most pediatricians and all ophthalmologists on patients as young as newborn infants. Every person who has taken a photograph with a camera with a built-in flash knows what is described as the "red reflex." It's the red-orange spot ("red eye") in the center in the pupil made by the light that reflects out of the eye.

The eye is an elongated sphere with the clear cornea over the coloured part of the eye (iris) in front connecting to the white of the eye (sclera). Light entering the front of the eye is focused by the cornea and the lens, onto the inside lining of the eye (retina). The retina is the colorless, tissue paper-thin layer of cells that turns the light energy into chemical energy and transmits the information to the brain via the optic nerve. The brain then reads the message as an understandable picture and vision is perceived. Underneath the transparent retina is another layer of the eye that provides the nourishment to the retina. This thin blood filled layer is called the choroid, and is reddish-orange in color.

Normally the center of the pupil is black, since there is no light coming from the inside of the eye. Imagine looking into someone's house from outside when there are no lights on inside. You can't see anything inside. As soon as the inside lights are turned on, outside observers can then see inside. When light is shined into the eye, it is reflected off of the red-orange choroid, and the reflected light produces a red-orange (or sometimes orange-yellow) light in the center of the pupil called the RED REFLEX.



The examining physician shines a light (from a tool called the direct ophthalmoscope) towards the patient's eyes from about 1 meter away. Then the doctor looks through a peep hole in the instrument that is shining the light at the quality of the color of the red reflex returning from each eye and simultaneously compares the reflexes of each eye to each other. It would be expected that this test would be performed in the newborn nursery by the pediatrician doing the child's first examination after birth. A pediatrician or family physician should usually perform a red reflex examination during every well-baby visit.

If anything interferes with the free and easy transmission of light through the front of the eye - and back again, the reflex is affected, producing either a white (light bouncing off something white inside the eye) or black (no light getting in to bounce back) reflex rather than red-orange.

A swollen cornea (a possible sign of glaucoma), cataract, tumour, retinal infection, or retinal detachment can all produce the abnormal white reflex. This would normally constitute an emergency referral to an eye specialist. A cataract, scar, or bleeding inside the eye can cause a black reflex due to blocking of the light. Sometimes, a difference in the red reflex quality between the two eyes is just due to an asymmetry in glasses prescription (e.g. one eye normal and the other very nearsighted).

In summary, the red reflex is a simple and valuable screening test performed by physicians to screen infants and children for serious eye disease. By simply shining a special light into the child's eye, the practitioner is able to determine if possible vision-threatening eye conditions exist.

# Development In Visually Handicapped Children

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Visual competence is a critical component of all aspects of child development. Blind children are disadvantaged in their learning as lack of vision affects mobility, sensory coordination and attachment. The evaluation of developmental progress in a blind child depends on the degree of visual deficit and on the presence or absence of other additional handicapping conditions – for example, hearing loss, cerebral palsy or cognitive delay. There are however, well recognized developmental variations in children with central or peripheral visual loss. The recognition of these patterns is important in the construction of therapy programs and parent counselling.

In the first 6 months of life, a child with visual handicap will often show absence or reduced eye contact and visual tracking (fixation and following). As small infants, these children tend to be very passive and will react by "freezing" to unfamiliar environmental sounds. There is a delay in the acquisition of gross motor skills and delayed reaching. Strategies for treatment use brightly colored objects and sound making toys or equipment. For example, placing a light wrist or ankle bracelet with small bells sewn onto the cloth will help a blind infant find the location of his or her limbs and allow a normal sequence of motor development to take place.

From the ages of 6-18 months, when normal motor milestones include sitting, crawling and walking, if there is an absence of vision, unusual patterns of motor development or delayed

motor skills are evident. Protective reflexes – for example, the parachute response (the reflex of extending the arms to catch oneself on falling forward) are delayed – and often not present until 18 months of age. In sighted children, this reflex is normally present by 8-9 months. This delay in visually handicapped children suggests the response is learned and requires visual input. Blind children do not develop a pincer (or fingertip) grasp until close to 2 years of age, but do use a "raking" grasp with all four fingers and a stationary receptive thumb – this is an efficient grasp and does not impede overall learning. Commonly, visually handicapped children show a dislike of, or are frightened of tactile experiences – for example they may be fearful of furry toys or animals.

Children from age 18 months through to 3 or 4 years of age develop confidence in movement and communication skills. Visually handicapped children show poor exploratory motor activities and seem most interested in near objects – or objects which can be held for inspection and manipulation. There is a marked reduction in self initiated mobility.

Developmental paediatric assessment must be modified for children with visual handicaps. Therapy must allow for opportunities for the blind child to understand their environment – the example above of "sound for sight" is useful. Teaching a blind child to reach for sound must be begun at the critical developmental period for reaching – and if successful can eliminate maladaptive "blindisms".

It is particularly important to encourage speech and communication skills as well as cognition for blind children. The child's position in the classroom, level of environmental lighting and sound are important in ensuring optimal developmental and learning progress.

## Practical Pearls Of Contact Lenses For Parents

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Contact lenses are used in children for various reasons: vision correction, improving appearance, and treatment of some eye diseases. The contact lenses present the usual considerations regarding the necessity to provide good vision, good comfort, and a proper maintenance of the health of the cornea. Contact lenses for children however, also bring along some more unique practical concerns and considerations.

Many of the lens wearers are not the one who is primarily in charge of lens care and compliance to the prescribed lens wearing system. It is up to the parent or guardian to learn insertion, removal, and care, while sometimes working with a child who is not a willing participant. In most cases, the contact lenses have been prescribed to correct for a vision problem that may not be effectively addressed with eyeglasses. It is important to remember that for most wearers, the lenses must be worn regularly in order to maintain visual function for the long term and prevent as much as possible the development of a "lazy eye" (amblyopia). It is for this reason that the commitment must be maintained and the wearing system adhered to as much as possible.

### LENS INSERTION

One of the most common difficulties for the parent is daily insertion of the lenses. Most are nervous initially when inserting and removing a child's contact lens for the simple reason

that they are not sure if the process is causing discomfort or pain. Be assured that the negative reaction that most kids express when inserting or removing their contacts is more psychological than physical discomfort. The entire process is, unfortunately, invasive of the child's space. Kids do not like to have their space invaded and will almost universally resist. With infants, it can be of help to wrap a blanket around them so as not to allow them to bring their hands to their face while you are trying to insert the lens. The same process may be repeated when removing the contact. It is important to be firm in your actions when restraining the child, holding their lids, and placing the lens on the eye. But at the same time, talk in a positive manner about the insertion of the lens and the fact that it is a very quick process in order that they keep in better perspective that the lens will be on the eye very quickly and you will be retreating from their space soon. Ultimately it is the person who is inserting the lens who must forge ahead and proceed with the insertion of the lens regardless of the protests of the child. In the end, we have yet to see a child "turn against" their parent. In fact, as a child gets older they begin to appreciate the vision offered by the contact lens and sometimes cry when the lenses are removed wanting them back in again!

