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Single system ectopic ureters and ureteroceles associated with dysplastic kidney*

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Abstract. Eight children forming an uncommon subgroup of renal obstructive dysplasia are presented. Each child had a nonfunctioning dysplastic kidney with a single collecting system with ectopic ureteral insertion and/or ureterocele. Five of the children had classic multicystic dysplastic kidneys, one had the hydronephrotic type of multicystic dysplastic kidney and two had hypoplastic kidneys. Other significant medical problems in 5 of the 8 children (63%) included VACTERL association, congenital heart disease and other genitourinary malformations. Unlike some children with unilateral multicystic dysplastic kidney, this subgroup of children has an increased risk of infection. They must be correctly identified on imaging so that tailored clinical management decisions can be made and associated anomalies detected.

We describe eight children with renal obstructive dysplasia including typical multicystic dysplastic kidneys resulting from a distal, less common site of obstruction. In each of these patients there was a single collecting system from a unilateral obstructive dysplastic kidney where the obstruction was at the level of a ureterocele and/or ectopic ureter. These 8 patients with this combination form an unusual group. Though it is recognized that the renal morphology of obstructive renal dysplasia can result from obstruction at any level of the urinary tract, multicystic dysplastic kidney have been most frequently described with proximal ureter atresia [1–4].

Subjects and methods

These 8 children each have one obstructive dysplastic kidney with a single collecting system with ectopic ureteral insertion and/or ureterocele. There were 4 girls and 4 boys with 2 left and 6 right kidneys involved. Age at diagnosis ranged from 25 weeks gestation to 5-years-old. Renal ultrasound was performed on seven of the eight patients. Four of these 7 patients also had antenatal ultrasonography. Radionuclide renal scans on 4 patients confirmed a single functioning kidney. All children had voiding cystourethrography (VCUG). The diagnosis was confirmed at the time of nephrectomy in five children, by cystoscopy and percutaneous nephrostogram in addition to other imaging studies in one child and solely with imaging studies in the remaining 2 children.

Results

Five of the children had a kidney with the classic multicystic dysplastic kidney morphology (Fig. 1). One child had the hydronephrotic type of multicystic dysplastic kidney (Fig. 2). Two children had tiny hypoplastic kidneys (Fig. 3). The results are summarized in Table 1. Two ectopic insertions were into the seminal vesicle close to the prostatic urethra, the others were in the bladder.

In addition to the renal obstructive dysplasia in the involved kidney, 5 of the 8 patients (63%) had problems including contralateral renal disease. The 2 boys with ectopic ureteral insertions into the seminal vesicle had distortion and malformation of the seminal vesicle and vas deferens on the same side which required excision in one patient due to recurrent infection. One girl with VACTERL association had vesicoureteral reflux into her one functioning kidney and esophageal atresia and tracheoesophageal fistula at birth. One boy with VACTERL association and vesicoureteral reflux into the one functioning kidney had imperforate anus, tethered spinal cord, hypopituitarism and ventriculoseptal defect in the heart requiring surgery. There were 2 children (one boy, one girl) who were otherwise healthy and normal.

Discussion

This uncommon subgroup of renal obstructive dysplasia is defined by the level and type of obstruction of the urinary tract. These cases are unusual since they have single collecting system kidneys as proved in the majority of cases at surgery. Whenever a ureterocele is identified a duplicated

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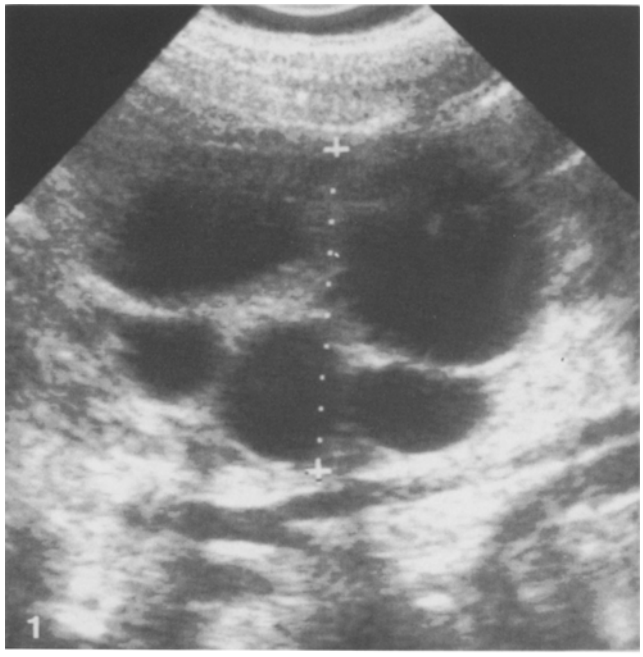

