Complete Duplication of the Ureter with Ureteropelvic Junction Obstruction of the Lower Pole of the Kidney: Imaging Findings

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OBJECTIVE. The purpose of our study was to identify the radiographic signs that aid in the diagnosis of obstruction of the ureteropelvic junction of the lower pole (or moiety) of the kidney in children with complete duplication of the ureter and to describe the imaging appearance of this unusual cause of lower-pole hydronephrosis.

MATERIALS AND METHODS. We reviewed the medical records and imaging studies of 16 children (11 boys and five girls) with complete ureteral duplication and ureteropelvic junction obstruction of the lower pole of the kidney over a 5-year period. Standard criteria for determining urinary tract obstruction were used.

RESULTS. Sonograms showed a lower-pole abnormality (hydronephrosis or cystic mass) in all 15 children who underwent sonography. Voiding cystourethrography, performed for all children, showed vesicoureteral reflux into the lower pole in addition to ureteropelvic junction obstruction in eight children (seven boys and one girl). For the other eight, the diagnosis of lower-pole ureteropelvic junction obstruction was made by excretory urography, at times complemented with diuretic renography or retrograde ureterography.

CONCLUSION. Ureteropelvic junction obstruction of the lower pole of the kidney in children with complete duplication of the ureter should be a diagnostic consideration when there is dilatation of the lower moiety. Imaging changes parallel those of ureteropelvic junction obstruction in a nonduplicated system. This anomaly, unlike others seen in duplication, appears to be more common in boys than in girls.

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Complete duplication of the ureter, the most common anomaly of the urinary tract, may be asymptomatic or recognized when complications develop as a result of reflux into the lower-pole ureter, obstruction of the upper pole with an ectopic ureterocele or a narrowed distal ureter with an ectopic orifice or, in females, dribbling of urine because of infrasphincteric insertion of the upper-pole ureter [1, 2].

Ureteropelvic junction obstruction has been reported to affect the lower pole in incompletely duplicated ureters [3–7].This condition also may occur in association with completely duplicated ureters and may be overlooked when reflux coexists or when the degree of dilatation produced is smaller than or similar to that produced by an obstructed upper pole. However, no one has reported the imaging findings for a series of patients with complete ureteral duplication with ureteropelvic junction obstruction of the lower pole.

Because surgery may be required to improve drainage and eliminate symptoms, it is important to recognize this anomaly. Familiarity with the imaging changes described in this paper may enhance detection of an obstructed lower pole.

Materials and Methods

Complete ureteral duplication and obstruction of the lower-pole renal pelvis were detected in 16 children (11 boys and five girls) seen at two pediatric hospitals over the last 5 years. At the time of diagnostic evaluation, their ages ranged from neonate to 9 years. All radiographic studies were reviewed to determine whether specific findings existed and whether an imaging

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protocol could be developed. The medical records were reviewed for information about clinical presentation, treatment, and surgical and pathologic findings, when available.

On voiding cystourethrography, coexisting ureteropelvic junction obstruction was diagnosed (1) when refluxed contrast material had difficulty entering the dilated renal pelvis, (2) when it was diluted, upon finally entering the renal pelvis, by the nonopaque urine trapped there, and (3) when it was shown to be trapped in the renal pelvis on delayed radiographs (Figs. 1 and 2).

Signs of ureteropelvic junction obstruction on excretory urography included visualization of caliceal crescents, prolonged nephrogram, or delayed excretion of contrast material. Secondary changes caused by the mass effect of the lower pole were (1) superior displacement of the upper-pole calices and obstruction and (2) medial displacement of the upper-pole ureter (Figs. 1 and 3).

Diuretic renography was performed with ^{99m}Tc-mercaptoacetyltriglycine. Standard criteria for obstruction were used [8, 9].

Sonography was not used to diagnose obstruction but rather to detect hydronephrosis or other abnormalities.

Ureteropelvic junction obstruction was diagnosed when any one test had multiple changes attributable to this condition; alternatively, when one study was suggestive of obstruction of the renal pelvis, another examination confirmed the findings. However, only one child underwent two examinations; the others underwent three or more before treatment.