Lens removal is usually easier although the level of resistance may still be high on the part of the child. For some it may be of help to wait until the child is asleep before attempting to remove the lens. With practice you and your child will become used to the process in time.

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## LENS LOSS

It is helpful with infants to check to make sure that the lens is in place before relocating the child. That is, look at the lens on the eye before removing the child from the car, going outside, etc. This will give a reasonable idea of the area in which the lens may be found if lost. Upon finding the lens, first check for damage or foreign substances on the lens. Rigid or silicone lenses can be inspected immediately. A soft lens should first be placed in soaking solution prior to handling or inspection. After the soft lens has had a sufficient amount of time to absorb the solution, re-hydrate and soften, it may be checked for any possible damage. A rule of thumb for re-hydration of a soft lens is to soak the lens 10 minutes for every hour that it has been out of the eye or storage solution. You should expect the lens to be as soft and pliable as it normally would be when it is removed

from the soaking solution after overnight storage.

After inspecting the lens it should be cleaned and disinfected in the normal manner that it would be treated at the end of a day of wear. Do not put a lost lens back into the eye until it has gone through the usual cleaning and soaking process. Be sure to inspect the lens once again before reinserting on the eye. Most of the time the lens will survive temporary loss and is fine to wear again.

## INFECTION

It is always necessary to see a doctor whenever the child's eye become red, painful, or develops discharge unless the condition goes away shortly after the contact lens is removed. If the eye does return to normal, clean the lens again and try reinsertion. If the red eye recurs, contact your eye doctor.

# Visual Impairment in School Life

## *Physical Education and the Child who is Visually Impaired*

*By Dr. Carol Farrenkopf  
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Should children who are visually impaired be allowed to participate in physical education class? Of course! Keeping active is essential to a child's health and well-being. However, sometimes school personnel assume that a child with low vision or a child that is totally blind cannot participate in gym class because he/she is visually impaired. With the assistance of specially trained teachers of students who are visually impaired and in some cases, other professionals working in the field of visual impairment (e.g., CNIB), schools can learn how to develop, accommodate, or modify the physical education program to meet the child's needs.

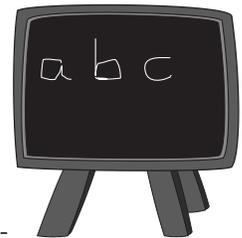
At some point early in the school year, a brief assessment of the child's physical ability should be assessed. This is best done by the physical education teacher (who has the expertise in physical development) in conjunction with the teacher of students who are visually impaired. If the child has additional physical disabilities, a physical therapist should also be included in the assessment. All children should be part of an active physical education program at school: it is inappropriate for the child who is visually impaired to be the scorekeeper all of the time or to sit on the sidelines while the rest of the class plays.

### **Prerequisite Skill Development**

If a child cannot throw or catch a ball, he/she will not be able to participate in a game of catch with classmates. Giving the child the opportunity to learn how to catch a ball (with or without a bell or some other sound-producing device inside the ball) in isolation, with direct 1:1 instruction by the vision specialist enables the child to perfect the skill prior to playing the game. Consequently, the child will likely have a more positive experience playing catch with his/her peers because he won't be seen as the child who can't catch a ball.

Another essential component to successful integration in physical education class is for the child to understand the rules of the

game. Sighted children can often pick up the rules simply by watching a game. However, children who are visually impaired need to have someone explain the rules and accompanying movements to them before participating in the game.



Finally, it is very important that the child build strength and endurance. For example, if a child with low vision quickly runs out of breath (keeping in mind that there are no other health considerations) and cannot endure jogging once around the track, playing a game of soccer will be difficult for the child. Parents can play a big part in helping their child develop the strength and endurance needed by encouraging physical activity at home after school and on the weekends. Taking family walks through the neighbourhood, hiking in the woods, cycling on a tandem bike or individually on a bike path, swimming at the local pool, or exercising at a fitness centre or in the basement will benefit everyone, not just the child who is visually impaired.

### **Modification of the Environment, of Teaching Techniques and Strategies, and of Activities**

In order for children who are visually impaired to participate as fully as possible in gym class, some modifications may be necessary. Many of these modifications can be made simply by applying coloured paint, tape, and other tactile materials or by ordering specially-made equipment through various suppliers.

- Environmental modifications are physical modifications made to gym equipment, materials, or the field of play. Brailled or enlarged print rules and score cards are examples of modified materials. Modified gym equipment can include things like bright orange base-markers, beeping balls, soft balls, and Velcro mitts (to catch a tennis ball). Modified fields of play can include using florescent pylons or tactile tape on the floor to indicate where the boundaries are.
- Teaching techniques and strategies can be modified so that the child who is visually impaired can benefit from the instruction. Students who are totally blind will require physical guidance, tactile modeling, and verbal direction through the movements of certain activities. For example, teaching a blind

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# Visual Impairment in School Life

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child to skip with a rope is a complicated sequence of movements. The child will need to be physically guided through each step (hold rope ends in both hands, place midpoint of the rope on the floor, step over the rope so it's behind the heels, swing both arms backward in circular motion over head, etc.). The vision teacher will likely have to model certain movements for the child (e.g., swinging the rope over his/her head) by allowing the child to feel the teacher's arms as the teacher performs the action. Finally, the teacher will have to provide constant verbal feedback and directions to the student so he/she knows what's happening and what to do next. For students with low vision, the same techniques described above can be used in addition to visual modeling by the teacher and/or peers. That is, the child can move closer to the teacher to observe the action at a distance he/she can see. The teacher can also reduce the speed with which the action is performed so the student with low vision can maintain visual contact with the teacher at all times.

- Modifications of activities allows children who are visually impaired to participate meaningfully in activities that might otherwise be unsafe or difficult to engage in without modification. For example, if a student playing in a game of soccer becomes disoriented, the referee can yell out a special command or blow a whistle three times, stop the game, and reorient the student so he/she doesn't get hurt. Or, a student running down the ramp to perform the long-jump runs off-course and is stopped by the teacher before making the leap into the sand-pit.

Listed below are some common strategies used with children who are visually impaired that enables them to participate in physical education activities. As long as the child is physically able to participate in the activity and the prerequisite skills needed for the activity are present, students who are visually impaired should be able to participate in most, if not all, of the physical education program.

- When running, the use of a sighted

guide or peer is often the easiest accommodation. Students may also choose to run toward a sound source independently or to use the tether method of holding one end of a rope held by a sighted runner.

