

CHD and Me – Talking With Adults With CHD

A concern that “heart parents” often have is how our children will do in the future. Will they be able to lead productive adult lives? Will their heart defects prevent them from doing activities or jobs that they want to do? In this edition, we feature the story of Shelagh Ross.



I was born 48 years ago in Toronto, the youngest in a family of four kids. The first clue that something was wrong was that I didn't have the strength to nurse, and I had “blue spells”. I was diagnosed in that first year with tetralogy of fallot, one of the most common congenital heart defects, made up of four related defects. I was also born with two sets of bowels and intestines and at first that was my biggest problem because I wasn't able to properly digest food and gain weight, but that was fixed with surgery early on and that part of me has been ok since.

At age two I had the Blalock-Taussig shunt as palliative surgery to help increase blood flow to the lungs (they took a major artery from my right arm and used it to re-route the blood. To this day I don't have a detectable pulse in my right arm). It was all they could do at the time and the hope was that I'd live long enough to survive open heart surgery when I was older and stronger. My parents were told I would likely have stunted growth and perhaps they were right. I'm “short” at 5'7” compared to my 5'10” sisters and 6'4” brother!

I was sick a lot with infections and I remember often being cold and tired. I'm told I used to squat a lot, which apparently kids with my heart defect do to cut off circulation

to the legs and divert the blood to the heart. I also used to get terrible headaches and my dad would hold me to his chest with my legs tucked up under me. But I think I was fairly oblivious of my problems, and like most kids I just wanted to keep up with my siblings. I think I can probably thank them for growing up “normal” because they certainly didn't coddle me.

In 1970, at age 8, I underwent nine hours of open heart surgery at Sick Kids Hospital for what was called “a complete repair”. In 1970 it was still considered very risky, with about an 80% mortality rate. The surgeons were Drs. Mustard and Trussler. The only things I really remember about Dr. Mustard is that he used to make me run up and down the stairs at the hospital before listening to my heart with a stethoscope (before the days of treadmills and stress echos) and to me he looked exactly like the wizard in the Wizard of Oz.

It says in my chart that Dr. Mustard hoped he never had another patient like me because it was the most difficult surgery he'd ever performed. I stayed at Sick Kids for about two months to recover and basically, besides the needles and being away from home, I enjoyed being treated like someone who just had something very special happen to them. Everyone was extraordinarily nice.

Life after surgery was a whole new world for me. I roared around on my bike, played tennis, became a ski instructor at 16, and in grade 13 won the Toronto schools tennis finals. I had lots of friends and did all the things normal/bad kids do, including things I shouldn't have done. I knew I had a “heart condition” but I felt normal. In addition to things heart-related, I have scoliosis, migraines, and am very allergic to peanuts; even though I'm very careful and carry an EpiPen, the peanut allergy has always been the main reason for visits to the hospital ER.

From age 18 to 33 I saw a cardiologist every year and every year he said “Things are great!” I'd been told I wouldn't be able to have kids but when I was about 19 the cardiologist said “I don't see why not”. I know now that no one really knew if it was safe or not; they just didn't know. I got pregnant at age 28 and despite not having any heart-related care besides an

ultrasound on the baby's heart, everything went smoothly and I felt really healthy and energetic. After my son, Graham, was born I thought I was fine but in retrospect I think I was unusually tired and during those first few years really struggled with fatigue and migraines. For several years we tried to have another baby but had no luck. I think it was actually a blessing because my health was great...until I was 33.

I woke up early one Victoria Day weekend in May with my heart pounding so hard my husband literally thought there was an earthquake. My son was 4. The big lilac tree outside the front door was in full bloom, along with the lily of the valley, and I was so sad to be leaving in an ambulance. I was diagnosed with ventricular tachycardia, a life-threatening heart arrhythmia. After three hours of constant pounding I was put under anesthetic and my heart was shocked back into rhythm. I woke up feeling like I'd been hit by a truck, but I was basically feeling normal again. For three weeks I underwent tests at the Toronto General Hospital, and became a patient for the first time at the adult congenital clinic – a clinic I didn't know existed. My pulmonary valve was leaky and my heart had become so enlarged that I would need a pulmonary valve replacement. This was in 1994, pre-internet, and I basically knew very little about my heart condition besides being told not “to overdo it” – whatever that meant. The lead-up to surgery was pretty scary for me and it was compounded by having to be all of a sudden away from my family. I was in the hospital for three weeks having tests and my husband and son would come to visit me and invariably my son's clothes would all be on backwards. It's funny how something that would normally bother me didn't seem important at all! (I've pretty much maintained that perspective ever since).

