Imaging of abdominal and thoracic manifestations of cystic fibrosis

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Learning objectives

Describe the radiological findings in cystic fibrosis (CF) with special attention to the Bransfield and Bhalla classification.

The imaging findings of the abdominal manifestations of cystic fibrosis will also be described.
Background

The Bhalla Scoring System is based on HRCT findings and assesses the severity and extent of bronchiectasis, peribronquial thickening, extent of mucus plugging, sacculations or abscesses, number of generations of bronchial divisions involved, number of bullae, emphysema and consolidation or collapse. Each one of this nine parameters get a score from 0 to 3. The total score is useful for comparing with subsequent studies.
Cystic fibrosis (CF) is the most common inherited fatal disease in whites, every year affecting approximately 1 in every 3500 children born. It is also the most common cause of pulmonary insufficiency in the first three decades. Inheritance is autosomal recessive, and the gene mutations have been identified at a single locus on the long arm of chromosome 7 where the CF transmembrane conductance regulator (CFTR) is produced. This protein causes abnormal chloride ion transport on the apical surface of exocrine gland epithelial cells and it results in viscous secretions which affect multiple organ systems.

Respiratory failure is the major cause of death.

Abdominal complaints are common and nearly all organ systems are affected, including the hepatobiliary system, pancreas, and gastrointestinal tract and less commonly the kidneys.

**PULMONARY MANIFESTATIONS**

The abnormal protein is responsible for the mucus get thicker and its low water content. The airway obstruction by thick mucus, facilitates the development of infections and inflammation of the bronchial tree and bronchiectasis.

On chest radiographs, signs of air trapping are the manifestation of the airway obstruction. Peribronchial wall thickening and cylindrical bronchiectasis, varicose or cystic changes depending on the time and severity of the disease, can also be seen. Nodular opacities are often, as a result of bronchiectasis occupied by mucus or pus, lobular consolidation, atelectasis and subpleural bullae. Fig. 1 on page  Fig. 2 on page 8

The most observed morphologic CT abnormalities are bronchiectasis and peribronchial wall thickening. Furthermore branched or v-shaped opacities, atelectasis and consolidation and subpleural bullae are seen. Fig. 3 on page 9 Fig. 4 on page 10 Fig. 5 on page 11 Fig. 6 on page 12 Fig. 7 on page 13 Fig. 8 on page 14

The small airways are especially vulnerable to infection, inflammation and obstruction that characterises CF lung disease. Branching or nodular centrilobular opacities (tree in-bud pattern) are frequently present and may be an early sign of disease. They reflect the presence of bronchiolar dilation with associated mucous impaction. Focal areas of decreased attenuation and vascularity on inspiratory CT and air trapping on expiratory CT are common. Fig. 9 on page 15 Fig. 10 on page 16
For monitoring the disease and differentiating radiological irreversible changes from others susceptible to treatment, several classification system have been developed. The most known are the Brasfield Scoring System based on plain film and the Bhalla Scoring System based on HRCT findings.

The Brasfield Scoring System take in account the air trapping, linear markings, nodular cystic lesions, large lesions and general severity.

The Bhalla Scoring System is based on HRCT findings and assess the severity and extent of bronchiectasis, peribronquial thickening, extent of mucus plugging, sacculations or abscesses, number of generations of bronchial divisions involved, number of bullae, emphysema and consolidation or collapse. Each one of these nine parameters gets a score from 0 to 3. The total score is useful for comparing with subsequent studies. Fig. 11 on page 17

**Hepatobiiliary Manifestations**

Liver disease is cause of death in patients with CF (3). It has classically been attributed to abnormally thickened secretions that accumulate within bile ducts, slowing biliary flow and concentrating caustic bile components in the hepatic tissues.

Fatty infiltration of the liver has been reported in 30% of all CF patients. It is rarely symptomatic unless associated with palpable hepatomegaly. The cause of steatosis in CF patients is unknown, and there has been no proven relationship between steatosis and the subsequent development of focal biliary fibrosis or cirrhosis.

