

Cystic renal diseases: role of ultrasound. Part I, non-genetic cystic renal diseases

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Abstract

Kidney cysts are quite common in adults. Though small simple renal cysts in an adult over 30-40 years of age are not too unusual, however, if the same cysts are seen in a child, and especially if there are additional findings, then several diagnostic possibilities may come to mind. The role of ultrasound, together with the help of intravenous contrast agents and Doppler mode, is very critical in describing the morphologic features and follow-up of the complex or multiple and bilateral renal cysts. Sonographic signs are occasionally specific for diagnosis, but in many cases they should be evaluated together with the other genetic and clinical data to reach a diagnosis.

The first part of this pictorial essay includes “non-genetic cystic renal diseases” and the second part will include “genetic cystic renal diseases”.

Keywords: cyst; cystic; Doppler; kidney; ultrasound (US)

Introduction

Cystic renal diseases form a large spectrum with many challenging and confusing cases which may have solitary and few cysts among those with multiple bilateral cysts. Ultrasound (US) has been the most valuable diagnostic tool used very early in patients, from the prenatal period. It has been also frequently used in the follow-up of patients and especially children. The high-resolution linear probes provide a better demonstration of tiny cystic changes in the fetal and neonatal period.

The current classifications of cystic renal diseases are variable and at least partly suffer the lack of experts’

consensus. Differential diagnosis by imaging, especially using US, maybe difficult in some of them and may need additional clinical and genetic clues. As many diseases are now being enlightened by new and previously unveiled genetic details, their classifications are being updated

Table I. Classification of cystic renal diseases

Cystic renal diseases are:

A) Non-genetic cystic renal diseases

- Simple cysts
- Complex cysts
- Cystic neoplasms
- Multicystic dysplasia / Obstructive cystic dysplasia
- Medullary sponge kidney

B) Genetic cystic renal diseases

- Autosomal dominant polycystic kidney disease
- Autosomal recessive polycystic kidney disease
- Juvenile nephronophthisis / Medullary cystic kidney disease complex
- Multiorgan syndromes with pluricystic kidneys (Tuberous sclerosis, von Hippel Lindau, etc.)

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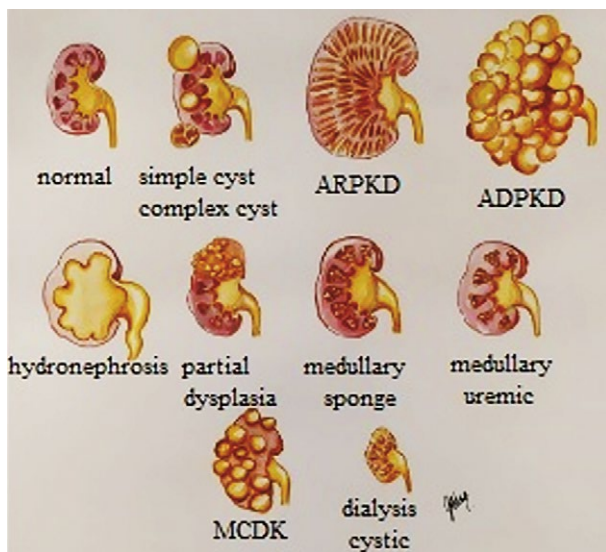


Fig 1. Schematic representation of frequent renal cysts or cystic diseases

accordingly [1]. For simplicity, we slightly modified the previous well-known classifications [2-4] as genetic and non-genetic (Table I). A schematic representation of frequent renal cysts or cystic diseases is shown in fig 1.

In this pictorial essay, we will demonstrate typical and atypical sonographic findings of cystic renal diseases reflecting our experience throughout the period 2000-2020. The first part covers non-genetic cystic renal diseases and the second part will focus on genetic cystic renal diseases.

Non-genetic cystic renal diseases

Simple renal cysts

The two well-known features of simple cysts are their thin or imperceptible wall and anechoic content without septae, internal echoes or mural nodules (fig 2a). The simple-complex cyst differentiation is very important in diagnosis, especially in terms of neoplasia. Although

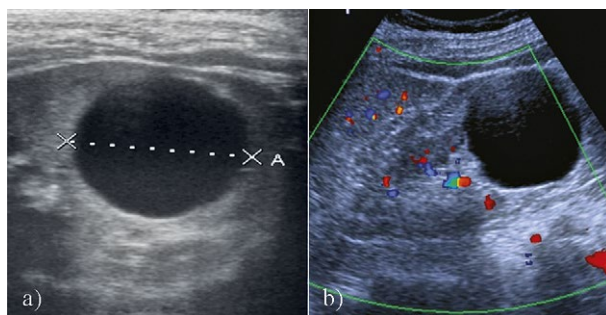


Fig 2. Simple cyst: a) US shows thin wall and anechoic content; b) no vascularity is detected by Doppler US.

valid in computed tomography (CT), the Bosniak classification may be roughly applied to sonography after careful color and spectral Doppler exam or contrast-enhanced US (CEUS) [5,6] (fig 2b). Any Doppler signal or contrast enhancement of the wall or internal structures, raises suspicions for neoplasia.

