https://nephropathol.com DOI: 10.34172/jnp.2023.21432 J Nephropathol. 2025;14(1):e21432

Journal of Nephropathology

An infant girl with bilateral multicystic dysplastic kidney: a case report

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ARTICLE INFO

Article type:
Case Report

Article history:
Received: 12 Jan. 2023
Accepted: 7 Mar. 2023
Published online: 10 Apr. 2023

Keywords:
Bilateral multicystic dysplastic kidney, Vesicoureteral reflux, Hypertension

ABSTRACT

Multicystic dysplastic kidney (MCDK) is a congenital kidney abnormality with an overall incidence rate of 1/4300 live births that is more prevalent in males than females. Here, we describe a rare case of bilateral MCDK complicated by hypertension, severe vesicoureteral reflux (VUR), and urinary tract infection (UTI), who stabilized during hospitalization with prospective kidney transplantation due to poor prognosis. Since bilateral MCDK is a rare disease with numerous complications and a poor prognosis, patient symptom management and treatment may be more challenging.

Case Report

Implication for health policy/practice/research/medical education:
The multicystic dysplastic kidney (MCDK) frequently results in death in infancy, our case report could be a new step toward managing complicated bilateral MCDK manifestations, especially for improved survival.

Please cite this paper as: Pourpashang P, Hosseini Tabatabaei SMT, Fallahi M, Mohajerzadeh L, Zahmatkesh A. An infant girl with bilateral multicystic dysplastic kidney: a case report. J Nephropathol. 2025;14(1):e21432. DOI: 10.34172/jnp.2023.21432.

Introduction

Multicystic dysplastic kidney (MCDK) is a congenital kidney abnormality with an overall incidence rate of 1/4300 live births that is more prevalent in males than females. It is the most common prenatal diagnosed cystic lesion of the kidney that occurs unilaterally or bilaterally and is associated with other structural ureteral anomalies, such as ureteropelvic junction obstruction (UPJO), vesicoureteral reflux (VUR), and ureterovesical obstruction (UVJO) (1). The failure of mesenchymal metanephros to differentiate into ureteral bud epithelial cells may be the cause of MCDK. The presence of numerous thin-walled cysts without connections within a kidney with an irregular outline is the classic MCDK ultrasound appearance. This appearance could be seen for the first time between 15 and 20 weeks of gestation (2). Most MCDKs are unilateral, more frequently affecting the left kidney, and have a favorable prognosis. Contrarily, bilateral MCDK is a rare condition with a poor prognosis. Infants with bilateral kidney disease frequently die during the neonatal period. External anomalies, anhydramnios, pulmonary hypoplasia, and chromosomal anomalies often are associated with bilateral MCDK (3). In a study on 21 bilateral MCDK cases, all patients terminate the pregnancy because of prognosis (2). In keeping with the lack of reports of bilateral MCDK that are still alive, we present in this study a 6-day-old girl infant who has bilateral MCDK.

Case Presentation

The only child of a mother with a gestational age of 36 weeks was referred us at the age of 6 days with a creatinine

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level of 6.5 mg/dL, anuria, and electrolyte abnormalities. The mother had mentioned the baby's kidney cysts during pregnancy screening, and there was a positive family history of kidney cysts in her aunt. She underwent an ultrasound at another hospital, where it revealed that her left kidney had dysplasia with numerous cysts, and her right kidney had a high calyx diameter (> 35 mm) and a few small cysts.

The patient's first symptoms in our center included edema, anuria, hypertension, and instability. She was initially diagnosed with UPJO in the right kidney and an MCDK in the left kidney. Because of her instability and electrolyte abnormalities, she underwent peritoneal dialysis. The laboratory results are shown in Table 1.

The patient had diuresis through a nephrostomy after receiving some dialysis and becoming stable. Then, she underwent pyeloplasty and a double J catheter for the right kidney, then her diuresis continued with a good flow. Initially, the glomerular filtration rate (GFR) was measured at about 5 mL/min. After these procedures, the GFR increased to 20 mL/min. The patient was in control in our hospital and the maintenance treatment prescribed for her, consequently overload issues were resolved. During hospitalization, she developed a urinary tract infection (UTI), which was treated with antibiotics. Subsequently, a voiding cystourethrography was performed, which revealed a grade 4 renal reflux (Figure 1).

Due to the recurrent urinary tract infections, a right urethrectomy was conducted. The infant's condition is now stable, she weighs 4 kg (up from 2 kg at birth), and no organ abnormalities are present. Supplements for chronic kidney disease and daily cephalexin are prescribed. She will become a candidate for a kidney transplant once she weighs 10 kg.

Discussion

Multicystic dysplastic kidney is one of the most common anomalies detected by prenatal ultrasound, and diagnosed earlier and earlier due to more accessible routine prenatal ultrasonography (US). According to a study, the prenatal diagnosis of MCDK is accurate in about 91.2% of cases, and the majority of earlier studies indicated that MCDK affects about 1:4300 newborns annually (4). Since bilateral MCDK frequently results in death, several studies have focused on unilateral MCDK instead. However, a significant population-based study conducted in Europe found that both unilateral and bilateral cases had a higher prevalence rate (5). Based on data from 64 included studies, MCDK is significantly more often found on the left side and has a male predominance (6). Additionally, MCDK has been reported with over 80 different syndromes and multiple congenital anomalies. We reported a girl with bilateral MCDK, in which the left kidney almost didn't function. A study found that girls with bilateral MCDK were twice as likely as boys to have non-renal associated anomalies and four times more likely to have chromosomal anomalies as boys, this could imply that male fetuses with severe malformations are less likely to survive (5). In cases of bilateral MCDK or associated anomalies, it makes sense to advise karyotype analysis. Moreover, MCKD is typically a sporadic anomaly rather than a familial one, then the formal screening of relatives is not advised (7).

