Multicystic Dysplastic Kidney

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Michela Maffi and Mario Lima

41.1 Introduction and Definition

Multicystic dysplastic kidney (MCDK) is the most common cystic renal anomaly found in children and the second cause of palpable mass in neonates after hydronephrosis. The kidney is characterized by the presence of multiple non-communicating cysts in absence of normal parenchyma.

41.2 Epidemiology

The estimated incidence of MCDK ranges from 1 in 1000 to 1 in 4300 live births, and males are slightly more affected than females (M/F = 2.4:1) [1]. The MCDK is generally unilateral with the left side more affected than the right one, but there are cases of bilateral MCDK which are mostly incompatible with life since renal tissue has minimal or no activity. In 60–80% of cases, it is diagnosed prenatally [1, 2].

41.3 Etiology

The etiology of MCDK is still not clarified; however, there are two predominant theories.

According to the first theory, pelvic-ureteral atresia leads to severe obstruction with massive hydronephrosis and MCDK, but according to others, the result would be a major hydronephrosis and not cystic dysplasia [2].

The second theory argues that MCDK derives from an altered interaction between the ureteric bud and metanephric blastema [3].

The interaction between the metanephros and the ureteric bud stimulates organogenesis that hesitates in the formation of the nephrons and collector systems. When this interaction is altered, the final architecture of the renal parenchyma will be subverted.

A recent study identified some genes to be mutated in children with MCDK: CHD 1L, ROBO2, HNF 1B, and SALL1 genes [4].

Another less accredited theory attributes a role to exposure to teratogens such as viral agents (cytomegalovirus, adenovirus, enterovirus) or medicaments during pregnancy (antiepileptics) that can cause malformations of the urinary tract among which MCDK [1].

41.4 Associated Anomalies

The incidence of associated anomalies ranges from 5 to 48% [1, 2, 6]. MCDK can be associated with several anomalies involving the contralateral urinary tract such as rotational or positional anomalies, hypoplasia, areas of dysplasia, vesicoureteral

M. Maffi · M. Lima (✉)
Department of Pediatric Surgery, S.Orsola Hospital, University of Bologna, Bologna, Italy
e-mail: mario.lima@unibo.it

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reflux (VUR) (7–26%), ureteropelvic junction obstruction (UPJO) (1.5–5%), ureterovesical junction obstruction (2%), ureterocele, horseshoe kidney, or genital anomalies [1, 2, 5–10].

Extrarenal associated anomalies include heart defects, esophageal or intestinal atresia, neural tube defects (myelomeningocele), and aneuploidy.

41.5 Diagnosis

41.5.1 Prenatal Diagnosis

About 60–80% of cases of unilateral MCDK are detected with prenatal ultrasound. The classical appearance of MCDK is an abdominal mass consisting of multiple thin-walled cysts of different size, not communicating with each other (Fig. 41.1). The kidney is usually enlarged and irregular, with no visible pelvis, atretic or absent ureter, and small or absent renal artery. Parenchymal tissue between the cysts is often hyperechogenic, and the kidney may reduce in size during pregnancy.

Sometimes the ultrasonographic appearance can be confused with an important hydronephrosis, but in this case, the cysts communicate each other and parenchyma can be seen. Rare case of Wilms tumor, multilocular cyst, or cystic mesoblastic nephroma can have a similar aspect to MCDK.

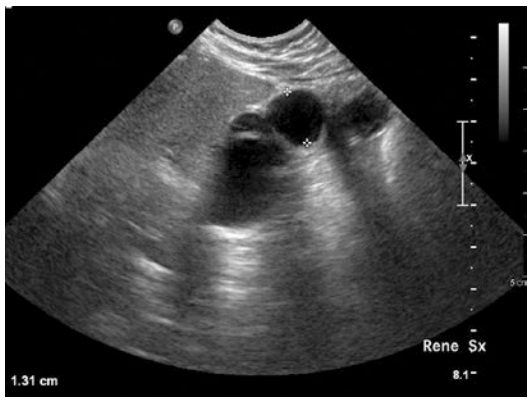

Fig. 41.1 RUS in MCDK showing multiple noncommunicating cysts

41.5.2 Postnatal Presentation

The patient may be totally asymptomatic, and the diagnosis can be done occasionally during an ultrasound examination performed for other reasons.

The most common clinical presentation is a palpable abdominal mass in the neonatal age. The surface is typically irregular in contrast to what happens in case of hydronephrosis or polycystic kidney disease. The contralateral kidney, if not dysplastic, is generally enlarged by compensatory hypertrophy.

41.6 Natural History

The natural evolution of the MCDK is the spontaneous involution. The complete involution may take place during fetal life and is greater in early life. The rate of involution ranges from 35 to 62% by 10 years of age, and if complete involution does not occur, the MCDK may decrease in size (30–44%) or remain stable (13–34%). Initial size less than 5–6 cm seems to be predictive of complete involution [1, 2, 6].

