



GASTROINTESTINAL DISEASE IN CYSTIC FIBROSIS

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Manifestations of Cystic Fibrosis

- General**
- Growth failure (malabsorption)
 - Vitamin deficiency states (vitamins A, D, E, K)

- Nose and sinuses**
- Nasal polyps
 - Sinusitis

- Liver**
- Hepatic steatosis
 - Portal hypertension

- Gallbladder**
- Biliary cirrhosis
 - Neonatal obstructive jaundice
 - Cholelithiasis

- Bone**
- Hypertrophic osteoarthropathy
 - Clubbing
 - Arthritis
 - Osteoporosis

- Intestines**
- Meconium ileus
 - Meconium peritonitis
 - Rectal prolapse
 - Intussusception
 - Volvulus
 - Fibrosing colonopathy (strictures)
 - Appendicitis
 - Intestinal atresia
 - Distal intestinal obstruction syndrome
 - Inguinal hernia

- Lungs**
- Bronchiectasis
 - Bronchitis
 - Bronchiolitis
 - Pneumonia
 - Atelectasis
 - Hemoptysis
 - Pneumothorax
 - Reactive airway disease
 - Cor pulmonale
 - Respiratory failure
 - Mucoic impaction of the bronchi
 - Allergic bronchopulmonary aspergillosis

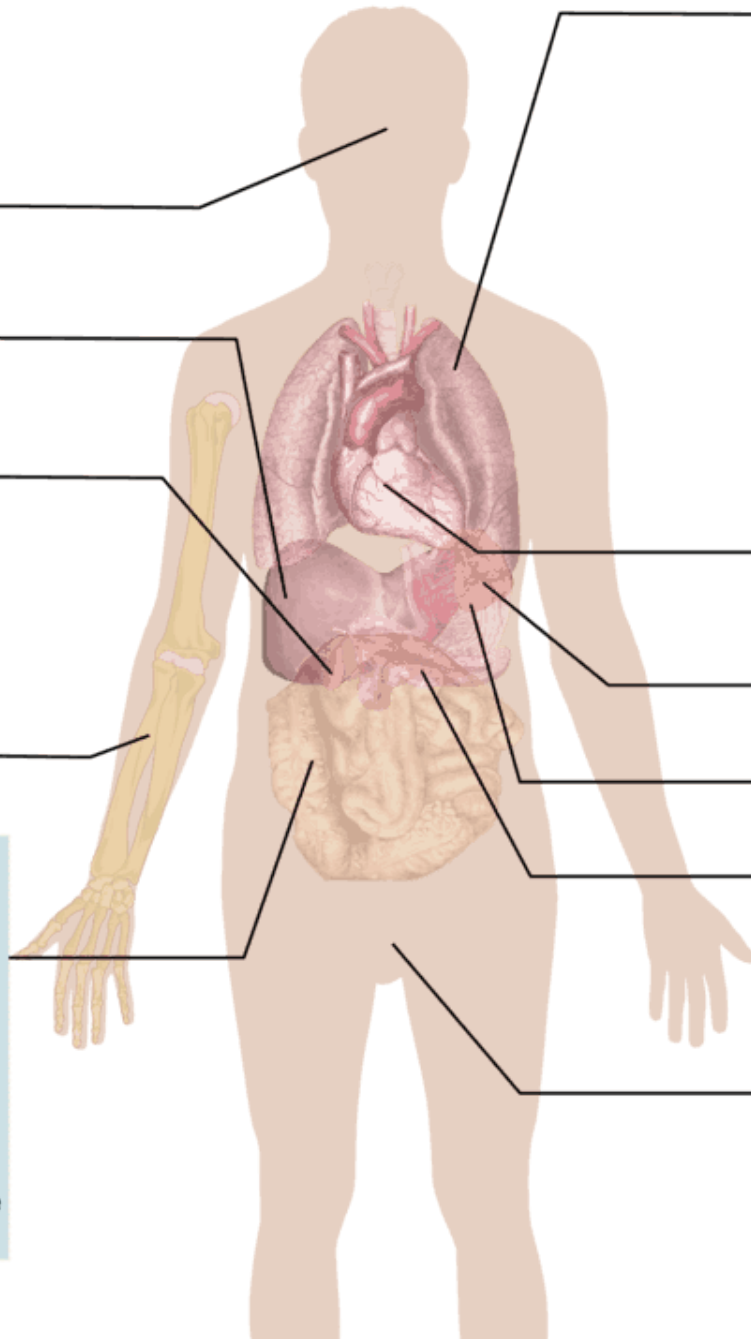
- Heart**
- Right ventricular hypertrophy
 - Pulmonary artery dilation

