

# Ultrasonography and magnetic resonance imaging findings of abdominal complications of cystic fibrosis

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**Abstract.** – In this study, our aim is to discuss the imaging findings of the abdominal complications seen in Cystic Fibrosis (CF), with ultrasonography (US) and magnetic resonance imaging (MRI). Our first case, a 22 year-old female showed changes characterized by severe biliary involvement such as cholelithiasis and cholangitis. Our second case was a 36 year-old male with severe hepatic manifestations. In this case signs of severe cirrhosis, portal hypertension and marked splenomegaly were seen. Micro-gallbladder, cholecystolithiasis and diffuse fatty infiltration of the pancreas were seen in both cases. In conclusion: US and MRI are effective and noninvasive imaging techniques to evaluate the abdominal complications of cystic fibrosis.

*Key Words:*

Cystic fibrosis, Abdominal manifestation, US, MRI.

## Introduction

Cystic Fibrosis (CF) is an autosomal recessive fatal disease more commonly seen in white people. Liver complications are the second most common cause of death in CF patients coming after pulmonary complications<sup>1</sup>. Number of the adult patients with CF has been increasing all over the world. That's why early diagnosis and treatment of the complications like diabetes mellitus, hepatobiliary disorders, cirrhosis and portal hypertension are important.

In this study, our aim was to discuss the imaging appearances of abdominal US, MRI/MRCP in these 2 cases of CF with severe hepatobiliary complications.

## Case Presentation

### Case 1

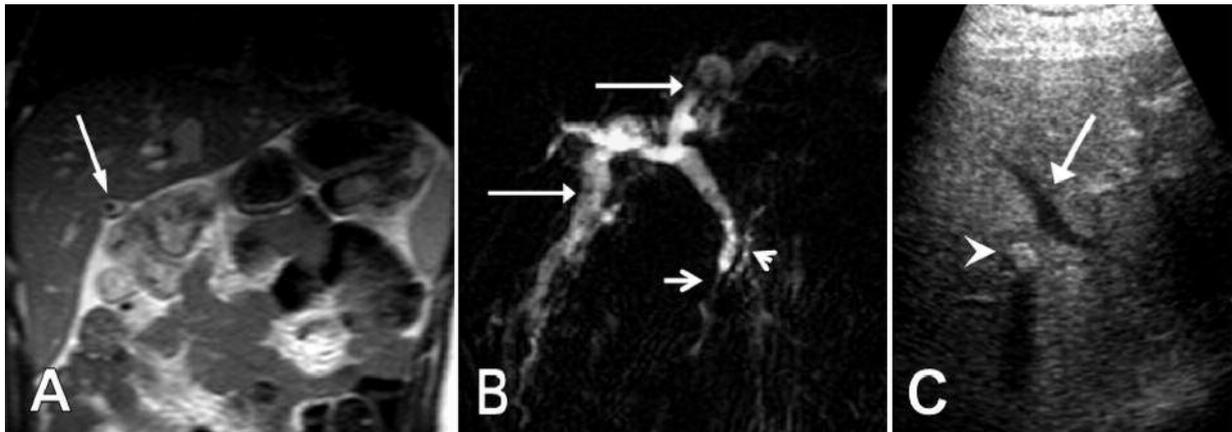
22 year-old woman had been followed in our Pediatric Clinic with CF diagnosis since 8 years. She admitted to our Clinic with the manifesta-

tions; right upper quadrant abdominal pain, soft colored gaita and dark colored urine which all appeared in last few days. Other diagnoses of this woman having cholangitis attacks during last one year period were; diabetes mellitus, chronic pulmonary infection, cholecystolithiasis, choledocholithiasis and papillary stenosis. She had undergone ERCP and during ERCP papillotomy and extractions of gallstones were performed twice in a year, 2 years ago.

