Lung transplant survival rates good for Canadians with cystic fibrosis

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The five-year survival rate for Canadians with cystic fibrosis who have received a lung transplant is 67 percent and half of those who have had transplants live beyond 10 years, according to new research by Dr. Anne Stephenson of St. Michael's Hospital. Credit: Courtesy of St. Michael's Hospital

The five-year survival rate for Canadians with cystic fibrosis who have received a lung transplant is 67 per cent and half of those who have had transplants live beyond 10 years, new research has found.

The encouraging statistics were published today in the *Journal of Heart Lung Transplantation* by Dr. Anne Stephenson of St. Michael's Hospital. She said this was the first time anyone had used the Canadian Cystic Fibrosis Registry, of which she is the director, to examine outcomes following lung transplantation at a national level in Canada. Previous studies have reported outcomes at single transplant sites.

"People think when you have cystic fibrosis and need a transplant it's a death sentence," said Dr. Stephenson. "But our paper shows that one-, three-, and five-year survival rates following lung transplantation are really quite high."

Cystic fibrosis is an inherited chronic disease that affects the lungs and digestive system. A defective gene and its protein product cause the body to produce unusually thick, sticky mucus that clogs the lungs and leads to life-threatening lung infections, obstructs the pancreas and stops natural enzymes from helping the body break down and absorb food. It is the most common fatal genetic disorder affecting Canadian children and young adults.

Lung transplants do not cure people of CF because the defective gene that causes the disease is found in all the cells of the body, with the exception of the newly transplanted lungs.

The transplant paper included 580 patients who received a lung transplant between 1988 and 2012. Overall post-transplant one-, five- and 10-year survival was 87.8 per cent, 66.7 per cent and 50.2 per cent.

Dr. Stephenson found a patient's age at the time of the transplant was a significant predictor of survival. The youngest and oldest patients had poorer survival rates compared to those in the middle age-range. In the first 10 years after transplant, survival rates were similar between those under and over age 18 years, but if someone survived beyond 10 years,

there was a four-fold increase in risk of death for those under 18 years of age.

Interestingly, factors associated with worse survival in the overall CF population, such as being female, early age of diagnosis, malnutrition and CF-related diabetes, did not significantly impact post-transplant survival.

Other factors such as pancreatic sufficiency or being infected with certain bacteria that can live in the lungs of people with CF, such as B. cepacia, had a detrimental impact on survival.

There was a trend toward improved survival in more recent decades as individuals transplanted in 2000 or later were 23 per cent less likely to die compared to those transplanted before 2000. However, this did not reach statistical significance.

Dr. Stephenson said that accurately predicting life expectancy after a transplant would allow CF caregivers to better quantify the risks associated with transplant so patients are fully informed when making decisions around transplantation.

This is the second piece of good news for cystic fibrosis patients that Dr. Stephenson has published recently. In November, a paper she published in the *European Respiratory Journal* showed that Canadians with cystic fibrosis are living almost 20 years longer than they did two decades ago.

The median survival age was 49.7 years in 2012, up from 31.9 years in 1990. Since the paper was written, Dr. Stephenson has updated the median survival age to include Cystic Fibrosis Canada data from 2013 and reported the median age of survival has in fact reached 50.9 years. Dr. Stephenson said there are few therapies that can impact survival as dramatically as lung transplantation, which has likely contributed to the

increasing survival in Canadians with CF.

Provided by St. Michael's Hospital