

METACHRONOUS PRESENTATION OF URETEROVESICAL JUNCTION OBSTRUCTION CONTRALATERAL TO A MULTICYSTIC DYSPLASTIC KIDNEY

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Multicystic dysplastic kidney is often associated with contralateral urinary tract anomalies (19% to 39% incidence).^{1,2} While obstructions at the level of the ureteropelvic junction or ureterovesical junction are uncommon, they are detected during the initial evaluation of the disease. To our knowledge we report the first case of a congenital intrinsic UVJ obstruction presenting metachronously during surveillance imaging of a multicystic dysplastic kidney.

CASE REPORT

Ultrasound of a 26-week-gestation fetus demonstrated a normal left kidney and findings consistent with a multicystic right kidney. Postnatal ultrasound on day 1 of life confirmed the multicystic nature of the right kidney, which measured $3.1 \times 1.5 \times 5.4$ cm., and a normal left kidney (fig. 1). Voiding cystourethrogram showed no reflux. Diethylenetetramine-pentaacetic acid renal scan at 1 month revealed no function in the right kidney thus, confirming the diagnosis of multicystic dysplastic kidney.

A surveillance imaging protocol was adopted to follow the involution of the right kidney. At 6 months the right kidney measured 1.8 cm. in greatest diameter and the left kidney was normal. At 9 months there was complete absence of the right kidney and new onset of marked left hydroureter and grade 1 hydronephrosis. An excretory urogram performed 2 months later demonstrated marked dilatation of the collecting system and ureter to the ureterovesical junction with prompt drainage of contrast material into the bladder. Similarly, the urine output had not changed.

Just prior to the next scheduled ultrasound, poor oral intake, lethargy and anuria developed. An emergent ultrasound

showed massive left hydroureteronephrosis. A percutaneous nephrostomy tube was placed and the patient was stabilized. A subsequent antegrade nephrostogram demonstrated no passage of contrast material beyond the ureterovesical junction (fig. 2). The patient then underwent tapered left ureteral reimplantation with resection of an adynamic segment of distal ureter. The final pathological evaluation of this distal segment demonstrated hypertrophic muscle proximal to the narrowed segment.

DISCUSSION

Obstruction early in renal development has been proposed as the etiology of multicystic dysplastic kidney. Concomitant obstruction in either the ipsilateral or contralateral urinary tract is not common in this disease, but in such reported cases the obstruction was discovered during the evaluation of the cystic renal lesion (ultimately the multicystic dysplastic kidney). Because of the vital need to preserve renal function in these cases, early discovery and intervention are necessary. Similarly, routine voiding cystourethrography to detect vesicoureteral reflux (18% to 43% incidence) is recommended in the initial evaluation of cases of multicystic dysplastic kidney so that prophylactic antibiotics can be started to prevent renal scarring and pyelonephritis.^{1,2}

To our knowledge this case is the first reported instance of metachronous presentation of a ureterovesical junction obstruction in a renal unit contralateral to an involuting multicystic dysplastic kidney. While ureterovesical junction obstruction of the contralateral kidney has been reported in only a few cases in the literature, these reports represent obstructions identified at the time of diagnosis of the multicystic dysplastic kidney and were treated accordingly.³ In our case the early detection of this lesion led to preservation of the renal unit when obstruction ultimately developed. Because of the crucial need to preserve renal function, we suggest periodic imaging of the remaining

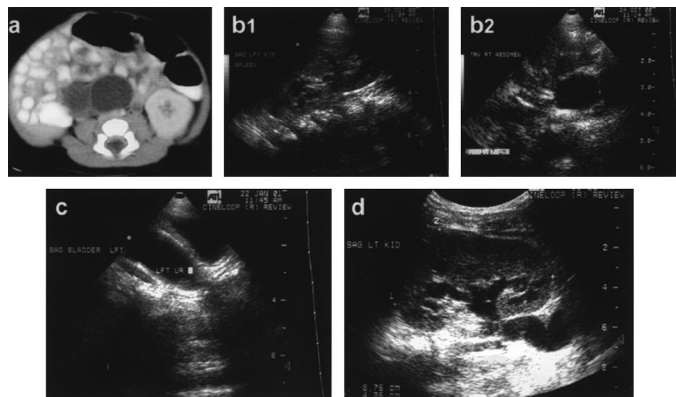


FIG. 1. *a*, computerized tomography with intravenous contrast performed on day 1 of life reveals normal left kidney and multiple cysts within right kidney. *b1* and *b2*, at age 6 months left kidney remains normal and right kidney shows progressive involution measuring 1.8 cm. in greatest diameter. *c*, involution is complete at age 9 months with new onset of marked left hydroureter. *d*, left side demonstrates hydroureteronephrosis and obstruction at age 1 year.

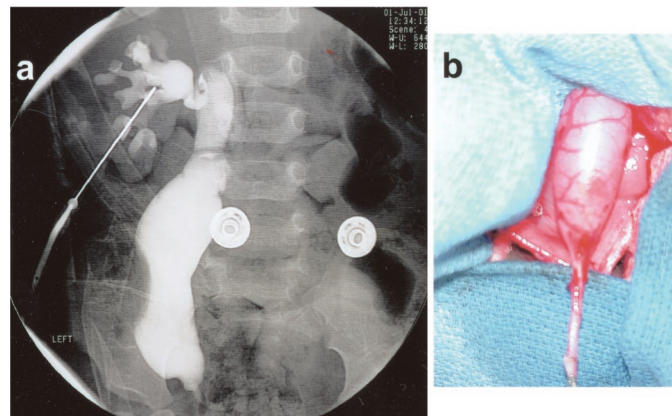


FIG. 2. *a*, antegrade nephrostogram performed several weeks after presentation with anuria reveals severe ureteral dilatation behind stenotic distal ureter. *b*, at surgery adynamic distal ureter is resected and reimplanted.

kidney, at the discretion of the clinician, even when there is evolving or clear involution of the multicystic dysplastic kidney.

REFERENCES

1. Atiyeh, B., Hussman, D. and Baum, M.: Contralateral renal abnormalities in multicystic-dysplastic kidney disease. *J Pediatr*, **121**: 65, 1992
2. Wacksman, J. and Phipps, L.: Report of the Multicystic Kidney Registry: preliminary findings. *J Urol*, **150**: 1870, 1993
3. Wu, C. Y., Chiou, Y. H. and Sung, P. K.: Left multicystic dysplastic kidney with contralateral ectopic kidney and ureterovesicle junction obstruction. *Zhonghua Yi Xue Za Zhi (Taipei)*, **64**: 129, 2001