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Cystic Fibrosis Research News

Title:

Validation of the French 3-year prognostic score using the Canadian Cystic Fibrosis registry.

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What was your research question?

A research group in France published a prognostic score developed using French Cystic Fibrosis (CF) registry data to predict the risk of death or lung transplantation in three years in the adult CF population. We wanted to determine if the proposed tool worked as well using the Canadian CF registry.

Why is this important?

The implementation of multi-disciplinary care, new treatments, newborn screening, and transplantation has resulted in changes in the CF population. National CF patient data registries, many of which have been around since the 1990s, are well-established in many countries and are increasingly used to answer clinically relevant questions. For instance, registry data are often used to predict how long people can be expected to live. However, before such tools become available worldwide, it is important to assess if they work outside of the population that they were developed on.

What did you do?

The validation was done on adults with CF who were awarded points based on their clinical results in 2011. A total of 8 clinical factors were included in the score: lung function, body mass index, *Burkholderia cepacia* colonization, frequency of intravenous antibiotic courses, whether they were hospitalized or not, oral corticosteroids use, long-term oxygen therapy,

and non-invasive ventilation. The score ranged from 0 to a maximum of 9.5 possible points. For instance, adults with a low lung function would receive 3 points, whereas those with a high lung function would not get any points. Hence, a lower score represents better health.

What did you find?

We found that the French prognostic score performed very well to predict death or lung transplantation in the Canadian CF population. In fact, for every one unit increase in the score, there was a threefold increased risk of death or lung transplantation in three years. The tool was able to accurately identify low, medium and high-risk individuals for death or lung transplantation. Only 1% of individuals with a low score died or needed a lung transplantation within three years compared to 54% with a high score.

What does this mean and reasons for caution?

Overall, the prognostic tool developed in France is an efficient tool that can predict the future health outcome of Canadian CF adults. We believe it worked so well because France and Canada share similar healthcare systems and access to lung transplantation. Confirmation of existing prognostic models will provide clinicians with a way to predict whether or not someone will die or need a transplant in the future. It also provides the CF team with an opportunity to modify treatments, council individuals about more aggressive therapies before lung transplantation, so the risk of death or transplant can be reduced.

What's next?

Since it is well known that Canada and France have comparable access to healthcare and lung transplantation, our next step would be to determine if the prognostic tool works efficiently in a CF population where the healthcare systems and access to transplant are different.

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