- Spleen**
- Hypersplenism

- Stomach**
- GERD

- Pancreas**
- Pancreatitis
 - Insulin deficiency
 - Symptomatic hyperglycemia
 - Diabetes

- Reproductive**
- Infertility (aspermia, Absence of vas deferens)
 - Amenorrhea
 - Delayed puberty



GERD

- More common in CF patients
 - ↑ intra-abdominal pressure
 - ↓ basal tone of LES
 - ↑ gastric pH
 - Delayed gastric emptying ± CFRD
 - Supine positioning during chest physiotherapy
- Acid suppression?

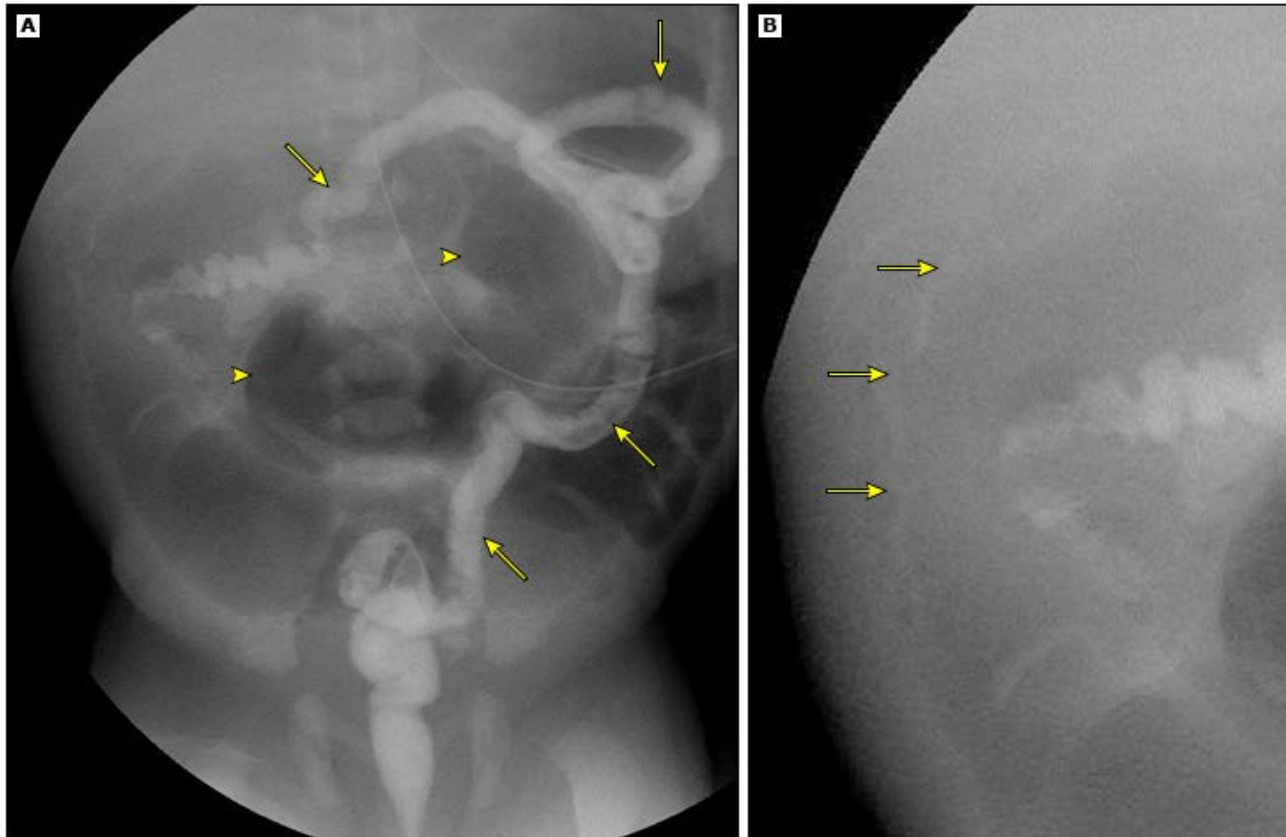


Meconium ileus

- Obstruction of the terminal ileum with inspissated meconium
 - 10% of CF patient present as MI
 - 80-90% of MI patients have CF
- Complications:
 - Simple MI
 - Complex MI
- Conservative management vs surgery



Cystic fibrosis and meconium ileus on contrast enema in infant



A contrast enema on a 9-hour-old infant with prenatal diagnosis of intestinal obstruction.

(A) A microcolon (arrows) and dilated gas-filled loops of small bowel (arrowheads), characteristic of meconium ileus.

(B) Calcifications (arrows) in the right upper quadrant, consistent with meconium peritonitis from in utero perforation.

Courtesy of Michael Callahan, MD.

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DIOS

- Acute complete/incomplete obstruction of the ileocecum
