Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

Cystic Fibrosis Research News

Citation:

Goss CH, Sykes J, Stanojevic S, Marshall B, Petren K, Ostrenga J, Fink A, Elbert A, Quon BS, Stephenson AL. Comparison of Nutrition and Lung Function Outcomes in Patients with Cystic Fibrosis Living in Canada and the United States. American Journal of Respiratory and Critical Care Medicine. 2018 Mar 15;197(6):768-775. PMID: 29099606.

What was your research question? (50 words maximum)

How can we compare survival differences in people with CF from Canada and from the United States by age, population changes in lung function and nutritional status, and rate of decline of both lung function and nutritional status?

Why is this important? (100 words maximum)

Being undernourished and having poor lung function are related to an increased risk of mortality with CF. Treatments aim to maintain good nutrition and preserve lung function. A 10-year gap in the median age of survival for CF patients was reported between patients living in Canada compared with patients living in the U.S. This study aims to help understand the changes over time and trends of lung function and nutrition between Canada and the U.S.

What did you do? (100 words maximum)

We looked at data from both the Canadian CF Registry (CFFR) and the U.S. CF Foundation Patient Registry (CFFPR) from 1990 to 2013. We created a dataset that blended variables collected in the two registries, and then compared outcomes between the two countries. We analysed differences in demographic (including gender, age, and race) and clinical factors (including genotype, whether patients were pancreatic sufficient, whether patients received newborn screening, and whether patients had CF-related diabetes).

Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

Cystic Fibrosis Research News

We adjusted for factors that might explain observed differences between the two countries.

What did you find? (100 words maximum)

The study included a total of 37,772 patients in the U.S. and 5,149 patients in Canada and confirms that patients with CF in both Canada and the U.S. have experienced significant improvements in nutritional status and lung function over time. Patients in the U.S. have experienced faster rates of improvement during the time frame for both outcomes. The survival gap between the two countries appears to be decreasing in younger birth groups. The survival gap remains in older patients and is likely a large factor for the observed 10-year survival advantage for patients living in Canada.

What does this mean and reasons for caution? (100 words maximum)

There are many reasons for observed improvements in nutritional status and lung function as well as the U.S. median survival catching up to Canada. These reasons include the use of high fat, high calorie diets in CF patients, introduction of newborn screening, and improved access to care for children in the U.S. Reasons for caution with these findings include: differences in the use of newborn screening among states and provinces, possible bias due to differences in survival, restricting the study population to ages 7 years and older, and differences in recorded medication use between the U.S. and Canada.

What's next? (50 words maximum)

Further analysis is needed to figure out whether the observed improvements in CF populations can be explained by access to CF therapies and treatment options such as lung transplants.