- When playing field games such as shot put, standing long jump, or running long jump, tactile ground markers that help the student figure out where to place his/her feet will be helpful. Also, verbal guidance by a sighted peer or teacher will help the student orient him/herself before following through on an action (e.g., throwing the shot put in the right direction).
- When participating in relay games, students who are visually impaired need to preview the relay course before going through it with a peer or independently. Verbal and physical prompts will help keep the student on track.
- When participating in a baseball game, the rules of the game may need to be modified, as will the equipment and the pace of the game. A batting tee or t-ball stand may be used rather than having a pitcher pitch the baseball. The student may also throw the ball (rather than hitting it with a bat), use a beeping baseball or an over-sized brightly coloured ball. Orange pylons can be used instead of the standard, flat bases.
- When participating in swimming, lane ropes can be used as a guide for the student to swim next to. If there are no lane ropes, swimming in the lane next to the edge of the pool is sufficient. A tapper (long pole with a sponge on the end) can be used for those students who swim competitively—a sighted person at each end of the pool gently taps the student with the tapper to indicate where the end of the pool is so he/she can flip at the appropriate moment.
- When participating in aerobics, students generally require few modifications of the movements once they have learned them. Physical guidance, tactile modeling, visual modeling, and verbal guidance are techniques that can be used to teach individual aerobic moves. It is important that the verbal directions given by the instructor are clear enough for the student to understand what type of movement should happen next (e.g.,

left leg kick forward). Orientation to the workout space is also important, as the student needs to know where he/she is relative to others working out and to the instructor.

- When wrestling, students who are visually impaired are on a level playing field with sighted peers. Wrestlers start the match in physical contact with each other and remain in contact with each other until the end of the match. As long as the wrestlers are wrestling on a mat, the student who is visually impaired will be able to feel when he/she has moved out of bounds.

## Considerations:

Although the goal is to have all children participate in physical education classes, it is sometimes unsafe or impractical for the child who is visually impaired to participate in an activity. In such cases, the student should be included in discussions as to how he/she can participate safely. Or, alternate modifications can be made to the activity that the entire class can enjoy that includes the child who is visually impaired. For example, instead of playing hockey, the class can play goalball (a popular blind sport where sighted players wear blindfolds). Finally, the student can participate in a parallel activity that is similar to what the other students are doing.

It should be noted that sometimes, the ophthalmologist may recommend that certain activities be avoided for fear of possible damage to the student's vision. For example, if a child has had a retinal tear repaired, a sudden, hard blow to the head might detach the retina again. Parents and teachers are urged to err on the side of caution and ask the child's ophthalmologist which gym activities pose a risk to the child's vision.

*\* Summarized from: Farrenkopf, C. & McGregor, D. (2000). Chapter 12: Physical Education and Health. In A.J. Koenig and M.C. Holbrook (Eds.), Foundations of Education (2nd Ed.), Volume II, Instructional Strategies for Teaching Children and Youths with Visual Impairments. New York: AFB Press.*

## Pharmacy Corner

### *Acetazolamide and Methazolamide*

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**A**cetazolamide (Diamox) and Methazolamide (Neptazane) are anti-glaucoma drugs called carbonic anhydrase inhibitors. When given by mouth or injection, they cause a reduction in eye pressure.

The front of the eye is filled with a watery fluid called aqueous humor. Aqueous humor is produced by tissue inside the eye (ciliary epithelium) with the help of a protein called carbonic anhydrase. Accumulation of aqueous humor (either due to abnormal drainage or overproduction) causes elevation in eye pressure (glaucoma). The major goal in glaucoma therapy is to keep the eye pressure in a range that permits optimal functioning of the eye. Eye pressure may be lowered medically by eye drops or pills that either assist the outflow of aqueous humor from the eye, or decrease the rate at which it is made. Diamox and Neptazane reduce the eye pressure by decreasing the production of aqueous humor. They do this by blocking the action of carbonic anhydrase.

Since their use may be associated with significant side effects especially in adults, Diamox and Neptazane are used cautiously in children with careful attention to dosing. These drugs are prescribed according to the child's body weight. Oral Diamox is usually given three or four times daily. Diamox can be made into syrup for children who do not swallow pills. Neptazane is a smaller pill with a longer duration of action, and is administered two or three times daily. Diamox also comes in a slow release form (Diamox Sequels) that requires only twice daily dosing. However, the pills are larger (harder to swallow) as is the dose per pill thus making it impractical for younger children.

When given by mouth, it takes about 30-90 minutes for peak effect on eye pressure. For more rapid onset of action, Diamox is given intravenously, following which the eye pressure drops within 15 minutes. If a dose is missed, it is advisable to give it as soon as possible. However, if it is time for the next dose, it is better simply to resume the medication schedule and not to double-up on dosage.

Adults can have serious side effects from Diamox and Neptazane, including a reduction in blood cells, kidney stones, blood electrolyte imbalances, confusion, and depression. The main side effects of Diamox in children include nausea/vomiting, tiredness, and loss of appetite. Less

commonly children may experience diarrhea, bedwetting, more frequent urination, and tingling and numbness of hands and feet. These side effects tend to lessen with continued use. Taking the medication with food may help minimize these problems. The inhibition of carbonic anhydrase can affect the blood chemistry in the body. In particular, it may result in loss of potassium. This effect is worse in children with preexisting kidney or liver disease, or children on other drugs which also affect potassium levels, such as steroids, drugs used for heart failure or hypertension. Caution and monitoring is therefore important while using carbonic anhydrase inhibitors in such children. These drugs contain a sulfur molecule. Therefore they are avoided, or used only with careful monitoring, in patients with sulfa allergy. Patients with sickle cell disease or related problems with their blood hemoglobin may also react adversely to these drugs. Although we have used Diamox safely in children for many years of their life, rarely growth can be adversely affected. All side effects discussed here are generally milder with Neptazane.

A general principle in glaucoma therapy is to use as little medication with the least amount of risk as possible, to achieve the desired eye pressure. Treatment is therefore always started with eye drops. Pills like Diamox or Neptazane are used when eye drops alone are unable to keep the eye pressure at the desired level. But when they are called upon, these are very good agents in the treatment of glaucoma.

## The Vision Therapy Debate

*The American Academy of Pediatrics (AAP), American Academy of Ophthalmology (AAO), and American Association for Pediatric Ophthalmology and Strabismus (AAPOS) published a joint statement, which you can find at*

***<http://www.aap.org/policy/re9825.html>***

*Also from the Institute for Clinical Systems Improvement  
***<http://www.icsi.org/knowledge/detail.asp?catID=107&itemID=753>****

# Parents as Advocates ... Not Adversaries

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Someone once stated that "it is very difficult to remain angry with someone who is being reasonable." Although I do not recall who made that statement, the reality and accuracy of it have consistently been confirmed in my observations and involvement with parents and staff at The Hospital for Sick Children.

According to the dictionary, an adversary is an opponent, but an advocate is a person who pleads for, or speaks up for another - a vocal supporter.

