



A child with bilateral multiple renal cysts presenting with ascites and pleural effusion: Questions

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Case summary

A 4-year-old girl was admitted to the Pediatric Gastroenterology Division with abdominal distention. Her family had noted progressive abdominal distention over the last 2 months. She was the first child of consanguineous parents. Her family history was unremarkable. Her weight was 14.5 kg (16th percentile), height was 97 cm (6th percentile), and blood pressure was 106/60 mmHg. Her cardiovascular and respiratory examinations were unremarkable. Her abdomen was distended; on percussion, there was periumbilical tympani with dullness in the flanks. There was no organomegaly or edema.

Her laboratory findings were as follows: white blood cell count $5100 \times 10^3/\text{mm}^3$, hemoglobin 12.7 g/dL, platelets $324 \times 10^3/\text{mm}^3$, urea 18 mg/dL, serum

creatinine 0.3 mg/dL, and electrolytes and urine analysis were normal. In addition, liver function tests and albumin were normal. The structure and diameters of the liver and spleen were normal on abdominal ultrasonography (US). Intra-abdominal-free fluid was present. The length of the right kidney measured 82 mm, the left kidney was 88 mm, and the parenchymal thickness was 10 mm. Renal echogenicity was increased in both the right and left kidneys, and corticomedullary differentiation was lost. Multiple, subcortical, anechoic cysts were present. The largest cysts measured 45 mm in diameter on the right kidney and 16 mm in diameter on the left kidney. There was no dilatation of the collecting system (Fig. 1). The patient was hospitalized with a differential diagnosis of ascites and renal cysts. Paracentesis of the ascites excluded infection (as culture was negative), chylous ascites (as fluid was clear with low triglycerides at 12 mg/dL), pancreatitis (amylase 9.3 IU/L), and urine leakage (ascites urea 23 mg/dL, creatinine 0.14 mg/dL). The fluid was a transudate (density 1008; ascites to serum protein ratio < 0.5 ; ascites to serum LDH ratio < 0.6). The serum ascites albumin gradient was > 1.2 g/dL. Cytology of the fluid revealed a few lymphocytes. Viral serology and echocardiographic examination was unremarkable.

Abdominal magnetic resonance imaging (MRI) revealed a normal hepatobiliary system and multiple subcortical cysts separated with septae, which were hypointense without enhancement in T1-weighted and hyperintense in T2-weighted images. The cysts resulted in indentations to the renal parenchyma. There was right-sided pleural effusion which was 13 mm thick and also abdominal-free fluid (Fig. 2).

The answer to this question can be found at <https://doi.org/10.1007/s00467-019-4198-1>.

