

Pediatrics

## Extrarenal calyces as a cause of non-functional kidney in a child: Case report



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### 1. Introduction

Originally described by Eisendrath in 1925, extrarenal calyces (ERC) are a very rare upper tract anomaly, with approximately 50 cases reported in the world literature, including this one (20% < 17 years).<sup>1</sup> This malformation usually consists of a discoid kidney with the major calyces and the renal pelvis located outside the renal parenchyma, and it has been associated with other renal anomalies, such as ectopic kidney and renal dysplasia.<sup>1,2</sup> Because of this rareness, we present the case of a female patient with severe right hydronephrosis and extrarenal calyces. The radiologic studies didn't revealed the exact diagnosis, but it was an intraoperative finding.

### 2. Case presentation

A 6-year-old female, asymptomatic, with history of bilateral renal cysts and oligohydramnios diagnosed prenatally and initially diagnosed with right multi-cystic kidney disease (MCKD). She was initially evaluated by the nephrologist at our hospital for

hyperphosphatemia and severe right renal ectasia. Afterwards, the patient was sent to our pediatric urology department for evaluation.

Renal ultrasound reported a normal left kidney and right kidney (Fig. 1) in normal location, enlarged, smooth and with regular edges, with loss of the cortex-medulla ratio due to the existence of an anechoic image, with smooth regular borders, which did not show vascularity to the color Doppler sonography, neither the presence of septa. Right kidney's measures were 81 × 46 × 52 mm, without visualizing renal parenchyma.

In our diagnostic approach, the 99mTc-DTPA renal scan reported a normal dynamic phase for the left kidney. The sequential phase showed a normal left kidney. The right kidney was not visualized during the study. Renographic curves showed the left kidney with ascending functional slope, function of 100%, time of maximum uptake at 2.5 min, with descending excretion slope with output efficiency >50%. The glomerular filtration rate for the left kidney was 128.0 ml/minute and the right one was not reported.

The patient was diagnosed with right non-functional kidney and ipsilateral hydronephrosis. No other studies were considered necessary, so she was submitted to nephrectomy. In the surgical procedure we identified a right hydronephrotic pouch with a volume of 80 ml of clear urine, with 1 artery and 1 vein. The ureter was normally inserted into a renal pelvis with no anatomical anomalies like pyelectasis. As an important finding, 3 major calyces emerge from the renal pelvis with an extrarenal path of approximately 1.5 cm each before entering the kidney (Fig. 2). No ureteropelvic junction obstruction was identified or stenosis in the ureter.

### 3. Discussion

The variations of the collecting system represent a complex and often confusing subset of urological variations, which may hinder preoperative diagnosis.

The presence of ERC is very rare and its etiology is not very clear. It has been hypothesized that the anomaly could be due to a disparity resulting from the slow development of the metanephric

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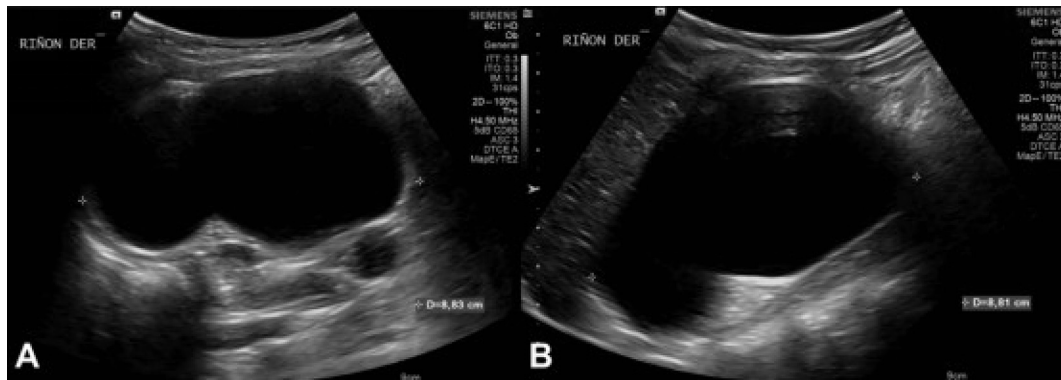


Fig. 1. Renal ultrasound. Both images from the right kidney showing hydronephrosis.