Simple cysts may be congenital or more often acquired with an increasing incidence in the elderly. They

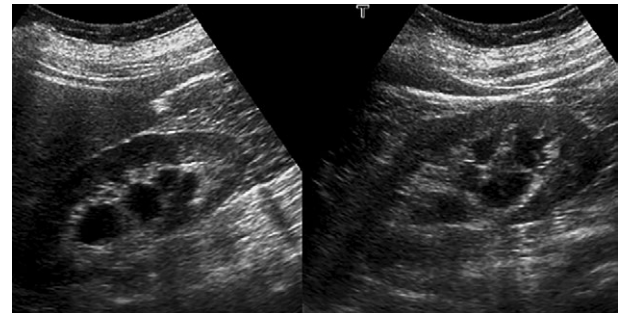


Fig 3. Bilateral parapelvic cysts mimicking hydronephrosis, in a 55-year-old female.

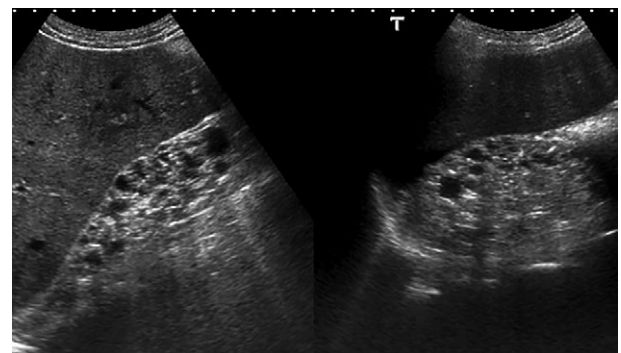


Fig 4. Bilateral small cysts in atrophic kidneys. The patient has been on dialysis for 10 years.

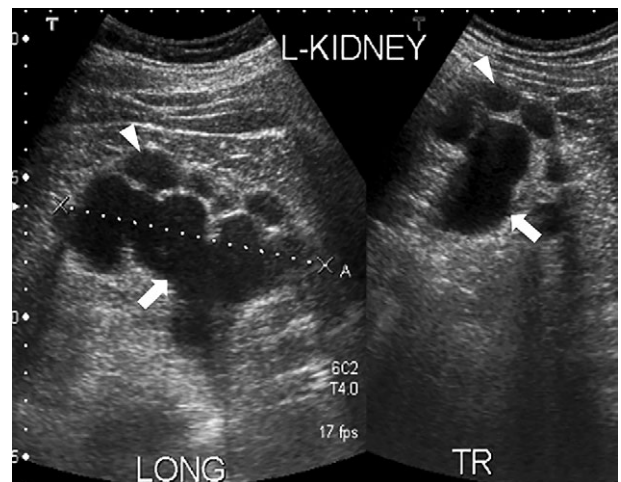


Fig 5. Left renal simple cysts (arrowheads) due to chronic obstruction (arrows) in a 65-year-old male with bladder cancer.

may have variable presentations; parapelvic cysts are usually seen bilaterally in the elderly (fig 3), and multiple cysts may be seen during hemodialysis (fig 4) or chronic urinary obstruction (fig 5).

Mimickers of simple cysts

Calyceal diverticula that may mimic simple cysts are rare, usually as congenital outpouchings of the minor upper pole calyces. They are frequently associated with stones and infection (fig 6). Another challenging simple cystic appearance is the dilated upper pole calyx of the duplex collecting system (fig 7).

