

HIGH RESOLUTION ULTRASONND FINDINGS OF LIVER AND KIDNEYS IN A CASE OF AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE IN A 4 DAYS OLD NEONATE

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ABSTRACT

Autosomal recessive polycystic kidney disease is characterized by renal collecting duct cysts, congenital hepatic fibrosis (biliary dysgenesis) and autosomal recessive pattern of inheritance. The typical sonographic appearance of autosomal recessive polycystic kidney disease (ARPKD) has been described as symmetrically enlarged, echogenic kidneys with occasional visualization of small cysts. We present a case of a 4 years old male neonate referred to our ultrasound department. High resolution Ultrasound revealed bilateral renal enlargement due to non-obstructive collecting duct ectasia in a bilateral and symmetrical fashion as well as changes due to hepatic periportal fibrosis and biliary dilatation.

Case Report

A four days old baby boy was referred to us for ultrasound abdomen with the complaints of breathing difficulty and poor feeding.

He was delivered at home after full term. He was third sib of consanguineous parents. On examination baby was not dysmorphic. His breathing was laboured and abdomen markedly protuberant with bilateral flank masses palpable. Rest of examination was normal.

His high resolution ultrasound revealed enlarged Kidneys which were echogenic with increased number of acoustic interfaces. Multiple renal cysts were seen ranging in size from 15 mm to 66 mm. (Fig 1). Acoustic enhancement was seen behind larger cysts. The cysts in the renal cortex were larger as compared to inner ones. There was loss of cortico-medullary differentiation. Central echogenic complex was not visualized. No hydronephrosis was seen. Right kidney measured

8 cm in length while left kidney was 8.5 cm. No solid mass was seen. Urinary bladder was not visualized.

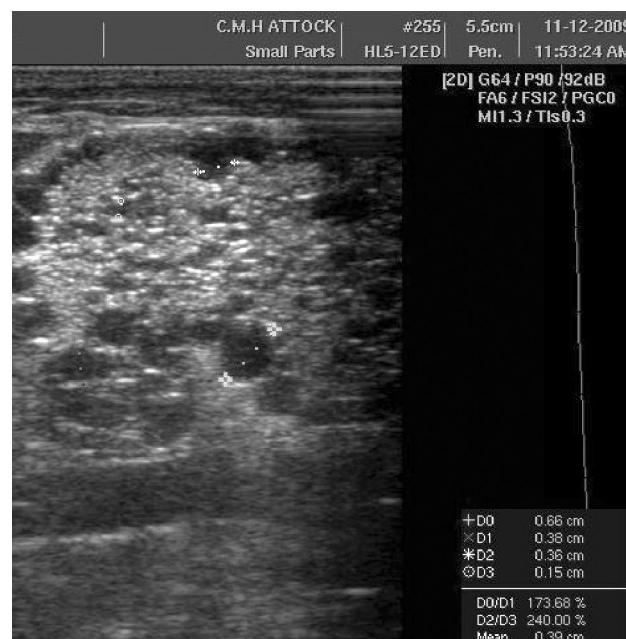


Figure 1A:

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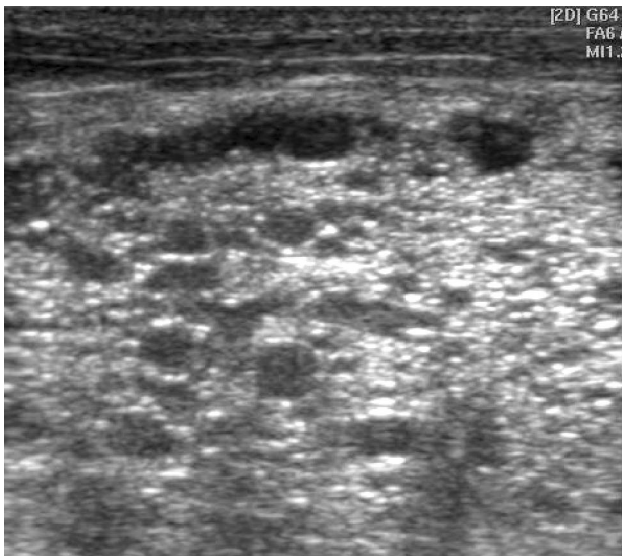


Figure 1B: High resolution ultrasound done with 10 MHz Linear probe (SONOACE -MEDISON) .Section of the kidney showing cysts ranging in size from 15 mm to 66 mm replacing renal parenchyma completely.

