Maria Willing remembers sitting in her Renfrew, Ont., kitchen at age 30 thinking, "I'm going to die". As a child, her life was saved by a surgical procedure performed by Dr. William Mustard. She has since been followed by adult congenital heart specialists at the Peter Munk Cardiac Centre.

Mustard Baby has twice defied the odds

Marina operator Maria Willing pays careful attention to her heart health with the help of congenital heart disease specialists

There is little doubt that Maria Willing has lived up to her name throughout her life.

Born with "blue baby syndrome" in June 1961, Maria was given six months to live. The syndrome is also known as the transposition of the great arteries – a form of cyanotic heart disease where the pulmonary and systemic circulation run parallel instead of in a series, resulting in a low blood-oxygen level. She had two holes made into her heart at only three weeks of age to allow mixing of the blue (non-oxygenated) and pink (oxygenated) blood.

A MUSTARD BABY

Blue babies had a less than 20 per cent survival rate back then, but Maria defied all odds, growing stronger each day. On May 16, 1963, Maria made medical history and became the poster girl for the groundbreaking surgical procedure performed by Dr. William Mustard at Toronto's Hospital for Sick Children. A heroic repair to the transposition of the great arteries that allows oxygenation of the blood in the lungs, it became known worldwide as the Mustard Procedure.

By 1987, the procedure had saved more than 500 babies at the Hospital for Sick Children and thousands worldwide. Survival to adulthood increased to 80 per cent for these "Mustard babies."

The hospital lost touch with Maria over the years – her family lived in Whitby, Ont., then Renfrew, Ont. It wasn't until she was 30 years old, facing another complication, that she was drawn back to Toronto.

She had developed atrial fibrillation and was experiencing irregular heart rhythms. She knew something was terribly wrong.

Maria vividly recalls sitting in her Renfrew kitchen: "I just sat there thinking, 'I'm going to die.'"

Not one to give up, but feeling she had nowhere to turn, she called the Hospital for Sick Children. "I told them I had had the Mustard Procedure. They immediately said, 'Can you get here tomorrow?'" Maria drove to Toronto, only to realize they were referring to the nearest children's hospital – the Children's Hospital of Eastern Ontario in Ottawa.

THE WRONG WAY, BUT THE RIGHT CHOICE

"I went the wrong way, but made the right choice," says Maria. "They saw me at Sick Kids and immediately referred me to Dr. Peter McLaughlin at Toronto General Hospital."

Maria had to be cardioverted three times, which involved converting an abnormally fast heart rate (arrhythmia) to a normal rhythm through electricity or medication.

Then she had a stroke, but within a week she was fitted with a pacemaker.

Maria attributes her survival to the fine work of the doctors at the Toronto General Hospital. Now 54 years old, she attends annual follow-ups at the Toronto Congenital Cardiac Centre at the Peter Munk Cardiac Centre (PMCC). "I always take a few days off and go on a mini vacation to Toronto with my girlfriends," she says.

Maria has developed a strong relationship with her former cardiologist, Dr. Vera Rose, who retired about three years ago, and now she's under the care of Dr. Erwin Oechslin, director of the Toronto Congenital Cardiac Centre.

"We have a good rapport," says Maria, a mother of three. "The joke is, [Dr. Oechslin] always wants me to have the stress test, and I always refuse."

"We see Maria once a year to monitor her heart function and heart rhythm," Dr. Oechslin says. "Her lifestyle is healthy, she is physically active and she watches her diet and her weight. She also has good genes."

FEELING TRULY BLESSED

Today, Maria and her husband, Brad Willing, own and operate a marina in Haliburton, Ont. Open from April to October, Maria manages a busy convenience store situated in the marina.

She attributes her youthfulness to the careful attention she pays to maintaining her health. "In August 2014 I got sick, and I thought I was going to have a heart attack," she says. "I didn't want to take the chance, so my friends graciously offered to lend their time to help out."

Maria and Brad often sneak in an evening cruise aboard their favourite pontoon in the summer.

"I like the calmness the water makes me feel," she says. "I have all the time in the world to enjoy the water. We are truly blessed."

SPECIAL EXPERTISE

In the 1980s, the Mustard Procedure was replaced by the Jatene Procedure, the arterial switch procedure to repair complete transposition of the great arteries. All survivors of the Mustard (Jatene) Procedure – like all patients born with congenital heart disease of moderate or great complexity – had to be transferred to adult congenital heart disease centres with special expertise.

According to Dr. Oechslin, "patients with a Mustard Procedure are at risk for long-term complications and require long-term follow-up in a specialized centre. Fifty per cent of patients with a Mustard Procedure who graduated from the Hospital for Sick Children are not associated with an adult congenital heart disease program and are lost to follow-up. We don't know where they are and how they are doing. Only an emergency brings some of them back to the health-care system."