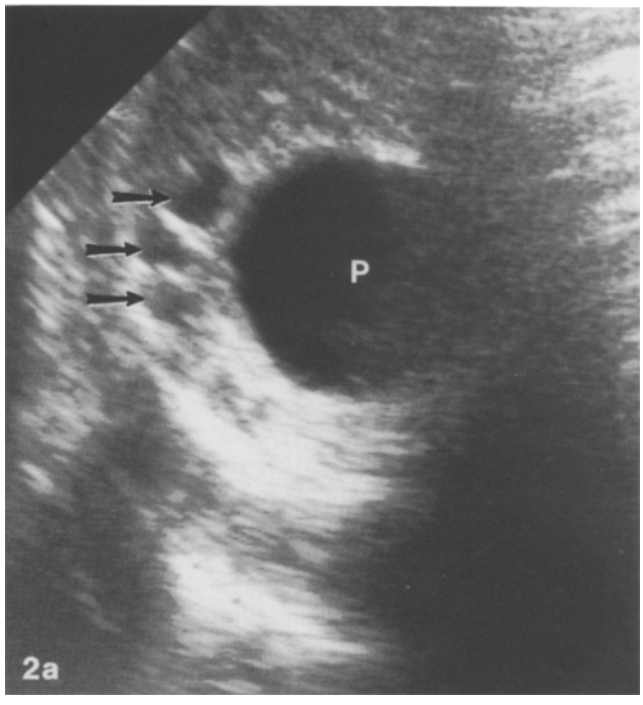

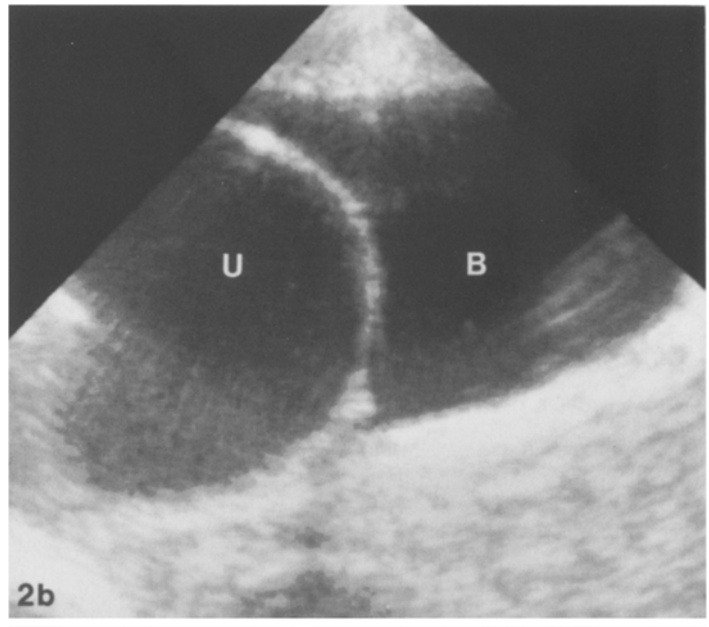

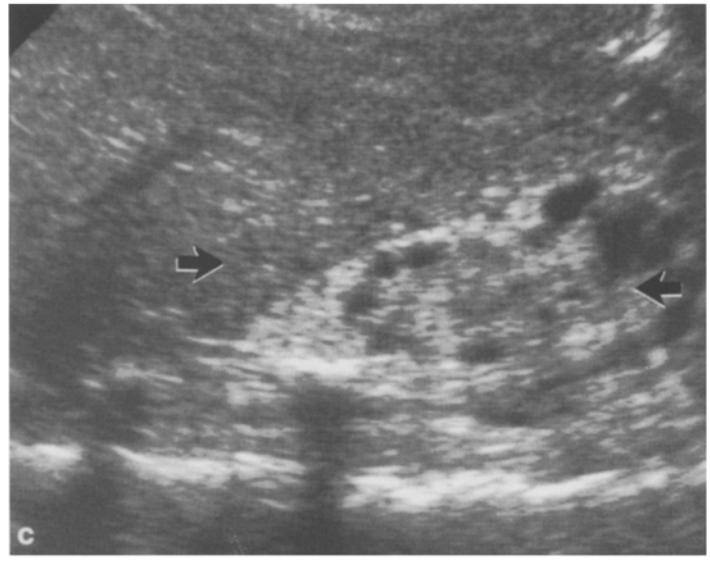

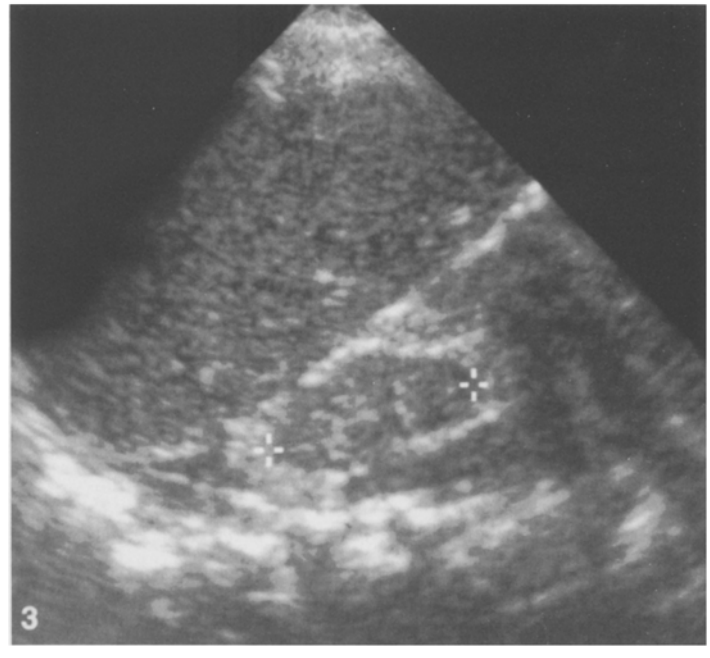

Fig. 1. A coronal ultrasound of the left upper quadrant in this 4-day-old girl reveals a characteristic appearance of multicystic dysplastic kidney

Fig. 2. **a** Transverse ultrasound of the right upper quadrant in this 1-day-old boy reveals a large central fluid collection in the pelvis *(P)* with peripheral smaller fluid collections, cysts (→)
b Longitudinal ultrasound through the bladder *(B)* reveals the ureterocele *(U)* at six days of age
c Longitudinal ultrasound through the right upper quadrant after drainage of the ureterocele reveals the dysplastic kidney (between →) with tiny cysts. The dilated renal pelvis is no longer identified

Fig. 3. Longitudinal ultrasound in the right upper quadrant reveals a tiny dysplastic kidney delineated by the markers in this young girl

Table 1. Patient data

Sex	Kidney	Age at presentation	Presenting problem	Obstruction	Imaging	Other problems	GU problems other than index kidney	Diagnosis
Girl	Rt Classic MCDK	6 wks	Abdominal mass	Ectopic ureterocele bladder	US VCUG DTPA CT	None	None	Nephrectomy
Boy	Rt Classic MCDK	25 wks Gestation	Routine US R/O sepsis birth	Ectopic ureter to Sem Ves	US VCUG IVP DTPA	None	Abnormal Sem Ves	Nephrectomy