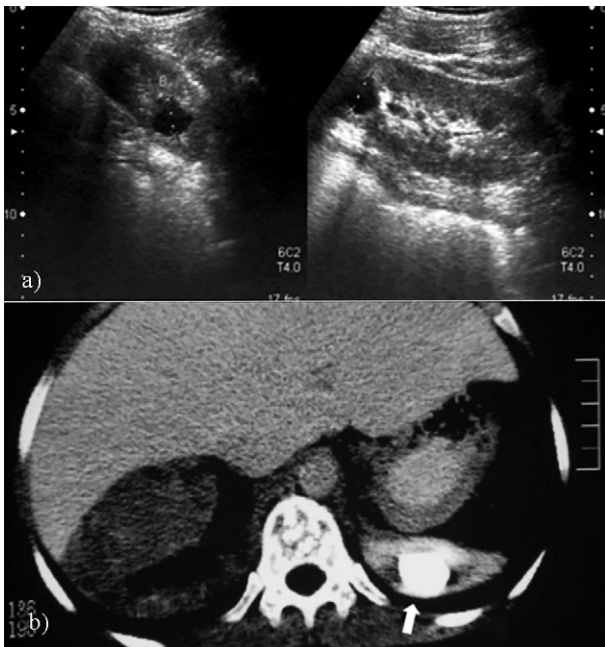


Fig 6. a) An 18 mm simple cyst in the left upper pole of kidney; b) CT with intravenous contrast agent shows filling of the lesion with contrast agent, and it is in fact a calyceal diverticulum (arrow).

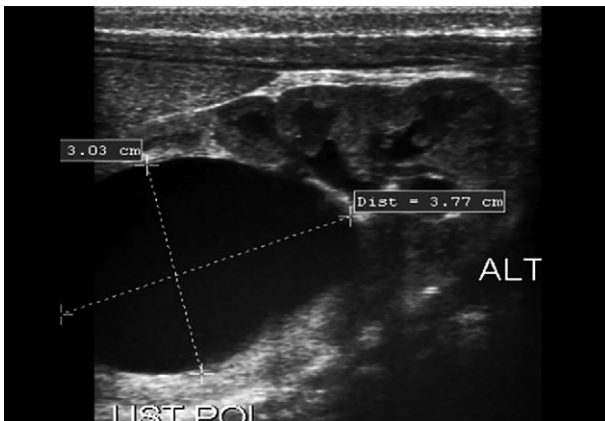


Fig 7. A simple cyst mimicker; dilated upper pole collecting system in a child.

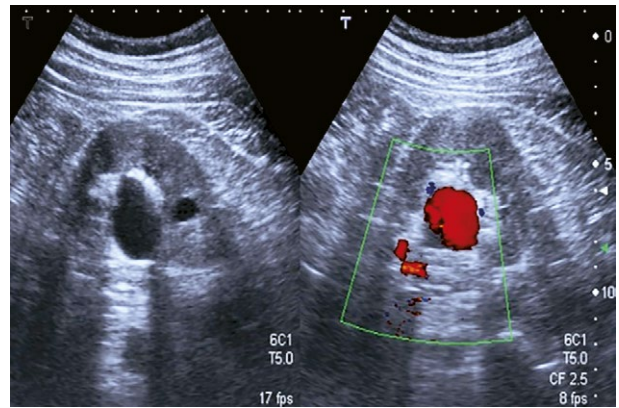


Fig 8. An aneurysm in a 47-year-old male. US shows a simple cystic structure.

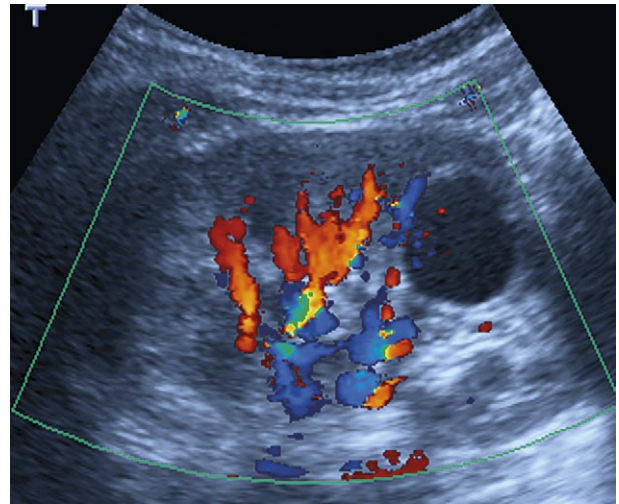


Fig 9. A lower pole simple cystic structure in a child after a post-traumatic hematoma has been followed-up for resolution. The wall is a bit thick for considering it as a simple cyst.

Several vascular lesions such as aneurysms may appear as simple cysts (fig 8). They can be differentiated by adding Doppler US findings. Thus, Doppler US should always be used in the evaluation of all cystic renal lesions. Post-traumatic renal hematomas may leave cystic cavities, as they resolve (fig 9).

Complex renal cysts

These cysts have some solid components, septae, or thick walls. Practically, the most important issue is the differential diagnosis from the malignant tumors. A few thin septae and internal echoes are usually not considered worrisome, but thick-calcified septae or walls need to be investigated or followed-up (fig 10-13). Bosniak classification is still frequently used to stratify complex cysts to choose the appropriate management.

