Multicystic dysplastic kidney

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Learning objectives

- Review the features of multicystic dysplastic kidney and the associated genitor-urinary abnormalities
- Recognise common imaging of multicystic dysplastic kidney by renal sonography in 8 patients visited in our hospital.

Background

Multicystic dysplastic kidney (MCDK), a variant of renal dysplasia, is one of the most frequently identified congenital urinary tract abnormalities. The incidence varies, but ranges from 1 in 3500 and 1 in 4300 live births.

The multicystic dysplastic variant has multiple non-communicating cysts separated by dysplastic parenchyma. Patients with bilateral MCDKs are at risk of fetal demise secondary to oligohydramnios sequence or end-stage renal disease in the pediatric age range, while renal function is often maintained by the contralateral kidney when MCDK is unilateral.

Although MCDK can be an isolated finding, it is often identified in association with other anomalies of the kidney and urinary tract.

Imaging findings OR Procedure details

EMBRIOLOGY

The urogenital tract is the most common site of all antenatally detected anomalies. As kidney and lower urinary tract have a complex pattern of development, these processes are often perturbed, making kidney and lower urinary tract malformations a relatively common occurrence. The earlier the disturbance of the normal development takes place the more complex is the extent of the disease. Fig. 1 on page 11, Fig. 2 on page 12.

Development of the Kidney, Fig. 4 on page 13.

- The definitive kidney (metanephros) is the third in a serie of excretory organs in the human embryo, forming after the pronephros and
mesonephros; the pronephros appear in the 4th embryologic week and are nonfunctioning.

- The mesonephros form late in the 4th week and function as interim kidneys until the metanephros develop (5th week) and begin to function (9th week).
- The metanephros develop from two sources: the ureteric bud and the metanephrogenic blastema. Their development depends on mutually inductive interactions.
- The ureteric bud is an outgrowth from the mesonephric duct, near its entry into the cloaca. It elongates and branches in a dichotomus pattern giving rise to the ureter, renal pelvis, calyces and collecting tubules. Through interaction with the metanephric mesoderm it induces the formation of nephrons.
- Initially the kidneys are found in the pelvis. The ascent of the kidneys occurs due to true migration and secondary to differential somatic growth of the lumbar portion of the body. During their ascent the kidneys rotate medially 90° so that the renal pelvis is directed anteromedially. Fig. 3 on page 12.
- The kidneys are in their adult location and position by the 9th gestational week.

**Development of the Urinary Bladder**

- In the 7th gestational week, the urorectal septum fuses with the cloacal membrane dividing it into a ventral urogenital sinus and a dorsal rectum.
- The bladder develops from the urogenital sinus; initially it is continuous with the allantois, but this connection soon constricts and becomes a fibrous cord called the urachus, which extends from the apex of the bladder to the umbilicus.
- As the bladder enlarges, the distal portion of the mesonephric ducts are incorporated as connective tissue into the bladder trigone and, at the same time, the ureters come to open separately into the bladder.
- An abdominal organ in infants and children, the bladder becomes a true pelvic structure only after puberty.

**Development of the Urethra**

- The epithelium of most of the male urethra and the entire female urethra is derived from the endoderm of the urogenital sinus. The urethral connective tissue and smooth muscle form from adjacent splanchnic mesenchyme.

**NATURAL HISTORY OF MULTICYSTIC DYSPLASTIC KIDNEY**

Tipically, the kidney has the appearance of "a bunch of grapes," with little stroma between the cysts. Renal size is highly variable, from slightly less than normal to enormous, filling most of the abdomen.
When the cysts are small, even microscopic, and stroma predominates, the condition is referred to as solid cystic dysplasia.

And when an identifiable renal pelvis is associated with what appears to be a multicystic dysplastic kidney, the condition is referred to as the hydronephrotic form of multicystic kidney.

The multicystic dysplastic kidney represents an active process, not an end result. Some areas of the kidney have increased expression of genes active in nephrogenesis and anti-apoptosis (e.g., *IGF2*, *WT1*, *PAX2*, *WNT4*, *BCL2*), whereas in other areas a lack of expression of these genes might be associated with increased cell death. The size of a multicystic dysplastic kidney and its cysts results from a balance (or imbalance) between the two. Whether a multicystic dysplastic kidney grows and dilates, shrinks, or stays the same size is dependent on this balance.

The fact that the contralateral kidney often has a dilated nonobstructed renal pelvis, and sometimes even a ureteropelvic junction obstruction, makes think if multicystic dysplastic kidney is the result of obstruction.

