

CLINICAL QUIZ (p82) ANSWER

1. Figure 1 shows the left kidney was enlarged with mild hydronephrosis and multiple small cysts (measured 1-3 mm in size) replacing the renal parenchyma. The normal corticomedullary demarcation was lost. Figure 2 showed the liver was grossly enlarged. Hepatic ductal ectasia was seen. Figure 3 showed a contrast nephrogram with striated pattern. The kidneys were enlarged. The striated nephrogram after contrast material administration represented the dilated collecting ducts in the kidneys¹ and was a typical radiological feature of ARPKD. The classical ultrasound findings of ARPKD in neonatal period are bilaterally enlarged echogenic kidneys with loss of corticomedullary differentiation. Cysts are usually very small and may not be detected easily in neonates and they are better characterised in older patients.
2. The diagnosis was autosomal recessive polycystic kidney disease (ARPKD).^{1,2} ARPKD is a heritable but phenotypically variable disorder. It is characterised by varying degree of non-obstructive renal collecting duct ectasia, hepatic biliary duct ectasia and fibrosis of both liver and kidneys. In severe cases, the dilated collecting ducts and interstitial fibrosis may significantly impair renal function and result in renal failure whereas periportal fibrosis accompanies the malformed and dilated bile ducts may result in portal hypertension.³

References

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2. McDonald AR, Avner ED. Inherited Polycystic Kidney Disease in Children. *Semin Nephrol* 1991, 11: 632-42.
3. Dische F. Structure and Development of the Kidney and Cystic Disease. In: *Renal Pathology*, second edition. Oxford Medical Publication, 1995:7-21.