Hemorrhage into cysts is not very frequent except in the autosomal dominant polycystic kidney disease

(ADPKD). Internal echoes or later fibrin septae may be seen in hemorrhagic cysts (fig 14, fig 15). Kidney abscess should be considered in complex cysts with thick walls, internal echoes, and peripheral vascularity (fig 16, fig 17). Hydatid cyst should be considered especially in endemic regions. Thick, double-layered walls raise suspicion. Multiple cysts in other sites, especially in the liver may indicate echinococcus (fig 18). A detached membrane makes the diagnosis almost always certain [7]

Fig 10. Complex cyst compatible with Bosniak 2F; there are a few thin septae.



Fig 11. Two different patients with complex hemorrhagic cysts: a) Cyst with intense internal echoes, in a 71-year-old male; b) a hemorrhagic cyst in a 46-year-old male with follow-up; Doppler US showing no vascularity.



Fig 12. A complex cyst with calcifications and internal echoes. The aspirate was negative for malignancy and echinococcosis.

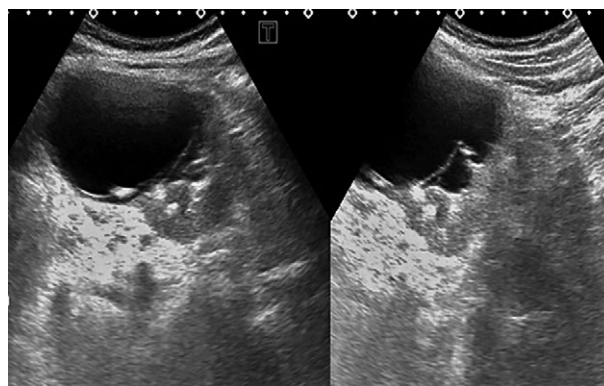


Fig 13. A complex cyst described as Bosniak 2F and followed up for 15 years without significant change.



Fig 14. Hemorrhage into a cyst in a patient with ADPKD.

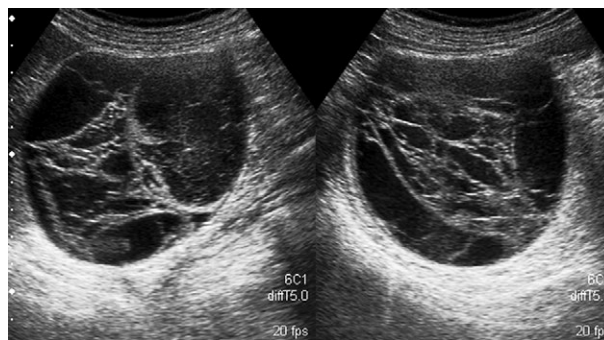


Fig 15. Fibrin septae during follow-up of a 55-year-old male with a hemorrhagic cyst.

(fig 19). More complex or aged or treated hydatid cysts may contain a solid matrix of collapsed membranes and calcifications (fig 20, fig 21).

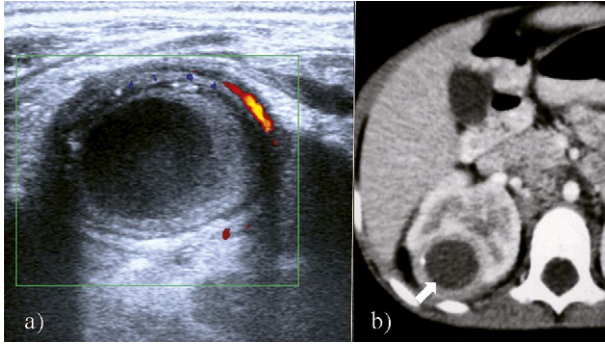


Fig 16. a) A 5-year-old boy with a thick-walled cystic renal structure with internal echoes and peripheral vascularity on Doppler US; b) CT shows a peripherally enhancing hypodense right kidney abscess (arrow).

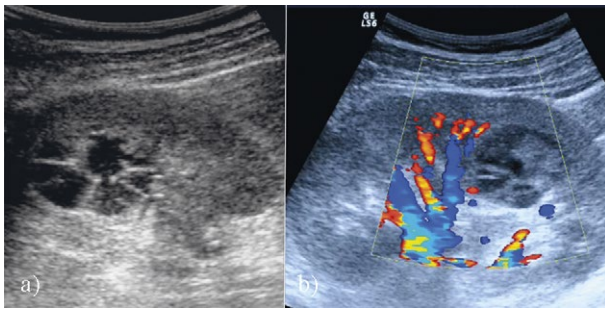


Fig 17. a) US and b) Doppler US images of a complex cystic structure proven to be an abscess after follow-up.