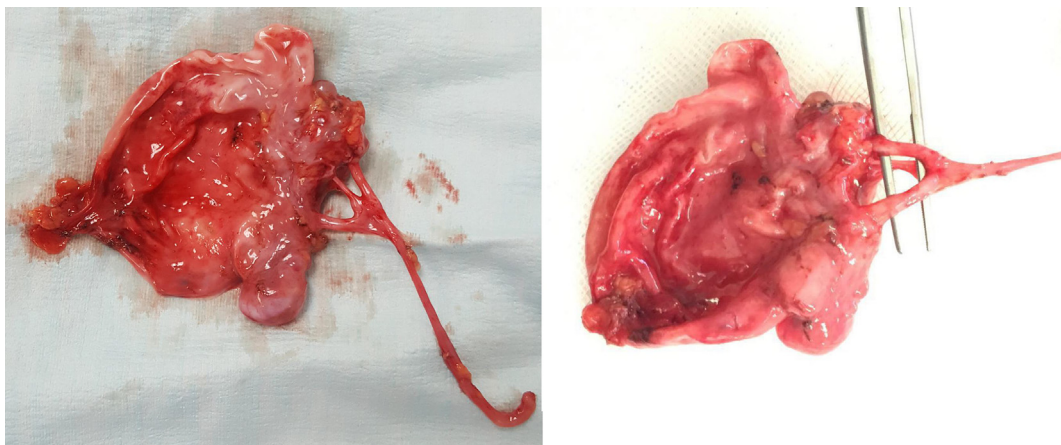


Fig. 2. Macroscopic piece of the non-functional kidney. There was no parenchyma macroscopically and pathology report didn't describe any "Right kidney measuring 6.0 × 4.0 cm, with a collapsed thin-wall and loss of cortex and medulla ratio." The hydronephrotic pouch was drained in order to do nephrectomy. Three major calyces can be observed joining the renal pelvis.

blastema or to a relatively rapid development of the ureteral bud.<sup>2</sup> If the latter has a rapid or early development, the calyx system may begin to divide before its coalescence with the metanephric blastema.<sup>3</sup> On the other hand, the delay in the development of the metanephric blastema could delay its adhesion to the collecting system allowing the extra renal development of the first or second order of the collecting system.<sup>1</sup>

In 1995, Kosinski and Oszukowski observed ERC in 3 kidneys of 300 autopsies, with an incidence of 1%.<sup>2</sup> To date, 49 cases of ERC have been reported in the world's literature. This abnormality has been associated with hydronephrosis, ureteropelvic junction obstruction, urinary tract infections, ectopic and horseshoe kidney and renal dysplasia.<sup>1,2,4,5</sup>

Since 1951, ERC has been reported to be twice as frequent on the left side.<sup>5</sup> In the review of the cases, 44.83% was reported on the left kidney vs 41.38% in the right one; 13.79% was bilateral. (In 40.81% of published data it was not possible to identify the affected side, because it wasn't mentioned).

Dretler reported that there appear to be two main configurations.<sup>1</sup> In one, there is a small renal pelvis that can be bifid. In the second, initially described by Malament as the radiographic appearance of a "small garden hand rake", there is no renal pelvis and the ureter is divided into 4 or 5 extrarenal branches.<sup>1</sup> However, the disposition varies and the renal pelvis can be composed of 2–5 ERC.<sup>1,2,5</sup>

Hydronephrosis and ureteropelvic junction obstruction were described in 24% of the cases. In the case that we present, a renal

pelvis and intact extrarenal calyces were observed, with severe hydronephrosis and no stenotic site. In no case reported to date, the association of extrarenal calyces with the dysfunction of the ipsilateral kidney has been described.

It is more common the unilateral ERC, but bilaterality has been reported (13.79%). They are usually asymptomatic, so they may remain undetectable for a long time or be diagnosed as autopsy findings.<sup>1,5</sup> In cases of patients with signs and symptoms such as pain, hematuria, pyuria, urinary tract infection with or without fever, the discomfort may be due to the complications arising from this abnormality such as urinary stasis, hydronephrosis, ureteropelvic junction obstruction and calculi.<sup>1–3</sup> Even dyspareunia has been associated to a female patient with extrarenal calyx and abdominal pain, that after nephrectomy she remained asymptomatic.<sup>1</sup> The most frequent symptom was the presence of lumbar or abdominal pain, located in iliac fossa, ipsilateral to the site of the abnormality.<sup>1–3</sup> It is frequent that the anomaly is not detected during the imaging studies and that they require surgery. It is at this point that the findings of the extrarenal calyces are reported.

#### 4. Conclusion

The present case report is a unique one, in which a hydronephrotic kidney drained into a pelvis with extrarenal calyces and no further obstruction, suggesting obstruction inside the collecting system. This may be explained as a malformation in the coalescence of the ureteral bud with the metanephric blastema. This anomaly

ended causing a non-functional kidney, which had never been reported. Exploration may be reserved for symptomatic patients and for those with obstruction and/or recurrent urinary tract infections.

### Consent

Because it is a retrospective review of a case, it does not require consent.

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