Boy	RT Hydro MCDK	Birth	Sepsis	Orthotopic ureterocele	US VCUG DTPA	VACTERL association	Lt vesicoureteral reflux	Nephrectomy
Girl	Lt Classic MCDK	33 wks Gestation	Other anomalies	Ectopic ureterocele bladder	US VCUG	VACTERL association	RT vesicoureteral reflux	Cystoscopy
Girl	Rt Hypoplastic	7 wks	Fever	Ectopic ureterocele bladder	US VCUG IVP	CHD	Lt vesicoureteral reflux	Imaging
Boy	Lt Hypoplastic	5 years	Enuresis	Ectopic ureter to Sem Ves	VCUG IVP	None	Neuro blad Rt hydro Abn Sem Ves	Nephrectomy
Girl	Rt Classic MCDK	28 wks Gestation	Routine US	Orthotopic ureterocele	US VCUG DTPA	none	None	Nephrectomy
Boy	Rt Classic MCDK	27 wks Gestation	Routine US	Ectopic ureterocele bladder	US VCUG	Premature 27 wks gestation	None	Imaging

Abbreviations used: Rt, right; Lt, left; MCDK, multicystic dysplastic kidney; Hydro, hydronephrosis; US, ultrasound; Sem Ves, seminal vesicle; VCUG, voiding cystourethrogram; DTPA, renal nuclear scan; CHD, congenital heart disease; Neuro Blad, neurogenic bladder; Abn, abnormal

collecting system should be suspected. Occasionally it is difficult to demonstrate the upper pole of a duplicated collecting system, so that the lower pole is inadvertently labelled as a single collecting system. This is particularly true when one of the segments is poorly functioning or not functioning [5].