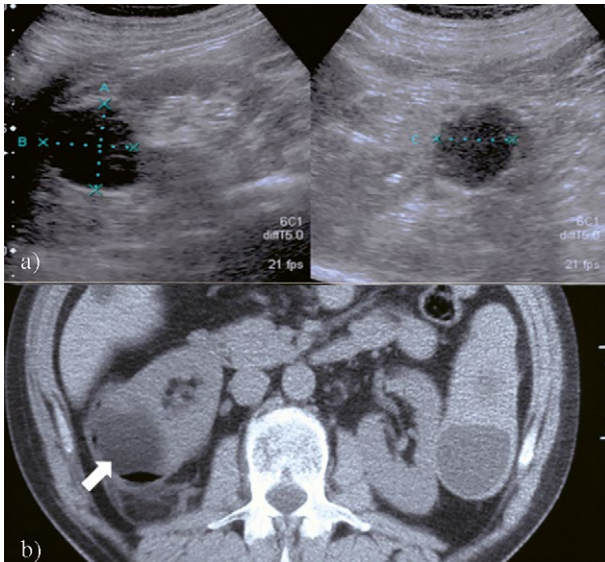


Fig 18. a) US shows a right renal cyst with thick walls b) The 65-year-old patient is known to have hepatic and splenic hydatid cysts. The treated cyst (arrow) is shown on CT.

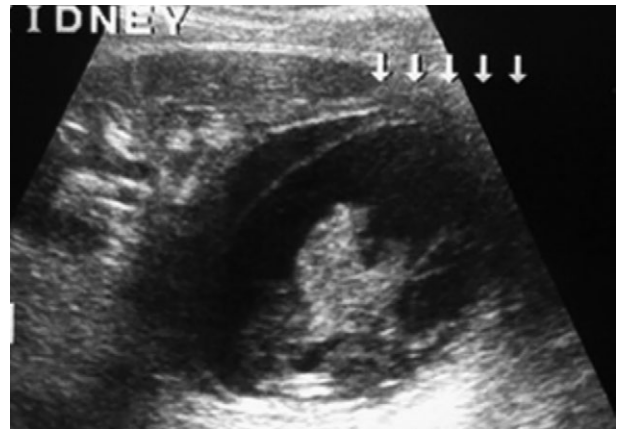


Fig 19. Detached membrane and solid matrix in a hydatid cyst (arrows) after percutaneous aspiration injection reaspiration (PAIR) procedure in a 17-year-old girl.

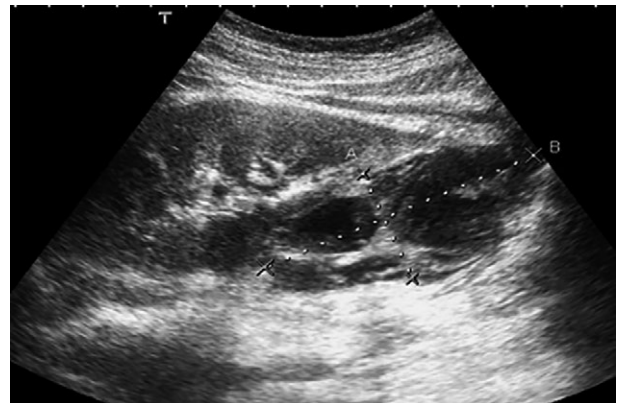


Fig 20. Collapsed membranes and solid components after PAIR of the lesion in a 13-year-old girl.

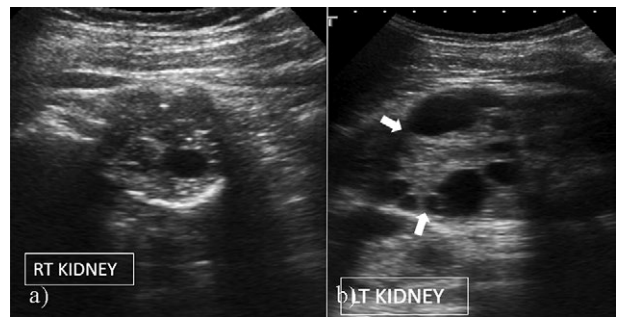


Fig 21. Multiple bilateral hydatid cysts in a 43-year-old male: a) US of the right kidney showing calcified cysts; b) the left kidney has a multilocular mass containing many daughter cysts (arrows).

