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Urologic Radiology

Congenital Midureteral Obstruction

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Abstract. Congenital midureteral obstruction, caused either by a ureteral valve or stricture, is an exceedingly rare entity. When encountered, it is generally misdiagnosed as either primary megaureter or ureteropelvic junction obstruction, leading to a less than optimal surgical approach. Seven children with midureteral obstruction were seen over the past 17 years. Two of the patients presented with hydronephrosis on prenatal ultrasonography, and five with urinary tract infection. In only three was the diagnosis suspected on intravenous urography and voiding cystourethrography alone. Five of the seven cases had either antegrade or retrograde pyelography, prior to or at the time of their operative procedure, and in four of these a correct diagnosis was made. Notably, in five of the seven cases, recognition of the midureteral lesion prior to the surgical incision allowed the approach to be modified and a more appropriate technique (ureteroureterostomy) to be performed. The etiology of midureteral valve and stricture has been attributed to improper recanalization, insufficient vascular supply, or persistence of ureteral folds. Three of our patients had contralateral renal dysgenesis, suggesting an underlying ureteral bud abnormality.

Key words: Ureter, stenosis or obstruction — Hydronephrosis — Genitourinary system, infants, newborn.

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Congenital midureteral obstruction on the basis of either valve or stricture is extremely rare. Children with this disorder often are misdiagnosed as having either ureteropelvic junction obstruction or primary megaureter. An awareness of this entity, combined with a systematic radiologic evaluation of children with hydronephrosis, will allow the correct preoperative diagnosis to be made, and thus a more appropriate surgical management. Over the past 17 years, seven cases of midureteral obstruction have been seen by the authors. The clinical findings, evaluation, and treatment of these children are listed in Table 1. Representative cases are presented below.

Materials and Methods

The records of the Division of Urology and Department of Radiology at the Children's Hospital were examined looking for all instances of congenital midureteral obstruction. Medical records and radiographs were reviewed in each case to confirm the existence of the lesions. Histopathologic slides were evaluated as available. In addition, cases from the personal experience of the urologists, while at other institutions, were reviewed in a similar manner.

Results

Since 1970, 7 cases of congenital midureteral obstruction have been treated by the authors (Table 1). The children ranged in age from 5 weeks to 12 years. Two were newborns who came for evaluation as a result of hydronephrosis on prenatal ultrasonography; the remainder presented with urinary tract infection. All underwent radiologic evaluation including intravenous urography (Figs. 1–4), voiding cystourethrography and, in all but two, retrograde

Table 1. Clinical findings, evaluation, and treatment

Patient	Age	Sex	Presentation	Side	Diagnosis made by	Pathologic diagnosis	Treatment
D.R.	5 wks	M	Hydronephrosis on prenatal ultrasound	Right	Intravenous urogram	Congenital stricture	Ureteroureterostomy
A.D.	5 wks	M	Hydronephrosis on prenatal ultrasound	Right	Retrograde ureteropyelogram	Congenital stricture Contralateral dysplasia	Ureteroureterostomy
S.L.	2½ yr	F	UTI	Right	Retrograde ureteropyelogram	Congenital stricture Contralateral dysplasia	Ureteroureterostomy
D.S.	3 mo	M	UTI	Left	Surgical exploration	Congenital stricture	Ureteral reimplant Psoas hitch
D.C.	12 yr	F	UTI	Right	Intravenous urogram	Ureteral valve	Ureteroureterostomy
K.P.	5 yr	M	UTI	Right	Intravenous urogram	Congenital stricture	Ureteroureterostomy
G.S.	6 yr	M	UTI	Left	Surgical exploration	Ureteral valve Contralateral dysplasia	Ureteral reimplant Psoas hitch