Animal experiments have demonstrated that obstruction early in utero can result in renal obstructive dysplasia [1, 2]. There is not similar documented experimentation to support the alternative theory that renal dysplasia is secondary to lateral ectopy of the ureter resulting in malalignment of the ureteral bud and metanephric blastema [6]. It would seem more likely that the degree of in utero obstruction in these eight cases and the gestational timing of the obstruction are responsible for the renal dysplasia. These cases demonstrate the variable morphology resulting in the kidney from early in utero obstruction. The kidney can have the classic imaging appearance of multicystic dysplastic kidney but without the usual proximal ureteral atresia. Large and/or small cysts or no cysts, dilated ureters and upper collecting systems or no identifiable collecting system can be seen.

Single system ectopic ureters and ureteroceles are not common. In general, ureteral ectopia with or without ureterocele, is more frequent in duplicated collecting systems in girls [7–9]. Single system ectopic ureters and ureteroceles though rare certainly account for a higher percentage of the boys with ureteroceles than the girls with ureteroceles [8, 10–12]. If we combine our cases to those in the literature, a total of 21 of 36 reported cases of single ectopic systems and ureteroceles are in boys (58%). There is certainly not the predominance of females seen in duplicated collecting systems and ectopia.

There is a report that boys with single system ectopic ureters and ureteroceles have an association with congenital heart disease and other genital abnormalities [11]. Others have indicated the association is not limited to boys [10]. Three of these 8 children, 2 boys and one girl in our group, had no other significant problem. The remaining 5 (63%), the majority, had other significant medical problems. This uncommon subgroup of renal obstructive dysplasia appears to have a high association with other anomalies including VACTERL association, congenital heart disease and genitourinary abnormalities. Whether or not this association is higher than that currently seen when renal obstructive dysplasia is identified on sonography in the general population of prenatal and childhood ultrasound studies is unknown, though it is certainly much higher than that reported in previous decades. In a review of 44 cases of unilateral multicystic dysplastic kidney published in 1975, only 4 (9%) had problems outside the urinary tract and 9 children (20%) had contralateral renal disease [3]. With the advent of almost routine antenatal ultrasound and improved neonatal survival, the population of patients that we are consulted on with renal obstructive dysplasia is changing. This change in age distribution of the recognized cases of renal obstructive dysplasia is also reflected in the increased incidence of associated anomalies both within the urinary tract and in other systems [13]. We are now consulted on cases that in the past may have been stillborn or died soon after birth, and in other asymptomatic cases that may have remained unrecognized during life.

Management of unilateral renal obstructive dysplasia, including multicystic dysplastic kidney, is controversial [14–16]. In the majority of patients with renal obstructive dysplasia that survive the neonatal period, the level of obstruction is at the junction of the renal pelvis and proximal ureter. The risk of hypertension and malignant degeneration has been shown to be very small in this group, and many physicians, both pediatricians and surgeons, believe routine nephrectomy is no longer appropriate [14–16].

Single system ectopic ureters and ureteroceles develop complications more often than the similarly anomalous duplicated systems [7, 8] or the asymptomatic obstructive dysplastic kidney [14–16]. The reported complications are bladder outlet obstruction related to the size and position of the ureterocele and ectopic insertion and infection [7, 8]. Thus in addition to the acknowledged low incidence of problems in the obstructive dysplastic kidney (hypertension and malignant degeneration), this subgroup of patients with ectopic ureteric insertions and ureteroceles do have the potential for additional problems [7, 8]. Management of this subgroup of patients must take these additional risks and potential complications into consideration. Thus these particular patients with renal obstructive dysplasia, independent of the morphologic appearance of a multicystic dysplastic kidney or hypoplastic kidney, must be recognized on imaging as comprising a different uncommon subgroup. Simple identification of a cystic dysplastic or hypoplastic kidney on ultrasound and non-function on a renal scan does not sufficiently document the anatomy of renal obstructive dysplasia. Recognition of the distal ureteral anomaly is important to indicate that surgical removal of these dysplastic units is necessary. Careful ultrasonography of the pelvis with a full bladder and VCUG should lead to the correct diagnosis in most cases. Since more than half of our patients in this unusual subgroup had serious other congenital problems physicians, particularly those involved in antenatal sonography, must include evaluation of other organ systems to look for associated anomalies.

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