Multicystic Dysplastic Kidney

Abstract

Multicystic dysplastic kidney (MCDK) is a renal abnormality that contains multiple cysts of various shapes and sizes in the kidney. Unilateral MCDK is very common in children and usually is associated with other contralateral anomalies. There are a few different etiology theories that are being evaluated, but the origination is still uncertain. Although there is no cure for MCDK, there are a few treatment possibilities that are determined by the severity of the patient. Certain imaging modalities such as Ultrasonography (US) are used to adequately evaluate and contribute to a diagnosis. In the two case reports, both females were diagnosed with MCDK on the right side; however, the patients underwent different treatment plans that effectively managed MCDK.

Introduction

Renal dysplasia also known as multicystic dysplastic kidney (MCDK) is a developmental irregularity that is composed of non-functioning cysts (see Figure 1a).\textsuperscript{1,4,7,9} It is the most documented renal cystic complication in children and occurs in 1 of every 4300 live births.\textsuperscript{2,4,7} MCDK may be detected prenatally by ultrasonography (US), but a post-delivery screening is needed to determine whether contralateral abnormalities exist or if other complications are present. Although the odds of having contralateral abnormalities are high, the prognosis for children with this disease are actually quite favorable if it only affects one kidney. The treatment options for MCDK include a conservative approach such as routine monitoring with US or a surgical approach such as nephrectomy. Either of these treatments are proven to be effective but they are determined by the urologist as well as the patient.
**General Information**

MCDK is a malformation that transpires during the development of a fetus. An abdominal mass may be felt as a protuberance once the child is born.\textsuperscript{1,4,9} The abnormality may be unilateral or bilateral, which means it may affect one or both of the kidneys. The kidney that is deformed is usually damaged and is unable to function as a healthy kidney. Renal function is then sustained exclusively by the contralateral kidney. Often times, the kidney opposite of MCDK undergoes compensatory hypertrophy which means the kidney increases in size to compensate for the loss of function from the dysplastic kidney.\textsuperscript{7} Unilateral MCDK is more prevalent in males while bilateral MCDK is more prevalent in females.\textsuperscript{1,5} If the condition is bilateral, the baby is usually aborted or a miscarriage occurs. Although there are certain treatments for this disease, there is still no cure.

**Anatomy of a Kidney**

The kidneys are positioned bilaterally in the upper, posterior portion of the abdomen. The bean-shaped structures are protected by the rib cage as well as a protective layer called the fibrous capsule. The renal parenchyma is the functioning portion of the kidneys (see Figure 2).\textsuperscript{10} It is divided into two main portions, the cortex and the medulla.\textsuperscript{11} The cortex contains small artery tufts called glomeruli.\textsuperscript{10} The glomerulus give the cortex a grainy appearance, and much of the blood is filtered through this location. The medulla contains 8 to 18 triangular masses called the renal pyramids.\textsuperscript{10} The apex portion of the renal pyramids, known as the renal papilla, direct urine to flow into the minor and major calyces. The major calyces unite to form the renal pelvis. The renal pelvis is funnel shaped and connects to the ureter.\textsuperscript{10,11} From there, the urine flows into the ureter and then down into the bladder (see Figure 3).
Function of a Regular Kidney

The kidneys are crucial organs that the body needs to survive. Some important functions that they perform on a daily bases include: producing and eliminating urine, balancing electrolytes, eliminating nitrogen wastes, adjusting water levels, and regulating pH levels in the body.²⁰ Urine is produced by filtering the blood that circulates through the kidneys. “At rest, more than 1 L of blood flows through the kidneys every 60 seconds, which results in removal of about 180 L of filtrate from the blood every 24 hours.”³⁰ Although 180 L of filtrate is removed, 99% of the filtrate is reabsorbed by the kidneys and returned back into the bloodstream.²⁰ The nephron, which is the basic working unit of the kidney, determines what must be excreted and what must be absorbed.¹¹ As the reabsorption process occurs, the pH levels, water levels, and the electrolytes are maintained by the kidneys. After reabsorption has taken place, certain materials such as creatinine, urea, uric acid, drugs, and unwanted electrolytes are excreted in the form of urine.

