A new beat of life

Aortic surgery is delicate work, but it becomes even trickier when your patient is eight years old

By **Renee Sylvestre-Williams**

Dr. Maral Ouzounian, right, and Dr. Chris Caldarone, left, were part of the team approach that helped treat Alex Latimer, far left, who was diagnosed with Loeys-Dietz Syndrome.

Peter Munk Cardiac Centre

It's news no parent wants to hear, but Brian and Jacqui Latimer were told that their son, Alex, may fall asleep and never wake up again.

They lived with that fear for nearly eight years, until a collaborative operation between Toronto's Hospital for Sick Children (SickKids) and the Peter Munk Cardiac Centre (PMCC) gave them the opportunity to plan for the future.

Alex is the Latimers' fifth child, and he was born at home in Bowmanville, Ont. The first 24 hours were fine. "The birth was normal and natural," says Jacqui. "He was big, 11 pounds, and we noticed that his feet were clubbed and his hands were curled. At first they thought it was because he was so long."

The next day was the start of their medical journey. The midwife didn't like how Alex was breathing. The family went to the Markham Stouffville Hospital for a series of tests and then went to their pediatrician, who said Alex was normal but a lot quieter. But he sent them to a pediatric cardiologist "just to make sure."

They were transferred to the Scarborough Hospital for an ultrasound and found that Alex's aortic root was a little bit dilated. That led them to SickKids for genetic testing, where Alex was diagnosed with Loeys-Dietz Syndrome.

Loeys-Dietz Syndrome (LDS) is a genetic disorder that affects the connective tissue in the body, similar in some ways to Marfan syndrome. "It may be inherited from one of the parents or may be the result of a new mutation," says Dr. Maral Ouzounian, Cardiovascular Surgeon and Surgeon Scientist at the PMCC.

The connective tissue is what provides strength and flexibility to bones, muscles and blood vessels. "There are multiple genes in the transforming growth factor (TGF)-beta family that may be effected in LDS." While the genes involved might be different, all of them are characterized by the enlargement of the aorta, the large blood vessel that moves blood from the heart to the rest of the body. With LDS, the aorta can weaken and stretch, which causes a bulge in the vessel wall known as an aneurysm. The stretching of the aorta can also lead to a sudden tearing of the layers in the aorta wall known as an aortic dissection – a catastrophic and sometimes fatal complication of this disorder. Once patients are diagnosed with LDS, says Dr. Ouzounian, they are closely followed with regular MRIs and CT scans of the aorta and all of its branches.

LDS is usually passed down through families, but the genetic testing done on the Latimers found that neither Brian nor Jacqui has it. Alex's LDS was a spontaneous occurrence. The Latimers learned a lot about LDS as a result of Alex's diagnosis.

"It was pretty daunting at first," says Brian. "I started researching the genetics and the collagen factor in his DNA and what it meant. Then we went from there to learning about the heart and the aorta and what imaging can show you, any symptoms to watch for. It's been a real journey trying to familiarize ourselves with the terminology and the medical community and how they work."

(Patients or families with LDS or

similar heritable aortic conditions such as Marfan syndrome or familial thoracic aortic aneurysms rely on support and education through organizations like the Genetic Aortic Disorders Association [GADA] of Canada.)

Alex had his first surgery when he was 18 months old – a valve-sparing aortic root replacement performed by Dr. Glen Van Arsdell – and everything was as fine as it could be under the circumstances, until January 17, 2016.

Earlier that day, Alex, his siblings and their parents were having a typical day out. They were tobogganing down the little hill at his school, but that night Alex started crying in pain, telling his parents that his shoulder, his jaw and his back were hurting. (They didn't know it at the time, but these were the very symptoms of an acute aortic dissection.)

Brian took his son to the Bowmanville hospital, and after X-rays and ultrasounds the hospital sent the pair straight to SickKids via ambulance.

At 4:30 in the morning, it was discovered that the aorta had dissected just above the site of the original graft. "Brian called me at 5:30," says Jacqui. "He wanted me to see [Alex] before he went into surgery." Dr. Van Arsdell was there again, seven years after his last time with Alex. He showed Brian the dissection, and Brian wrote in his journal: "And then, in quiet, measured tones, he walks me through a horror show of possible complications." Alex had to have surgery.

The Latimers got through the surgery, which repaired his aortic arch, but were still faced with the fact that they could lose their child. They slept on mattresses on the floor in his bedroom, reluctant to be away from him during the night. It wasn't over. Alex returned with pain and the scan showed an impending rupture of his aorta.

"I remember it was a Friday, and Glen called me to say he was leaving town and was leaving me with [Dr.] Chris [Caldarone, Surgeon-in-Chief, Department of Surgery, Staff Cardiovascular Surgeon at SickKids]," says Dr. Ouzounian.

"[A team meeting took place] with Glen, Chris, the radiologists and Alex's cardiologist, and immediately upon seeing the scans, we knew that Alex needed an urgent operation."

Dr. Ouzounian had a complicated aortic surgery already booked that day, so the team decided to do Alex's surgery the next day, a Saturday.

"I've never operated on a kid before," says Dr. Ouzounian, who had done a pediatric cardiac surgery rotation in training, but had never done a thoracoabdominal repair on an eight-year-old. In fact, there have been very few previous thoracoabdominal aortic repair cases at SickKids, as the condition is extremely rare in children.

"That's why the team approach was so essential," she says. "Chris understands the pediatric physiology and doing major aortic surgery on a child, while I had the technical expertise of actually doing the thoracoabdominal repair." This is an extremely high-risk surgery in the emergency setting, says Dr. Ouzounian, with a significant chance of death or leaving the patient paralyzed. It was an eight-hour operation that involved repairing his aorta from his arch down to his bowel and kidney vessels.

Alex's second surgery was a success. He went back to school in June, and he's playing with friends and eating a lot. "He takes it easy in gym," says Jacqui. But the effect of eight years and three surgeries did take their toll.

"It's a little of PTSD [posttraumatic-stress disorder]," says Brian. "It definitely is a bit of hospital hangover," he says of their stay at SickKids. Alex spent (just over) a week in the intensive care unit (ICU), and Jacqui says that it wasn't a comfortable stay for their son.

"He had so many things attached to him, and you want so badly to be there for him, but part of you dreads it because you feel so helpless," says Jacqui. "He would look at me; he couldn't even speak. I would say, 'Are you comfortable?' and he would shake his head no, and I would get the nurse to help me turn him over on the other side. He would doze for five or 10 minutes, and his eyelids would flutter and he would be uncomfortable again."

"It was difficult and draining," says Brian, but now the Latimers are approaching a life of normalcy. "Dr. Ouzounian is off the charts. In our last e-mail, she said to get him outside. He'll be tired and sore, but it's good for him," says Jacqui.

Dr. Ouzounian adds: "Even though the majority of his aorta has been replaced, Alex could still have recurrent dissections or aneurysms in other blood vessels, and thus must be monitored for life."

Alex Latimer's surgery was an extremely high-risk procedure, performed in an emergency setting that required a co-ordinated team approach, PMCC Cardiovascular Surgeon Dr. Maral Ouzonian, above, says.