Cystic Fibrosis Research News

Title:
Cirrhosis associated with decreased survival and a 10-year lower median age at death of cystic fibrosis patients in the Netherlands

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What was your research question?
The goal of our research project was to evaluate the effect that CF-related cirrhosis (scarring of the liver) has on life expectancy and cause of death in people with CF in the Netherlands

Why is this important?
In people with cirrhosis normal liver tissue is replaced by scar tissue, and this may lead to reduced liver function. About10% of people with CF develop cirrhosis, mostly around the age of 10 years. Those with CF-related cirrhosis may suffer from bleeding in the gut from varices (abnormal swollen and enlarged veins) due to high pressure in the in vein between the gut to the liver (portal vein). Some people with CF-related cirrhosis do need a liver transplant. To see if the care of people with CF-related cirrhosis needs to be improved, we first wanted to establish if cirrhosis influences life expectancy and cause of death in people with CF.

What did you do?

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The study was done in all CF care centres in the Netherlands. From the medical charts, we identified all people with CF who had cirrhosis and were alive in 2009 and followed them up to 2016. If an individual died during the observation period, we collected the age of death, the cause of death and the lung function and nutritional status in the year leading up to the date of death. We compared these results to the data from National CF patient registry of the Netherlands over the same period. In the Netherlands, liver transplantation is generally available to patients if indicated and CF-related is an established reason for liver transplantation.

What did you find?

We found that for people with CF who died, the age at death of those with CF-related cirrhosis was on average 10 years lower than others with CF (27 vs. 37 years). Also, the proportion of people with CF-related cirrhosis who died in the observation period was higher. This difference could not be explained by a difference in lung function or nutritional status. When we looked at the reported cause of death, we found that, compared to other people with CF, most people with CF-related cirrhosis reportedly died due to respiratory issues and only a minority due to liver failure.

What does this mean and reasons for caution?

CF-related cirrhosis is a significant risk factor for early death. Based on our study we could not identify those at risk for early death based on their lung function and nutritional status. Also, liver failure, as a cause of death, was a rare event. Based on our study we could not identify the cause of the risk of early death in this patient group. Neither can we prove that for instance, earlier liver transplantation would improve the survival chances of these individuals in the long-run.

What's next?

Prospective studies are needed to provide answers to these critical questions. Also, collaboration between the large patient registries, using the same or similar definitions for CF liver disease are needed to unravel the natural history of the disease to provide solutions for improving the medical care and to provide a better outcome for these patients.

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