On upper abdominal US and MRI/MRCP imaging studies; hepatomegaly (cranio-caudal length 18 cm), calcular images in intrahepatic and extrahepatic biliary ducts, dilatations and contour irregularities were observed. Gallbladder was observed as micro gallbladder (25×13 mm) and there were two milimetric calculi in the lumen of the gallbladder (Figure 1A). Increased echogenicity of the liver associated with hepatosteatosis and increased echogenicity of the pancreas associated with fatty change observed on US studies. Contours of the pancreas were difficult to assess with MRI. Pancreatic ductus was observed with irregular borders, some parts mildly dilated and stenotic with MRCP (Figure 1B). Dilatations of intrahepatic and extrahepatic biliary ducts, stenoses in some parts, and multiple milimetric calculi observed on ERCP and US study (Figure 1C). Multiple cholesterol stones were taken out by papillotomy. A stent replaced to distal part of common bile duct. Acute cholangitis and cholangiolithiasis diagnosis made with these imaging studies. Acute cholangitis of this patient was treated with intravenous antibiotic treatment.

### Case 2

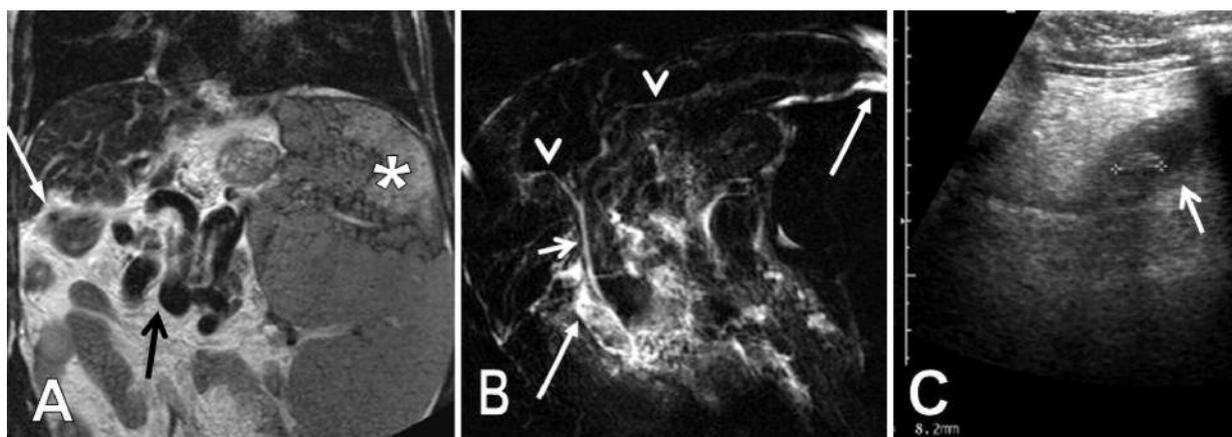
36 year-old men who had undergone partial splenic embolization three months ago because of splenomegaly and hypersplenism. Besides CF, other diagnosed problems of this patient, who