For nine children, prenatal sonography had detected a renal abnormality. The other seven children were referred for radiologic studies because of urinary tract infections.

Two children had bilateral lower-pole ureteropelvic junction obstruction; one of these two children had bilateral complete duplication (Fig. 1), and the other had contralateral incomplete duplication. The other 14 children had unilateral lower-pole obstruction. Thus, a total of 17 obstructed lower poles in completely duplicated systems were examined. Eleven of the 17 affected lower poles were on the left.

Two children had contralateral ureteropelvic junction obstruction of a single renal pelvis (Fig. 3). The other children had no contralateral abnormality.

The ipsilateral upper pole was normal in all but two children. One child had an ectopic ureterocele (Fig. 1), and the other had obstruction of the upper-pole ureter by the dilated pelvis of the lower pole. One child had coexisting ureterovesical junction obstruction of the affected lower pole.

Treatment varied and depended on the degree of obstruction and the degree of function in the lower pole, whether another site of obstruction was present, and whether vesicoureteral reflux coexisted. Lower-pole pyeloplasty was done for 10 kidneys. Three kidneys are being monitored clinically and radiologically. Two lower poles have been treated with lower to upper pyeloureterostomy; in one of these two cases, coexisting reflux and distal ureteral obstruction were present. Two poorly functioning renal segments have been excised; one was an obstructed upper pole, and the other was a lower pole with vesicoureteral reflux.

Results

Sonography was performed for 15 children and demonstrated dilatation of the pelvicaliceal system of the lower pole in each. In some children, this condition had the appearance of typical hydronephrosis, and in others, the dilated system simulated a large cyst (Fig. 1). This finding was nonspecific.

Voiding cystourethrography was performed for all children. In seven boys and one girl, the lower-pole ureteropelvic junction obstruction was complicated by ipsilateral lower-pole vesicoureteral reflux (Figs.1 and 2). In these eight children, the refluxed contrast material showed that the duplication was complete and additionally revealed the obstructive nature of the renal pelvis.

Excretory urography demonstrated changes of lower pole obstruction in all 11 children for whom it was performed (Figs. 1 and 3). Diuretic renography, performed for five children, was a

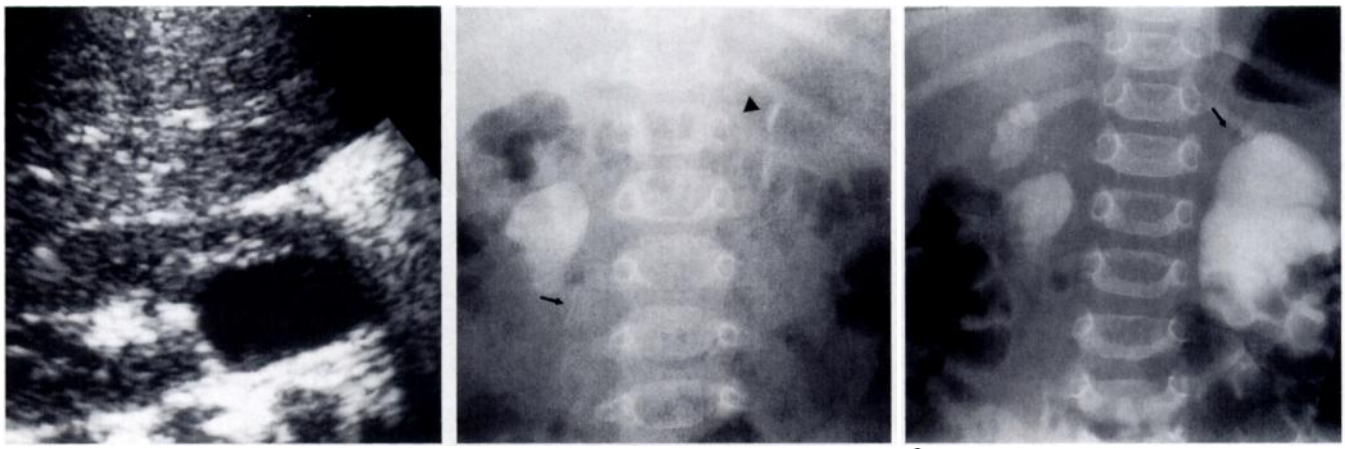

Fig. 1.—14-month-old boy with bilateral duplication and lower-pole ureteropelvic junction obstruction.

