# The Cystic Fibrosis Survival Gap: Why Do Canadians Fare Better Than Americans?

The Cystic Fibrosis Foundation Patient Registry (CFFPR) has chronicled considerable improvement during recent decades in the median predicted survival of U.S. patients with cystic fibrosis (1). Increased survival has been attributed to better nutrition, particularly in infants and young children; the approval and use of new medications for cystic fibrosis complications; and continual improvement in quality of care, a powerful tool made possible by CFFPR data transparency. Comparisons of outcomes among U.S. care centers have been a driving force in recognizing and adopting best-care practices (2).

In recent years, the strategy of comparing outcomes in different settings to recognize improvement opportunities has been applied across countries through their local cystic fibrosis patient registries. For example, a comparison of registry data from the United States and United Kingdom attributed geographic differences in pulmonary function in children and young adults to greater use of mucolytic therapies in the United States (3). In their article, Stephenson and colleagues report a sobering comparison of cystic fibrosis survival between Canada and the United States that shows a marked advantage for Canadians beginning around 1995 and increasing to the current gap of approximately 10 years (4).

One's first reaction to these data may be to wonder whether this survival gap is real or the result of fundamental differences in how the registries collect and analyze information. However, the researchers took great pains to assure commonality in how expected survival was calculated from each of the databases, and we believe their results are real. Whereas predicted median survival is a theoretical number with which we might quibble, this study also showed a striking disparity in median age of death that favors Canadians with cystic fibrosis, a hard reality we must accept.

To use this study to improve care, we must ask the obvious question: What are the bases of this disparity and, in particular, the survival trend divergence in the mid-1990s? Some "usual suspects" associated with elevated cystic fibrosis mortality risk (such as differences in *CFTR* mutations or infectious airway pathogens) seem inadequate to account for the observed differences, particularly because we have no reason to believe that these factors have disproportionately changed during the past few decades. Three possible explanations considered by the authors are differences between the 2 countries in lung transplantation procedures, nutrition, and health care systems.

Lung transplantation became a viable procedure in the 1980s, and transplant units in both Toronto and Montreal were early adopters of double-lung transplantation for patients with cystic fibrosis. Lung transplants confer a survival advantage to eligible patients (5). Further, a survival advantage for Canadian versus U.S. transplant recipients has been reported (6, 7). Although a greater proportion of Canadians than Americans with cystic fibrosis receive transplants, they still represent small fractions of the overall populations with this disease, and the survival advantage following transplantation does not approach 10 years. Although lung transplantation clearly contributes to overall survival in both populations, whether transplant differences can fully account for geographic survival disparities is unclear.

The authors also considered the survival disparity described in the 1980s between a Canadian and a U.S. cystic fibrosis center that was attributed to differences in nutritional support protocols (8) as a potential contributor. In the decades since that report, the U.S. Cystic Fibrosis Foundation has placed a strong emphasis on childhood nutrition. Although the benefits of these changes may not be completely realized in today's survival numbers, how better nutrition in the United States could contribute to an increasing survival difference between the United States and Canada in subsequent decades is unclear.

An important observation in this study was the association between insurance status and survival among U.S. patients with cystic fibrosis. In particular, when U.S. patients insured by Medicaid or Medicare and those without health insurance were excluded from analyses, survival differences between the 2 countries disappeared. Although decreased access to care is associated with poorer health outcomes, the current study indicates a greater use of chronic therapies and more clinic visits among U.S. patients with cystic fibrosis than their Canadian counterparts. Insurance status might possibly be a surrogate marker of poverty, which is known to have an adverse effect on health (9). Although poverty levels in the United States and Canada are not strikingly different, the infrastructures for delivering health care in the 2 nations have markedly dissimilar effects on disadvantaged patients (10). In fact, median predicted survival for *all* Canadians is higher than that of U.S. citizens, and this disparity also has increased during the past 2 decades.

In summary, cystic fibrosis registries have again demonstrated their power to identify important opportunities to improve the health and well-being of our patients. The U.S. Cystic Fibrosis Foundation and Cystic Fibrosis Canada are to be commended not only for supporting these valuable tools but for being bold enough to compare the data within them to demonstrate such a significant survival difference between the 2 populations. Now we are faced with the more difficult task of trying to identify and implement solutions to bridge this survival gap, which seems to be based on

This article was published at Annals.org on 14 March 2017.

 

fundamental differences in the 2 nations' health care systems.

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**Disclosures:** Dr. Flume reports grants and personal fees from the Cystic Fibrosis Foundation outside the submitted work. Dr. VanDevanter reports personal fees from the Cystic Fibrosis Foundation outside the submitted work. Disclosures can be viewed at www.acponline.org/authors/icmje/ConflictOfInterestForms.do?msNum=M17-0564.

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*Ann Intern Med.* 2017;166:599-600. doi:10.7326/M17-0564

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