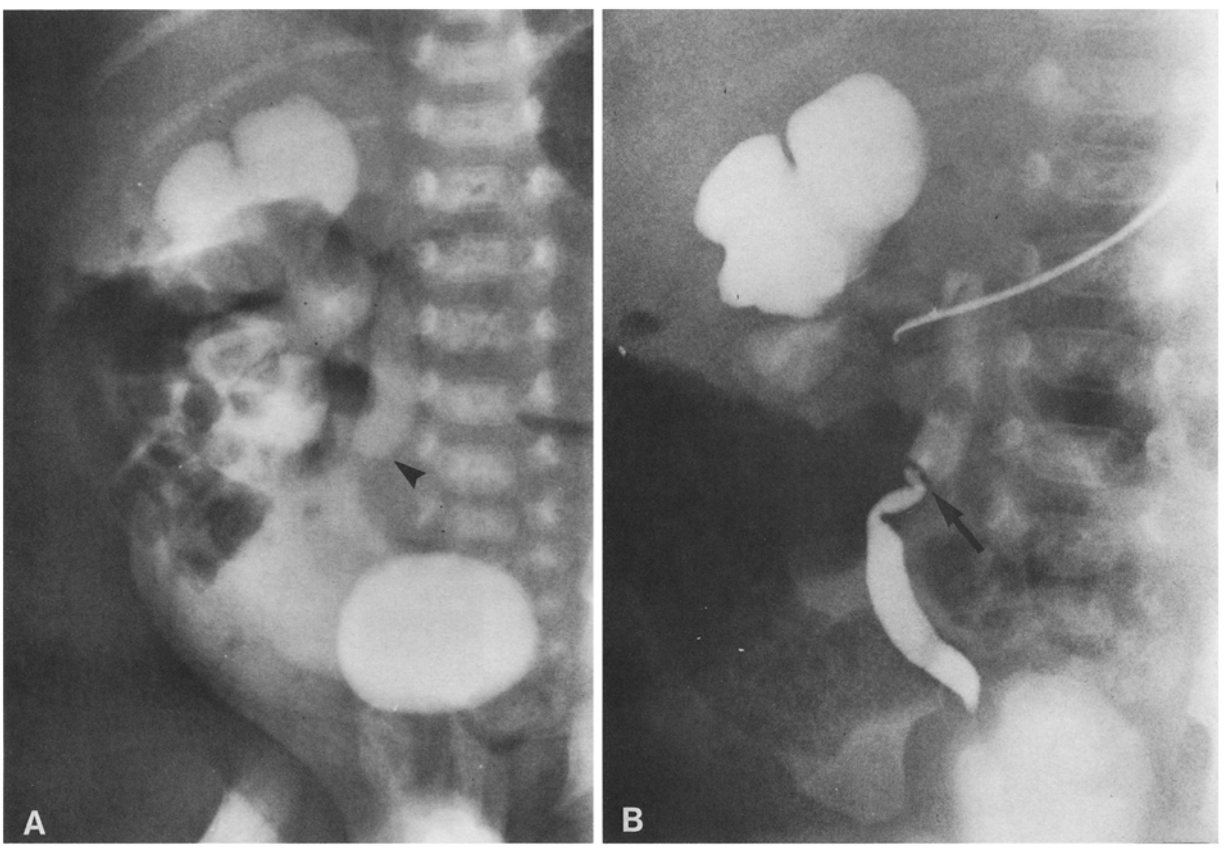

Fig. 1. *Case 1.* **A** Late film from intravenous urogram demonstrating dilated renal pelvis and proximal ureter. The point of obstruction is in the midureter (*arrow*). The distal ureter is not well visualized. **B** Retrograde ureteropyelogram reveals presence of a midureteral stricture (*arrow*). Nasogastric tube is superimposed over the upper ureter.

(Fig. 1B) or antegrade pyelography. Two of the cases had a multicystic dysplastic contralateral kidney; one had a blind-ending ureter on retrograde ureterography, and presumed dysplastic kidney. There were no other associated genitourinary abnormalities. Five cases were correctly diagnosed as midureteral lesions prior to the surgical incision; planned ureteroureterostomies were thus performed. In two, an initial transvesical approach to a presumed megaureter was eventually modified to correct the lesion discovered at exploration. Surgical results have been satisfactory in all.

Histologic sections were available for review in three cases. All of these were consistent with a diagnosis of congenital midureteral stricture. There was only one case (D.S.) for whom longitudinal sec-