Biliary abnormalities range from cholelithiasis and sludge to ductal strictures and sclerosing cholangitis. A common abnormality is the microgallbladder and cholelithiasis. Gallbladder wall thickening is nonspecific, especially in the setting of cirrhosis, ascites, hypoalbuminemia, and nutritional deficiencies.

Focal biliary fibrosis is considered a characteristic histopathologic lesion in CF and has been estimated to be present in 78% of patients older than 24. The classicsonographic appearance is hyperechoic periportal thickening due to fibrosis or focal fat, with diffuse increased hepatic echogenicity.

The incidence of progression from focal biliary fibrosis to multinodular cirrhosis is unknown. Imaging findings are similar to those in other adult patients with cirrhosis and include a small, nodular, heterogeneous liver with portal hypertension, varices, and ascites. Fig. 12 on page 18
PANCREATIC MANIFESTATIONS

Endocrine gland dysfunction thought to result from fibrosis and gland atrophy.

Complete fatty replacement is the most common pancreatic finding at imaging in adult CF patients. Sonographic findings include hyperechoic and atrophic pancreatic parenchyma. A gland that is markedly enlarged with fatty replacement has been termed lipomatous pseudohypertrophy of the pancreas. Fig. 15 on page 21

Pancreatic calcifications are infrequent and they are usually found along the course of pancreatic ducts.

Pancreatic cysts are relatively common in CF patients, are usually small, measuring 1-3 mm. Occasionally, aggregates of true epithelium-lined cysts completely replace the pancreas, a condition referred to as pancreatic cystosis. The imaging appearance can be similar to those of cystic pancreatic neoplasms, von Hippel-Lindau disease, and autosomal dominant polycystic kidney disease. Fig. 13 on page 19

GASTROINTESTINAL MANIFESTATIONS

Gastroesophageal reflux is seen in up to 27% of CF patients younger than 5 years. Complications related to chronic reflux disease, namely, esophagitis, strictures, and the development of Barrett metaplasia, can be seen in the adult.

An increased frequency of peptic ulceration of the gastric and duodenal mucosa, celiac sprue and giardiasis has been reported.

Distal intestinal obstruction syndrome or meconium ileus equivalent is a well-known complication and is thought to be caused by pancreatic insufficiency, thickened intestinal secretions, undigested food remnants, poor motility, and fecal stasis with resultant impaction of mucofeculent material in the distal ileum and right colon. The most common radiographic finding is a bubbly soft tissue mass in the right lower quadrant.

Another cause of right lower quadrant pain is intussusception. It is most frequently ileocolic and is related to an inspissated fecal mass that acts as a lead point. Radiologic appearances of intussusception range from a doughnut or pseudokidney appearance at US to the target sign of edematous bowel and intermixed mesenteric fat at CT. Although a watersoluble contrast enema may allow successful reduction, recurrence rates are high.
Acute appendicitis is somewhat unusual despite the fact that the appendiceal lumen is often swollen and filled with inspissated secretions at autopsy. Differentiating acute appendicitis from a chronically distended appendix with imaging alone is difficult if not impossible.

The colon is also often abnormal in patients with CF. Proximal colonic wall thickening, pericolonic fat proliferation, and mesenteric fat infiltration have all been reported.

Fibrosing colonopathy is a known cause of colonic stricture and obstruction and typically involves the right colon. It is thought to be related to high-strength pancreatic enzyme replacement (5) and is almost exclusively reported in children.

Pneumatosis intestinalis is usually confined to the colon in patients with CF and often coincides with the development of obstructive lung disease (5,35). It is thought to result from the dissection of air from the pulmonary interstitium to the perivasculatc connective tissue planes below the diaphragm. It is most often clinically silent.

As survival increases, gastrointestinal malignancies are now more commonly reported as CF patients live into adulthood and the diagnosis is often delayed. Causes are unclear but may be related to differential expressions of the CFTR gene or chronic pathologic changes to the colonic mucosa.