Cystic renal neoplasms

Benign multiloculated cystic nephroma (MLCN), cystic Wilms tumor and cystic renal cell carcinoma (RCC) are the most frequent neoplasms. MLCN is seen in a bimodal age of presentation: in children below 2

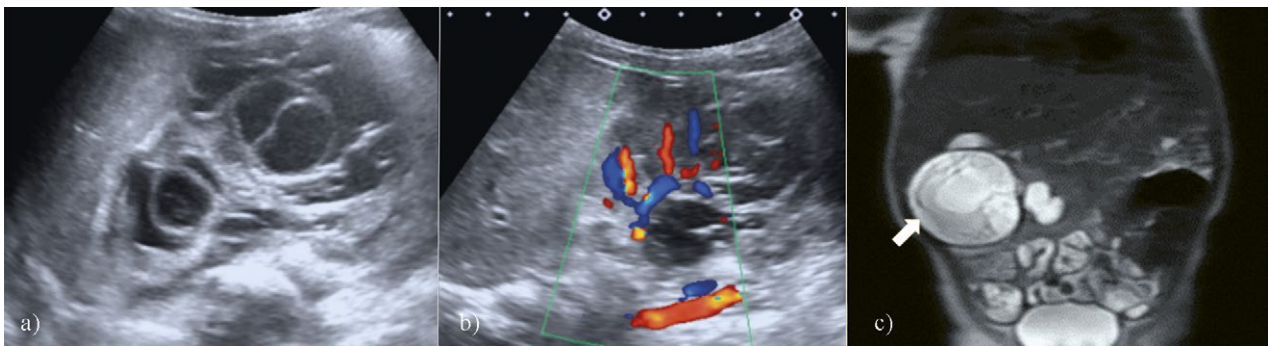


Fig 22. A large cystic mass of right renal lower pole with multiple septae in a 1-year-old female; a multiloculated cystic nephroma: a) US b) Doppler US and c) Coronal T2-weighted MR image showing the lesion (arrow).

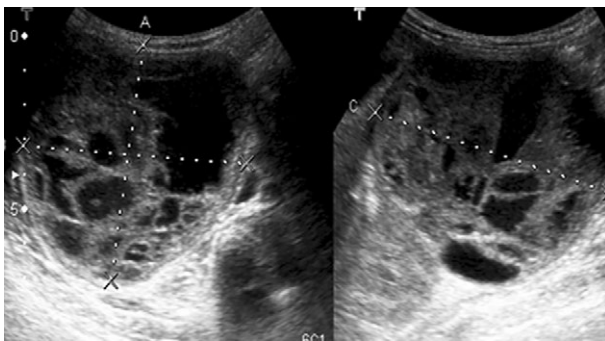


Fig 23. US shows an 8 cm mass. In the pathology report of this cystic Wilms tumor, large necrotic areas, corresponding to the cystic spaces were described.

years of age and middle age (40-69) adults. It is a cystic neoplasm with multiple septae showing vascularity (fig 22). Although a capsule has been emphasized as an important feature, it is usually not possible to differentiate benign MLCN from the malignant cystic Wilms tumor (nephroblastoma) based solely on the presence of a capsule (fig 23).



Fig 25. Cystic immature teratoma in a 2-month-old boy. Solid calcific components (arrow) are present on one side.

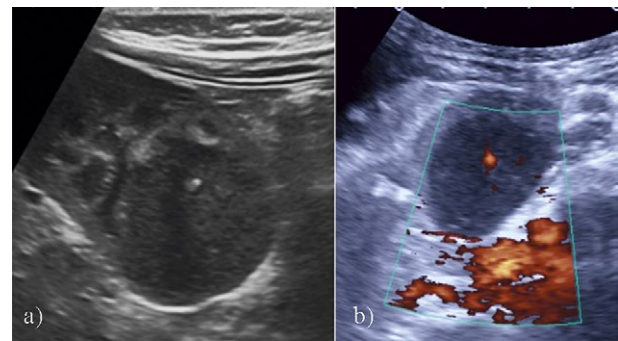


Fig 24. Mesoblastic nephroma in a 5-month-old female. a) US shows a complex cyst; b) power Doppler US image showing the vascularity of a septum.

Mesoblastic nephroma is a rare neoplasm that comprises about 5% of childhood kidney tumors and more than 90% of cases appear within the first year of life (fig 24). More than 15% of the cases are prenatally detected [8]. It is the most common kidney tumor found in infants younger than 6 months. Most of them are cystic-necrotic [9].

Extremely rare cystic teratomas should also be kept in mind, especially if there are associated calcifications (fig 25).

