

# Cystic Fibrosis Research News

**Title:**

Cystic fibrosis transmembrane conductance regulator modulators and the exocrine pancreas: A scoping review

**Lay Title:**

An overview of CFTR modulators and pancreas function

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**What was your research question?**

We know that CFTR modulators greatly improve pulmonary (lung) function. We wanted to find out whether they would also improve pancreatic function. We used standard measures of pancreatic function to measure this.

**Why is this important?**

Currently, CFTR modulators are expected to improve life expectancy for people with CF because they greatly reduce the lung disease component of CF. This research is important because it tells us whether CFTR modulators are useful for improving pancreatic function. If

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so, patients with CF and severe pancreatic dysfunction may further benefit from CFTR modulator treatment, even in the absence of lung disease.

## What did you do?

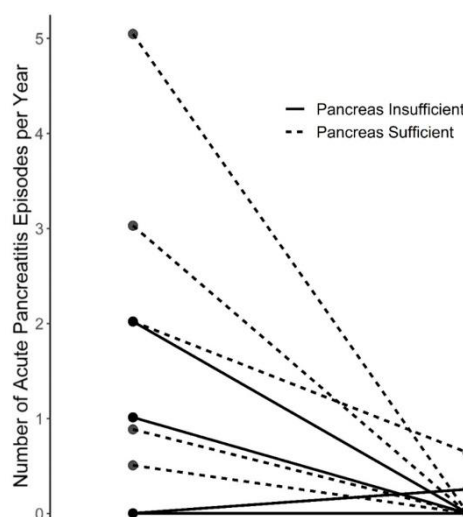
We performed a “scoping review” which is a systematic and rigorous way of studying the published literature. To do this, we first searched two large medical literature databases with an exhaustive strategy to get as many possible studies as we could. Then, we reviewed all of these studies in order to find high-quality, relevant studies. Last, we extracted the data from these studies, grouped the data together, and summarized our findings.

## What did you find?

We found that CFTR modulator therapy is associated with improvement in a number of pancreas function measures. The rate of pancreatitis (inflammation of the pancreas) decreased by 85% overall (**Figure**). Measures of pancreatic injury and stress (trypsinogen, amylase, lipase) decreased in subjects who were using CFTR modulators. Many subjects experienced an increase in fecal elastase (measures the ability of the pancreas to produce digestive enzymes), and 50% (16 out of 32 subjects in this arm of the study) regained pancreatic function (switching from pancreatic insufficiency to sufficiency).

## What does this mean and reasons for caution?

This data shows that CFTR modulators have a positive impact on the pancreas. Based on the few studies, small number of subjects, and variety of CF mutations and medications, the estimates of the impact are not precise. As such, our findings will need to be confirmed in bigger, better-controlled studies. Lastly, there have been reports that CFTR modulators might increase the risk of pancreatitis for some subjects. This is an area of caution that requires further study.



**Figure:** Number of acute pancreatitis episodes per year before and during CFTR modulator therapy in subjects with cystic fibrosis.



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## What's next?

We did not encounter any studies that linked pancreas function measures to gastrointestinal/stomach symptoms (like bloating, constipation). The next step is to study gastrointestinal symptoms and pancreas function measurements simultaneously in order to see whether these laboratory changes are linked to improvements in gastrointestinal symptoms.

## Original manuscript citation in PubMed

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