Cystic fibrosis is the most common genetic disorder affecting the white population. This autosomal recessive disease is caused by a mutation in the gene for the protein cystic fibrosis transmembrane conductance regulator (CFTR) found on chromosome 7. CFTR is located in epithelial cells of the airway, gastrointestinal tract, sweat glands, and genitourinary system. The CFTR gene mutation causes a deficiency of chloride ion secretion, which leads to sodium retention and fluid absorption causing increased viscosity of luminal secretions manifesting as malfunction of electrolyte transport (Figure 1). This presents as multiorgan system exocrine abnormalities. Manifestations of this disease include sinusitis, nasal polyps, recurrent lung infections, chronic lung disease, bowel obstruction, pancreatic insufficiency, and infertility. Radiologists play a key role in diagnosing and monitoring disease progression in patients with cystic fibrosis.

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Sinonasal Manifestations
The prevalence of rhinosinusitis in patients with cystic fibrosis borders on 100%. Viscous secretions impair mucociliary clearance, leading to infection and inflammation. Nasal polyps may be identified on endoscopic evaluation in up to 86% of patients with cystic fibrosis. The most frequent findings on CT are nonspecific and include bilateral medial displacement of the lateral nasal wall and uncinate process demineralization (Figure 2).

The prevalence of rhinosinusitis in patients with cystic fibrosis approaches 100%.

Pulmonary Manifestations
Abnormal chloride transport causes thick viscous secretions with reduced mucociliary clearance, which in turn causes airway obstruction and recurrent infections. Inflammation of the airways causes gas trapping that leads to hyperinflated lungs, the earliest pulmonary radiographic finding with or without atelectasis. Airways become filled with thick, viscous mucus causing mucoid impaction, which has a “fingerglove” appearance (Figure 3). Cystic fibrosis is characterized by recurrent infections, and it has an upper lobe predominance. Colonization occurs with bacteria such as Haemophilus influenzae, Staphylococcus aureus, Pseudomonas aeruginosa, and Burkholderia cepacia complex.

The earliest pulmonary radiographic finding of cystic fibrosis is hyperinflated lungs.

Chronic inflammation of the airways leads to progressive irreversible dilatation of the main bronchus or bronchi (bronchiectasis). The diameter of the normal bronchus should measure approximately 0.65 to 1.0 times that of the adjacent pulmonary artery branch. Bronchiectasis causes a bronchoarterial
Gastrointestinal Manifestations

Gastrointestinal (GI) manifestations range from obstructive diseases such as meconium ileus and intussusception to an increased risk of Crohn disease. Approximately 15% to 20% of infants with cystic fibrosis present with distal small bowel intestinal obstruction secondary to meconium ileus. This finding is the earliest GI manifestation of cystic fibrosis, which occurs because of abnormally thick inspissated meconium. Contrast enema with a water-soluble contrast medium is often diagnostic and therapeutic, demonstrating a microcolon (unused colon due to impaired passage of succus entericus) and a dilated terminal ileum containing filling defects representing impacted meconium (Figure 10). Complications of meconium ileus include volvulus, bowel necrosis, perforation, meconium peritonitis, and meconium pseudocyst formation.

GI manifestations of cystic fibrosis range from meconium ileus and intussusception to an increased risk of Crohn disease.
Distal intestinal obstruction syndrome is the meconium ileus equivalent in older children and adults, occurring mainly in the second and third decades of life. It is seen in up to 10% to 15% of patients with cystic fibrosis. Bowel obstruction occurs in the distal ileum and right colon. Causes are hypothesized to be secondary to inspissated secretions, pancreatic insufficiency, undigested food residue, disordered intestinal motility, fecal stasis, and dehydration. Treatment usually is conservative and may include a therapeutic water-soluble (Gastrografin) enema.

**Pancreatic Manifestations**

Cystic fibrosis represents the most common cause of inherited pancreatic insufficiency. This manifestation most often presents as exocrine insufficiency, with endocrine insufficiency less frequently. Fat deposition in the pancreas is the most common imaging finding. Pancreatic ductal obstruction can be caused by calcium-rich, thickened secretions. Other ductal abnormalities include strictures with beaded appearance seen on cholangiopancreatography (Figure 11).
Complete pancreatic fatty replacement is the most common pancreatic abnormality seen on CT or MRI and is found in 56% to 93% of patients with cystic fibrosis. However, pancreatic fibrosis also may occur. The average age of fatty replacement is 17 years. The imaging appearance varies according to the amount of fat or fibrosis, with fat appearing T1-hyperintense on MRI and hypoattenuating on CT (Figure 12). Fibrosis is hypointense on both T1- and T2-weighted MR images.

Lipomatous pseudohypertrophy of the pancreas is a rare disorder associated with cystic fibrosis, but may occur as a distinct entity. The exact etiology is unknown. It is characterized by pancreatic enlargement, fatty replacement of exocrine tissue, and preservation of the ducts and islets.