***A*, Coronal renal sonogram shows that upper pole appears normal. Obstructed lower-pole pelvis appears as echolucent cystic structure.**

***B*, Radiograph made 10 min after conclusion of voiding cystourethrogram shows that refluxed contrast material in right lower pole has not drained because of obstruction of right ureteropelvic junction. In addition, right renal pelvis and calices are disproportionately larger than adjacent ureter (*arrow*). Contrast material also is present in small left upper pole (*arrowhead*) because ureterovesical junction was made incompetent when distal ureterocele was incised to permit drainage of upper pole. Obstructed left lower pole is not seen because no reflux into this segment occurred.**

***C*, Because the child had known reflux, bladder was catheterized and left open to drainage during excretory urography. Contrast material is present in all four moieties. Left lower pole is grossly dilated, and right lower pole is less so. Air seen in left upper pole (*arrow*) is attributable to reflux despite bladder catheterization.**

Fig. 2.—8-year-old boy with vesicoureteral reflux into obstructed left lower-pole pelvis. Voiding cystourethrogram shows contrast material in left ureter forming discrete jet (*arrow*) as it enters dilated renal pelvis.

valuable adjunct to excretory urography (Fig. 3) rather than an alternative imaging technique for three children and was used for preoperative diagnosis for two children. Retrograde ureterograms were obtained in five children, all at one hospital [10], to exclude obstruction at a site other than the ureteropelvic junction or obstruction of unusual etiology, such as ureteral valves or polyps.

Discussion

Lower-pole ureteropelvic junction obstruction was suspected in eight of the 16 children tested because of specific changes seen when coexisting lower-pole reflux occurred on a voiding cystourethrogram. For the other eight children, more than one radiographic examination was necessary before the diagnosis could be made. Familiarity with the radiologic and clinical findings of both ureteropelvic junction obstruction and duplication was valuable in making the diagnosis.

Children with lower-pole ureteropelvic junction obstruction may display the same problems as those with ureteropelvic junction obstruction of a single collecting system: flank mass, urinary tract infection, or hematuria. In this clinical setting, sonography usually is the first examination performed to evaluate the kidneys and bladder.

The radiologic findings of a dilated lower-pole pelvicaliceal system are not unique to ureteropelvic junction obstruction. The most common complication of duplication, vesicoureteral reflux into the lower pole, is a more likely cause [1]. Simple reflux can be differentiated readily from reflux coexisting with ureteropelvic junction obstruction by use of the criteria described in the Materials and Methods section. Simultaneous dilatation of the upper and lower poles may occur if an upper-pole ureterocele occludes the ureteral orifice of the lower pole or if a dilated upper-pole ureter causes extrinsic compression and obstruction of the lower-pole ureter where they cross. Thus, in children with ureteral duplication, the bladder should be examined carefully for a ureterocele. In the first setting, both the upper- and the lower-pole ureters would be dilated. In the second, only the ureter entering the ureterocele would be dilated, and careful sonographic technique might show dilatation of a small proximal segment of the lower-pole ureter. An upper-pole ureter also may extrinsically compress a lower-pole ureter if it becomes dilated from distal intrinsic narrowing. This condition may produce findings that may be difficult to understand: dilatation of both renal pelves and visualization of one ureter in the retrovesical region.

