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Case Report

# A Case of Unilateral Multicystic Dysplastic Kidney and Bilateral Vesicoureteral Reflux With Hyponatremia and Hyperkalemia

**Paniz Pourpashang[1], Arefeh Zahmatkesh[2*], Masoumeh Mohkam[1]**

*1. Pediatric Nephrology Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran.*
*2. Department of Pediatrics, School of Medicine, Shahid Beheshti University of Medical Sciences, Tehran, Iran.*

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**Corresponding Author:**
Arefeh Zahmatkesh
Address: Department of Pediatrics, School of Medicine, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
E-mail: arefeh.zhmtksh@gmail.com

## ABSTRACT

Multicystic dysplastic kidney (MCDK) is defined as a variant of renal dysplastic with multiple non-communicating cysts separated by dysplastic parenchyma. Contralateral vesicoureteral reflux (VUR) is the most frequent coincidence genitourinary anomaly in children with unilateral MCDK. Here, we described a case of unilateral MCDK (right side) with bilateral third-grade VUR who first, underwent conservative therapy because of electrolyte disturbance and metabolic acidosis.

**Keywords:** Multicystic dysplastic kidney (MCDK), vesicoureteral reflux (VUR), Electrolytes

## Introduction

Multicystic dysplastic kidney (MCDK) is a subtype of congenital anomalies of the kidney and urinary tract (CAKUT), which is the most common cause of abdominal mass in the neonatal period and is the most common cystic malformation of the kidney in infancy. The incidence of MCDK is 1 in 2000-4000 births and is usually sporadic. Although the cause of MCDK is unclear, it may be due to the failure of the union of the ureteric bud with the renal mesenchyme resulting in a non-functioning kidney that is replaced by non-communicating cysts of different sizes with no renal cortex and atretic ureter [1-5]. The condition occurs more commonly in males than females (2.4:1) and the left kidney is more affected than the right kidney. MCDK is mostly an isolated condition; however, the contralateral urinary tract could be affected by other abnormalities, such as vesicoureteral junction obstruction (UPJO) and ureterovesical obstruction (UVJO). Complications of MCDK include urinary tract infection (UTI), hypertension (HTN), and renal malignancies, which recently are managed primarily conservatively. MCDK is accepted to be a benign condition but early identification of associated abnormalities and appropri-

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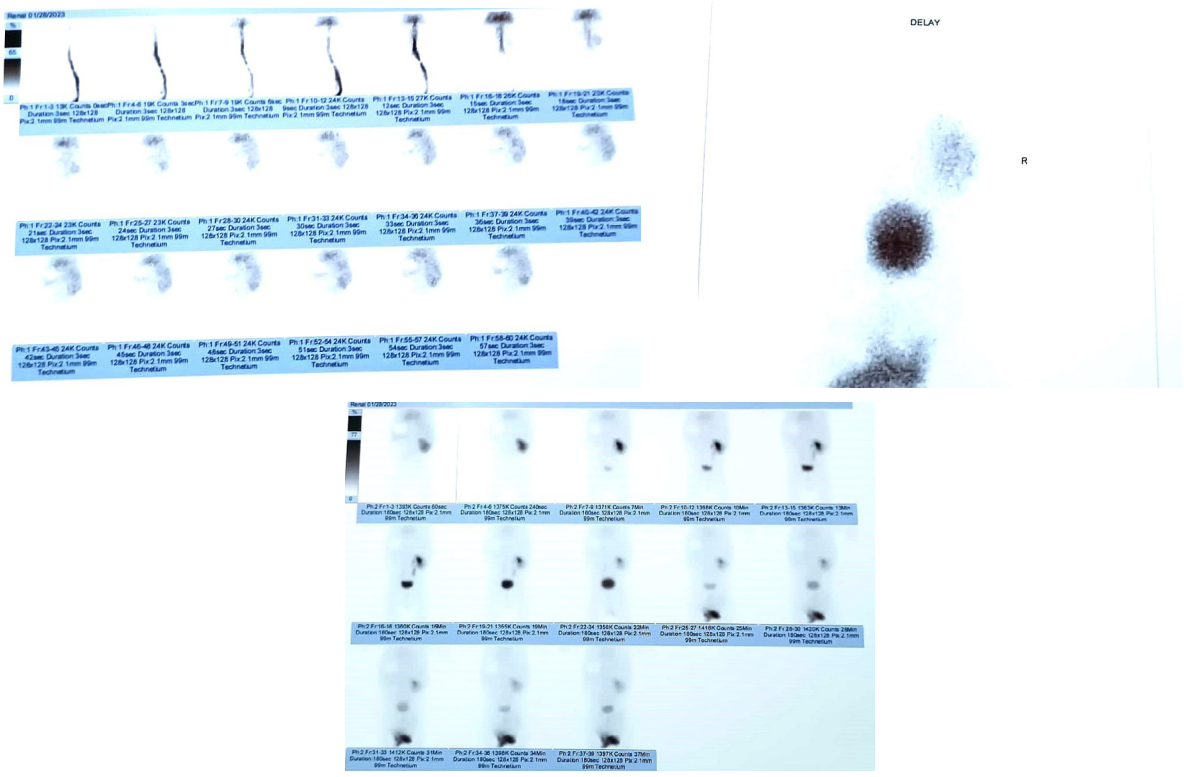