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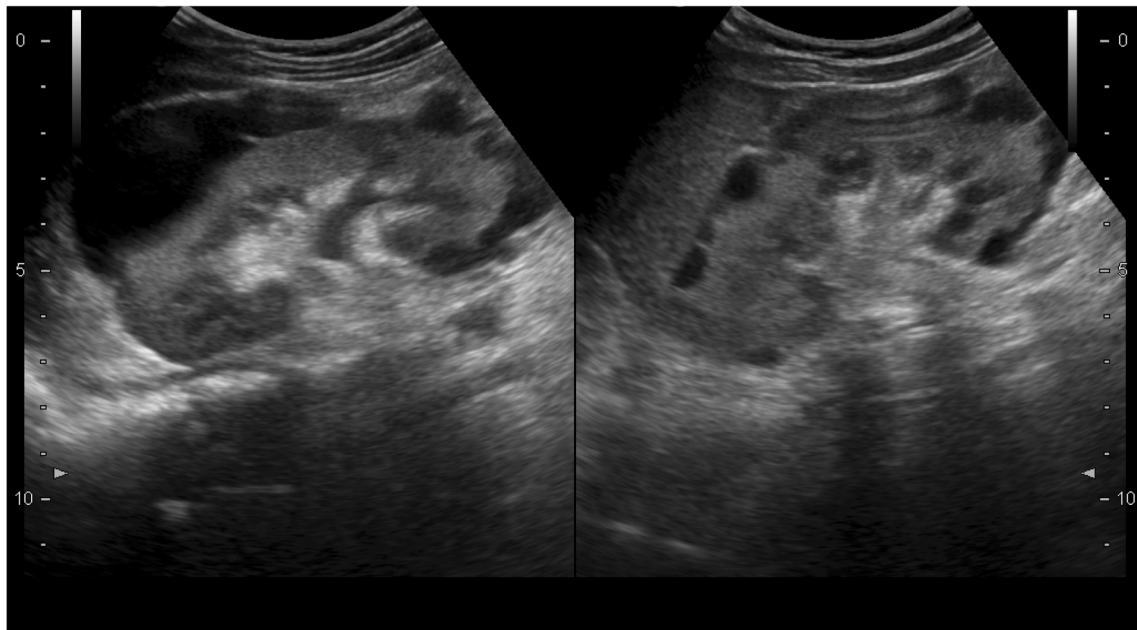
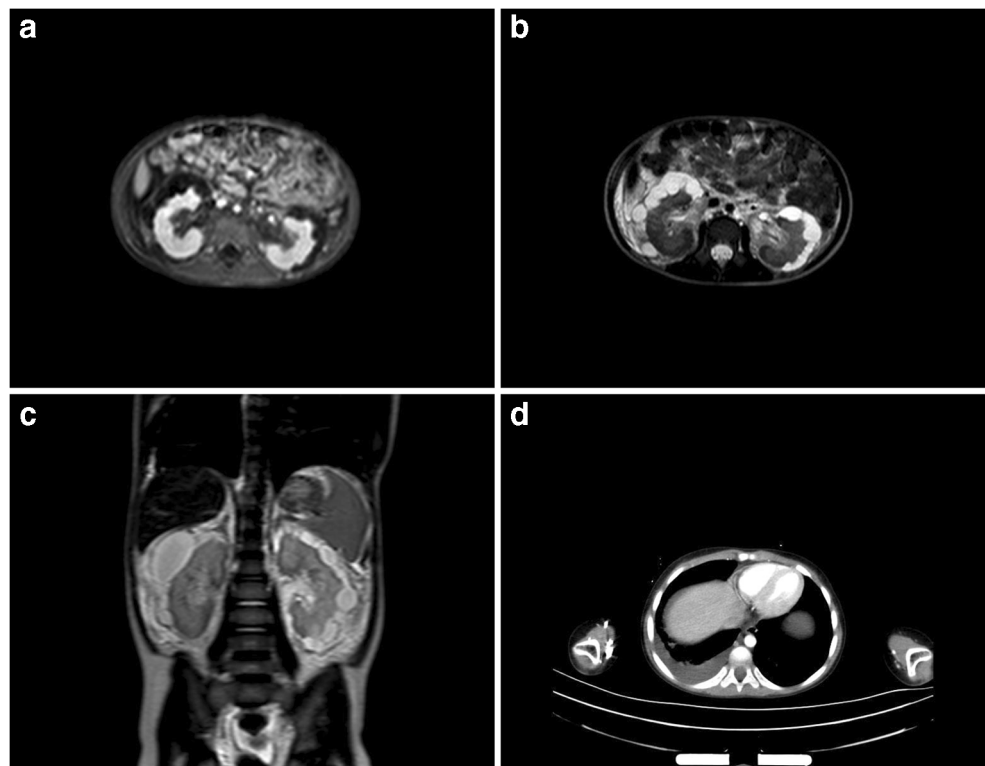


Fig. 1 Renal US on admission. Bilateral renal echogenicity were increased. Subcortical anechoic cysts were separated by septa, and causing indentation on renal parenchyma. Maximum cyst diameter was 45 mm on the right kidney, 16 mm on the left kidney

Fig. 2 Abdominal MRI. **a** Axial T1-weighted (hypointense) and **b** T2-weighted (hyperintense) images demonstrating subcortical cysts. **c** Coronal T2-weighted image of the cysts. **d** Right-sided pleural effusion



Questions

1. What is your diagnosis?
2. Is further investigation or intervention required for the diagnosis?
3. What is the treatment and prognosis?

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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