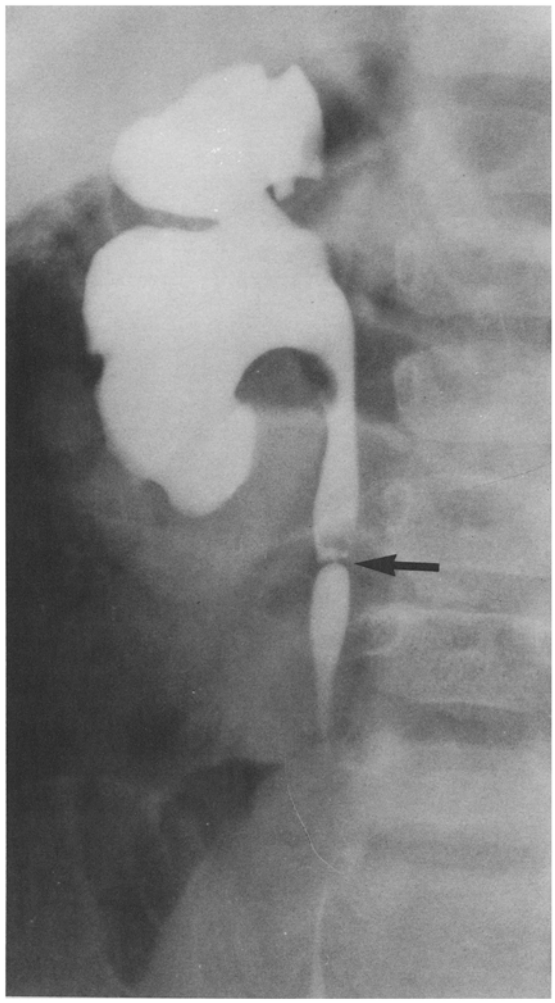

Fig. 2. *Case 2.* Left retrograde ureteropyelogram demonstrating midureteral stricture (*arrow*). (Reproduced with permission from Young DW, Lebowitz RL: Congenital abnormalities of the ureter. *Semin Roentgenol 21:*172, 1986.)

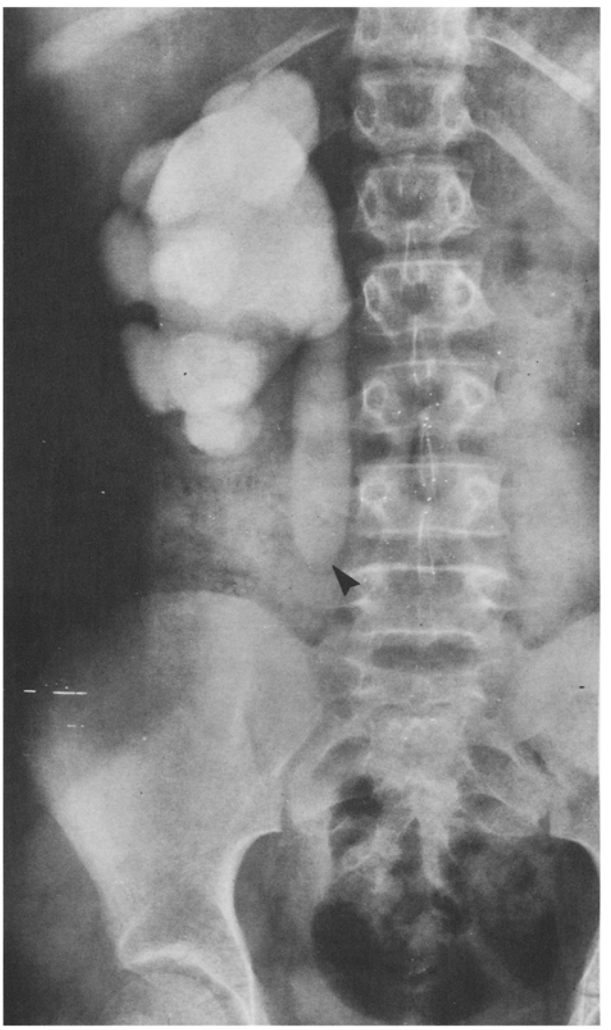

Fig. 3. Intravenous urogram in a 12-year-old girl who presented with urinary tract infection, demonstrating midureteral obstruction (*arrow*). Exploration revealed a midureteral valve. (Reproduced with permission from Young DW, Lebowitz RL: Congenital abnormalities of the ureter. *Semin Roentgenol 21:*172, 1986.)

tions were on file; such sections are considered necessary for the accurate differentiation of stricture and valve [1]. Three cases had written pathology reports on record. One (G.S.) had a longitudinal section noted, on which the diagnosis of congenital ureteral valve had been made. The other two had been diagnosed as strictures, with no mention of longitudinal sections. One child (D.C.) had neither slides nor a report for review, but notes in the chart indicated that the pathology was consistent with congenital ureteral valve.

Case Reports

Case 1

A.D., the male product of a term gestation and uneventful delivery, was noted on prenatal ultrasonography to have a small, abnormal-appearing left kidney and right-sided hydronephrosis. A postnatal sonogram confirmed the findings. Voiding cystourethrography was normal. An intravenous urogram revealed right-sided hydronephrosis with proximal ureteral dilatation and nonvisualization of the left kidney (Fig. 1A). Nuclear renal imaging revealed nonfunction of the left kidney and hydroureteronephrosis with delayed uptake and excretion on the right. At 5 weeks, serum creatinine was 0.3 mg/dl.

The child was taken to the operating room at 5 weeks of age with a preoperative diagnosis of right megaureter or midureteral obstruction and left dysplastic kidney. A right retrograde ureteropyelogram was performed and clearly revealed a midureteral stricture (Fig. 1B). The operative procedure was therefore carried out through a right lower abdominal oblique incision and retroperitoneal approach. The stricture was excised and primary ureteroureterostomy performed. During the same procedure a left nephrectomy was performed through a separate left flank incision.