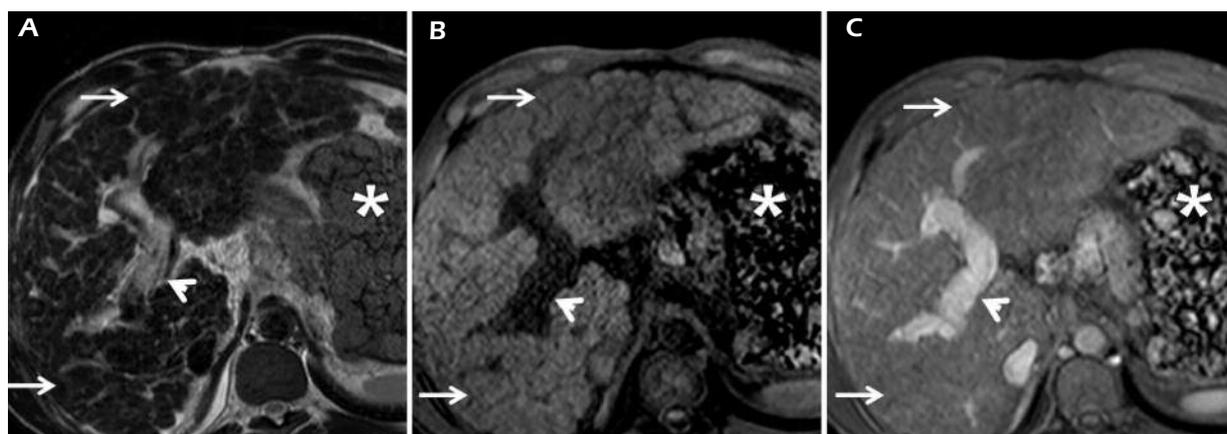
**Figure 1.** Case 1 (**A-C**). Coronal T2-weighted MR images (**A**) shows, hepatomegaly, micro gallbladder and a milimetric calculus in the lumen (*long arrow*). MRCP image (**B**) shows marked stenosis of the common bile duct at papillary level (*short arrow*). Irregular dilations of the intrahepatic and extrahepatic biliary channels with milimetric calculi (*long arrow*), sporadically decreased arborization at peripheric areas, irregular dilations and stenosis of the pancreatic channel (*arrow heads*). US image (**C**) of the same case shows dilatation of the intrahepatic biliary channels (*short arrow*) and a biliary stone in the channel (*arrow heads*).

was hospitalized for control were multilobular biliary cirrhosis, portal hypertension, diabetes mellitus, chronic bronchitis and pancytopenia. With US and MRI; contours of the liver were found marked and lobular; the liver parenchyma was heterogenous with diffuse nodularity associated with cirrhosis. Marked dilatation and tortuosity of the vena porta (28 mm) and its branches, and also varices at the perigastric area close to the abdominal wall were observed. Size of the spleen was markedly increased, measured 30×17×15 cm at the widest point. On MR images, signal changes observed at the upper pole

of the spleen with clear and irregular borders, consistent with an infarction due to partial splenic embolization (Figure 2A, 3A-C). Also on MRCP, decreased arborization of intrahepatic biliary channels and narrowing of all biliary channels were observed. Common bile duct width was measured 2.4 mm (Figure 2B). Few cystic lesions were observed at pancreatic lodge; biggest one measured 9 mm on MRCP. Increased echogenicity observed at US correlates well with diffuse lipid infiltration. Contours of the pancreas could not be clarified with MRI. Also increased periportal echogenicity observed on US. On US,



**Figure 2.** Case 2 (**A-C**). Coronal T2-weighted MR images (**A**) show micro-gallbladder (27 × 14 mm), thickened walls and hypointense calculus in the lumen (*long arrows*). Dilated portal vein branches at hilar area (*short arrow*) and splenomegaly through pelvic inlet. Heterogenous infarct area at the upper pole of the spleen secondary to partial embolization (\*). MRCP image (**B**) shows narrowing of the biliary channels and decreased arborization (*arrow heads*). Common bile duct (*short arrow*) looks thin (2.4 mm). Pancreatic channel can not be visualized clearly. Ascites in the perihepatic area (*long arrows*). US images (**C**) of the same patient, thick walled gallbladder and 8.2 mm calculus in the lumen of the gallbladder (*arrow*).



**Figure 3.** Case 3 (A-C). Axial T2-weighted image (A), T1-weighted fat-saturated precontrast (B) and portal venous phase contrast-enhanced images (C) show irregular contours of the liver due to cirrhosis, multiple regeneration nodules and left lobe hypertrophy. Hypo-isointense nodules seen separated with hypointense fibrous septas (arrow heads) on T1-weighted images and they do not hold contrast material. Dilated portal vein branches secondary to portal hypertension (arrow heads). (\*) Infarct area at the upper pole of the spleen secondary to embolization.

decreased size of the gallbladder (27×14 mm), thickened walls and 8 mm calculus in the lumen of the gallbladder observed (Figure 2C).

