Cystic Fibrosis Research News

Title:
Acute ST-elevation myocardial infarction in two young women with cystic fibrosis and cystic fibrosis-related diabetes

Lay Title:
Two cases of heart attacks in young women with cystic fibrosis and diabetes

Authors:
Sirisha Reddy Thambuluru[a], Sayyad Kyazimzade[b], Katherine A. Despotes[c,d] , Deepa Kirk[a], Jennifer L. Goralski[c,d]

Affiliations:
[a] *Division of Endocrinology & Metabolism, University of North Carolina, Chapel Hill, NC, USA*
[b] *Department of Internal Medicine, University of North Carolina, Chapel Hill, NC, USA*
[c] *Division of Pulmonary Diseases and Critical Care Medicine, University of North Carolina, Chapel Hill, NC, USA*
[d] *Division of Pediatric Pulmonology, University of North Carolina, Chapel Hill, NC, USA*

What was your research question?
To report the first two cases of people with CF (PwCF) and cystic fibrosis related diabetes (CFRD) who had a ST-elevation myocardial infarction (STEMI), a dangerous type of heart attack due to blockage of the heart's blood vessel by plaque.

Why is this important?
Heart disease and heart attacks have been rare in PwCF. PwCF were thought to be at low risk for heart disease because of low cholesterol, specifically low LDL ("bad cholesterol"). PwCF typically died due to lung disease at younger ages, while heart disease usually takes a longer time to develop. For this reason, when PwCF have symptoms of chest pain or shortness of breath, doctors generally focus their evaluation on the lungs. These two patient reports are important because they show that heart attacks and heart disease can happen in PwCF, and they may become more common because treatment for PwCF is changing.

What did you do?
We looked into possible risks of heart disease in the CF population. We looked to see if these two patients had known risk factors for heart disease in the general population such as high cholesterol, diabetes, family history of heart disease, smoking history, high blood pressure

(hypertension), and obesity. We also looked into potential risk factors for heart disease that are unique to PwCF.

What did you find?
Both of these patients with CF have diabetes and other risk factors for heart disease. Patient 1 had a history of high cholesterol. She had also gained weight and her LDL had increased more after starting modulator therapy (medications that fix the protein defect in CF). Patient 2 had high blood pressure and had had a lung transplant. Heart disease is a possible long-term complication of lung transplantation. We also discussed characteristics specific to PwCF such as high fat diets, inflammation, decreased antioxidant levels, and increased stiffness of heart vessels that may contribute to heart disease.

What does this mean and reasons for caution?
As PwCF live longer, and with treatments like modulators that improve not just lung function but also nutrition (and how fat is absorbed from food), doctors need to be aware of a potential increase in heart disease in PwCF, especially in those who already have classic risk factors. We suggest that providers think about heart disease when caring for PwCF who have chest pain and shortness of breath, so serious medical disease like a heart attack is not missed.

What's next?
We need more studies to understand how heart disease occurs in PwCF and how to screen for high cholesterol in PwCF. We also do not know the best way to treat high cholesterol in PwCF given possible medication interactions with other treatments, and this will also need further study.

Original Manuscript Citation in PubMed
https://pubmed.ncbi.nlm.nih.gov/34911664/