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

CYSTIC FIBROSIS FOUNDATION
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Cystic Fibrosis Research News

Citation:

Knapp EA, Fink AK, Goss CH, Sewall A, Ostrenga J, Dowd C, Elbert A, Petren KM, Marshall BC. The Cystic Fibrosis Foundation Patient Registry. Design and Methods of a National Observational Disease Registry. Annals of the American Thoracic Society. 2016 Jul;13(7):1173-9. PMID: 27078236.

What was your research question? (50 words maximum)

We wanted to explain how the Cystic Fibrosis Foundation Patient Registry (CFFPR) data is collected, describe the patient population, and detail the challenges of this research.

Why is this important? (100 words maximum)

The CFFPR is one of the United States' oldest data registries. It contains data on people living with cystic fibrosis (CF) with records as far back as 1986. CF is a life-threatening chronic genetic disease. Collecting high-quality observational data is important, as its helps us to improve research and clinical care efforts. The registry is also an important tool in tracking the changing cystic fibrosis population.

What did you do? (100 words maximum)

We conducted an audit of the 2012 registry to compare medical records on file to data found in the registry. All people diagnosed with CF and associated disorders who receive care at a CF Foundation (CFF) care center and give their informed consent are included in the CFFPR. The CFF has a network of 121 CF care centers, all of which are required to participate in the CFFPR. The data for the CFFPR is collected through a web-based portal and entered by care center staff using medical records and patient forms. The data is grouped into 5 categories – demographic, diagnosis, encounter, care episode, and annual

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review. All data is kept secure, and subject to validation testing which includes data audits.

What did you find? (100 words maximum)

In 2012, the CFFPR captured 81-84% of all people with CF in the United States (US). Approximately 6% of people seen at a CF care center did not consent to the registry. The CFFPR showed that over 90% of people in 2009 stayed in the registry through 2014. The audit comparing patient records to the CFFPR in 2012 showed that there was minimal missing data and high accuracy on many important variables such as demographics, lung function, nutritional status, and hospitalizations.

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

The CFFPR is a vital tool for research and clinical care efforts, as well as national observation. However, there are still reasons for caution. For example, missing data remains a large challenge. Other challenges include: no central reporting of new cases of CF which creates difficulty in assessing the true size of the US CF population; differences in disease progression from person to person, which can bias data analysis; and increased newborn screening, which creates earlier diagnoses and makes current demographics hard to compare to older decades.

What's next? (50 words maximum)

We will continue doing research using CFFPR data. Important next steps include enhancing current CFFPR data by linking to other data sources to increase utility and data quality by comparing US observational data to other countries' registry data. Finally, we continue to try to improve the quality of our data collection.