E.g. a parent who speaks up for his or her child.

By its very definition, advocacy is dependent on communication, which can be either the glue that holds the world together, or the gunpowder that blows the world apart. The more effective the communication is, the more effective the advocacy will be.

The communication process involves three basic components as follows:

- ☒ INTENT
- ☒ CONTENT
- ☒ IMPACT

- INTENT - the information or message which you intend to convey to another person.
- CONTENT - what you actually say, both verbally and in body-language.
- IMPACT - the effect or result of your message, on the recipient

When the content of our message matches our intent, that is direct communication.

When we are uncomfortable about something we want to say (usually because we

are anxious about the response we will receive), we often tend to dilute or disguise the real message. This is indirect or evasive communication, and it often results in the recipient not 'getting the message' which was intended. For this reason, when we have something that we clearly want another person (e.g. doctor) to know about what we are thinking or feeling, it is important to prepare our thoughts before speaking, by :

- being clear about what it is that we want the person to know
- being thoughtful about how to convey this fact and/or feeling

## *Aim for an 'A'*

- ☒ ATTITUDE ...
- ☒ APPROACH ...
- ☒ ACTION ...
- ☒ APPRECIATE ...

### **ATTITUDE:**

- be committed to a constructive process in interacting with your child's team members
- separate your feelings from the facts e.g. "Is my frustration a generalized response to my child's condition, or is it a focused response to a specific situation?"

### **APPROACH:**

- be clear about what your concerns, needs and questions are
- be an advocate for your child, not an adversary of your child's team

Sometimes when we are anxious, frustrated, or upset about something, we may think that we are being advocates in what we are trying to achieve, but if the approach we take is adversarial, we will be experienced as being adversaries - and we will likely be responded to that way. Because most human beings do not feel like co-operating with someone who is criticizing or 'attacking' them, their focus and energy are then used defensively for themselves rather than being used constructively for addressing the concern. Ask yourself... "If I was on the receiving end of my own behavior or approach toward this person right now, how would I feel like reacting or responding?" , and then adjust your approach accordingly if necessary.

### **ACTION:**

- participate as a partner in the solution - when expressing a concern or dissatisfaction,

be specific about why this is a concern and offer suggestions for changes or actions which you think would help to improve the situation

- present accurate information (avoid overstating or understating details of a concern)
- partialize the concern into manageable amounts, and be specific; rather than generalizing the problem by stating that "communication is the problem", state what has prompted you to feel this way e.g. "the staff doctor and the resident give me conflicting information about my child, and now I don't know what is accurate."
- prepare ahead by sending/faxing your prioritized concerns/questions (and reasons for them) to the doctor's office ahead of your child's appointment with a note to say that you are looking forward to discussing these points at the next appointment on [date]; bring your own copy of this same documentation to the appointment, as this helps to maintain focus during the appointment, and it shows that you are participating as a partner in sharing the responsibility for the solution

### **APPRECIATE:**

- express appreciation for what has been done and/or for what you have been told will be done to address your concerns

### **In summary...**

- be Respectful
- be Reasonable
- be Ready

### ***Build bridges, not walls.***

Bridges can provide a means for people and purposes to come together.

Walls can separate and provide barriers.

In your interactions with you child's team, be advocates building bridges, rather than adversaries building walls. Be committed to making each of the smooth and the rough stones along the path of your child's course of treatment, stepping stones in a bridge that brings you together rather than building blocks in a wall that separates you from your partners in your child's team.

***"Coming together is a beginning; keeping together is progress; working together is success."***

- (Henry Ford)

## PATCHING PANEL

### Alternatives to Patching

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As discussed many times in this newsletter, perhaps the most difficult part of treating a child with glaucoma or cataract, is the patching required to treat amblyopia ("lazy vision" due to the brain preferring the better eye).

Children resist having their good eye covered. Behavioral intervention is sometimes useful. Consult with your pediatrician or family doctor to see what techniques might be useful. For example, positive reinforcement works much greater than negative reinforcement. Your use of the PGCFA website and newsletters to hear how other parents have dealt with the difficult behavior issues involved in patching is highly recommended.

In addition, patches sometimes cause skin irritation and allergic reactions. Are there other options that can be used instead of patching?

1. There have recently been papers published in the medical literature talking about the use of eyedrops to treat amblyopia. The drop is called atropine. They dilate the pupil of the good eye and cause blurring of vision in that eye. This is the experience that most adults are accustomed to when they get their pupil dilating drops at their routine eye doctor appointments. Various dosing regimens can be used from 1 or 2 doses daily to 1 dose weekly. Atropine does sting and sometimes the stinging of the atropine is enough to con-

vince the child to wear the patch: the patch is the "lesser of two evils"! The research shows that atropine can work as effectively as patching in selected children (although atropine takes longer to work than patching). Atropine drops will only work in children where the drops are able to blur the good eye worse than the vision in the bad eye. In other words, even if the good eye is blurred, if the vision in the bad eye is still worse than the blurred good eye with atropine, then the brain will still prefer to use the good eye even with the atropine blurring. In order for amblyopia treatment to work, the brain must start to prefer the bad eye and ignore the good eye temporarily. In addition, atropine only works on eyes that have a lens. If the child has had cataract surgery in both eyes and has amblyopia in one eye, then atropine treatment will not work for the good eye. Lastly, one must consider that atropine will make glaucoma worse in some children if the good eye also has glaucoma. Careful planning with the help of your eye doctor is important in deciding whether atropine eye-drops will work for your child in the treatment of amblyopia.

2. Some parents have found that restricting the motion of a child's arms so they cannot take off the patch is useful. In infants, simply putting mittens or a sock over the hands might prevent them from being able to peel off their patch. In toddlers and older children, some restricting device at the elbow may be useful so that the child cannot bend their arms to let their hands get to their face. The simplest technique is water wing flotation devices usually placed around the upper arm for swimming. These can be inflated over the elbows. More rigorous treatments include firm restraints built into special vests or jackets (or even homemade) to keep the elbows from bending. I am aware of one situation in

which a child was given casts on both arms! One must be concerned that at some level the restriction becomes too great and too disruptive of the child's life style to be justified. Careful consideration must be given to these techniques when consulting with your eye doctor.

3. Sometimes, simply making the patch stick better to the face can solve the problem. This can be done through a variety of dressings or tape applied over the patch to the skin. Very thin adhesive dressing such as Tegaderm® (or other similar products) is very difficult to peel off the skin. Some doctors have even considered stitching the patch to the face or using glue to stick the patch to the face. A mild form of glue that is commonly used in medicine is called Tincture of Benzoin. This helps the patch stick on better as well. Be careful not to get glue preparations into the eye itself.