Voiding cystourethrography is commonly performed for infants with prenatally diagnosed hydronephrosis (as in nine of the children), as reflux is responsible for 10–30% of such cases of hydronephrosis and also may occur in conjunction with obstructive lesions, such as ureteropelvic junction obstruction and primary megaureter [11–15]. Reflux may simultaneously clarify whether the duplication is complete or incomplete. Because the surgical options (pyeloplasty or pyeloureterostomy) are often the same, it may not be necessary to perform additional imaging. Visualization of the ureter via reflux also may allow detection of any additional obstruc-

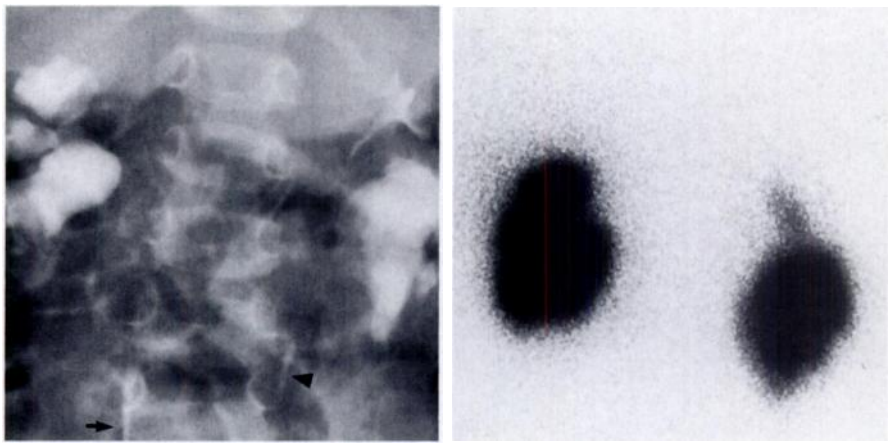

Fig. 3.—2-year-old girl with lower-pole ureteropelvic junction obstruction and contralateral ureteropelvic junction obstruction. Since no reflux was demonstrated in this child, it was impossible to differentiate complete from incomplete duplication before cystoscopy.

***A*, Excretory urogram shows that right renal pelvis and calices are dilated. Small segment of nondilated right ureter is identified (*arrow*). Left kidney has dilated lower-pole pelvicaliceal system. Small segment of left upper-pole ureter is visible (*arrowhead*).**

***B*, On this diuretic renogram, isotope has readily washed out of left upper pole but has remained in obstructed left lower pole and right renal pelvis.**

tion, as for the child in our study who had a simultaneous ureterovesical junction obstruction, a lesion previously described for the ureter of the lower moiety [16].

The frequency of ureteropelvic junction obstruction of the lower pole is uncertain. We have no data base of hospital admissions or surgery for ureteral duplications; however, in an earlier review of 100 children with complications of ureteral duplication (usually complete), not one case of this anomaly was reported [2]. Similarly, in our experience, lower-pole ureteropelvic junction obstruction is much less frequent than is reflux, ectopic ureterocele, or ectopic ureter. We believe that ureteropelvic junction obstruction of the lower pole has been overlooked because of its relative infrequency. We also believe that because of the way that radiologic services are delivered in our two institutions, we have had a unique chance to make these observations and collect these data.

It is interesting that 11 of the 16 children tested were boys, as most complications of ureteral duplication have a pronounced female predominance [2]. For example, ectopic ureters are seen in six to eight times as many girls as boys. The unusual distribution that we observed may have resulted from our small sample or may have been a manifestation of the facts that the lower pole behaves as a single system and ureteropelvic junction obstruction of single collecting systems is more common in boys than in girls. However, vesicoureteral reflux was present in half of the lower poles, a pattern quite typical of duplicated systems and not of single systems, which are reported to have coexisting ureteropelvic junction obstruction and reflux in only 10% of patients [12].

What is the best imaging method for children with ureteropelvic junction obstruction of the lower pole? Many children being studied for other, more common abnormalities already will have undergone sonography and voiding cystourethrography before the diagnosis of lower-pole ureteropelvic junction obstruction is considered. These two studies together may be diagnostic, especially if vesicoureteral reflux occurs and a delayed film is obtained. In the absence of reflux, excretory urography or renal scintigraphy can be used to obtain anatomic or physiologic evaluation of the lower pole. Retrograde ureteral studies should be used only for the few children whose anatomy has remained unclear.

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