



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, these 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) at 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

01 Dr. Erwin Oechslin and Dr. Rachel Wald are members of the PMCC team providing comprehensive treatment and care for adults with ACHD.

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.

“Within ACHD, the ties 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 further 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 vantage points,” 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 medicine, where there are many people with a certain disease who can be studied to help inform future medical decisions, with ACHD there aren’t huge numbers of patients who have reached adulthood.

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.



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Dr. Erwin Oechslin, Director, Adult Congenital Heart Disease program, the Peter Munk Cardiac Centre

Before then, this birth defect was fatal.

“Now we’re seeing our first wave of adults surviving in their 20s, and we don’t really know what’s going to happen when this cohort survives another 10 years [and more], into their 30s, 40s and 50s,” explains Dr. Wald. “You’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 be aware of.”

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 cardiac interventionalists (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, Director of the Adult Congenital Heart Disease program at the PMCC and Bitove Family Professor in ACHD. “Frequently, all of them are involved in the treatment of an ACHD patient.”

Together, the ACHD team is recognized for its long-term outcome studies, care of pregnant women with heart disease, attention to psychological aspects and end-of-life questions in congenital heart disease patients and state-of-the-art adult congenital heart surgery

and congenital catheter-based interventions. The introduction of a dedicated heart failure clinic for ACHD patients led by Dr. Lucy Roche, cardiologist, is one of the latest innovations at the PMCC. “The hearts of congenital heart patients are structurally abnormal from the very beginning, and their heart muscle is deemed to fail,” says Dr. Oechslin.

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 trainees in ACHD education with global impact.

“These patients are born with an abnormal heart from the very beginning, and after the surgeons remodel the heart, you end up with a very complex anatomy,” says Dr. Oechslin. “What’s important to remember is that these patients are not fixed, they’re not cured,” he adds. “They’ll never be cured, only repaired. Most patients with a scar in their chest need lifelong specialized care.”

And although their hearts have been repaired, Dr. Oechslin explains that adults with congenital heart disease are at increased risk for stenosed or leaky valves, significant arrhythmias, heart failure and early death, so it’s vital to follow them to prevent a catastrophic outcome.

These patients are “survivors of modern medicine,” according to

Dr. Oechslin, as it was modern medicine that created this cohort of patients. And he says it’s now the duty of the medical community to care for these patients into adulthood.

“There have been great advances in pediatric medicine in the last 50 years, and now we have a new generation of patients,” he explains. “So we as physicians and as a society have an ethical responsibility to deal with the consequences of Western medicine. I predict a further 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 occurring 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 collaborate with the specialists in these areas, as well,” explains Dr. Oechslin.

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

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.”