- Unique to CF
- Management
 - Oral/NG therapy (ORS/Gastrografin)
 - Enemas (Gastrografin)
 - Surgery
- Osmotic laxatives 6-12 mo



Cystic fibrosis, distal intestinal obstruction, and therapeutic Gastrografin enema



Therapeutic enema with diatrizoate meglumine and diatrizoate sodium (Gastrografin) in a patient with cystic fibrosis and small bowel obstruction due to distal intestinal obstruction syndrome (DIOS). A spot view of the cecum shows a contrast-filled cecum (asterisk). Two days following the enema, the patient's symptoms improved with normal stooling and ability to tolerate solid foods.

Courtesy of Michael Callahan, MD.

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Constipation

- Gradual onset of fecal impaction of the colon: sigmoid → proximally
- 25-50% of CF patients: large colonic fecal load
 - abnormal intestinal fluid composition
 - dysmotility
 - pancreatic insufficiency
- Osmotic laxatives ± stimulant laxatives
- Lubiprsotone, Linaclotide



Fibrosing colonopathy

- Severe intestinal fibrotic process → strictures ± ascites
- Large doses of PERT > 10.000 lipase units/kg/day
- Methacrylic acid copolymer in the enteric coating?

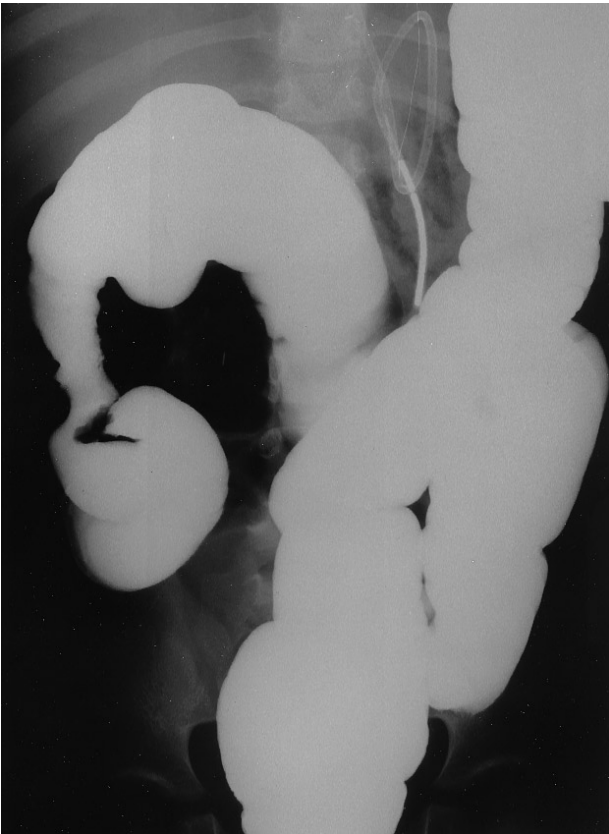


Imaging :

