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CAUSES OF MULTICYSTIC DYSPLASTIC KIDNEY DISEASE AND ITS CLINICAL FEATURES IN CHILDREN

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Annotation. Multicyctic dysplastic kidney disease (MCDK) is a genetic disorder characterized by the development of multiple cysts in the kidneys. Multicystic dysplastic kidney is a condition in which the kidney has been essentially replaced by multiple cysts. It is the result of abnormal fetal development of the kidney. There is little or no normal function to this kidney.

Keywords: multicyctic dysplastic kidney disease (MCDK), cysts, stroma, hydronephros, obstructiom, vesicoureteral reflux (VUR).

Introduction

In 1936, Schwartz was the first to describe a “unilateral multicystic kidney” in a specimen removed from a 7-month old boy suspected to have Wilms’ tumor or massive hydronephrosis. He described an irregular lobulated mass with various-sized cysts resembling a “bunch of grapes”.

The morphologic appearance of an MCDK varies depending on the predominance or absence of the cystic component. Three morphologic types of MCDK exist. A kidney with small cysts and abundant dysplastic stroma is a solid cystic dysplastic kidney. A kidney with large cysts and minimal stroma is the typical MCDK. Finally, an identifiable renal pelvis within dysplasia is referred to as the hydronephrotic form of MCDK. Two phenotypes of dysplasia are associated with urinary tract abnormalities: obstructive and multicystic. Obstructive renal dysplasia associated with lower urinary tract obstruction is characterized by peripheral cortical cyst formation. In contrast, MCDKs are characterized by an absent or atretic pyelocalyceal system and multiple deformed cysts. Shibata and colleagues investigated the pathology of nine MCDKs using detailed morphologic analysis. They found that the incidence of glomerular cysts in MCDKs decreases with gestational age. Three-dimensional reconstruction also showed that the cysts were connected to the collecting ducts through tubules, and that basic nephron structures could be found in the cystic nephron. The three-dimensional analysis by Shibata and colleagues supports a theory that cysts develop from early induced nephrons.

The etiology of MCDK disease is unclear. Many investigators have proposed that obstruction is the primary factor involved in the development of dysplasia. Felson and Cussen in 1975 viewed the multicystic kidney as an extreme form of hydronephrosis that occurs secondary to atresia of the ureter or renal pelvis. Cystic dysplasia of an entire upper pole can be seen in kidneys with duplicated collecting systems, especially associated with ureteroceles. Bilateral cystic dysplasia also may be seen secondary to posterior urethral valves. Animal models involving ligation of the ureter during early gestation have shown the induction of dysplasia. Multicystic dysplasia secondary to obstruction has not been simulated in an animal model, however. Shibata and Nagata performed detailed morphologic studies of fetal dysplastic kidneys to show that early in utero obstruction causes urine retention in the developing nephrons, leading to multiple cyst formation. They reported that nephron induction with filtrating function occurs before the development of cysts; early fetal urinary tract obstruction causes cystic formation in the developing nephrons, which subsequently disrupts nephron induction and tubular development. Shibata and Nagata also cited the importance of further investigation into abnormalities in the activity of transcription factor PAX2 and antiapoptosis protein BCL2 in the pathogenesis of renal dysplasia. There are many theories regarding the origin of this disease, below we will discuss the current research focusing on genes.

More recent studies have focused on finding the genes involved with renal development whose abnormal expression is responsible for renal dysplasia. Various genes have been identified, including BMP4, FOXC1, KAL, EYA1, and AGTR2; mutations in these genes have been identified in ectopic budding of the ureter from the wolffian duct and the subsequent urogenital anomalies. One of the most studied is PAX2, a member of the PAX gene family that encodes transcription factors crucial in embryogenesis. It is expressed in mesenchymal/epithelial conversion and branching morphogenesis during nephron formation, and is involved with eye, ear, and central nervous system development. Its persistent expression has been shown to cause epithelial proliferation and cyst formation, as seen notably in renal coloboma syndrome, an autosomal dominant disorder that involves ocular and renal malformation, commonly vesicoureteral reflux (VUR) and renal hypoplasia. PAX2 also is abnormally expressed in patients with Wilms' tumor and renal cell carcinoma owing to its relationship to WT1 expression, which may be the link between dysplastic kidneys and malignant conversion. Winyard and associates showed that the PAX2 protein and BCL2, an antiapoptosis protein, are abnormally expressed in MCDK, and proposed that cyst formation in dysplastic kidneys is caused by overexpression of PAX2 and ectopic expression of BCL2; MCDK is a result of aberrant temporal and spatial gene expression. A case report has been published that described a family with renal coloboma syndrome and a novel mutated PAX2 gene with autosomal dominant inheritance whose phenotype showed MCDK, VUR, UPJ obstruction, and renal hypoplasia. Although the etiology of MCDK disease remains unclear, theories of intrarenal obstruction and failure of ureteral bud–metanephros interaction predominate. Future research is necessary to elucidate further the complex genetic events that are responsible for renal dysplasia and the specific events that lead to multicystic renal dysplasia.