Cystic RCCs make up almost 5% percent of all RCCs and are usually of clear cell type (fig 26, fig 27). They have a better prognosis than the solid RCCs [10].

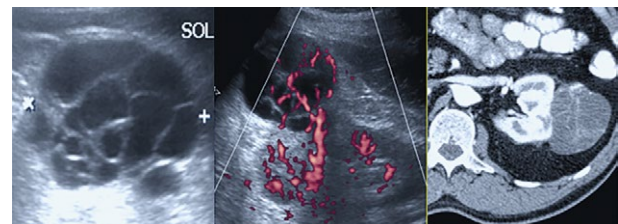


Fig 26. Cystic RCC in a 45-year-old male. The septae are not thick on US, but highly vascular by Doppler US and show post-contrast enhancement on CT.

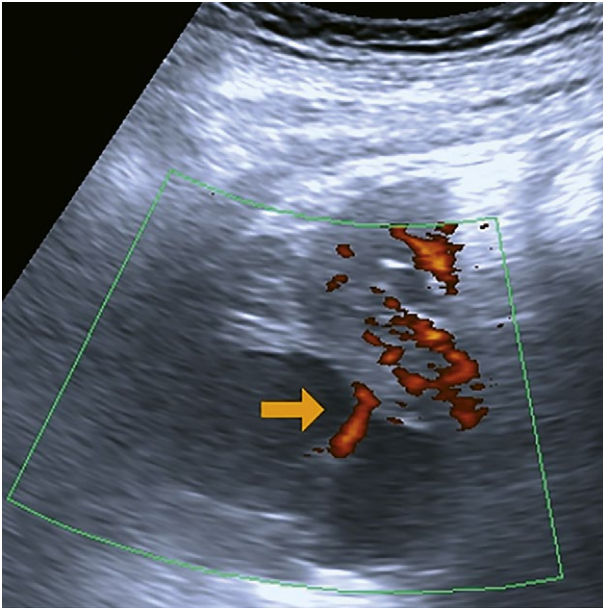


Fig 27. Cystic clear cell RCC in a 34-year-old female. The septae (arrow) and solid components show hypervascularity on power Doppler US.

Multicystic dysplastic kidney / Obstructive cystic dysplasia

These two common entities are usually taken as two separate titles, but there are certainly similarities in etiology, obstruction being the common cause. Some consider “Dysplastic Diseases” divided into 4 subtypes of dysplasia, including “segmental dysplasia” [2]. Multicystic dysplastic kidney (MCDK) results from an early defect in the connection between the ureteral bud and blastema whereas “obstructive cystic dysplasia” is associated with urinary tract dilatation and various congenital uropathies [11]. In MCDK, there is practically almost no functioning nephron and the kidney consists of “a bunch of cysts” (fig 28). Bilateral MCDK is rarely compatible with life. Renal agenesis on one side and dysplasia on the other side may also be seen. In unilateral form, the other kidney is prone to other abnormalities, uretero-pelvic junction obstruction being the most frequent one [12].

Cysts usually get smaller by time or remain stable for years (fig 29). It is no longer believed that MCDK has an increased risk of malignancy.

MCDK may be segmental; most of the segmental cases are believed to have duplex collecting systems. However, several researchers interpret these cases differently and label these as “segmental cystic disease” or “unilateral cystic disease” (fig 30).

Obstructive cystic dysplasia usually starts in the prenatal period and is associated with severe obstructions such as posterior urethral valves, ureteropelvic obstruction,

or vesico-ureteral reflux. The parenchymal echogenicity is increased (fig 31).

Medullary sponge kidney

Although this is not a very rare disease, many patients are asymptomatic or discovered incidentally with stone formation within the dilated tubules. The disease may be bilateral, unilateral, or even segmental. The medullary cystic structures are actually dilated distal collecting ducts. The cysts are usually smaller than 1 cm and rarely exceed 2 cm (fig 32a). Medullary nephrocalcinosis

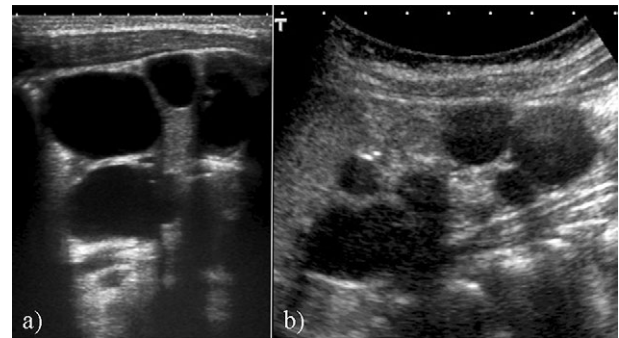


Fig 28. a,b) Two pediatric cases with typical MCDK.