The accurate prenatal assessment of MCDK and related malformation is crucial for determining prognosis because prenatally identified findings are linked to poor neonatal outcomes, particularly with contralateral renal abnormalities (8). Different variables were studied to find prognostic factors for MCDK. The complete involution of MCDK has been associated with positive outcomes. Gender, the size of the MCDK, and the size of the kidney on the opposite side had no predictive value (5). Our patient's GFR after pyeloplasty and double J catheter reached 20 mL/min and this problem shows that the prognosis of our patient is poor unless she undergoes a kidney transplant in the future.

Table 1. According to the laboratory results, the serum creatinine and serum uric acid levels are dramatically higher than they were during the first visits and hospitalization

Test	First	Follow-ups
Serum creatinine (mg/dL)	6.5	1.1
Urea (mg/dL)	53.6	25
Uric acid (mg/dL)	20	2.3
Calcium (mg/dL)	7.2	10.5
Phosphorus (mg/dL)	10	4.3
Magnesium (mEq/L)	1.9	2.3
Sodium (mEq/L)	110	138
Potassium (mEq/L)	6.9	4.6

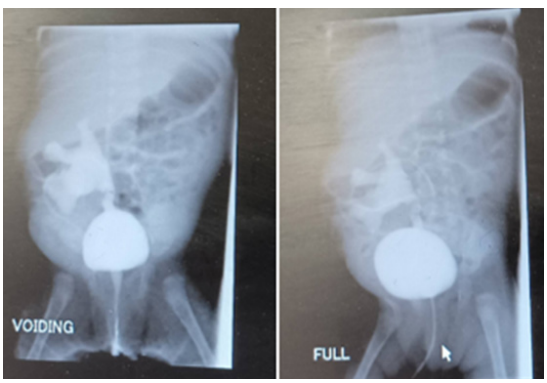

Figure 1. After catheterization urinary bladder is filled with contrast media, which showed bladder distensible with regular and normal wall thickness, no filling defect or outpouching is seen, no residue is seen after voiding, grade IV VUR is seen in right side.

MCDK can occur alone, but it frequently coexists with other kidney abnormalities, urological abnormalities of the urinary tract, such as VUR, obstruction at the ureterovesical junction (UVJ), and obstruction at the ureteropelvic junction (9). VUR is the most frequent coincident disorder, with a reported prevalence varying from 10% to 50% (10). According to some studies, between 26% and 50% of children with unilateral MCDK also had high-grade VUR (grade III, IV, or V) in the kidney on the opposite side, and a few of these patients needed surgery to treat their reflux (1). Despite cysts in the right kidney following voiding cystourethrography performance in a stable condition, our case had high severe reflux. Due to the low survival rate of bilateral MCKD, no studies have reported severe VUR.

The rate of hypertension in MCDK is not well-determined and has published rates of 0.6% to 17.7% (11). The presence of any accompanying urinary anomalies, such as renal dysplasia, UPJO, and pyelonephritic scar secondary to VUR in the contralateral kidney, has been identified as a potential cause of hypertension. Children with MCDK should undergo routine blood pressure monitoring to detect hypertension and should be treated accordingly (12). Another significant complication of MCDK was reported as UTI, which occurred twice while our patient was hospitalized and was treated with antibiotics. Urinary tract infection prevalence rates in MCDK ranged from 5% to 34% (7).

The contralateral kidney typically undergoes compensatory hypertrophy, which is estimated to range from 16.6% to 89.8% (9). It is believed that, if there is no hypertrophy, the practitioner should be suspicious of the contralateral kidney having an abnormality (13). In our case study, due to bilateral MCDK's poor prognosis, kidney transplantation should be considered.

Conclusion

The prevalence of MCDK is higher than previously believed, due to the advancements in prenatal diagnosis over the past two decades. Since bilateral MCDK is a rare disease with numerous complications and a poor prognosis, patient symptoms management and treatment may be more difficult.

Authors' contribution

Conceptualization: Paniz Pourpashang.
Data curation: Seyed Mohammad Taghi Hosseini Tabatabaei and Minoo Fallahi.
Formal analysis: Paniz Pourpashang and Seyed Mohammad Taghi Hosseini Tabatabaei.
Investigation: Paniz Pourpashang.
Methodology: Paniz Pourpashang, Seyed Mohammad Taghi Hosseini Tabatabaei.
Project administration: Paniz Pourpashang and Arefeh Zahmatkesh.
Resources: Paniz Pourpashang.
Supervision: Seyed Mohammad Taghi Hosseini Tabatabaei and Minoo Fallahi.
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Visualization: Seyed Mohammad Taghi Hosseini Tabatabaei and Minoo Fallahi.
Writing–original draft preparation: Paniz Pourpashang.
Writing–review and editing: Arefeh Zahmatkesh.

Conflicts of interest

The authors declare that they have no competing interests.

Ethical Issues

This case report was conducted in accord with the World Medical Association Declaration of Helsinki. Patient's parents have given us a written informed consent for publication as a case report. Ethical issues (including plagiarism, data fabrication, double publication) have been completely observed by the authors.

Funding/Support

The authors received no specific funding for this research.

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