Rx: thickened colon

Contrast irrigography: colonic strictures, reduction in length, abnormal haustra

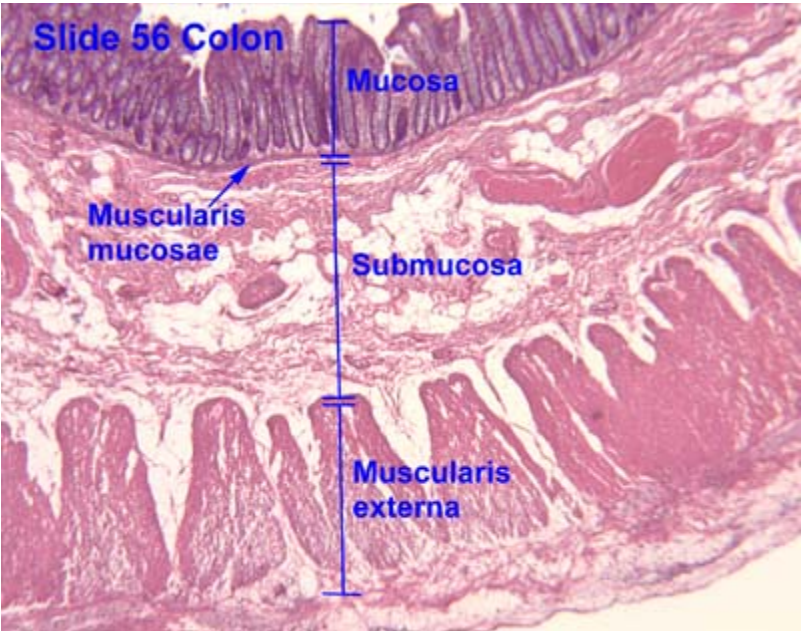
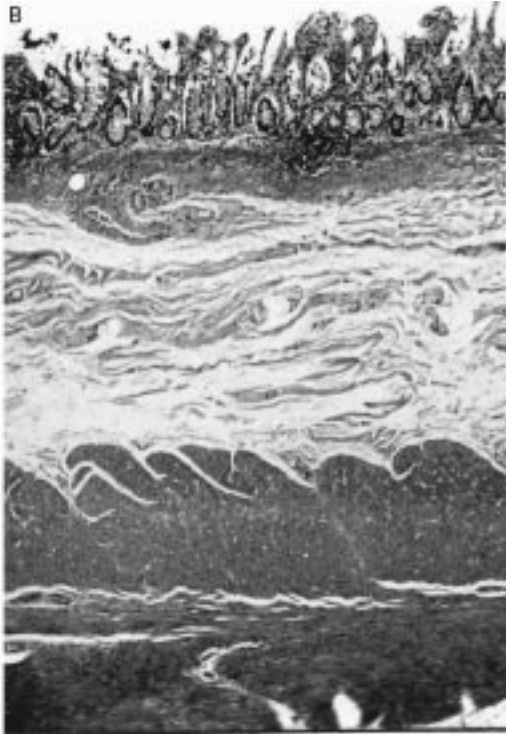
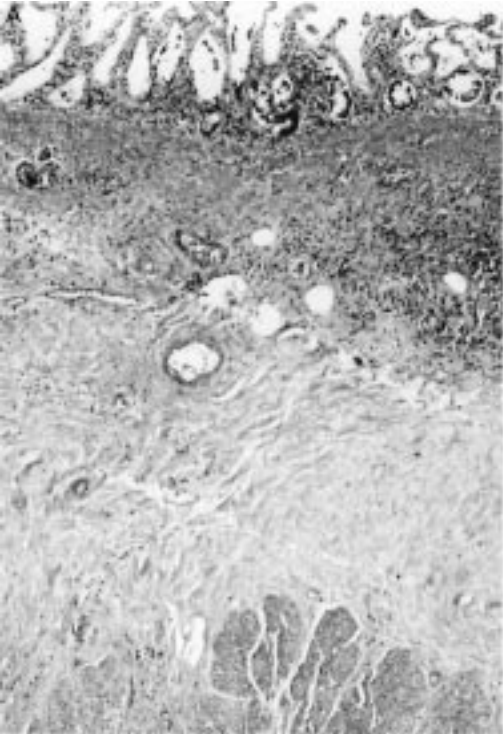
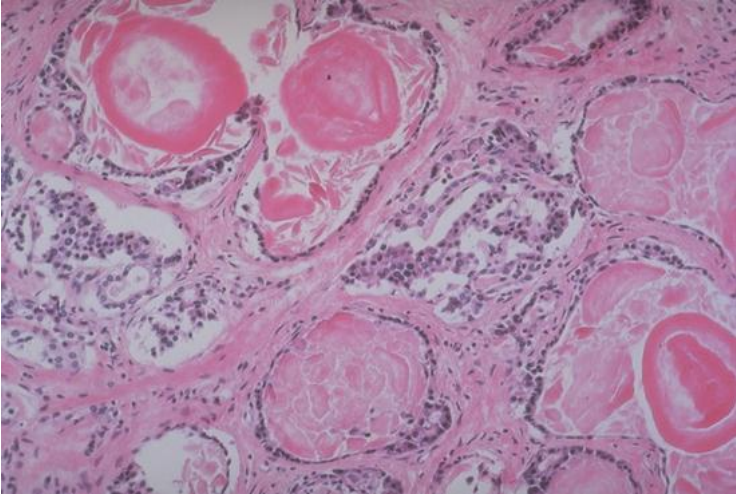
Ultrasound: thickening of the intestinal wall by more than 2 mm, reduction of peristalsis and accumulation of fluid in the affected area.