The RVU, frequently present in the contralateral kidney, is in 90% of cases of low grade (<of grade III) and very rarely requires treatment [11].

The MCDK was once accused of being the cause of hypertension. In fact different studies showed that only a very small proportion of patients develop hypertension with a rate ranging from 1.5 to 6% [2, 10, 12–14], but above all, nephrectomy resulted in the resolution of hypertension in 25–50% of cases [3].

Among the theoretical risks of MCDK is malignant degeneration. In particular it has been suggested an association with the occurrence of Wilms tumor. Again, although some cases have been reported, several reviews reported no malignancies in patients with MCDK [2, 11, 15]. We can argue that the total risk of Wilms tumor in MCDK is not increased if compared to the general population and not sufficient to indicate a prophylactic nephrectomy in a healthy patient.

41.7 Management

According to these data, the most correct approach in case of MCDK is a conservative management based on ultrasound monitoring of the kidney and the management of associated anomalies of the contralateral kidney. Currently the debate regards imaging modality, frequency, and duration of follow-up. In particular, the questioned topics involve the need for a confirmatory renal scan and for a routine voiding cystourethrogram (VCUG) and the frequency and duration of ultrasonographic follow-up.

In the past years, a routine renal scan was performed to confirm absence of function of the affected kidney (Fig. 41.2). Recently, in order to avoid radiation exposure, several studies have investigated the benefits of carrying out this investigation, concluding that renal ultrasounds (RUS) are able to diagnose almost all cases of MCDK, so renal scan should be reserved for doubtful cases [16].

Similarly, VCUG should be reserved to patients with contralateral hydroureteronephrosis or signs and symptoms of a UTI as most cases of VUR in MCDK with normal RUS are not clinically significant [2, 5, 17].

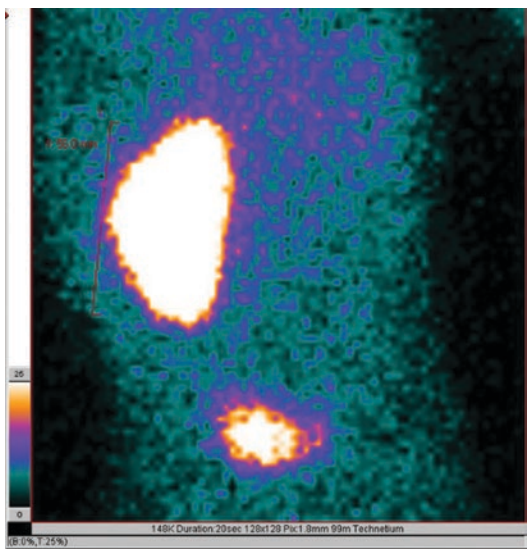

Fig. 41.2 DMSA renal scan showing absence of function of right kidney affected by MCDK disease

A possible algorithm recommends a postnatal RUS and one at 1 year of age with any other imaging investigation guided by any abnormality of the contralateral urinary tract, abnormal blood pressure, or increase in dimension of MCDK [2]. Another more widely accepted algorithm recommends performing RUS at birth, 4 weeks, 2 years, 5 years, and 10 years of age [18]. In all cases, follow-up should be completed by routine blood pressure measurement, urinalysis to detect proteinuria, and renal function studies (e.g., serum creatinine) especially in patients with contralateral abnormalities that may develop chronic renal disease [19, 20].

Currently, nephrectomy is not part of the normal management of MCDK; however, it can be indicated in selected cases:

- Symptoms caused by compression: large or increasing in size MCDK, can lead to intestinal compression. Although it has been called into question the possible decompression, nephrectomy is still one option for treatment.
- Other symptoms: in a minority of cases, the patient may complain of pain, hematuria, recurrent infections, and hypertension. This can also be an indication for surgery [21].
- Equivocal diagnosis: in case of uncertainty, it should be considered that the patient may have another potentially life-threatening disease such as cystic Wilms tumor [22].

41.8 Nephrectomy

The approach to the kidney is traditionally extraperitoneal although it can also be approached transperitoneally. The nephrectomy can be performed with open or minimally invasive surgery, laparoscopic or retroperitoneoscopic, and robotic.