**Figure 1.** Diuretic DTPA renal scan

Right kidney showed mild decreased perfusion and excretory function. Prominent retention of activity was observed in the pyelo-caliceal system with fair response to the diuretic injection.

ate management of these patients are crucial in children due to having solitary kidneys. Here, we presented a unilateral MCKD with bilateral VUR (grade III), which is treated conservable.

## Case Presentation

A two-month-old boy with distant parents was admitted to the hospital with chief complaints of nausea, vomiting, poor feeding, fever, and diarrhea. He had not been circumcised before and had no urinary tract infections (UTIs). MCDK was diagnosed in the right kidney in another facility with multiple cysts after ultrasonography (US) was performed. A DTPA scan revealed a hydronephrotic right kidney with no obstruction as well as no discernible renal functioning tissues on the left side (Figure 1).

His initial laboratory results showed severe hyponatremia and hyperkalemia due to septicemia (Table 1), which were treated with appropriate liquid therapy

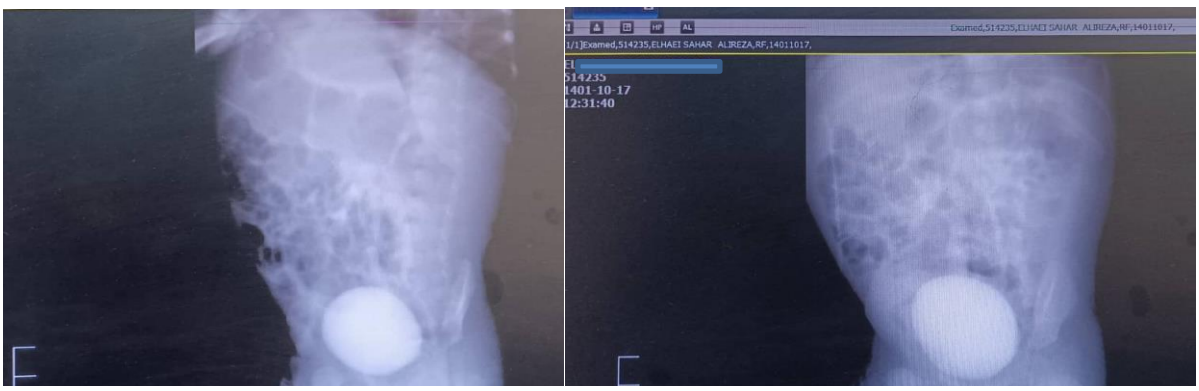


**Figure 2.** After catheterization, the urinary bladder is filled with contrast, and the medial bladder is distensible with regular and normal wall thickness

No filling defect or outpouching is seen. No residue is seen after voiding bilateral. Grade three right VUR was reported.

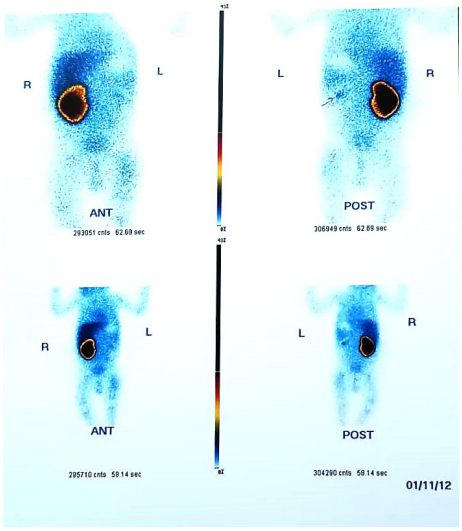


**Figure 3.** Right kidney showed rather uniform radiotracer uptake

A distinct cortical defect was not found. The renal contour was intact. A small focus of radiotracer was noticed in the left kidney.

and antibiotics. Positive blood culture and the patient's symptoms during hospitalization led to the diagnosis of septicemia. Due to septicemia and diarrhea, metabolic acidosis developed but was treated subsequently.

After the patient's stabilization, voiding cystourethrogram (VCUG) was done in our center due to hydronephrosis, and grade three right vesicoureteral reflux (VUR) was reported (Figure 2).