RENAL MANIFESTATIONS

The most prevalent entity is nephrolithiasis which is estimated to occur in 3.0%-6.0% of CF patients. the CFTR protein is expressed in abundance in the kidney, and abnormal gene expression may result in subtle alterations in the concentrating and diluting of urine. Stones are usually composed of calcium oxalate, and patients are often found to have hyperoxaluria, decreased levels of urinary citrate, and depressed urinary volumes.

Secondary renal complications, such as interstitial nephritis due to antibiotic therapy and amyloidosis, are likely to become more prevalent. Fig. 16 on page 22
Fig. 1: Frontal chest X ray in CF patient shows diffuse interstitial disease with bronchiectasis, peribronchial wall thickening and nodular densities of mucoid impaction. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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**Fig. 2:** Frontal chest X-ray in CF patient shows nodular densities. Lateral chest X-ray shows a middle lobe consolidation with loss of volume separate by fissures.

References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Fig. 3: HRCT scan in CF patient demonstrates peribronchial wall thickening. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Fig. 4: HRCT scan in CF patient demonstrates cylindrical bronchiectasis and peribronchial wall thickening. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Fig. 5: HRCT scan in CF patient demonstrates varicose bronchiectasis.

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**Fig. 6:** HRCT scan in CF patient demonstrates cystic bronchiectasis. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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**Fig. 7:** Fig. 7: HRCT scan in CF patient demonstrates a cystic bronchiectasia and a consolidation area. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Fig. 8: HRCT scan in CF patient demonstrates branched and v-shaped opacities and bronchiectasis occupied by mucus. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Fig. 9: HRCT scan in CF shows mosaic pattern with air trapped areas and nodular centrilobular opacities. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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**Fig. 10:** HRCT scan in CF shows centrilobular opacities. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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<table>
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<th>Severeidad de las bronquectasias</th>
<th>Ausentes</th>
<th>Medio (diámetro de la luz discretamente mayor que la arteria acompañante)</th>
<th>Moderado (2-3 veces mayor).</th>
<th>Grave (más de 3 veces mayor).</th>
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<th>Engrosamiento peribronquial</th>
<th>Ausente</th>
<th>Medio (enrojecimiento de la pared 1-2 veces el diámetro del vaso).</th>
<th>Moderado (enrojecimiento de la pared 1-2 veces el diámetro del vaso).</th>
<th>Grave (enrojecimiento mayor de 2 veces el diámetro del vaso).</th>
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<th>1.95</th>
<th>6 a 9</th>
<th>Más de 9</th>
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<td>Más de 9</td>
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<tr>
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<td>-5a generación</td>
<td>-6a generación</td>
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<td>Bilateral -4.</td>
<td>-4</td>
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<td>5</td>
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<td>Ausentes</td>
<td>Subsegmentarias</td>
<td>Segmentarias/lobares</td>
<td></td>
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</tbody>
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**Fig. 11:** Modified Bhalla scoring system. References: Turcios N, Aponte V, Jenkins M, Leitman, Maccauley. Cystic fibrosis: scoring system with thin section CT. Radiology. 1991;179:753-8.

Fig. 12: Fig. 12: US image in a patient with CF demonstrates diffusely increased hepatic echogenicity with periportal increased echogenicity. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Fig. 13: US image of pancreas in a patient with CF shows an heterogeneous echogenicity due to small cysts

References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Fig. 14: US image of the right iliac fossa in a patient with CF. The cecal appendix is distended due to persistent thickened secretions. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Fig. 15: US image of pancreas in a patient with CF shows a normal size gland with diffusely increased pancreatic echogenicity. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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**Fig. 16:** US image of the right kidney shows a non-obstructive stone located in the middle caliceal group. References: Radiology Department, Hospital Universitario Central de Asturias. Spain

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Conclusion

CF is a multisystemic disease and the radiologist plays an important role. Awareness of its pulmonary manifestations is important to successfully guide management of cystic fibrosis.
References