Etiology

The origination of MCDK is still unclear, but certain associations have been made such as hereditary disturbances, teratogens, infections, and urinary tract obstructions.⁷ Mutations in the EYA1, SIX1, and PAX2 genes have been correlated to many renal complications because of the significant role they have on renal development.⁷ “A family with a novel PAX2 mutation had affected members with MCDKs (as well as other renal anomalies) that occurred across three generations.”⁷(p.234) Certain antiepileptic medications have been linked to renal malformations and are considered contributing factors to the development of MCDK. Other contributing factors
of MCDK that have been associated with fetal viruses may include: the enterovirus, cytomegalovirus, and adenovirus.\textsuperscript{7}

**Conservative Approach**

A conservative approach is a form of treatment by which patients are routinely monitored to see how the MCDK is progressing and to verify that the healthy kidney is functioning properly. US has been the modality of choice to monitor and evaluate MCDK. The reason being is that US can provide prenatal detection of MCDK and may also monitor the progression or reduction of the cysts without using ionizing radiation to the infant. Many researchers and physicians have noticed that a substantial amount of patients with MCDK have experienced involution, which is shrinkage of the cysts within the kidney.\textsuperscript{1-5,9} “Complete involution rates vary from 19–74% over 9 months to 10 years, depending on the study.”\textsuperscript{7} The amount the cysts shrink is very important in determining the natural course of the disease and whether or not surgical intervention is needed. In a study conducted by Tiryaki et al\textsuperscript{2}, forty-six patients with MCDK were evaluated for four years. They found that a kidney length of 62 millimeters on a US was linked with faster involution.\textsuperscript{2,7} The authors also stated, “The majority of the non-involuted kidneys were in the big and left-sided group, and in fact the chance of involution for a large kidney on the left was zero; however, the involution risk for a small right-sided kidney was 67\%.”\textsuperscript{2} With this type of data, treatment selection may be made easier and more effective than ever before.

**Surgical Approach**

“Historically, until 1980’s, nephrectomy was regarded as the standard treatment to avoid complications of infection, bleeding, flank pain, hypertension, and possible malignant
In recent years, the treatment options have been debated due to the noninvasive conservative approach. However, the newly developed laparoscopic nephrectomy procedure has changed the outlook on surgical intervention. "The benefits of this technique are a shorter hospital stay, faster recovery, a smaller wound, fewer wound-related complications, and less postoperative pain compared with the open technique or the mini-incision technique." Many studies have leaned towards this surgical procedure because the natural progression of the disease is unknown and surgical removal is inexpensive compared to multiple scans throughout the patient’s lifetime. In a case report conducted by Nishio et al they found that when analyzing US results, a clear scan doesn’t necessarily mean that MCDK has involuted completely because an ultrasound cannot detect small remnants from an involute cyst.

**Contralateral Abnormalities**

Contralateral abnormalities are defects that affect the healthy kidney and are commonly associated with unilateral MCDK. Some of these abnormalities include:

- Vesicoureteral reflux.
- Hydronephrosis (see Figure 1b).
- Ureterocele.
- Crossed ectopia.
- Echogenic kidney.
- Ureteropelvic junction obstruction.
- Ureterovesical junction obstruction.

Vesicoureteral reflux is the most common irregularity and causes urine to flow backwards from the bladder into the ureter or even the kidney. This abnormality can cause pain and discomfort to the patient. It may also scar the kidney or quite possibly disrupt the function of the kidney. The second most common defect is the ureteropelvic junction obstruction. This occurs when a blockage is formed between the renal pelvis portion of the kidney and the ureter.
The blockage usually emerges when the kidneys are developing in the womb. If these abnormalities are not detected prenatally with ultrasound, frequent urinary tract infections (UTIs) will occur and further testing will be required.

**DMSA and VCUG**

A dimercaptosuccinic acid scan (DMSA) is a nuclear medicine scan that generates tomographic and three-dimensional pictures of the kidneys.\(^1\,^6\) This scan can detect the cortical scarring caused by contralateral abnormalities and how the kidneys are functioning. DMSA scans are often used to distinguish between upper and lower UTIs when certain tests and physical symptoms are often uncooperative.\(^1\,^6\) Unfortunately, DMSA scans are not useful in differentiating contralateral abnormalities. A voiding cystourethrogram (VCUG) however, is beneficial in determining which abnormality is causing the UTIs.\(^1\,^6\,-\,^7\) A VCUG is a fluoroscopy exam that determines how the bladder is filled and if reflux occurs. It also detects the severity of the reflux on a scale of 1-5, with five being the worst possible.

