Growing into adulthood when you are born with heart disease

Specialized medical teams at the Peter Munk Cardiac Centre lead the way globally in the treatment and care of adults born with a heart defect.

By Daina Lawrence

FIFTY YEARS AGO, BABIES BORN WITH COMPLEX CONGENITAL HEART DEFECTS had a less than 5 per cent rate of survival to adulthood, as surgical strategies that would eventually treat many of these patients were still in their infancy or not available. Today, more than 90 per cent of these babies make it into adulthood, creating a whole new patient demographic where there are currently more adult patients living with congenital heart disease than children – and medicine must now do its best to keep up.

The rapidly growing field of adult congenital heart disease (ACHD) deals with patients who had one or more structural abnormalities of the heart present at birth. Thanks to advances in diagnosis and treatment, those people survive to adulthood despite their birth defect, but need continued follow-up and care throughout their adult lives.

"In the last few decades, this specialty [ACHD] has really taken off," explains Dr. Rachel Wald, a cardiologist with the Toronto Congenital Cardiac Centre for Adults (TCCCA) and the Peter Munk Cardiac Centre (PMCC). "Initially, these children simply didn’t survive, and that’s just not the case anymore. Now this expanding patient population requires a specialized team of physicians and nurses to look after them.

To treat the rapidly growing number of ACHD patients in Canada, the TCCCA and the PMCC were at the forefront of the creation of a multidisciplinary team of specialized cardiologists and congenital heart surgeons who work to marry the pediatric and adult cardiac worlds. The clinic is one of the oldest and largest of its kind in the world, with more than 9,500 active patients. “With the move from primary care to the pediatric world are exceptionally strong because congenital heart disease was once strictly a pediatric condition,” explains Dr. Wald, originally a pediatric cardiologist who later became sub-specialized in the care of adults with congenital heart disease. "Treating congenital heart defects in adults often requires an understanding of the unique complexities of each patient, as many have more than one medical issue. "We believe the best way to balance out this team is to have a diverse array of physicians with complementary strengths,“ adds Dr. Wald.

When an ACHD patient comes to the centre for surgical or advanced medical treatment, members of the team gather to discuss the best approach for that individual. There is a limited amount of medical evidence to guide therapies, as these patients are often among the first of their kind to survive into adult life. Unlike other areas of congenital heart surgery, where there are many people with a certain disease who can be followed to help inform future medical decisions, with ACHD there are too few numbers of patients who have reached adulthood to draw conclusions.

For instance, a surgical technique to help treat babies born with hypoplastic left heart syndrome – which occurs when the heart’s left side isn’t able to effectively pump blood, forcing the right side of the heart to pump blood to the entire body – wasn’t available until the 1990s. Before then, this birth defect was fatal.

"Now we’re seeing our first wave of adults surviving in their 20s,” she says. “We don’t really know what’s going to happen when this cohort survives another 20 years (and more,) into their 30s, 40s and 50s.”

Dr. Wald says, “We’re trying to come up with an educated guess of which complications might happen, and how these can be prevented, however management is very much an evolving process, as our patients teach us what we need to know.

The specialties of the team members make the treatment at the centre truly unique. "Not only are there the congenital heart surgeons and ACHD cardiologists who are experts in ACHD, but the team also consists of cardiology intensivists (specialists trained in catheter-based treatment), electrophysiologists with expertise in heart rhythm problems, cardiac imaging specialists and several others with congenital heart disease expertise to help provide the most balanced patient care.

"Each colleague contributes his or her expertise to find the best treatment option for the individual patient,” says Dr. Erwin Oechslin, an ACHD cardiologist who was a surgical resident in the early 1990s. "I predict a future growth of ACHD patients in numbers and complexity in the next 10 to 15 years. Leaders in health care need to find answers to the rapidly increasing disconnect between available resources and patient demand. "To provide the highest level of personalized care, the collaboration between these multidisciplinary specialists doesn’t stop with the heart. Many ACHD patients have multiple health issues involving other organs and conditions simultaneously, and they require the care of dedicated specialists outside of cardiology.

"Many patients have psychosocial issues, liver problems or pulmonary arterial hypertension, which is high blood pressure in the lungs, so we need a pool of experts who are also experts in these specialties in these areas, as well,” explains Dr. Oechslin.

"The treatment and care for ACHD patients is about navigating uncharted waters, so we require ideas from a multitude of experts to develop dynamic treatment options and map out guidelines for future treatment.

For Dr. Oechslin, “it’s this team approach and a pool of experts with passion, empathy and dedication who can find and provide the best care for these patients.”

"What’s important to remember is that these patients are not fixed, they’re not cured. They’ll never be cured, only repaired. Most patients with a scar in their chest need lifelong follow-up and care, consolidation rather than dilution of experiences and a highly dedicated, multidisciplinary team make the TCCCA at the PMCC a provincial, national and international reference and resource centre. The wealth of expertise has also made TCCCA a hub for training in ACHD education with global impact.

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It takes all of these experts working together to chart out the treatment and management for these patients, as the work is ongoing. The comprehensive approach and provision of patient-centred care, consolidation rather than dilution of experiences and a highly dedicated, multidisciplinary team make the TCCCA at the PMCC a provincial, national and international reference and resource centre. The wealth of expertise has also made TCCCA a hub for training in ACHD education with global impact.

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