

Cystic Fibrosis Research News

Title:

Cardiac structure and function in people with cystic fibrosis.

Lay Title:

Cystic fibrosis and the heart

Authors:

Lisa Steen Duus*, MD^a, Maria Dons*, MD^{a,e}, Rebekka Faber Thudium, MD, PhD^d, Susanne Dam Nielsen, MD, DMSc^{b,c,d}, Mette F Olsen, MSc, PhD^{d, h}, Tavs Qvist, MD, PhD^d, Mats Højbjerg Lassen, MD^{a,e}, Kristoffer Grundtvig Skaarup, MD^{a,e}, Niklas Dyrby Johansen, MD^{a,e}, Thomas Mørk-Strøm Bluhme, BSc^d, Terese L Katzenstein, MD, PhD, DMSc^d, Tacjana Pressler, MD, DMSc^d, Daniel Faurholt-Jepsen, MD, PhD^{c,d}, Tor Biering-Sørensen, MD, MSc, MPH, PhD^{a,e,f,g}

Affiliations:

- a) Cardiovascular Non-Invasive Imaging Research Laboratory, Department of Cardiology, Copenhagen University Hospital - Herlev & Gentofte. Hospital, Gentofte Hospitalsvej 8, 2900 Hellerup, University of Copenhagen, Denmark
- b) Viro-immunology Research Unit, Department of Infectious Diseases, Copenhagen University Hospital Rigshospitalet, Blegdamsvej 9, 2100 Copenhagen, Denmark
- c) Department of Clinical Medicine, Faculty of Health and Medical Sciences, Blegdamsvej 3B, 2200 Copenhagen, University of Copenhagen, Denmark
- d) Cystic Fibrosis Centre, Department of Infectious Diseases, Copenhagen University Hospital Rigshospitalet, Blegdamsvej 9, 2100 Copenhagen, Denmark
- e) Center for Translational Cardiology and Pragmatic Randomized Trials, Department of Biomedical Sciences, Blegdamsvej 3B, 2200 Copenhagen, Faculty of Health and Medical Sciences, University of Copenhagen, Denmark
- f) Steno Diabetes Center Copenhagen, Borgmester Ib Juuls Vej 83, 2730 Herlev, Denmark
- g) Department of Cardiology, Copenhagen University Hospital Rigshospitalet, Blegdamsvej 3B, 2200 Copenhagen, Denmark
- h) Department of Nutrition, Exercise and Sports, Faculty of Science, Nørre Allé 51, 2200 Copenhagen, University of Copenhagen, Denmark

What was your research question?

Cystic Fibrosis Research News

We set out to investigate whether cystic fibrosis (CF) affects the structure and/or function of the heart by comparing the hearts of people with CF to those without. The study also sought to identify any factors that might negatively impact the heart in people with CF.

Why is this important?

Cystic fibrosis (CF) primarily affects the lungs and digestive system, but its impact on the heart is poorly understood. With the recent advances in treatment, people with CF are living longer, healthier lives, making it increasingly important to understand whether CF or CF-related factors poses any risks to the heart.

What did you do?

We examined 104 adults with CF alongside 104 people from the general population matched for age and sex. The examination included an ultrasound of the heart (echocardiogram) to measure heart size and function such as pumping efficiency. We also performed a lung function test, blood tests, general clinical evaluation and recorded genetic mutations as well as medical treatment. We then compared the heart status in people with CF to people without.

What did you find?

The findings revealed that 44% of the people with CF showed early signs of abnormal heart function, compared to just 20% in those without CF. People with CF had smaller hearts, probably due to smaller body size. Although the heart function in individuals with CF remained within normal limits, the hearts showed early signs of working less efficiently, with reduced pumping function in both the left and right heart chambers when compared to those without CF. Poorer lung function and male sex were associated with higher risk of early heart function abnormalities.

What does this mean and reasons for caution?

While heart problems are not typically the primary concern for people with CF, this study suggests that heart health should not be overlooked. Progressive lung disease can strain on the heart, and long-term inflammation and/or infections may also contribute to these heart changes. With the advent of highly effective treatments called CFTR modulators, people with CF are living longer and healthier lives. However, this new landscape also brings attention to possible long-term heart issues and calls for assessment on whether CF as well as CFTR modulators affect heart health in unexpected ways.



Cystic Fibrosis Research News

What's next?

While CF primarily affects other organs, this study shows that it might also have a significant impact on the heart. The findings suggest that regular heart monitoring could become an important part of CF care, especially for people with poorer lung function or males, who seem to be at higher risk. Early detection and intervention may help improve long-term heart health in people living with CF. However, further research is needed to understand the long-term effects of CF on heart health and to determine the best ways to protect the heart as people with CF live longer.

Original manuscript citation in PubMed

<https://pubmed.ncbi.nlm.nih.gov/39343639/>