Canadians with cystic fibrosis living 20 years longer than they did two decades ago

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Canadians with cystic fibrosis are living almost 20 years longer than they did two decades ago, according to a research paper published today by Dr. Anne Stephenson, a respirologist and research at St. Michael's Hospital. Credit: Courtesy of St. Michael's Hospital

Canadians with cystic fibrosis are living almost 20 years longer than they did two decades ago, according to a research paper published today.

The median survival age was 49.7 years in 2012, up from 31.9 years in 1990, Dr. Anne Stephenson, a respirologist and research at St. Michael's Hospital wrote in the *European Respiratory Journal*. 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.

In addition, the median age at which Canadians with cystic fibrosis die has risen to 32 years in 2012 from 21.7 years in 1990, Dr. Stephenson said. The death rate declined from 1.4 deaths per year in 1990 to 0.99 deaths per year in 2012.

"By analyzing national data, we can confirm what we are seeing clinically, specifically that our cystic fibrosis patients are living longer, well into adulthood and middle age, which is very exciting," said Dr. Stephenson.

"Although improved survival is very positive news for the CF community, we must remember that people with cystic fibrosis still die prematurely. In fact, in 2012, half of the deaths due to cystic fibrosis happened before the patients reached their 32nd birthday. This reinforces the need for continued research in this area in order to optimize health for all individuals living with this progressive disease."

Dr. Stephenson's paper is based on data of about 5,000 patients in the Canadian Cystic Fibrosis Registry, of which she is the director.

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 disease affecting Canadian children and young adults.

Dr. Stephenson said improvements in cystic fibrosis survival are due to

several factors, including multi-disciplinary care, rigorous infection control practices, the availability of new medications such as inhaled antibiotics and therapies to thin mucus, as well as the option of lung transplants.

In addition, in the 1970s and 1980s, she said Canadian clinicians focused on improving nutrition in CF patients with the expectation of normal growth. Individuals born at that time were exposed to optimal nutrition from birth, which has likely had a huge impact on their health now that they are in their 30s and 40s. Malnutrition and lung infections are associated with increased risk of death in people with cystic fibrosis, so better nutrition and minimizing hospitalizations due to infections are important to further improve survival in CF.

Dr. Stephenson said the data showed increasing survival for all groups of CF patients, even people with the most severe mutations, indicating the improvements in median age of survival were not simply due to detecting milder cases or the inclusion of those milder cases in the CF Registry.

Dr. Stephenson said it was too soon to evaluate the effect of newborn screening or newly available protein modulator therapies on survival; however, she anticipates these will contribute to improved survival in the years to come.

Newborn screening for cystic fibrosis is now available in seven Canadian provinces. In 2012, Health Canada approved the drug Kalydeco for use in CF patients. St. Michael's was the only site outside of the United States involved in the Phase 2 clinical trial for the drug and had the largest number of patients enrolled in the Phase 3 trial. It is the first drug that targets the underlying cause of CF, a faulty gene and its protein product.

Provided by St. Michael's Hospital