I was scheduled for surgery in November (six months away) but my heart rate was so low that they advanced the date and in July I had surgery for a pulmonary pig valve replacement. I stayed in the hospital for all of five days, and was at my sister's wedding two weeks later (not the life of the party but

(continued on page 10)

I was there!). About six weeks after surgery I resumed my regular activities, including tennis, bike riding, walking with my dog, and working.

Despite everything I feel very healthy and generally live a pretty ordinary life. I have my B.A. in English from U of T, have travelled a lot (including hitch-hiking across Canada, walking down and up the Grand Canyon, and backpacking through South America). I work in publishing, my son is now 20 and at university, and I generally can do most things I want to do. I have my parents, and particularly my mom, to thank for all the care she gave me through those years – and rewarding my hospital visits with an Eskimo pie from the vending machine! She's 81 and she still worries about me. If I'm lucky I will live long enough to have another surgery to replace the pulmonary valve because at some point it will wear out. I know that I'm actually "old" for a CHD patient and of course I worry about the future and sometimes struggle with anxiety. I have to make an effort to manage stress otherwise I experience arrhythmias. I try to deal with it by walking and staying busy, and working from home, for the most part, allows me to manage my own time. Our second Border Collie named April, now 9, keeps me walking fast.

In 2004 I attended an information night at the Toronto General Hospital adult cardiology clinic. Dr. Gary Webb, the director at the time, told a group of us that we needed to advocate for our own health, because the doctors and nurses couldn't do it all. Following that meeting four of us formed the **Canadian Congenital Heart Alliance** (www.cchaforlife.org), a registered non-profit whose mission it is "To improve the quality of life and health outcomes for individuals with congenital heart defects by raising awareness, providing peer support and mentoring, and advancing research." Since 2004 one of the founding members died at age 40 of the same heart defect as mine, and a particularly dynamic executive member died following a heart transplant at age 34. Many others who help have had their own medical and psycho-social struggles. We have been working as volunteers

to try and educate patients and the public that congenital heart defects last a lifetime: they can be repaired but are never fixed. Most patients will require follow-up surgeries, medications, and/or devices like pacemakers and implantable defibrillators. When I was a kid I was told I was fixed, because back then they didn't know any better. Back then only 20% of us survived – now it is 98%! In fact, there are now more adults with CHD than kids (about 130,000 adults vs. 50,000 kids in Canada). The problem is that the healthcare system hasn't kept pace and there are too few cardiologists specially trained in adult CHD, too few hospital beds, too long wait times for surgery, no social workers, only one specially trained psychologist(!), and basically no support system in place to help patients. There are many more patients out there who are like I was; they aren't aware that they should be seen regularly by a specialist in adult CHD, often until it's too late.

It's been a real struggle for us to communicate this message to parents because to many of them age 18 seems forever away, but we know that it's important to lay the groundwork for life with CHD after age 18, when there will be a huge need for follow-up care. Though I'm doing well thanks to the outstanding medical care I've received all my life, many patients I know are unable to work, some are on disability, and many really struggle to get by. In my view, we owe them the same level of care as they received as children. I know we're not as cute, but we still have a lot to offer.

Share Your Story

We invite you to share with us your experience with congenital heart disease. We would like to hear from parents, as well as children, teens and adults who themselves have a CHD. Your story may provide the encouragement and support someone else needs. For assistance in preparing your story, or to submit your story, contact the Newsletter Coordinator at jenb@heartbeats.ca

Three cheers for grandparents!

Our recently published article in a prestigious nursing journal, *Journal of Advanced Nursing*, highlights the key contribution that grandparents made in the families who participated in the Sano study. Thanks to the parents and grandparents who participated in this study!

AIM. This paper is a report of a study of the process of grandparent involvement with siblings of preschool children with hypoplastic left heart syndrome.

METHOD. Individual interviews were conducted in 2007 with 15 grandparents of six preschool children with complex congenital heart disease. The interviews were conducted in home settings or by telephone.

KEY FINDINGS. 'Stepping in as needed' and 'safeguarding relationships' were identified as two core categories related to grandparenting siblings of children with heart disease. Grandparents stepped into a parent role with toddler and preschool-aged siblings by attending to their daily care routines, recreational and play times, and relational needs while parents were occupied with their sick and hospitalized infants. Grandparents' concerted efforts to sustain parent-child and child-sibling relationships were also striking.

Reference: Ravindran, V., & Rempel, G. (2011). Grandparents and siblings of children with congenital heart disease. *Journal of Advanced Nursing*, 67(1), 169-175.

If you would like more information about the study findings, please contact us.

Safeguarding the Heart Child Research Program

Faculty of Nursing, University of Alberta

Lead Investigator: Dr. Gwen Rempel (780-492-8167)

Research Coordinator: Louisa Fricker (780-492-6279)

Email: safeguarding@ualberta.ca

