

APPENDICITIS IN CYSTIC FIBROSIS PATIENTS, A DIAGNOSIS DILEMMA.

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Background: Cystic Fibrosis (CF) is the most common inherited lethal disease in the Caucasian population. It is caused by mutations in the CFTR gene encoding the anion channel CF transmembrane conductance regulator leading to abnormally viscous mucus and secretions in lungs, gastrointestinal (GI) tract, pancreas, and hepatobiliary system. As lung function and survival improves with CF the extrapulmonary sequelae of CF will become an increasing driver of morbidity and mortality. Thus, increasing our understanding of the gastrointestinal manifestations of CF is of great importance. Abdominal pain is a frequent complaint amongst CF patients and it is difficult to differentiate CF-related vs non-CF-related causes. One of the more difficult diagnoses is appendicitis. The appendix in CF is at baseline distended due to inspissated secretions making radiographic diagnosis difficult. Also, clinical manifestations of appendicitis overlap with common GI sequelae of CF, such as distal intestinal obstruction syndrome, confounding the diagnosis.

Materials/Methods: We present a series of three patients with CF and abdominal pain concerning for appendicitis. We describe their presentation, management, and outcomes.

Results: Three patients with CF presented to Texas Children's Hospital with abdominal pain and concern for appendicitis given thickened and/or enlarged appendices noted on imaging. Patient 1, an 18-year-old female, had no fever or lymphocytosis and was discharged home on bowel regimen, but eventually had surgical appendectomy at an outside hospital. Patient 2, an 11-year-old male, presented with fever, no lymphocytosis, in the context of a respiratory infection. He was medically managed with antibiotics with resolution of abdominal pain. Patient 3, a 12-year-old female, had acute onset of severe abdominal pain and CT scan noted diffuse intestinal thickening and enlargement of appendix. She was also treated medically with resolution of pain.

Conclusions: This series illustrates the difficulty in diagnosing appendicitis in CF. It also highlights the importance of knowing common abdominal ultrasound and CT findings in CF in order to avoid false positive appendicitis diagnoses and unnecessary surgical treatment.