Also, according to an endocrinology consultation, hydrocortisone started and tapered off during hospitalization, and aldosterone and renin levels needed to be checked again because they were reported to be high (in a supine position). Although he had no adrenal insufficiency signs (e.g. genital hyperpigmentation), his 17-hydroxy progesterone levels were measured, which were in the normal range. Additionally, DSMA renal scan was performed, which reported a normal-looking right kidney and no appreciable functioning tissues on the left side (Figure 3).

**Table 1.** Laboratory test results of the patient

| | | | |
|---|---|---|---|
| **Electrolytes** | | | |
| Na (mEq/L) | | 119→131 | |
| K (mEq/L) | | 7→5.6 | |
| **Urea Biochemistry/Urine Random** | | | |
| Na (mEq/L) | | 105→55 | |
| K (mEq/L) | | 15.5→11 | |
| **Biochemistry** | | | |
| BS (mg/dL) | 100 | Mg (mg/dL) | 2.4 |
| Ca (mg/dL) | 10.2 | BUN (mg/dL) | 36.5 |
| P (mg/dL) | 5.5 | Cr (mg/dL) | 0.72 |
| Alkp (U/L) | 408 | Uric acid (mg/dL) | 2.9 |
| AST (U/L) | 25 | ESR (mm/hr) | 4 |
| ALT (U/L) | 16 | CRP (mg/L) | 3 |

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Finally, the patient was discharged with no symptoms. Kayexalate was prescribed due to hyperkalemia. Routine follow-up was recommended as a single kidney can cause several complications.

The follow-up laboratory results also showed hyponatremia and metabolic acidosis, which confirm renal tubular acidosis type IV, and medical treatment was considered.

## Discussion

MCDK has an incidence of 1 in 4300 fetuses with 94% of all cases being detected using ultrasonography. Several studies have revealed a predominance of MCDK in the left kidney and more in males than females, which is consistent with our study. If MCDK is unilateral, it may be associated with a contralateral kidney anomaly approximately 30%-40% of the time. A 15-year study on MCDK patients revealed that VUR was the most commonly associated abnormality and high-grade VUR was found in all patients with contralateral VUR. Some authors revealed the rate of the high-grade VUR (grades III, IV, and V) in the affected contralateral kidney varied from 26% to 50% and only a few patients required antireflux surgery. This issue has been increasingly questioned by many authors because VCUG is an invasive technique, and low-grade VUR is more common and is resolved early in life. Two successive normal renal US findings in infants clinically rule out the significant contralateral anomalies and thus, VCUG may not be necessary. A meta-analysis of children born with MCDK reported that contralateral kidney abnormalities, such as VUR were found in 19.7% of patients and most patients with significant VUR developed hydronephrosis. Although VUR is resolved spontaneously, VCUG should be performed in the presence of contralateral hydronephrosis or a history of UTI. Thus, using extra modalities was not necessary in this study [5-8].

Water, electrolyte, and acid-base disturbance are more common in infants and children with renal diseases, like MCDK, and can present with complex clinical pictures. Symptoms include dry skin and mucous membranes, elevated pulse, nausea, vomiting, diarrhea, decreased fluid intake, irritability, lethargy, weight loss, seizure, and comma. Our patient presented with electrolyte disturbance and metabolic acidosis due to acidosis and responded to accurate fluid and antibiotics therapy. Also, primary patients' symptoms, such as diarrhea, nausea, and vomiting resolved pursuantly. Not surprisingly, as a result of severe dehydration and septicemia, we observed that metabolic acidosis may be induced by type IV RTA (renal tubular acidosis).

The complications of MCDK include UTI, HTN, and renal malignancies. However, none of these complications existed in our case. Renin and aldosterone levels were reported high, which may be due to the plasma renin activity, and angiotensin raised in the decreased renal perfusion following unilateral MCDK. Also, for more investigation, aldosterone should be measured in both supine and upright positions. The management of MCDK has changed greatly over time. Previously, nephrectomy was performed to avoid infection, pain, HTN, and malignancy, but recently, management is primarily conservative due to favorable outcomes of patients. The prognosis of patients with MCDK is generally related to the function of the contralateral kidney, and abnormalities of the contralateral urinary tract and bladder. Although the risk of HTN is high in MCDK, it is not higher than in the general pediatric population, periodic follow-up visits every six months, and then annually, are prudent to prevent the development of HTN or hyperfiltration damage [9-13].

## Conclusion

The prevalence of MCDK is higher than previously believed thanks to advancements in prenatal diagnosis over the past two decades. Early and accurate diagnosis of MCDK and related abnormalities could help a practitioner to avoid using extra modalities and straight management.

## Ethical Considerations

### Compliance with ethical guidelines

Informed consent was obtained from the patient and his parents prior to being included in the study.

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### Authors' contributions

All authors contributed equally to the preparation of this article.

### Conflict of interest

The authors declared no conflict of interest.

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