MCDK disease is one of the most common causes of an abdominal mass in infants.13,32-35 The incidence of MCDK disease is estimated to be 1:3100 to 1:4300, a much higher number than was reported in the era before routine antenatal ultrasound. Previously, the diagnosis was most often made in infancy in the presence of a palpable abdominal mass, which is found in 22% to 37% of cases.35-37 It is difficult to ascertain what percentage of prenatally diagnosed MCDKs would have been identified by palpation during routine neonatal examination had the diagnosis not been made previously. Diagnosis may be made at 15 weeks of gestation on routine prenatal screening ultrasound; mean age of diagnosis is 28 weeks (range, 21 to 35 weeks). MCDK is more common on the left, with a slightly greater incidence in boys. Postnatal presentation includes palpable abdominal mass, flank pain, urinary tract infection (UTI), or hypertension.

Renal agenesis and MCDK disease share similar clinical features, including male predominance, left-sided predominance, and a similar incidence of contralateral renal anomalies. Segmental multicystic dysplasia may appear in horseshoe kidneys and in single moieties of kidneys with duplicated collecting systems. Borer and associates reviewed the literature and found 18 cases with MCDK disease as a component of a horseshoe kidney. Laterality of the diseased segment was reported in 12, with equal occurrence in the right and the left kidneys. Segmental disease in a horseshoe kidney may present as a midline abdominal mass. A review of the literature by Corrales and Elder62 found segmental MCDKs in a duplicated system to be more common in girls, with the left side involved more often than the right. Renal scintigraphy is essential in evaluation because ultrasound evaluation can overlook a functional segment that is obscured by the large cystic dysplastic portion. Anomalies in the lower, nondysplastic moiety include lower pole VUR, lower pole UPJ obstruction, and orthotopic ureterocele.

In 20% to 43% of MCDKs, contralateral upper or lower urinary tract abnormalities are encountered, with the most common being contralateral VUR, occurring in 15% to 28% of cases. Contralateral UPJ obstruction is found in 3% to 12% of patients.

In addition to anomalies of the contralateral kidney, patients with MCDKs can have malformations of the ipsilateral internal genitalia owing to the close embryologic relationship of the urinary and reproductive systems. Ipsilateral anomalies of the testes and vas may be associated with renal anomalies, including MCDKs, owing to their embryologic association with the wolffian duct. During development, the rete testes become continuous with the vas deferens and the epididymis, which are distal derivatives of the wolffian duct. The wolffian duct gives rise proximally to the ureteral bud, which induces the metanephros to form the kidney. Insult to the wolffian duct can potentially cause renal and testicular defects. The most common abnormality of the wolffian duct is an absence of the vas deferens and ipsilateral renal agenesis. Wolffian duct anomalies associated with MCDK disease include absence or ectopia of the ipsilateral vas deferens and cystic dysplasia of the testis.

In other situations in which patients are left with solitary kidneys, the remaining kidney undergoes compensatory

hypertrophy. In infants and children, this growth occurs in addition to the rapid growth and changes associated with normal renal maturation. Compensatory hypertrophy of the contralateral kidney has been reported in multiple series of patients with MCDKs.

Conclusion, It is important to remember that the risk factors that accompany genetic factors in the development of this disease also need to be taken into account. Regardless of how MCDK manifests itself in the body, it is a serious threat to the child's life.

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