## Discussion

Secondary to increased life span of patients with cystic fibrosis, pancreatic and hepatobiliary complications appear more often<sup>1</sup>. The overall rate of pancreatic manifestations is 85-90% in the patients with CF and appears as the exocrine and endocrine insufficiency of the pancreas<sup>2</sup>. Pancreatic manifestations in CF are characterized by fat deposition or fibrosis. It is difficult to differentiate the pancreas with lipid infiltration from the retroperitoneal fatty tissue and measure the size. Increased echogenicity of the parenchyma or atrophy are the typical US findings. Findings of MRI change according to the amount of fat deposition and the degree of fibrosis. Fat deposition is seen as increased signal on T1-weighted images, fibrosis as decreased signal both on T1 and T2-weighted MRI. Pancreatic cysts are a relatively common finding and probably appear secondary to obstructed ducts because of thickened secretions. These cysts are typically 1-3 mm in size, however, can reach 1-2 cm in some cases and best visualized with MRCP<sup>3</sup>. Pancreatic channel may show narrowing or dilatation<sup>4</sup>. Hepatic involvement shows itself as hepatosteatorosis, hepatomegaly, and focal biliary cirrhosis; in some of the patients with hepatic involvement, diffuse cirrhosis and portal hypertension will further de-

velop. The prevalence of hepatic involvement is below 20-50%; the prevalence of development of cirrhosis and portal hypertension is below 5% in CF patients. Cirrhotic liver on US shows a coarse and nodular echo structure. Atrophy in the right lobe and hypertrophy in the caudate lobe and left lateral lobe may be observed. Regenerative nodules secondary to cirrhosis are observed as isointense or hypointense nodules separated from each other with hypointense fibrotic bands on T1-weighted MR images and they do not hold contrast material. Hepatosteatorosis stands out with parenchymal hyperintensity on T1-weighted MR image<sup>3-5</sup>. Dilatation of the portal vein, portosystemic shunts, splenomegaly, hepatofugal flow and ascites may be seen with portal hypertension. Spleen may reach to huge sizes and signs of splenomegaly develop secondary to portal hypertension. Partial splenic embolization is one of the treatment modalities used for thrombocytopenia secondary to hypersplenism<sup>6</sup>. Liver manifestations secondary to CF starts at the biliary ducts. Obstructions occur with the thickening of bile and cytotoxic damages appear in biliary ducts. This damage progresses to periportal fibrosis, bridge fibrosis and focal biliary cirrhosis<sup>2</sup>. Changes in the biliary ducts were reported with using US and MRCP in 24-50% of the cases in the literature<sup>7</sup>. Involvement of intrahepatic and extrahepatic biliary channels presents itself in various forms and signs described in different forms. This can be like progressive thinning of the distal intrahepatic biliary ducts, focal or diffuse dilations, strictures, narrowing and string

of pearls. Intrahepatic or extrahepatic biliary stones, diffuse narrowing of the common bile duct or focal stricture may also be observed<sup>8</sup>. Narrowing of the biliary channels and decreased arborization may observe when cirrhosis develops<sup>9</sup>. Visualization of the gallbladder is generally difficult in patients with cystic fibrosis. Gallbladder may be seen as normal or very small or even may not be seen. Sometimes walls get thickened and contracted, sometimes gall bladder contains a stone. Gallbladder size less than 30×15 mm described as micro-gallbladder, measurement performed when patient in hunger<sup>10</sup>.

### Conclusion

US can be used as a screening technique to determine hepatobiliary changes as early as possible. MRI should be the second choice. Especially, with the combination of MRI and MRCP, spread and severity of biliary complications of CF besides hepatic and pancreatic complications can also be evaluated in detail without using invasive techniques.

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