4. In selected children a black opaque contact lens can be placed in the good eye. One must be careful that there are no other risk factors for contact lens use in the good eye. In addition, all contact lenses have some risk (albeit usually very small risks) but these must be considered when placing a child's only good eye at risk.

Sometimes, patching is just too disruptive for the family and child, or leaves the child with such bad vision in their bad eye that they cannot function normally. In these situations, one must consider alternate schedules of patching (e.g. patching only when not in school) or occasionally abandon patching altogether. There are indeed times when the stress and emotional effects of patching are outweighed by the benefits for selected families and selected children. However, the consideration to stop patching must be carefully discussed with your eye doctor.

### DOG GUIDE VERSES CANE; WHICH IS BETTER?

Karen Watts  
Orientation and Mobility Instructor  
Toronto District CNIB

Depending on their needs some people would find benefits from a dog guide and others from a cane. The long cane acts as a probe and identifies obstacles that are in the person's line of travel. When an unexpected obstacle is contacted with the cane, the person must investigate whether it is safe to proceed beyond the obstacle; this can often make cane travel a slow process. In inclement weather the cane may not be able to contact certain landmarks such as a grass border, and this can become a disorienting experience for some travelers. When crossing an intersection a person must follow a straight line of travel, line up with the traffic audibly, and cross the street by analyzing the traffic flow. If the person veers they must learn skills to recover from veering. With a guide dog the person will follow the straight line of travel, line up with the traffic audibly, analyze the traffic flow and command the dog to go forward with the appropriate surge of traffic. As long as the person

was correctly lined up towards the opposite curb, the dog will travel straight towards that curb, avoiding veering into cars or obstacles in the path.

A guide dog from a recognized guide dog school is trained to avoid obstacles in the path of the person who is blind or visually impaired. A person who uses a guide dog must be an independent traveler and be able to travel with a cane safely and independently. A guide dog must be treated differently than a pet dog. While in harness a dog should not be distracted, including being petted by passers by. A common misconception is that a dog guide, actually guides the person who is blind or visually impaired, however, it is actually the person who directs the dog by using a basic set of commands such as forward, left, right. A possible disadvantage of dog guide travel would be the upkeep and everyday needs of the dog. It is important that a person has cane skills to safely travel when the dog may be sick. The person who has a guide dog must be active to keep the dog in shape and working effectively. If the owner does not uphold the proper training, the dog could become a safety hazard for the person. The choice of guide dog or cane very much depends on the travel needs and lifestyle of the blind person.

## World View

### Seva Canada

Maggie Westhaver, Executive Director

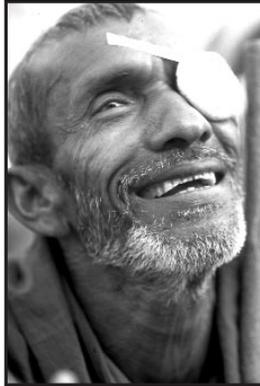
For over two decades Seva Canada Society ([www.seva.ca](http://www.seva.ca)) has contributed to the reduction of avoidable blindness in developing countries such as India, Nepal, and Tibet by supporting locally-run programs. Recently Tanzania became a partner country. Seva is guided by principles that respect cultural, racial, spiritual, ethnic, and other forms of diversity and is driven by the desire to reduce suffering due to blindness through compassionate action.

Over the years Seva Canada has developed unique strengths. These include special attention to projects that help balance eye care delivery equally for male and female (including male and female children). There can be barriers that prevent females from gaining equal access to eye care. To create equity, 64% of people seen have to be female. Without carefully tested approaches to overcome these barriers females will continue to lag behind, a situation that simply isn't ethically acceptable – or helpful at all to the family and community.

Another strength of Seva is in the area of outcome assessment and evaluation, which means checking to see if the work being done is successful. Seva considers follow-up care and evaluation essential. Seva's work is sustainable not relief-based. Systems that help to ensure the work being done is in fact giving people excellent visual outcomes after surgery are high priorities, as are all the blindness prevention programs.

The gender work and evaluation work together create a very interesting network full of possibilities and opportunities for Seva and its various partners. Dr. Nirmalan's work is one example of this. Dr. Praveen Nirmalan in south India heads up The Katiapatti Pediatric Eye Evaluation Project which provided baseline data of children aged 15 or younger in south India. The American Journal of Ophthalmology has accepted an article on this project for publication. Dr. Nirmalan will be in Canada this spring, giving presentations and speaking to the media.

One of our board members, Dr. John Pratt-Johnson, a retired pediatric ophthalmologist is a foremost advocate to vaccinate our



world's children against rubella. This is what he has to say about the continuing problem of rubella in developing nations:

*The implementation of the rubella vaccine in Canada soon after it was available in 1969 was so effective that rubella is rarely seen in Canada or the USA anymore. Although rubella itself is a mild illness with a rash and low fever-like symptoms, its significance becomes horribly apparent if a woman is infected within the first 3 months of pregnancy. There is a 90% chance that the fetus will be infected too. 50% of children are born with cataracts, 80% are deaf, 50% have heart abnormalities, 50% have mental abnormalities from infection of the brain, 10% develop congenital glaucoma and there is a 400% increase in the number of children with crossed eyes and other eye alignment problems. So it is a very serious problem we can't ignore.*

*One of the many problems is that most developing countries need money and help to implement an effective immunization program. Seva has recently made a breakthrough in Nepal and soon we may see nation-wide immunization in that country. Since the vaccine is 98% effective in conveying lifelong immunity we are very hopeful that we can make a positive impact on the future of those children and others who need our help.*

All of this said, the staggering numbers of untreated cataracts (millions and millions of them) remain the highest priority for all of us because among all chronic eye diseases it is the one that is the easiest to correct and usually has very good outcomes. Furthermore, the cost is low – currently \$50 per surgery (or less) is what it takes to save someone's sight. It is truly amazing!

In Canada, Seva has an Active Citizenship project which invites learners of Grades 6-11 to combine compassion, initiative and critical thinking in their engagement with effective social action.

Our Canadian public engagement show, Himalayan Visions ([www.seva.ca/tour](http://www.seva.ca/tour)) is currently a huge success.

If you would like to discuss our programs with us or receive a free Gifts of Service catalogue, please call or email our office in Vancouver ([admin@seva.ca](mailto:admin@seva.ca)/ toll free 1-877-460-6622). The catalogue is a cherished Seva tradition and hundreds of people use it every year. It is filled with alternative gift ideas that give sight and echo your compassion.