Widespread of prenatal ultrasound evaluation has greatly increased the frequency with which the condition is identified. Although the pathogenetic process leading to a multicystic kidney probably is operative by the 8th week in utero, the mean age at the time of antenatal diagnosis is about 28 weeks, with a range of 21 to 35 weeks. The reason is not apparent. In less severely affected patients, the condition may be an incidental finding during evaluation of an adult for abdominal pain, hematuria, hypertension, or an unrelated condition. At any age, the condition is more likely to be found on the left.

Males are more likely to have unilateral multicystic dysplastic kidneys (2.4:1), whereas bilateral multicystic kidneys appear twice as often in females. Unilateral multicystic kidneys, when not associated with other renal or nonrenal anomalies, rarely involve a chromosomal disorder. Bilateral disease, however, is associated with other anomalies as well as chromosomal anomalies.

The contralateral system frequently is abnormal as well. Contralateral ureteropelvic junction obstruction is found in 3% to 12% of infants with multicystic kidney and contralateral vesicoureteral reflux is seen even more often, in 18% to 43% of infants.

Because the high incidence of reflux, voiding cystourethrography usually has been considered advisable in all newborns with a multicystic kidney.

When a diagnosis of multicystic kidney is made in utero by ultrasound, the disease is found to be bilateral in 19% to 34% of cases.
Those with bilateral disease often have other severe deformities or polysystemic malformation syndromes.

Involution sometimes occurs in the multicystic kidney, either antenatally or postnatally. This involution may be so severe that the affected kidney disappears from subsequent sonograms. In such cases, the kidney may be only a "nubbin," and the condition is referred to as "renal aplasia" or "aplastic dysplasia.

**HISTOPATHOLOGY**

Multicystic kidneys with large cysts tend to be large with little stroma, whereas those with small cysts generally are smaller and more solid.

Usually the ureter is partly or totally atretic, and the renal pelvis may be absent.

The most of the studies have shown that MCDKs tend to involute. The velocity at which MCDKs involute appears to be greater early in life. Fig. 5 on page 14.

**ASSOCIATED ABNORMALITIES OF THE GENITO-URINARY TRACT AND THE CONTRALATERAL KIDNEY**

The most frequent anomalies found in the contralateral kidney and urinary tract or in the ipsilateral mid-kidney in case of renal duplication are:

**Vesicoureteral reflux**

The most common and significant urologic reflux that is seen is VUR to the contralateral kidney. Since the contralateral kidney is the only functional kidney, VUR with ascending infections could be detrimental, from renal scarring, leading to chronic renal insufficiency and hypertension.

**Obstruction**

Obstruction are often seen in patients with MCDKs. Ureteropelvic junction obstruction (UPJ) and ureterovesical junction (UVJ) obstruction are reported up in 15% and 6% of the patients.

**Megaureter:** primary megaureter or secondary megaureter.
Ureterocele.

Blindureter.

Renal duplication

Nephroblastomatosis (not reported in the literature)

DIAGNOSTIC IMAGING EVALUATION

Renal masses in infants most often represent either multicystic kidney disease or hydronephrosis, and it is important to distinguish between the two, especially if the surgeon wishes to remove a nonfunctioning hydronephrotic kidney or repair a ureteropelvic junction obstruction while leaving a multicystic organ in situ. In newborns, ultrasonography usually is the first study performed. In a few cases, it is difficult to differentiate multicystic kidney disease from severe hydronephrosis.

In general, however, the multicystic kidney has a haphazard distribution of cysts of various sizes without a larger central or medial cyst and without visible communications between the cysts. Frequently, very small cysts appear between the large cysts.

In comparison, in ureteropelvic junction obstruction the cysts or calyces are organized around the periphery of the kidney, connections usually can be demonstrated between the peripheral cysts and a central or medial cyst that represents the renal pelvis, and there is absence of small cysts between the larger cysts. When there is an identifiable renal sinus, the diagnosis is more likely to be hydronephrosis than multicystic kidney.

In these difficult cases, radioisotope studies may be helpful.

Hydronephrotic kidneys usually show some function on DMSA scan, whereas renal uptake is seldom seen in multicystic kidneys.

Voiding cystourethrography is indicated in the workup because of the high incidence of reflux into the single functioning kidney.

EVOLUTION

The following events may occur during the evolution of MCDK:
COMPENSATORY HYPERTROPHY OF THE CONTRALATERAL KIDNEY

The kidney contralateral to the MCDK undergoes compensatory hypertrophy to offset partially the loss of functional renal tissues in the dysplastic kidney. While the mechanism is unclear, compensatory hypertrophy likely begins in utero and is defined as a renal length greater than +2 standard deviations of the mean.