**A Surgical Case Report**

A 5-month-old girl presented with frequent episodes of vomiting. There was not a previous history of UTIs or appendicitis. The urinalysis that was performed came back normal, and the culture findings were negative.\(^3\) First she underwent a computed tomography (CT) that discovered a right sided mass and no visualization of a right kidney. The next test that was performed was a magnetic resonance imaging (MRI) which confirmed the CT results of a 20 x 30 mm cystic mass on the right side.\(^3\) A VCUG as well as DMSA were also performed which showed no right kidney and no known vesicoureteral reflux. A ureteral orifice was not identified
in a cystoscopy or a vaginoscopy. The patient then underwent an observational laparoscopic procedure to diagnose the cystic mass.

For surgery, the patient was placed on her side with a 10-mm incision at her umbilicus so a small camera could be placed inside. Then, two more incisions were made to allow surgical tools to enter inside the patient. After the incisions were made, carbon dioxide pressure was used to distended the abdomen and provide the best working environment for the surgeon. The surgeon took only 89 minutes to remove the cystic mass as well as the right ureter, renal vein, and renal artery. The pathological conclusions revealed a MCDK with unusual glomeruli and tubules. The patient’s post-operative recovery was almost nonexistent and no further treatment was required. In this patient study, a laparoscopic nephrectomy was the best course of action due to the unknown cystic mass. Although further studies could have been conducted such as a US to evaluate and diagnose MCDK sooner, surgical intervention was still necessary due to the persistent vomiting.

**Conservative Case Report**

A 28-year-old woman that was 20 weeks pregnant came in for a routine US. After the fetus was examined, the radiologist noticed something unusual on the US and diagnosed the fetus with Down syndrome. However, he referred the mother to a hospital that specialized in pediatric care. At this facility, the mother underwent another US that revealed unilateral right-sided MCDK and not Down syndrome. The MCDK was greater than 6.2 mm in length and demonstrated thinning of the renal cortex. The left kidney presented normally on the US. After birth, the little girl was set on a conservative treatment plan. This included an annual US, blood work involving the kidneys, and a urinalysis. The patient was also required to monitor her blood pressure every 6-months. As the patient has grown, the MCDK has involuted normally and the
left kidney has displayed compensatory hypertrophy. There have been no signs of hypertension or malformation of the cysts. The patient is otherwise healthy as can be.

**Conclusion**

In conclusion, MCDK is a common renal abnormality that presents with multiple cysts within the kidney. Many imaging modalities are used to analyze and evaluate MCDK; however, US is the modality of choice due to the involution rate of the cystic kidney and because of the prenatal detection. Although there are only two types of treatment plans for MCDK, researchers are frequently debating which treatment plan provides a more reliable outcome for patients. Fortunately, both of these treatment plans have proven to be successful as demonstrated in the case reports above. The only deciding factor is what action will best adhere to the patient’s needs and wants. Although the etiology of this disease is unclear and there is no cure, this disease can be overcome. With the advancement in medicine, this disease can be fought and children each day can live normal lives without many complications.
References


Figure 1. Ultrasound images of multicystic dysplastic kidney and differentiation from severe hydronephrosis. A. Multicystic dysplastic kidney in a 1-day-old male. By ultrasound, MCDK appears as a mass of non-communicating cysts of variable sizes and with no recognizable pattern. B. Severe hydronephrosis secondary to ureteropelvic junction (UPJ) obstruction in a 2-year-old male. Hydronephrosis can present an ultrasound appearance similar to that of MCDK. However, in hydronephrosis, the largest cystic structure (the renal pelvis) lies in a central location and is surrounded by and communicates with dilated calices. Reprinted with permission from Hains DS, Bates CM, Ingraham S, Schwaderer AL. Management and etiology of the unilateral multicystic dysplastic kidney: a review. Pediatr Nephrol. 2009;24;233-241. doi:10.1007/s00467-008-0828-8.