## Advance Notice

The Annual General Meeting (AGM) and Education Day for the Pediatric Glaucoma and Cataract Family Association will take place in Toronto at The Hospital for Sick Children on Saturday, November 15, 2003 from 9:00 a.m. until 3:00 p.m. and is open to the public. The day will feature a series of presentations from doctors and health care professionals on a variety of subjects related to pediatric glaucoma, pediatric cataract, and vision in childhood. Registration begins at 9:00 a.m. followed by a brief AGM. Lunch is included. Registration is \$10.00 per person. Cheques, payable to PGCFa, can be sent to Leslie Mowat, 9 Muirhead Crescent, Brantford, Ontario N3R 7S5, or pay by Visa on the web site [www.pgcf.org](http://www.pgcf.org). For more information or registration please contact Leslie Mowat (519) 753-8840 or [lesliemowat@rogers.com](mailto:lesliemowat@rogers.com).

## PGCFA Research Award 2003



The PGCFA was proud to present the second annual award for the Best Presentation on Pediatric Glaucoma or Cataract at the 2003 meeting of the American Association of Pediatric Ophthalmology and Strabismus (AAPOS). This is the largest and most important meeting in the field which each year brings together researchers from around the world to present their work. After the AAPOS Program Committee selects which papers deserve presentation from the hundreds of applications they receive, the PGCFA Scientific Advisory Board receives an abstract synopsis of each accepted paper. These are submitted to a review committee convened by our SAB. If a reviewer is one of the candidate authors they must remove themselves from the committee. The top 5 abstracts are selected and the authors invited to submit their entire manuscript for review. From that group a winner is chosen: clearly the "cream of the crop"!

The PGCFA would like to thank the international reviewers who volunteered their time to select our winner.

*David Walton, MD (USA)*

*Sharon Freedman, MD (USA)*

*Terri Young, MD (USA)*

*Ronald Keech, MD (USA)*

*Monte Del Monte, MD (USA)*

*Edward Wilson, MD (USA)*

*Gregg Lueder, MD (USA)*

*Ken Nischal, MD (England)*

*Alex V. Levin, MD (Canada)*

This year's winner was "Opacification of the Visual Axis after Cataract Surgery and Single Acrylic Intraocular Lens Implantation in the First Year-of-Life" authored by Drs. Rupal H. Trivedi,

M. Edward Wilson Jr., Garima Lal, M. Millicent Peterseim, and Luanna R. Bartholomew from Charleston, South Carolina. The lead author receives a monetary prize and an invitation to present to the PGCFA. Congratulations to the winners!



## AcrySof Intraocular Lens Implantation in the First Year-of-Life

*Rupal H. Trivedi, M.D., M. Edward Wilson, Jr., M.D.,  
Garima Lal, M.D.,*

*M. Millicent Peterseim, M.D., Luanna R. Bartholomew, Ph.D.*

*Miles Center for Pediatric Ophthalmology, Storm Eye Institute,  
Department of Ophthalmology, Medical University of South  
Carolina, Charleston, SC.*

*Supported in part by an unrestricted grant to MUSC-SEI from  
Research to Prevent Blindness, Inc, New York, NY.*

*The authors have no financial or proprietary interest in any product  
mentioned herein.*

It is very important to provide some kind of artificial focusing device (See *MORIN* information issue January, 2001) following cataract surgery to develop visual acuity in children. The two most common ways to provide this focus is with contact lens use or intraocular lens (IOL) implantation. Use of IOLs in children has gradually increased and is now widely accepted in children beyond their second birthday. However, the use of IOLs in children younger than 2 years-of-age is still controversial. Increases in intraoperative difficulties, as well as a higher rate of postoperative complications, have been the issues of primary concern. One of the most common complications following cataract surgery in children is the development of scarring/ cloudiness of the line of vision is sometimes called a "secondary cataract" or an "after-cataract" even though there is not really a new cataract.

At the 2003 pediatric meeting in Hawaii, We reviewed our experience with acrylic IOL implantation in the first year-of-life. These newer acrylic IOL materials now available allow the IOL to be folded before entry into the eye and then unfolded within the eye. This allows the pediatric eye surgeon to use a smaller incision. Thirty eyes of 21 pediatric patients were included in our study. In each case, a foldable, acrylic IOL was implanted at the time of surgery. One third of these eyes (33.3%) developed a "secondary cataract" that required surgical intervention to remove it. The occurrence of a "secondary cataract" was 7.7-times greater when the size of the eye was shorter than normal for age and had one or more other eye abnormalities and 2.2-times greater when surgery was performed within the first 6 months-of-life.

Scarring of the pupil to the IOL was seen in 26.7% of eyes but did not require surgery for correction. Two eyes (6.7%) developed glaucoma. Only one of these eyes required glaucoma surgery.

The use of IOLs in the first year-of-life remains controversial. "Secondary cataract" is still a significant complication. It is very important that these young patients have regular follow-up visits with their eye doctor, to detect any unwanted complications following surgery. A multicenter, randomized clinical trial in the U.S. is being planned to better understand to these issues.

## Recent Events

### Boston workshop 2003

Liz McManus

A very successful workshop was hosted in Boston, USA by Dr Walton, The Children's Glaucoma Foundation and the PGCFA in April 2003. In the morning Dr. Sharon Freedman from Duke (and a member of our Scientific Advisory Board) spoke about the medical care of childhood glaucoma and gave some tips for parents, Beth Arnold, a social worker at the Massachusetts Eye and Ear Infirmary gave a useful list of resources for parents, Dr. Steven Kane, from Columbia University in New York talked about trabeculectomy and Susan Murray-Campbell shared her family story (see page 12 of this newsletter).

After lunch Dr. Ching-Lin Ho, an ophthalmologist from Singapore now studying pediatric glaucoma with Dr. Walton, talked about goniotomy surgery, Dr. Alex Levin, Chair of the PGCFA Scientific Advisor Board, spoke about glaucoma implant (tube) surgery. The

winners of the 2002 PGCFA award were represented by Dr. David Weakley, from Texas, who talked about their work on eye growth in aphakic children (see MORINformation summer 2002). The PGCFA was represented by our Co-Chairs Mike and Dawn Atwell, who spoke about the history and future of the organisation and I was there to talk about the newsletter. The day wrapped up with a question and answer session.

Around 55 parents and grandparents participated in the day and found it very valuable



indeed. Much of the information shared on the day will appear in future issues of MORINformation.

### Family picnic – Toronto June 2003

The day, June 14, 2003, could not have been nicer for the 4th Annual PGCFA Picnic. The clouds of early morning gave way to bright sunshine for a fun filled day. It was great to see old friends from past picnics, but even better to see new faces at the event. The food was fantastic, the soccer game exciting and a great time was had by all. The crowd consisted of about 30 people including lots of kids. It was a wonder why the food didn't taste like the bubbles that kept floating around (thanks kids). A big thank you to the organizers, burger flippers, toy givers etc. for another successful day. See you same weekend next year!