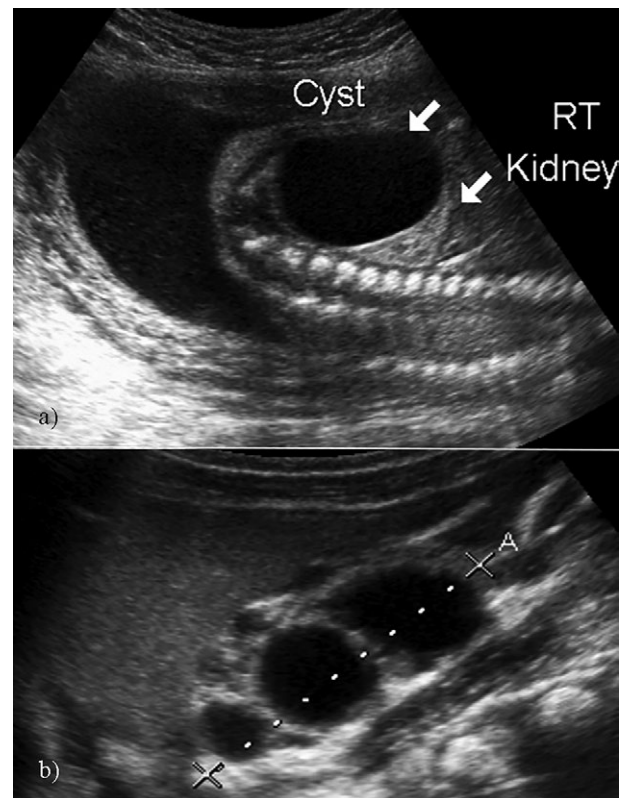


Fig 29. a) Intrauterine appearance of MCDK with a big and multiple tiny cysts. b) After 5 months, the same kidney has different-sized cysts in the newborn.

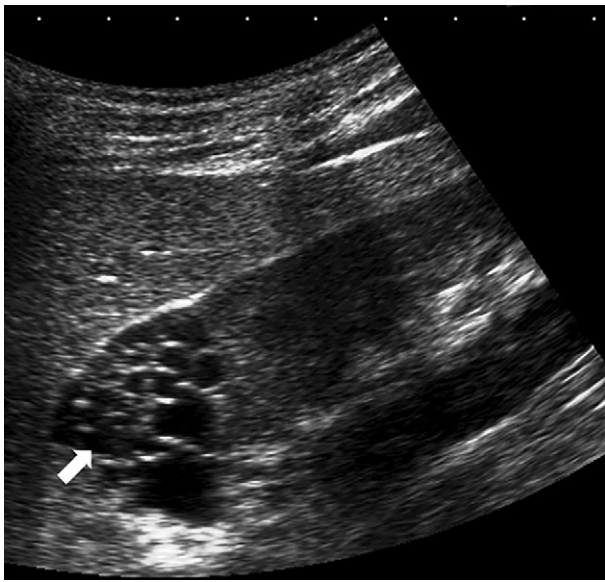


Fig 30. Segmental MCDK in a 17-year-old girl. A group of cysts (arrow) is seen in the upper pole, while the lower pole has a normal structure.



Fig 31. The cortex-medulla differentiation cannot be made in this 3-month-old boy with obstructive cystic dysplasia. There is a dilated collecting system with tiny scattered cysts.

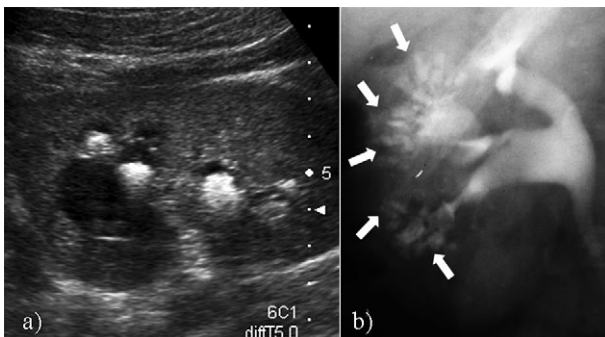


Fig 32. a) Small medullary cysts and stones in a medullary sponge kidney; b) the typical medullary contrast filling (arrows) during excretory urography in medullary sponge kidney.

may accompany. The excretory urography in these patients is pathognomonic with contrast pooling within the dilated collecting ducts restricted to the medulla-“brush appearance” (fig 32b). The main differential diagnosis is nephronophthisis where kidneys are small, but the medullary sponge kidney is normal in size [7].

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