

Pediatric Urology Associates, Ltd.

& Pediatric Enuresis Center of Arizona

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MULTICYSTIC DYSPLASTIC KIDNEY

Multicystic dysplastic kidney (MCDK) is the term used to describe a congenital renal anomaly that consists of a kidney completely replaced by multiple cysts that are held together by connective tissue. The kidney looks like a bunch of grapes. The ureter is often atretic or absent. The cysts often do not communicate and this condition must be distinguished from hydronephrosis.

With increasing utilization of prenatal ultrasound, the common mode of presentation has changed dramatically. At the present time, 47% are detected prenatally, 30% present with a flank mass, 5% urinary tract infection, 18% other presentations. It is more common in males and there is a slight left-sided predominance.

The diagnostic evaluation includes a renal ultrasound which shows several characteristic features that helps to distinguish a multicystic kidney from a hydronephrotic kidney. To substantiate the appearance of the renal ultrasonogram, a renal scan MAG-3 is usually obtained. This test usually confirms the absence of renal function. In addition, this test helps to evaluate the contralateral kidney. In 18% of the cases, a contralateral renal abnormality may be encountered: ureteropelvic junction obstruction, ureterovesical junction obstruction. In some cases, there might also be contralateral vesicoureteral reflux. For this reason, a nuclear cystogram or voiding cystourethrogram is also recommended.

If the condition is detected prenatally, it is important to visualize the opposite kidney to see whether it is abnormal. At the same time, the amniotic fluid volume should be assessed to make sure that it is adequate. If the amniotic fluid volume is markedly decreased and if there is bilateral renal disease, further steps should be taken to assess the viability of the baby. Luckily, the disease is rarely bilateral and the patients usually do well with one kidney.

The treatment of this condition is controversial. One group favors surgical removal of the kidney and the other favors observation. The risks of leaving the kidney in place include hypertension (rare) and the development of a renal tumor. Only 6 cases of renal tumors have been reported, 3 in childhood. The childhood tumors occurred at 10 months, 4 years, and 15 years; whereas, the adult tumors occurred between 26 and 68 years. Based on the incidence of this disease, the risk of developing a tumor is low. If the kidney is left in place, the patients must be observed yearly with renal ultrasound and blood pressure measurements.

We favor the surgical removal of this kidney for two important reasons. In the first place, the procedure can be accomplished electively with a short postoperative stay. Secondly, if observation is chosen, the patients must be followed until the ultrasound confirms complete involution of the kidney.

In our experience, in 10 to 15% of the cases, the kidney involutes (can no longer be found on ultrasound). For this reason, usually the patient is followed with ultrasound during the first 6 to 12 months of life. If the kidney disappears on ultrasound, no surgical exploration is indicated. However, if after this period of time the MCDK persists, surgical removal of the kidney will be offered. In rare occasions, the diagnosis of MCDK will be inconclusive despite the radiologic workup and in this situation, surgical exploration is indicated.