Dawn Atwell



## Book Review

### The Eye Book

by Dr. Seuss writing as Theo. LeSieg,

illustrated by Joe Mathieu

ISBN 0-375-81240-7

In the typical rhythmic and rhyming style of Dr. Seuss this book is a celebration of sight, listing a host of everyday things we can see. The illustrations of the boy and the rabbit are comical (though the one where they are looking at the sun could be used to initiate a discussion about not doing this). Available in various formats (board, paperback and hardback), this book is suitable for adults reading aloud to the very young as well as beginner readers to tackle themselves. The final line, "Hooray, hooray, hooray ... for eyes!" is one to be remembered.

Liz McManus.

### Ten tips for being a successful childhood glaucoma parent

Extracts from Susan Murray-Campbell's story

Presented at the Children's Glaucoma Foundation/PGCFA Boston Workshop

April 12, 2003

Susan and Todd have three sons, two of whom have glaucoma. Their full story can be found on our web site at [www.pgcf.org](http://www.pgcf.org).

**One:** Align yourself with a successful childhood glaucoma doctor. Our ophthalmologist, Dr Walton, shows brilliance, tirelessness, concern, and tenderness toward his patients. When you're in that exam room, you get the feeling that your child is the focus of all his efforts, and that no one's outcome is as important. That kind of dedication must come at great personal cost, and witnessing it inspires me to do the best I possibly can as a parent. Dr. Walton demands no more of anyone else than he does of himself, so the least I can do

is make sure my kid wears a lousy patch for an hour or two a day!

**Two:** Identify your allies. Seek out people who will support you in fighting for the success of your child. These people may include pediatricians, optometrists, therapists, social workers, friends, and family members. Our own list includes my mother (a double bonus because she's also a registered nurse), the teachers at our sons' Montessori school, and Dr Walton's assistant Karen Sicher, who with

*continued on page 11*

patience and humor (and candy dispensers) sees to it that our family finds a way to enjoy every visit to Dr. Walton's office.

**Three:** Supply the extra love and attention that these children require. When you're handed a post-surgical four-week old infant with shields over his eyes and hear that you should keep his head elevated and prevent him from crying for a couple of weeks, the request may seem daunting, indeed. But I found that the two went together quite well: the baby was both upright and quiet as long as he stayed attached to a loving adult's shoulder. If you are a mother who has the option of continuing to breastfeed, I would recommend it, as it offers familiar and reassuring comfort for a baby recovering from the trauma of surgery. If your child is a little older and you're offered the opportunity to accompany her to the operating room for the anesthesia procedure, go – your anxiety at seeing her there pales in comparison to her need for your reassuring presence in that scary environment.

**Four:** Keep good written records of your child's progress. Everything's on file in your doctor's office, of course, but we've found it helpful to have our own notes at home. For the first few months of Seth's life, we kept calendar pages posted over his changing table so that we could keep track of which eye was receiving which drops on which days of the week. Take your notes with you to the baby's regular pediatric checkups so that you can discuss any concerns. Be the clearinghouse for your child's information, rather than relying on behind-the-scenes file shuffling between medical offices. (Now, I'm not saying that you need to produce a three-color graphic representation of the progression of your child's intraocular pressures before and after surgery, as my husband did – it's just his way of coping!)

**Five:** Try to see the humor. There are lots of times, frankly, when there isn't any. So seize the opportunity to loosen up when you can. I recall one day when Seth was struggling with that first trabeculectomy. Dr. Walton had put some yellow-green drops into his eyes to better inspect a corneal abrasion that had developed. Seth napped on the way home, and when he awoke we saw that the drops had saturated the blue opacity of his irises to give his big eyes an absolutely unearthly green glow. While my mother and I found this really kind of cool, Tod was not at all amused, and felt uncomfortable displaying the baby in public. However, we'd been cooped up in the hospital for days and knew we'd be back there soon, so Mom and I won out and the bunch of us went to a local café for dinner. You know what? No one noticed.

**Six:** Don't neglect your other relationships. Good friends will understand why they don't see you anymore, at least for the time being; but if you have a spouse or other children, they must not be ignored.' It's hard to find the time and energy to keep the lines of communication open, but do your best. It may seem like more trouble that it's worth to dress up (or even shower) and go out to dinner or a movie, but it's important to take a break from the stress surrounding your child's treatment for just a few hours. The summer that Seth was born, our annual week-long stay at a cabin in the New Hampshire mountains shrank to a few days squeezed between surgeries, but Dr. Walton insisted that we go. Sometime' it's a real effort to give your life some semblance of normality, but it's important to try.

**Seven:** Count your blessings. Call them lucky breaks if you like, but I found a half-dozen in Seth's first hours alone. First, Seth was first. No siblings to care for or to find care for while we shuttled around seeking medical advice and treatment. Second, he was born on his due-date: if he had been premature, there would likely have been other complications, and if he had been allowed to stay in utero for up to two more weeks, irreparable damage may have occurred to his optic nerves by the time he was delivered. Third, his eyes showed signs of opacity that may not have shown up in some babies with similar pressures, calling attention to an insidious condition; and fourth, my obstetrics team was sharp enough to raise concerns so that he could be treated immediately. Finally, we live in the back yard of Boston, a city that is home to some of the finest medical institutions in the world, and the practice of Dr. Walton. Most important, I was grateful that Seth's condition was not life threatening. Although he happened to have been born with an unrelated laryngeal defect that affected his breathing for a while, it was assumed that he would outgrow it. There are complicated diseases and congenital syndromes of which glaucoma is just one aspect, so I feel fortunate that with us, the eyes are as far as it goes.

Sometimes' it's all in how you look at it. For generations, Tod's family has made a special cause of the needs of the visually impaired, with no occurrence of blindness in their own history. Tod's mother is a registered orthoptist who has served as vice president of the board of the Maryland Society for the Prevention of Blindness, and his late father served as president of the board of the Maryland School for the Blind for nearly two decades. Tod at first found it bitterly ironic that his father's namesake and first grandson, our Seth, should be born with severe congenital glaucoma. I decided that, perhaps, the good work that

Seth's grandfather had done on the behalf of others struggling with blindness might have indirectly influenced Seth's own positive outcome; a kind of posthumous gift to his unknown grandson.

Some of you may think that musing on the "what ifs" of a situation are a dubious use of time, but I find it to be a valuable coping mechanism. There's nothing like a trip to the pediatric ophthalmologist's office to put my family's position in perspective. (Currently, my heart goes out to those shell-shocked parents emerging from the examining room, grasping an infant carrier in one hand and their first bottle of bright-pink diamox in the other.)

Not only are childhood glaucoma rates significantly higher in some other parts of the world, but access to quality treatment is far more limited. It is only very recently that children born in our own country with glaucoma as persistent as Seth's had even a chance at retaining their vision. Be grateful that your children were born at this time, in this place, with you for parents.