In the open approach, the patient lays in the lateral decubitus with the shoulders rotated slightly forward and the pelvis rotated slightly back. The incision is performed at the apex of the 12th rib. Parietal muscles are divided by blunt dissection following fibers direction. Then the Gerota fascia is opened exposing the kidney. Vessels and ureter are then isolated and dissected. The evacuation by puncture of the cysts can facil-

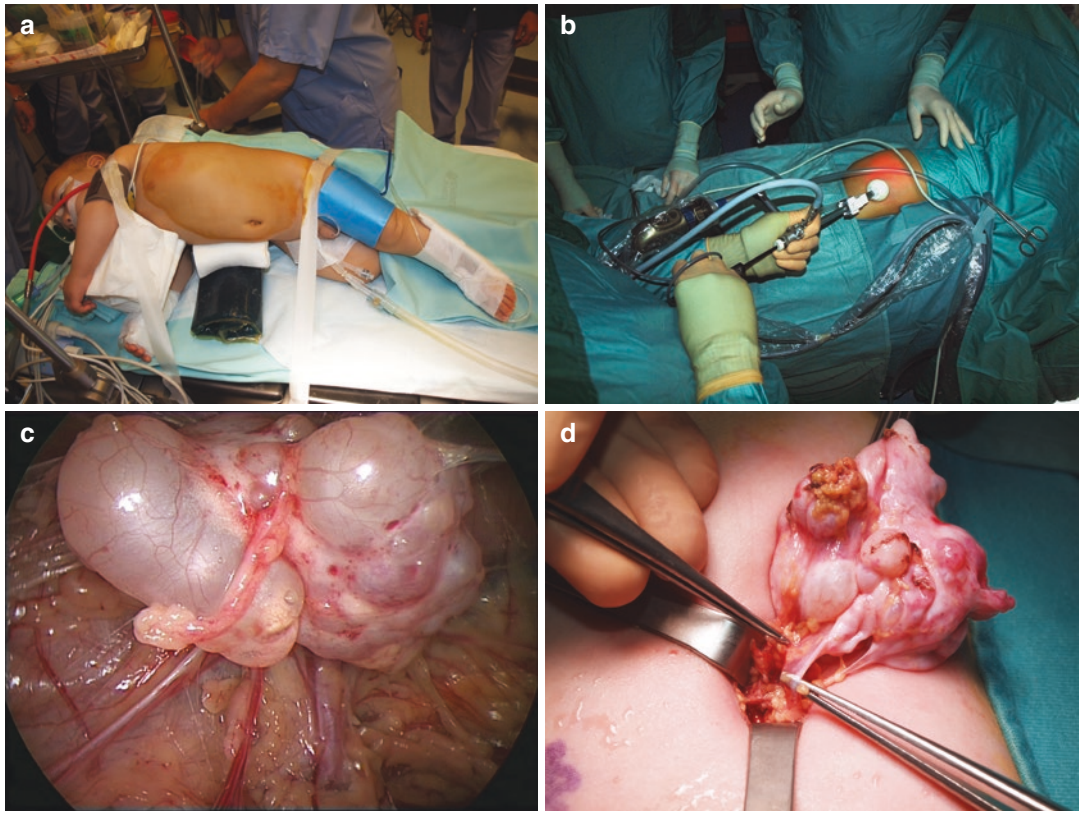

Fig. 41.3 One-trocar-assisted nephrectomy: (**a**) patient position; (**b**) operative setup and trocar placement; (**c**) retroperitoneoscopic appearance of MCDK; (**d**) exteriorization of the MCDK

itate the identification of the hilum and thus the removal of the kidney.

The minimally invasive approach can be retroperitoneoscopic with the patient in the prone position or laparoscopic. In our institute we usually perform a video-assisted retroperitoneoscopic approach through a single access (Fig. 41.3) [23]. The patient position is the same of the open approach, and the incision is performed in front of and above the apex of 12th rib but has minimum extension, just to allow the insertion of 10 mm self-anchoring trocar. The working space is created by blunt dissection with the help of a laparoscopic swab. The kidney is identified and, after emptying the cysts, pulled out through the incision and removed as in open surgery. It should be noted that in case of ipsilateral RVU, it is essential to remove the ureter as distal as possible and suture the residual stump.

References

1. Cardona-Grau D, Kogan BA. Update on multicystic dysplastic kidney. Curr Urol Rep. 2015;16:67.
2. Eickmeyer AB, Casanova NF, He C, et al. The natural history of the multicystic dysplastic kidney—is limited follow-up warranted? J Pediatr Urol. 2014;10(4):655–61.
3. Hains DS, Bates CM, Ingraham S, Schwaderer AL. Management and etiology of the unilateral multicystic dysplastic kidney: a review. Pediatr Nephrol. 2009;24(2):233–41.
4. Hwang DY, Dworschak GC, Kohl S, et al. Mutations in 12 known dominant disease-causing genes clarify many congenital anomalies of the kidney and urinary tract. Kidney Int. 2014;85:1429.
5. Ismaili K, Avni FE, Alexander M, Schulman C, Collier F, Hall M. Routine voiding cystourethrography is of no value in neonates with unilateral multicystic dysplastic kidney. J Pediatr. 2005 Jun;146(6):759–63.
6. Onal B, Kogan BA. Natural history of patients with multicystic dysplastic kidney-what followup is needed? J Urol. 2006;176(4 Pt 1):1607–11.