The absence of compensatory hypertrophy may indicate a pathological condition such as hypoplasia in the contralateral kidney.

BLOOD PRESSURE

Historically, hypertension has been considered a potential complication of MCDK and has been provided as a reason for nephrectomy of the affected kidney.

Nowadays, most studies don’t show an increased risk of hypertension in MCDK patients compared with the general population.

DIAGNOSIS AND FOLLOW-UP STUDIES

Historically, MCDK was a rare finding, usually diagnosed by palpation of an abdominal mass. Identification of MCDK in newborns has increased with the use of fetal ultrasound.

Unilateral MCDK is relatively evenly divided between the right and left kidney and occurs slightly more often in male patients.

The differential diagnoses of MCDK include: polycystic kidney diseases and glomerulocystic disease. Fig. 7 on page 28.

ULTRASOUND. Fig. 6 on page 30.

If MCDK is suspected on prenatal US, a postnatal US will confirm the diagnosis and screen for other urinary tract abnormalities. If normal postnatal US findings are obtained in the first 48h of life, a subsequent US should be obtained at 6 weeks, due to the high rates of false negatives secondary to relative oliguria shortly after birth.

The indications for follow-up US in unilateral MCDK have included screening for Wilms tumor and evaluating for involution of the MCDK, and confirming compensatory hypertrophy of the contralateral kidney.
Given the low incidence of Wilms tumor, the frequent testing needed to reduced tumor stage, and the lack of evidence for improvement in mortality rates with monitoring, the routine screening for Wilms tumor is not yet indicated.

**VOIDING CYSTOUREROGRAM**

Undiagnosed high-grade VUR to the contralateral normal kidney could lead to pyelonephritis and scarring in the only functioning renal unit in patients with MCDK. Given the risk of scarring in a functional solitary kidney, a VCUG is recommended in MCDK patients that exhibit an abnormal contralateral upper urinary tract and/or kidney on US.

**NUCLEAR MEDICINE STUDIES**

If a renal pelvis is identified in the suspected MCDK, a renogram should be obtained to confirm that a hydronephrotic, obstructed kidney is not present. Since patients with MCDK are at risk of acute renal failure and associated complications with a UPJ or UVJ obstruction in the contralateral kidney, it is important to diagnosis these conditions.

If a renal pelvis diameter of >5mm in the contralateral kidney is present with a negative VCUG finding or >10mm with a positive VCUG finding, a diuretic renogram should be obtained.

**BLOOD PRESSURE MONITORING**

Routine blood pressure checks in the primary care setting should be sufficient, since patients with MDCK do not appear to have an incidence of hypertension greater than that of the general population.

Currently, it is recommended that pediatricians check blood pressures in clinical encounters after the age of 3 years, with the blood pressure measured for special circumstances under the age of 3 years.

**MEASUREMENT OF SERUM CREATININE**

Serum creatinine should be measured at the initial examination to confirm normal renal function. While compensatory hypertrophy is encouraging, patients with unilateral MCDK and apparently normal contralateral kidney still have a 12-50% incidence of at least stage 2 chronic renal failure at 10 years of age, according to recent studies.

Nowadays it is recommended the creatinine measurement at the 2-year and 5-year follow-up examinations, so that progressing chronic renal failure may be rapidly diagnosed.
REMOVAL OF THE MCDK

The latest studies suggest that non-surgical management of MCDK is a viable option in treating the patients with MCDK.

CASES

Case 1, Fig. 8 on page 27, Fig. 9 on page 26.

Asymptomatic female patient with multicystic dysplastic kidney suspected by prenatal ultrasound, whose diagnosis was confirmed after delivery.

In the first pictures were seen cystic lesions on the upper pole of the kidney; the more caudally upper right renal parenchyma was preserved. There was an image suggesting a duplex kidney and another one suggesting cystic juxtavesical right ureterocele.

Left kidney unchanged.

In the next control, persist the image of the renal duplication and theoretical multicystic renal dysplasia in the upper pole. We see pelvicalyceal ectasia of the lower pole of the kidney probably associated with a megaureter. The left kidney persists unchanged.

Two months later the patient presented with symptoms of pyelonephritis. An US was performed that showed the ureterocele at the bladder and a right ureteral and pelvicalyceal dilatation. There are also cystic images in the upper pole of an hypertrophic left kidney not seen previously.

The image of acute pyelonephritis in the upper pole was correlate with DMSA.

Case 2, Fig. 10 on page 25, Fig. 11 on page 24.