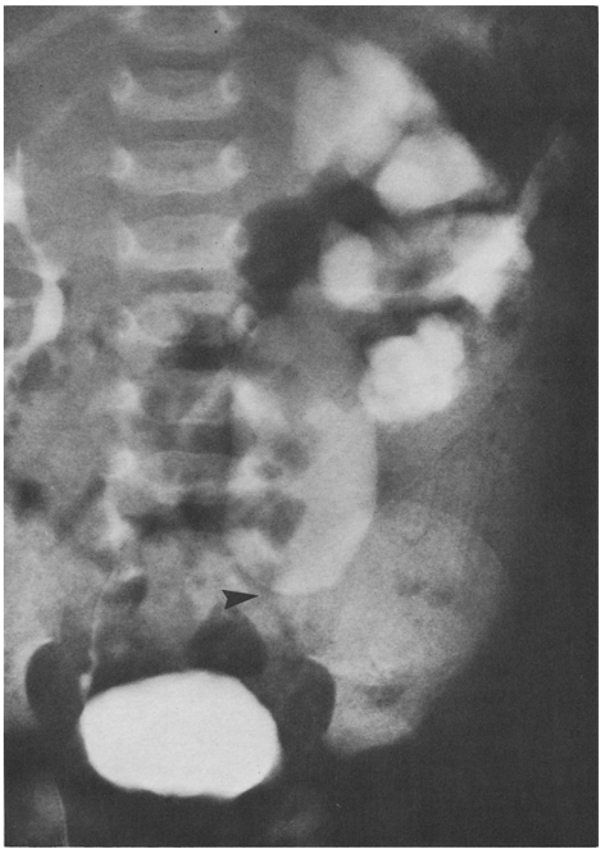

Fig. 4. Intravenous urogram in a 3-month-old boy who presented with urinary tract infection, demonstrating midureteral obstruction (*arrow*). Longitudinal section of the excised ureter revealed a midureteral stricture. (Reproduced with permission from Young DW, Lebowitz RL: Congenital abnormalities of the ureter. *Semin Roentgenol 21:*172, 1986.)

The child has done well over the 6 months since the procedure. Histology confirmed a multicystic dysplastic left kidney and a right ureteral stricture with focal muscular thinning. There was no fibrosis at the site of the stricture.

Case 2

S.L., a 2½-year-old girl was in good health until she had two febrile urinary tract infections. Physical examination was normal. Voiding cystourethrography was normal. An intravenous urogram revealed right hydroureteronephrosis and nonvisualization of the left kidney. Left retrograde ureterogram demonstrated a blind-ending left ureter (suggesting a multicystic dysplastic kidney) and a right retrograde study showed a discrete narrowing of the right mid-ureter consistent with a stricture (Fig. 2).

The right ureter was approached via a right flank incision. The upper ureter was widely dilated to a point 4 cm below the ureteropelvic junction. The ureter below this point was of normal caliber. The area of stricture was excised and ureteroureterostomy was performed. Histology revealed an irregular ureteral stricture.

Fifteen years postoperatively, the child is doing well except for mild hypertension controlled without medications. Her serum creatinine is 0.8 mg/dl.

Discussion

Congenital ureteral valve and stricture are rare entities; there are only 18 previous cases of well-documented ureteral valve in the pediatric literature [1]. The differentiation of valve and stricture can generally only be made on longitudinal sections of the excised portion of the ureter. The embryologic etiology of these anomalies has been elusive, though several theories have been presented. The presence of physiologically insignificant ureteral folds in fetal life and infancy has been well-documented [2], and the abnormal persistence of these folds has been postulated to account for obstructive ureteral valves [3, 4]. Lower ureteral valves have been considered remnants of Chwalla's membrane [5], an obstructing membrane in the lower ureter which is present at 6 weeks of gestation and is presumed to rupture under hydrostatic pressure at 8 weeks. Congenital ureteral stricture has been attributed to intrauterine ureteritis [6], extrinsic compression by blood vessels [7, 8], and improper progression of the normal process of ureteral obstruction and recanalization during fetal life [9]. Abnormal ureteral embryogenesis as a cause of these entities has been suggested by the high rate of occurrence of associated genitourinary abnormalities [10]. In the current series, two of the seven cases of midureteral obstruction had proven contralateral renal dysplasia, and one had a blind-ending ureter suggestive of dysplasia.

As is demonstrated in this series, a systematic progression of imaging tests in the evaluation of hydronephrosis such as ultrasonography, voiding cystourethrography, intravenous urography, and, especially if the diagnosis is in question, percutaneous antegrade pyelography or retrograde ureteropyelography will allow preoperative diagnosis in the majority of cases of congenital midureteral obstruction. The resulting increase in diagnostic accuracy will allow specific surgical intervention without the need for extending the original incision or performing a ureteral anastomosis with suboptimal exposure.

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