**Eight:** Be diligent; you've signed up for life. Though it may be brought under control, your child will have glaucoma forever. You'll need to be watchful, and to have his eyes and his pressures monitored carefully. If you have concerns about how your child sees as he grows, seek out the resources to help. At the same time, try to refrain from making assumptions about what your vision-impaired child may or may not be able to do. As the saying goes, "Prepare the child for the path; not the path for the child."

**Nine:** The "red-eye eliminator" pen. If conventional flash photographs of your child look anything like ours, this pen can really help you out when you're working on your family photo albums. Seth's pupils give our camera remarkable access to those highly reflective retinas. This particular pen is made by Fiskars; Dot Line Corporation makes one with two different-sized nibs. You can pick these up at a photo shop. They contain special blue-colored ink that filters out the redness when you dab it onto photos of your child's eyes.

**And, finally, ten:** Share your stories. I'm willing to bet that no one else living on your block has a child with congenital glaucoma; it can be a lonely existence. There can be tremendous comfort in simple company. So let's support each other and share our stories.

\* \* \*

*If you would like to share your story in the PGCEA newsletter please contact the Editor through [www.pgcf.org](http://www.pgcf.org).*

## Family Stories

### *Mackenzie's Story*

**M**y little 8 pound 11.5 oz angel was born on October 19th 1999. My pregnancy was normal. After an emergency C-section all was fine until I noticed that her right eye was not opening. "Oh, don't worry, some babies are like that" was what I heard repeatedly. The day I left for home I was told she needed to see a pediatric ophthalmologist because her right eye still refused to open and was smaller. At the time I thought all was going well – Mackenzie seemed to be following my voice with her head, but she was blind. I thought she was looking at me and smiling! Looking back, I should have seen signs but that is only because I am more educated on eye problems than I was then. She never wanted to be far from me and refused to leave my arms for months.

The pediatric ophthalmologist confirmed that both eyes had extremely dense posterior anterior cataracts, and that the right, slightly microphthalmic eye would not have vision so only recommended surgery for the left eye. I immediately asked for a second opinion. The following day I was told that my three-week-old little angel was to have bilateral cataract surgery the following day in our local area and would need contacts or glasses.

Mackenzie needed constant drops in her eyes, which she did not tolerate very well. At the same time we were investigating the possible outcome of her curled toes and apparent low set ears. All the normal testing had been done for sexually transmitted diseases two times with no avail as to why this little wonder had been born with cataracts. At this point Lowe syndrome had been ruled out as well as galactosaemia. I persisted with her drops in hopes that glaucoma and infection would be kept at bay. None-the-less I saw her pressures rise in her right eye; thankfully her left remained normal.

During this time Mackenzie was not feeding properly and it was apparent some palate problem existed. I have now found out that she has a sub-mucus cleft and one fused tooth. Alongside all of this was the constant collection of fluid in her ears. Tubes were inserted and have been a god-send to me since her hearing has been dependant on this marvel of medicine. This miracle of a girl also experiences higher muscle tone in her lower body without explanation. We had a first CT scan and all looked good. Genetics have no clues in regard to her vision or any of her other issues. We were told that some children are undiagnosed.

At approximately 18 months Mackenzie was wearing bifocal glasses and we were

monitoring her pressures carefully.

Around 26 months her pressure became so high in the right eye that it was slightly damaged so we inserted a tube to ease the pressure. Thankfully it worked. She became a pro at EUA's [examination under anesthesia], for they cannot come near her any other way! No Ketamine for this little fighter. Instead of relaxing her it gets her so fired up, that she runs wildly up and down the hallways annoying the many nurses that must never have had children, or forgotten.

So then we re-evaluated the situation. Mackenzie's left eye dominated her vision for the first year of life. But remember the right eye that wouldn't have vision? Well now it is so much stronger that dominance has switched and the left eye may have refused to work unless we patched. For a year we watched the vision in the left eye deteriorate with no real cause. She had lens cell regeneration but not to the extent that it would prevent vision. It was decided that it would not be removed for fear of glaucoma. I am unsure how I feel about this.

Even with patching, which she hated, it seemed to make no difference. Because I was unable to patch it was decided to remove the lens from her glasses. Her left eye had slight nastagmus and was turning into her nose. No surgeries were being discussed as I was told this was purely cosmetic, not helpful. Patching was later tolerated so I did so all the time, allowing only little breaks when I washed her hair etc. She was so terrified of the patch I had to literally sit on her arms and body and hold her head to put it on and take it off. She hated having her hair washed because of the fear of the patch. She even slept with her glasses on. If you tried to take them off she would wake up! Thank heavens she is little – I hope she won't remember any of this.

When we first went to the children's hospital I was told she had marks on her retinas that looked like 'bear tracks', though this was inconclusive. My family found out that these 'bear tracks' may indicate gastrointestinal track problems, possibly cancerous. The diagnosis was then retracted and we were told this may or may not have anything to do with Mackenzie's apparent yet undiagnosed syndrome.

With every new doctor we are asked what her diagnosis is and we have to say 'nothing'. It is infuriating and worrisome. I feel helpless and irritated. However, with every

new development, and the maintenance of the issues that already exist, this little wonder keeps a constant smile on her little face. She is absolutely amazing.

At the present time the results of the CT scan revealed no brain problem. GREAT! We are now able to patch but it is heart-breaking as she screams and tells me she cannot see. However my persistence has done well and she now refrains from pulling it off. We will visit genetics again to be tested again for Lowe syndrome and maybe further our knowledge. In the mean time Mackenzie eagerly awaits the arrival of her sunglasses!

Mackenzie has been dubbed sensory assertive and sensory defensive, which means she seeks out certain stimuli as well as avoids them so her emotions swing from daredevil to timid mouse. I was told this does not have anything to do with her vision – but I wonder. However, she still maintains her own unique balance and is the most wonderful creature on two feet. She is a very aggressive, forthcoming little girl who goes for what she wants and stops at nothing until she gets it. What a great trait!

As our story continues we will keep monitoring and praying all stays well. I wrote her story so others will see it and maybe have insight into their situation or possible insight for me on ours. Please feel free to write to me – I would love to hear from you.



*Mackenzie and Sylvia Vunak*

### PGCFA SUBCOMMITTEES

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Contact us by e-mail through the web site at [www.pgcfca.org](http://www.pgcfca.org) or at our postal address at the office c/o Dr. Levin at The Hospital for Sick Children, 555 University Avenue, Department of Ophthalmology Suite # M158, Toronto, Ontario, M5G 1X8, Canada.

Please contact us regarding anything you would like to see in the newsletter. We welcome new members to the Board or any of the subcommittees if you would like to get involved.

*Medical information and advice provided by the PGCFA or physicians acting at their request, does not represent a prescription and should not replace the information and advice given by your own physicians and other medical professionals.*

*This is a Canadian based newsletter. Comments by Canadian physicians are intended only for residents in Canada in accordance with the principles mentioned above.*

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