Involuted dysplasia (9-year old girl with involuted MDCK of the left kidney, IV grade VUR and right nephropathy).

US: empty left renal fossa after involution of MCDK. Right kidney with hyperecogenity of the cortex associated with multiple, small, refractive images of the pyramids that resemble the medullary collecting duct wall due to cystic dilatation or fibrotic changes secondary to reflux nephropathy.

In the following controls persisted the hyperechogenity of the right renal cortex.

Case 3, Fig. 12 on page 23, Fig. 13 on page 22.

Left kidney of normal morphology.

Case 4, Fig. 14 on page 21, Fig. 15 on page 15.

Prenatally US suspected of MCDK.

First US: cystic lesions and loss of the renal parenchyma in the right, upper mid-kidney and pyelocalyceal dilatation of the lower pole of the kidney, supporting the diagnosis of multicystic renal dysplasia and renal duplication.

Contralateral kidney with multilobed, hyperechoic images that resemble a nephroblastomatosis, not yet confirmed.

Two months later, the two US remained unchanged.

Case 5, Fig. 16 on page 16, Fig. 17 on page 17.

Two years old boy with cystic lesions of the upper mid-kidney with total loss of the renal cortex, that suggests partially involuted MCDK.

Two years later the next US shows the involution of the cystic lesions and the compensatory hypertrophy of the contralateral kidney.

Case 6, Fig. 18 on page 18.

Five years old girl:

First US showed two cystic lesions in the right renal fossa and atrophy of the renal cortex. Left kidney was normal.

In the next US the cystic lesions had been decreased and the contralateral kidney was hypertrophied compensatory.

Case 7, Fig. 19 on page 19, Fig. 20 on page 20.

Two years old boy:
Prenatal US: right pelvicalcilar mild ectasia and left cystic lesions of maximum 8mm diameter.


The next US: right compensatory hypertrophy and left blind ureter.

Case 8

Four years old boy:

Prenatal US: pelvicalyeal ectasia and cystic lesions in the right renal fossa.

Control US: involuted dysplasia of the right upper mid-kidney, image suggestive of renal duplication and right pelvicalcilar mild ectasia suggestive of junction stenosis.

Images for this section:
**Fig. 1:** Fig.1

The green portions of the drawing are uretic bud derivatives, the red portions are the metanephric bud derivatives.

**Fig. 2:** Fig.2
the kidney does maintain its lobulated appearance throughout development. Note the change in size of the kidneys and adrenal glands relative to one another.

Fig. 3: Fig.3
Fig. 5: Fig.5

Cut surface of a nephrectomy specimen from a patient with a multicystic dysplastic kidney (MCDK).
Contralateral kidney with multilobed, hyperechoic images that resemble a nephroblastomatosis, not yet confirmed.

Fig. c) Contralateral kidney with multilobed, hyperechoic images that resemble a nephroblastomatosis, not yet confirmed.
Fig. d) Follow-up US: the hyperechoic lesions persisted unchanged.

Fig. 15: Fig.15
Two years old boy with cystic lesions of the upper mid-kidney with total loss of the renal cortex, that suggests partially involuted MCDK.

Fig. a) The arrow shows partially involuted MCDK.
Fig. b) Compensatory hypertrophy of the right kidney.

Fig. 16: Fig.16
Two years later the next US:

Fig. c) Compensatory hypertrophy of the right kidney (23 mm)
Fig. d) Persisted small residual cystic lesions. The arrow shows partially involuted MCDK

**Fig. 17:** Fig. 17
Five years old girl First US showed two cystic lesions in the right renal fossa and atrophy of the renal cortex.

Fig. a), b) Two cystic lesions in the right renal fossa and atrophy of the renal cortex.
Fig. c) Left kidney was normal. In the next follow-up was hypertrophied compensatory.

**Fig. 18:** Fig.18
Two years old boy:
Prenatal US: right pielocalcilar mild ectasia and left cystic lesions suggestive of MCDK.
Control US: involated dysplasia.

Fig. a) Empty left kidney fossa (S)pleen.
Fig. b,c) Normal right kidney. compensatory hypertrophy
Fig. 20: Fig.20

Fig d), e) Image suggestive of blind ureter (u)
Case 4

First US: cystic lesions and loss of the renal parenchyma in the right, upper mid-kidney and pyelocalyceal dilatation of the lower pole of the kidney, supporting the diagnosis of multicystic renal dysplasia and renal duplication.

Fig. a, b: Cystic lesions and loss of the renal parenchyma in the right, upper mid-kidney and pyelocalyceal dilatation of the lower pole of the kidney.