The differential diagnosis of pancreatic fatty replacement also includes Shwachman-Diamond syndrome, obesity, diabetes, and age-related fatty infiltration.

Shwachman-Diamond syndrome is the second most common cause of inherited pancreatic insufficiency after cystic fibrosis. It is a rare autosomal recessive disorder caused by a mutation in the Schwachman-Diamond-Bodian gene on chromosome 7. It is characterized by exocrine pancreatic insufficiency (similar to cystic fibrosis), cytopenias, and skeletal abnormalities such as metaphyseal dysostosis (Figure 13).

Pancreatic cysts are relatively common in patients with cystic fibrosis, but pancreatic replacement by cysts greater than 1 cm, as seen in pancreatic cystosis, is a rare manifestation of cystic fibrosis-related pancreatic disease (Figures 14 and 15). The etiology may relate to ductal dilatation resulting from blockage by abnormally concentrated pancreatic secretions. The differential diagnosis of pancreatic cystosis includes...
von Hippel-Lindau disease, polycystic kidney disease, and lymphatic malformations.

**Hepatic Manifestations**

The onset of liver disease in patients with cystic fibrosis is usually in early childhood. Hepatic steatosis presents as an echogenic liver on ultrasound and demonstrates decreased parenchymal attenuation on CT (Figure 16). Focal biliary cirrhosis occurs in 40% of patients with hepatic involvement, with 1% progressing to end-stage liver disease. In hepatic cirrhosis, the liver appears nodular with splenomegaly, portal vein enlargement, and varices seen in the setting of portal hypertension. Biliary involvement in patients with cystic fibrosis includes microgallbladder, cholelithiasis, gallbladder wall thickening and contraction, and involvement of intra- and extrahepatic bile ducts.

**Conclusion**

Radiography is the core diagnostic and staging imaging examination tools used in the evaluation of patients with cystic fibrosis. Given that these patients will likely require multiple imaging studies as their disease progresses, one has to be mindful of the amount of ionizing radiation used in this pediatric population. This CME activity emphasizes that with thorough understanding of this multisystem disease, early identification, and appropriate therapy can improve the overall life expectancy and quality of life of affected patients.

**References**

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1. Which one of the following is the earliest GI manifestation of cystic fibrosis?
   A. Gastric volvulus
   B. Anal atresia
   C. Meconium ileus
   D. Intestinal malrotation
   E. Hirschsprung disease

2. Which of the following is/are complications in a transplanted lung for a patient with cystic fibrosis and end-stage lung disease?
   A. Infection
   B. Acute rejection
   C. Main stem bronchus stricture
   D. Bronchiolitis obliterans
   E. All of the above

3. Which one of the following is the earliest pulmonary radiographic finding of cystic fibrosis?
   A. Bronchiectasis
   B. Hyperinflated lungs
   C. “Finger in glove” appearance
   D. Abscess
   E. Ground-glass appearance

4. Which one of the following statements regarding the distal intestinal obstruction syndrome is true?
   A. It occurs only in newborns.
   B. It usually requires surgical resection of the involved bowel.
   C. It can be treated with a water-soluble enema.
   D. The intestinal obstruction occurs at the level of the rectosigmoid colon.
   E. It occurs in 50% of patients with cystic fibrosis.

5. All of the following should be included in the differential diagnosis of pancreatic fatty replacement, except
   A. cystic fibrosis
   B. Shwachman-Diamond syndrome
   C. diabetes mellitus
   D. pancreatic fibrosis
   E. obesity

6. All of the following are hepatobiliary manifestations of cystic fibrosis, except
   A. hepatic steatosis
   B. microgallbladder
   C. cholelithiasis
   D. hepatic adenoma
   E. cirrhosis

7. Which one of the following is the most common pancreatic abnormality seen on imaging in a patient with cystic fibrosis?
   A. Pancreatic fatty replacement
   B. Pancreatic fibrosis
   C. Pancreatic duct strictures
   D. Pancreatic cysts
   E. Pancreatic duct obstruction

8. Patients with cystic fibrosis have an increased risk of
   A. ulcerative colitis
   B. Crohn disease
   C. Hirschsprung disease
   D. asplenia
   E. congenital diaphragmatic hernia

9. Which one of the following is the cause of the “finger-in-glove” appearance in the lungs on chest radiographs of a patient with cystic fibrosis?
   A. Interstitial emphysema
   B. Segmental atelectasis
   C. Mucoid impaction
   D. Recurrent pneumonia
   E. Pulmonary infarction

10. All of the following are manifestations of cystic fibrosis, except
    A. sinusitis
    B. pancreatic insufficiency
    C. infertility
    D. bowel obstruction
    E. ventricular septal defect