7. Guarino N, Casamassima MG, Tadini B, Marras E, Lace R, Bianchi M. Natural history of vesicoureteral reflux associated with kidney anomalies. Urology. 2005;65(6):1208–11.
8. Merrot T, Lumenta DB, Tercier S, Morisson-Lacombes G, Guys JM, Alessandrini P. Multicystic dysplastic kidney with ipsilateral abnormalities of genitourinary tract: experience in children. Urology. 2006 Mar;67(3):603–7.
9. Atiyeh B, Husmann D, Baum M. Contralateral renal abnormalities in multicystic-dysplastic kidney disease. J Pediatr. 1992 Jul;121(1):65–7.
10. Moralıoğlu S, Celayir AC, Bosnalı O, Pektaş OZ, Bulut IK. Single center experience in patients with unilateral multicystic dysplastic kidney. J Pediatr Urol. 2014 Aug;10(4):763–8.
11. Cambio AJ, Evans CP, Kurzrock EA. Non-surgical management of multicystic dysplastic kidney. BJU Int. 2008;101(7):804–8. https://doi.org/10.1111/j.1464-410X.2007.07328.x. Epub 2008 Jan 8
12. Sarhan OM, Alghanbar M, Alsulaihim A, Alharbi B, Alotay A, Nakshabandi Z. Multicystic dysplastic kidney: impact of imaging modality selection on the initial management and prognosis. J Pediatr Urol. 2014;10(4):645–9. https://doi.org/10.1016/j.jpurol.2014.03.004. Epub 2014 Mar 29.
13. Doğan ÇS, Torun-Bayram M, Aybar MD. Unilateral multicystic dysplastic kidney in children. Turk J Pediatr. 2014;56(1):75–9.
14. Narchi H. Risk of hypertension with multicystic kidney disease: a systematic review. Arch Dis Child. 2005;90(9):921–4. Epub 2005 May 4.
15. Narchi H. Risk of Wilms' tumour with multicystic kidney disease: a systematic review. Arch Dis Child. 2005;90(2):147–9.
16. Whittam BM, Calaway A, Szymanski KM, Carroll AE, Misseri R, Kaefer M, Rink RC, Karmazyn B, Cain MP. Ultrasound diagnosis of multicystic dysplastic kidney: is a confirmatory nuclear medicine scan necessary? J Pediatr Urol. 2014;10(6):1059–62. https://doi.org/10.1016/j.jpurol.2014.03.011. Epub 2014 May 2.
17. Calaway AC, Whittam B, Szymanski KM, Misseri R, Kaefer M, Rink RC, Karymazn B, Cain MP. Multicystic dysplastic kidney: is an initial voiding cystourethrogram necessary? Can J Urol. 2014;21(5):7510–4.
18. Aslam M, Watson AR, Trent & Anglia MCDK Study Group. Unilateral multicystic dysplastic kidney: long term outcomes. Arch Dis Child. 2006;91(10):820–3. Epub 2006 June 5.
19. Westland R, Schreuder MF, van der Lof DF, Vermeulen A, Dekker-van der Meer IM, Bökenkamp A, Van Wijk JA. Ambulatory blood pressure monitoring is recommended in the clinical management of children with a solitary functioning kidney. Pediatr Nephrol. 2014;29(11):2205–11. https://doi.org/10.1007/s00467-014-2853-0. Epub 2014 June 9.
20. Mansoor O, Chandar J, Rodriguez MM, Abitbol CL, Seeherunvong W, Freundlich M, Zilleruelo G. Long-term risk of chronic kidney disease in unilateral multicystic dysplastic kidney. Pediatr Nephrol. 2011;26(4):597–603. https://doi.org/10.1007/s00467-010-1746-0. Epub 2011 Jan 15.
21. Kumar B, Upadhyaya VD, Gupta MK, Bharti LK, Rao RN, Kumar S. Early nephrectomy in unilateral multicystic dysplastic kidney in children cures hypertension early: an observation. Eur J Pediatr Surg. 2017;27(6):533–7. https://doi.org/10.1055/s-0037-1599837. [Epub ahead of print].
22. Minevich E, Wacksman J, Phipps L, Lewis AG, Sheldon CA. The importance of accurate diagnosis and early close followup in patients with suspected multicystic dysplastic kidney. J Urol. 1997;158(3 Pt 2):1301–4.
23. Lima M, Ruggeri G, Molinaro F, Gargano T, Gregori G, Randi B. One-trocar-assisted nephrectomy (OTAN): initial experience and codification of a technique. Surg Endosc. 2012;26(4):1165–9. https://doi.org/10.1007/s00464-011-1998-1. Epub 2011 Nov 4.