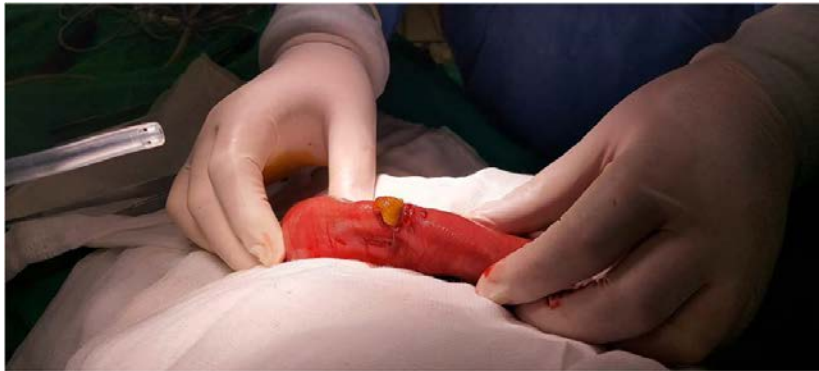


N Engl J Med. 1997 May 1;336(18):1283-9

Pediatr Radiol (1997) 27: 315-316

Histopathology: thickening of the submucosa through fibrosis of the connective tissue → reduction of the lumen of the large intestine (at any level), with normal epithelium / minimal inflammatory changes / eosinophilic infiltrate





SIBO

- ↑↑↑ bacterial content in the upper GI tract
- Normal nutrients → toxic substances → enterocyte damage → malabsorption → malnutrition
- Nonspecific clinical symptoms
- 35% CF patients
- Rifaximin



Va multumesc!