Fig. 14: Fig.14
The image of the pelvis shows a diastal megaureter and ureterocele. (VCUG indicative of the VUR in the right, lower mid-kidney).

Fig. d) Right megaureter (Mgu)
Fig. e) More distally, ureterocele (u)

**Fig. 13:** Fig. 13
Case 3

Fig. 12: Fig.12

7 months old boy with right renal duplication. MCDK in the upper mid-kidney and pelvicalyceal ectasia of the upper mid-kidney. Megaureter and distal ureterocele. Normal left kidney.
VCUG demonstrates VUR in the right, lower mid-kidney

Fig. a, b) Right renal duplication with cystic lesions in the upper mid-kidney and total loss of the renal parenchyma and pelvicalyceal ectasia of the upper mid-kidney.
Fig. c) Left kidney of normal morphology
The next controls demonstrated resolution of the reflux and the persistence of the renal cortex nephropathy secondary to this one.

Fig. c), d) In the next control persisted the hyperechogenicity of the right renal cortex.

**Fig. 11:** Fig.11
Case 2

Fig. 10: Fig.10

*Fig. a*  Girl diagnosed of left MCDK by prenatal US.

*Fig. b*  Postnatal US shows involution of the left kidney and an axial image of the right kidney with loss of cortico-medullary differentiation secondary to reflux nephropathy. VCUG demonstrated IV grade VUR

Fig. a) Empty left renal fossa after involuted MCDK.

Fig. b) Axial image of the right kidney with loss of cortico-medullary differentiation secondary to reflux nephropathy.
Two months later the patient presented with symptoms of pyelonephritis and the U.S. shows a dilated pelvicalyceal megaureter and a urethrocele.

Fig 1c) Pyelocaliceal ectasia of the right kidney associated with megaureter (Mgu)  
Fig 1d) Juxtavesical ureterocele (arrow).  
B = bladder.

**Fig. 9:** Fig.9
Case 1

**Fig. 1a**

**Fig. 1b**

*MCDK in duplicated right kidney with posterior disgenesis megareter with urterocele of inferior unit. VCUS was negative for RVU.*

Fig. 1a: Right renal fossa confluent cystic lesions occupying the apical pole of the kidney. Fig. 1b: Left kidney of 5.3 cm in length with preserved ecostructure. It shows not significative calyceal estasia of the urinary tract.

**Fig. 8:** Fig.8
THE FOLLOW-UP OF THE PATIENTES WITH MCDK:

- An US at birth, 1 month, 2, 5 and 10 years of age to screen initially for findings suggestive of VUR and to monitor the contralateral growth.
- A cystourethrogram if the patient develops a urinary tract infection or has abnormality of the contralateral kidney on US.
- Routine blood pressure and serum creatinine monitoring.

**Fig. 7: Fig. 7**
CONCLUSIONS

- MCDK is associated with active expression of genes involved with nephrogenesis.

- MCDK has a changing morphology.

- Kidneys usually get smaller or disappear from imaging studies.

- There is no clear indication for removal of the kidney.

- Some kidney abnormalities associated with MCDK indicates the need of ultrasound or other imaging procedures (VCUG, DMSA) follow-up.

Fig. 21: Fig.21
-Kidney grossly enlarged with loss of reniform shape and ureter atretic or absent.

-Multiple variably sizes noncommunicating cysts separated by little or no echogenic parenchyma.
Conclusion

Multicystic dysplastic kidney is associated with active expression of genes involved with nephrogenesis, and has a changing morphology.

Kidneys usually get smaller or disappear from view on imaging studies (i.e., renal aplasia), very occasionally increase in size, and very rarely are associated with Wilms’ tumor.

There is no clear indication for removal of the kidney unless an increased amount of solid tissue is identified.

The follow-up of the patients with MCDK include:

- an US at birth, 1 month, 2 years, 5 years and 10 years of age to screen initially for findings suggestive of VUR and to monitor contralateral growth.

- a cystourethrogram if the patient develops a urinary tract infection or has abnormality of the contralateral kidney on US;

- routine blood pressure and serum creatinine monitoring. Fig. 21 on page 32.

Images for this section:
CONCLUSIONS

-MCDK is associated with active expression of genes involved with nephrogenesis.

-MCDK has a changing morphology.

-Kidneys usually get smaller or disappear from imaging studies.

-There is no clear indication for removal of the kidney.

-Some kidney abnormalities associated with MCDK indicates the need of ultrasound or other imaging procedures (VCUG, DMSA) follow-up.

**Fig. 21:** Fig.21
Personal Information

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