

Cystic Fibrosis Research News

Title:

Successful treatment of distal intestinal obstruction syndrome with N-acetylcysteine and polyethylene glycol via colonoscopy.

Lay Title:

Successful treatment of a blockage in the lower portion of the small intestine in patients with cystic fibrosis using a solution administered during a colonoscopy.

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What was your research question?

In patients with cystic fibrosis who develop a blockage in the lower portion of the small intestine, a disorder known as “distal intestinal obstruction syndrome” (DIOS), what other medical remedies exist if they fail to improve with oral laxatives?

Why is this important?

Patients with cystic fibrosis may develop a blockage in the lower portion of their small intestine due to thick mucus plugs in that region. Although most patients improve with oral laxatives, a number of patients may not respond to medications alone. Surgery is often required for these patients, but this carries increased risks of complications. Therefore, it is important to explore alternative treatments for this disorder that may eliminate the need for surgery.

What did you do?

A young woman with cystic fibrosis presented to the hospital with abdominal pain, abdominal distension, and constipation. Her evaluation revealed that she had a blockage in the lower portion of her small intestine due to thick mucus plugs, confirming a diagnosis of “distal intestinal obstruction syndrome.” She was treated initially with different types of oral laxatives, but her condition worsened despite these treatments. The surgery team was consulted to consider taking her to the operating room. However, given the risks associated

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with surgery, the gastroenterology team was consulted to consider treating with a colonoscopy instead.

What did you find?

There have been reports that the use of a solution created by mixing “N-acetylcysteine” and “polyethylene glycol” can be effective in breaking down thick mucus plugs in the intestine in patients with cystic fibrosis. In this case, the patient was taken emergently for colonoscopy during which a camera scope was gradually advanced from the rectum to the first portion of the large intestine. Within the valve between the small and large intestines, thick mucus and stool were visualized causing complete obstruction. The solution was injected at this site, leading to breakdown of the mucus plug and relief of the obstruction.

What does this mean and reasons for caution?

Given the effectiveness of treating distal intestinal obstruction syndrome with colonoscopy and injection of N-acetylcysteine and polyethylene glycol solution, many patients with this condition may be able to avoid surgery. Furthermore, surgery may be reserved for patients who ultimately fail to improve even after colonoscopy. Caution must be taken with colonoscopy, however, as this typically involves inflating the intestine with air or water during the procedure, which carries an increased risk of complications such as bowel perforation (formation of a hole in the intestine due to pressure from within).

What's next?

While there have been multiple individual reports of success with treatment of DIOS via colonoscopy, there are currently no studies to support this approach. Future directions should involve the conduction of well-designed studies to assess the safety and efficacy of colonoscopy with N-acetylcysteine and polyethylene glycol solution for treating DIOS.

Original manuscript citation in PubMed

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Image:

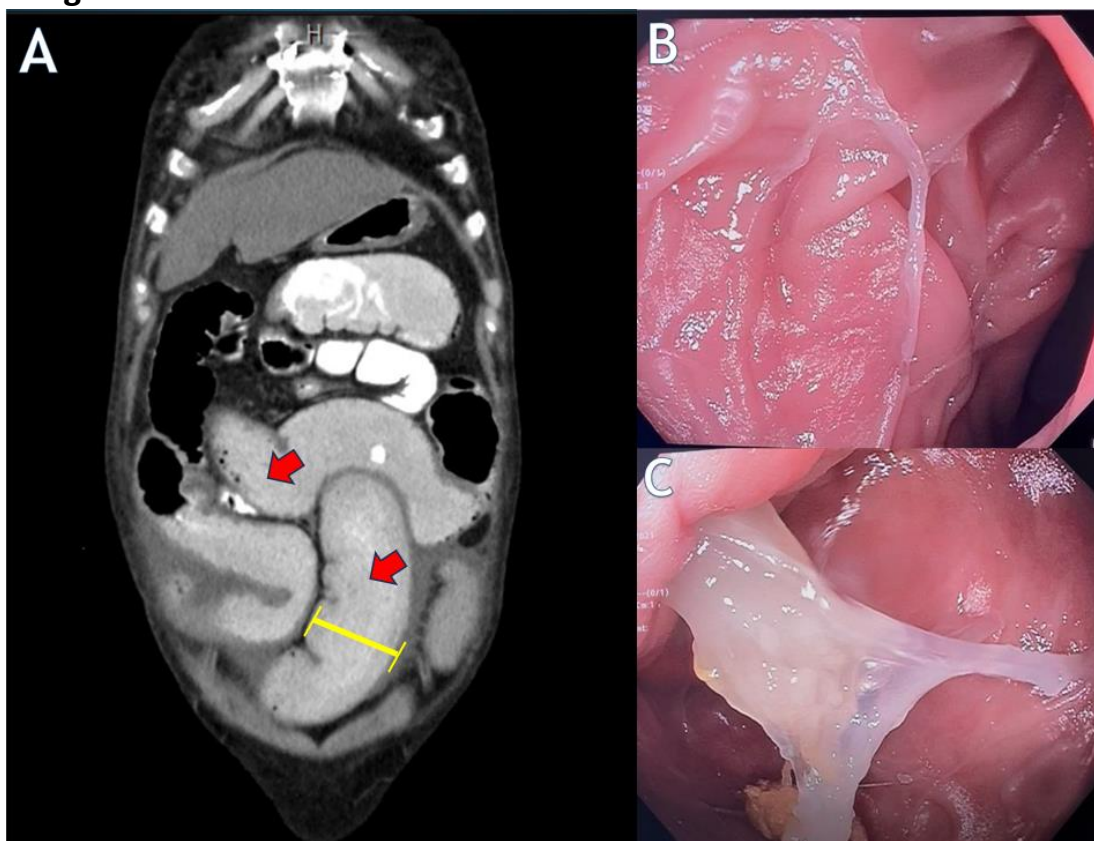


Figure 1. (A) Abdominal CT scan showing dilated loops of bowel (yellow line) and extensive debris in the lower portion of small intestinal (red arrows). (B) Matted, mucoid-adherent folds in the large intestine. (C) A mucus plug extending from the valve between the small and large intestine to the walls of the large intestine.