Liver was enlarged and showed marked periportal fibrosis and bile duct proliferation. Periportal fibrosis was seen as multiple echogenic areas scattered throughout the liver (Fig 2).

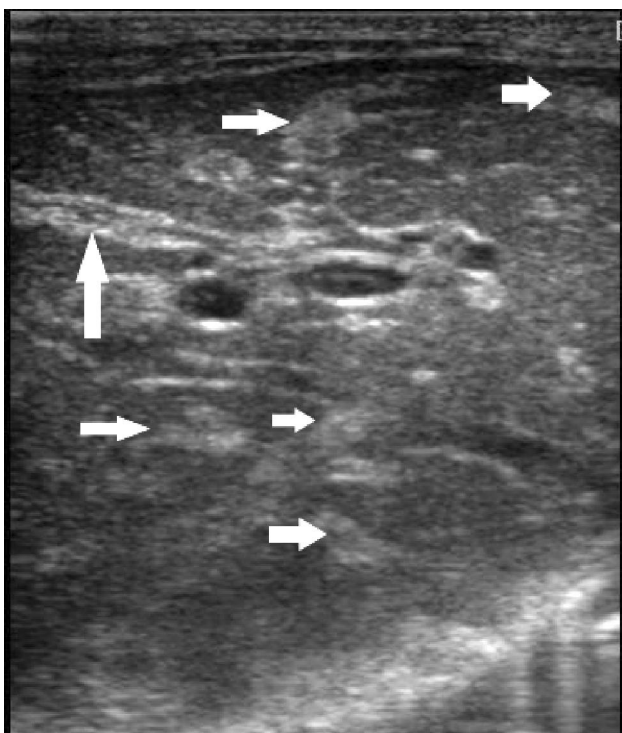


Figure 2A:

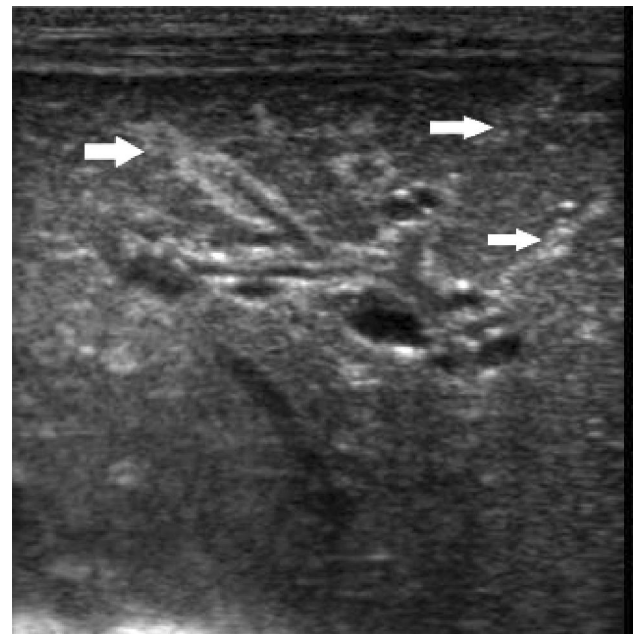


Figure 2B: High resolution ultrasound done with 10 MHz Linear probe (SONOACE -MEDISON) .Section of the Liver showing periportal fibrosis (arrows) seen as echogenic lesions diffusely spread in the liver mm replacing renal parenchyma completely.

Few of these lesions had a branching pattern. These areas represented the thickened fibrosed portal tracts that were characteristics of hepatic fibrosis. Tubular cystic dilatation of intrahepatic biliary channels was also seen (Fig 3).

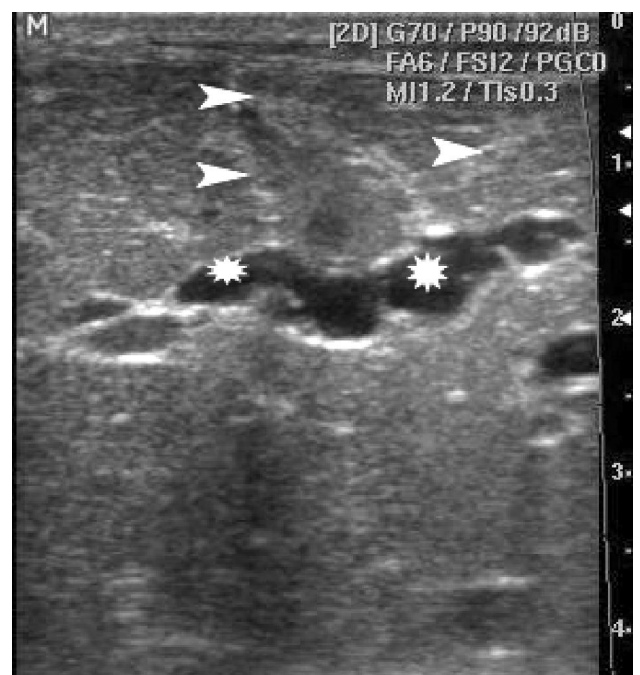


Figure 3A:

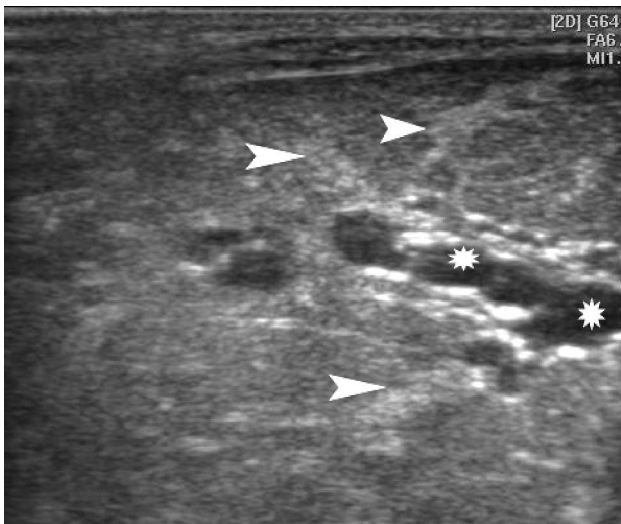


Figure 3B: High resolution ultrasound done with 10 MHz Linear probe (SONOACE -MEDISON) .Section of the Liver showing dilated biliary channels (asterisk) as well as periportal fibrosis (arrowheads).

The echogenic areas representing periportal fibrosis were seen extending upto the peripheral margins of the liver and were diffusely scattered in the enlarged liver. No splenomegaly or varices were seen. Color Doppler revealed normal hepatic and portal venous flow without varices (Fig 4). Patient was lost to follow up and didn't return.

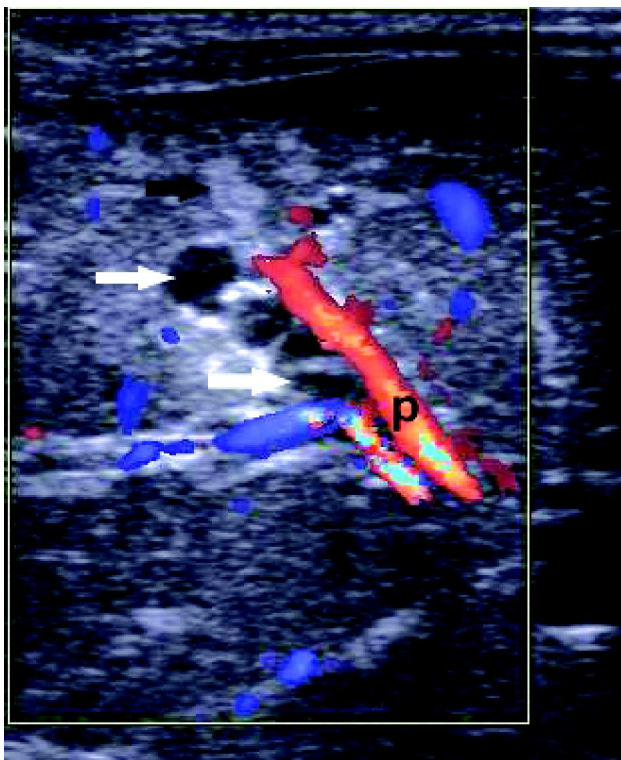


Figure 4A:

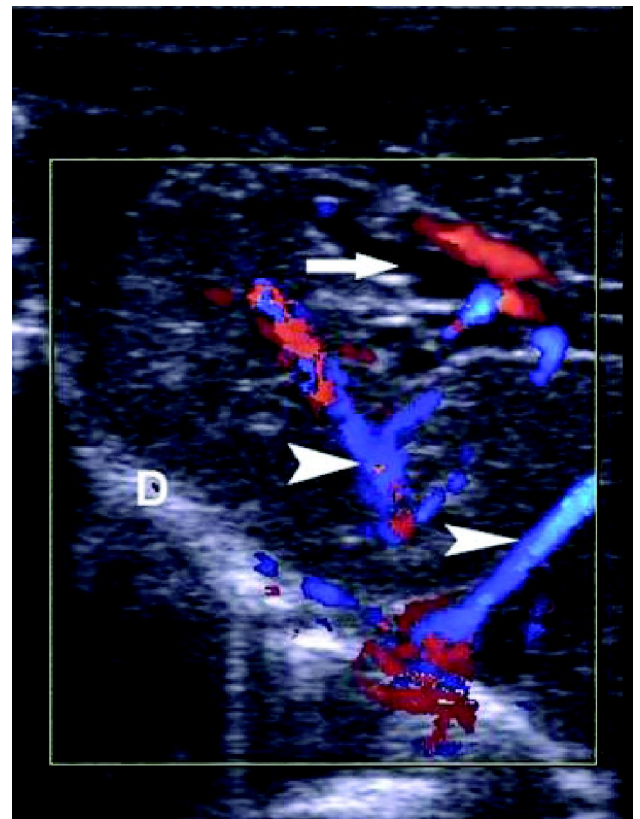


Figure 4B: High resolution color Doppler ultrasound done with 10 MHz Linear probe (SONOACE -MEDISON) .Section of the Liver showing dilated biliary channels (white arrows) as well as periportal fibrosis (black arrows).White arrowheads = hepatic veins. P= portal vein branch. D = Diaphragm.

Discussion

Two predominant features characterize the liver in ARPKD: abnormality of the biliary tree and fibrosis of the portal tracts. Abnormality of the biliary tree consists of irregularly formed, dilated, and usually too numerous intrahepatic bile ducts (these are contiguous with other portions of the biliary tree and therefore nonobstructive). Despite the abnormal bile ducts and the fibrosed portal tracts, the hepatic parenchyma is normal. Thus, it is not surprising that hepatocellular function is almost always normal in affected patients, even when they have relatively severe portal tract disease. The liver disease is referred to as congenital hepatic fibrosis (CHF), which is always found in patients with ARPKD.^{1,2} The incidence of ARPKD is 1: 40,000.³ According to clinical appearance, ARPKD is classified into 4 separate groups: prenatal, neonatal, infantile, and juvenile forms.

There is considerable variation in the imaging findings in patients with Autosomal Recessive Polycystic Kidney Disease. In all cases nephromegaly is a constant imaging finding at the time of diagnosis.

In neonates and infants with moderate to severe renal disease, the kidneys are smoothly enlarged because of the numerous dilated collecting ducts. The degree of enlargement is directly proportional to the number of dilated ducts.⁴

Magnetic Resonance Cholangiopancreatography (MRCP) is superior to ultrasound in the assessment of cystic liver disease. In addition to higher sensitivity, MRCP provides information on the anatomic relationship between the cysts and the biliary tree.⁵ The hepatic disease progresses to develop portal hypertension associated with splenomegaly and esophageal varices. Congenital hepatic fibrosis (CHF) is characterized by the intrahepatic form of portal hypertension, which is caused by the intrahepatic obstruction that affects the blood supply to the liver and subsequently leads to the development of cavernous transformations of the portal vein with arise in portal venous pressure. ARPKD is a disease of tubular malformation and ectasia. In the kidney, the disorder manifests as nonobstructive collecting duct ectasia. The ducts are dilated and elongated, with 10%–90% of them being involved, usually in a bilaterally symmetric fashion. When a large number of ducts are involved, the kidneys are typically enlarged. Fibrosis develops in the renal interstitium, and, when the amount of ductal ectasia and fibrosis is considerable, renal functional impairment may result. Hypertension, diminished urinary concentrating ability, and renal insufficiency and failure may ensue, requiring dialysis or renal transplantation. End-stage renal disease occurs in approximately 800 children per year in the United States, and ARPKD accounts for about 5% of these cases. Patients with ARPKD also have liver involvement, which consists of abnormal biliary ducts and portal tracts. The bile ducts are abnormally formed, often increased in number, and dilated; the portal tracts are enlarged and fibrotic. This pattern is referred to as congenital hepatic fibrosis (CHF) and is always present in ARPKD.⁶

Interestingly, when the bile ducts are macroscopically dilated, ARPKD in the liver may be indistinguishable from Caroli disease as seen in our case. Further

complications include ascending cholangitis, secondary to entry of nonsterile gastrointestinal contents into the dilated intrahepatic bile ducts.

References

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