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Cystic Fibrosis Research News

Title:
Technological advances shed light on left ventricular cardiac disturbances in cystic fibrosis

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What was your research question?
It is well accepted that cystic fibrosis (CF) mostly affects the lungs. Heart disease in CF is typically thought to be a result of having lung disease. We asked whether CF causes a problem in the heart and if this is related to the lung disease experienced.

Why is this important?
With new therapies showing promise for increased life expectancy beyond the current 40 years, the proportion of those living with CF into middle-age is increasing. The increasing lifespans and activity levels of individuals with CF will likely be putting significantly more strain on their hearts over time. If individuals with CF are at increased risk for heart problems relative to the general population, identifying and understanding this risk will be vital to the caring for adults with CF.

What did you do?
In this review, we examined why previous studies examining heart function did not identified a primary problem in the heart of individuals living with CF. We delved deeper into the results from older studies that use methodologies such as post-mortem analysis, radionuclide angiography, and traditional heart echocardiography. By taking a closer look at the data collected from the left side of the heart, we found evidence for an intrinsic defect in the CF heart that has previously been overlooked. In addition, we provided evidence from new advancements in imaging technology that further support this.

What did you find?
Early studies investigating the heart in CF have mostly concluded that heart problems arise as complications of having lung problems. The conflicting reports on heart dysfunction provided by these earlier studies were likely due to an incomplete understanding of the incidence of primary heart disease in CF, especially since heart disease has not previously been considered

a significant complication in CF. However, more recent advances in heart imaging—in particular, echocardiography and magnetic resonance imaging that are highly sensitive reveal early signs of heart dysfunction in adults living with CF with no known heart problems. Collectively, these new findings provide evidence that defects in the CF heart is not only related to lung disease.

What does this mean and reasons for caution?

It is not surprising that the focus of CF clinical care and research has been on the lungs and gastrointestinal system since these complications comprise the majority of problems in CF. However, there is an increasing amount of research showing that individuals with CF may have abnormalities in the heart even without having lung problems. More data from advanced techniques in echocardiography and magnetic resonance imaging are needed Additionally, we do not know if the detection of heart dysfunction prior to the onset of symptoms ultimately affects the overall health of individuals with CF.

What's next?

Studies should continue to characterize heart disease in individuals living with CF patients and clinically examine the health of hearts in people with CF using the latest techniques. This will help establish health care practices specifically targeting the optimization of heart health in individuals with CF, leading to improved quality of life associated with this disease.

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