Chapter 37. Disorders of the Ureter & Ureteropelvic Junction

Barry A. Kogan, MD

Disorders of the Ureter & Ureteropelvic Junction: Introduction

The ureter is a complex functional conduit carrying urine from the kidneys to the bladder. Any pathologic process that interferes with this activity can cause renal abnormalities, the most common sequels being hydronephrosis (see Chapter XXX) and infection. Disorders of the ureter can be classified as congenital or acquired.

Congenital Anomalies of the Ureter

Congenital ureteral malformations are common and range from complete absence to duplication of the ureter. They may cause severe obstruction requiring urgent attention, or they may be asymptomatic and of no clinical significance. The nomenclature can be confusing and has been standardized to prevent ambiguity (Glassberg et al, 1984).

Ureteral Atresia

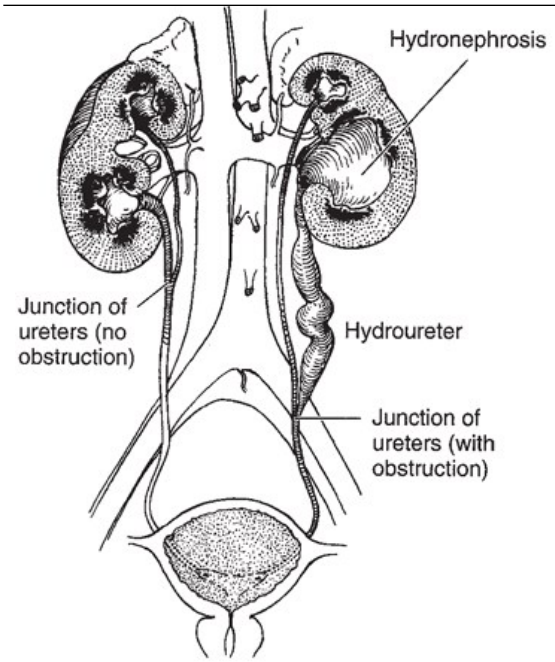
The ureter may be absent entirely, or it may end blindly after extending only part of the way to the flank. These anomalies are caused during embryologic development, by failure of the ureteral bud to form from the mesonephric duct or by an arrest in its development before it comes in contact with the metanephric blastema. The genetic determinants of ureteral bud development and the causes of bud abnormalities are being elucidated and it is known that GDNF signaling via the RET receptor is generally required (Michos et al, 2010). In any event, the end result of an atretic ureteral bud is an absent or multicystic dysplastic kidney. The multicystic kidney is usually unilateral and asymptomatic and of no clinical significance. In rare cases, it can be associated with hypertension, infection, or tumor. Contralateral vesicoureteral reflux is common, and many clinicians recommend a voiding cystourethrogram as part of the initial workup. There has been a concern about the risk of malignancy in these cases; however, the preponderance of evidence now suggests that no treatment is necessary and indeed no follow-up is needed from a urological standpoint (Onal and Kogan, 2006).

Duplication of the Ureter

Complete or incomplete duplication of the ureter is one of the most common congenital malformations of the urinary tract. Nation (1944) found some form of duplication of the ureter in 0.9% of a series of autopsies. The condition occurs more frequently in females than in males and is often bilateral.

Incomplete (Y) type of duplication is caused by branching of the ureteral bud before it reaches the metanephric blastema. In most cases, this anomaly is associated with no clinical abnormality. However, disorders of peristalsis may occur near the point of union (Figure 37–1).

Figure 37–1.



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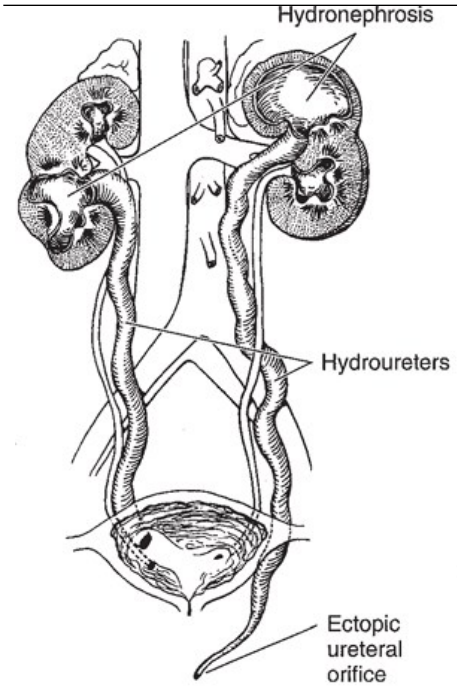
Duplication of the ureter. Incomplete (Y) type with hydronephrosis of lower pole of left kidney. Ureteroureteral (yo-yo) reflux can also occur and account for the radiographic appearance.

In complete duplication of the ureter, the presence of two ureteral buds leads to the formation of two totally separate ureters and two separate renal pelves. Because the ureter to the upper segment arises from a cephalad position on the mesonephric duct, it remains attached to the mesonephric duct longer and consequently migrates farther, ending medial and inferior to the ureter draining the lower segment (Weigert-Meyer law). Thus, the ureter draining the upper segment may migrate too far caudally and become ectopic and obstructed, whereas the ureter draining the lower segment may end laterally and have a short intravesical tunnel that leads to vesicoureteral reflux (Figure 37–2) (Tanagho, 1976). More recent studies have suggested that apoptosis of the common nephric duct cells is essential for separation of the ureter from the Wolffian duct; failure of this likely is the root cause of an ectopic ureter (Mendelsohn, 2009). Moreover, it is the urogenital sinus that induces this apoptosis and separation, and it appears that the upper ureteral bud may, in some cases, be too far from the urogenital sinus to receive the signal, thereby failing to separate from the Wolffian duct.

Figure 37–2.



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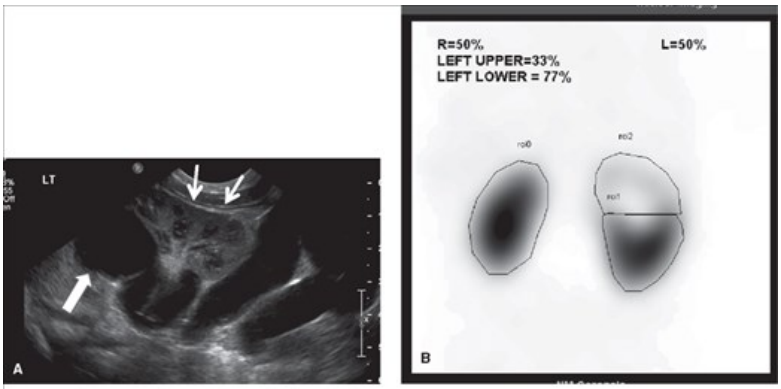
Duplication of the ureter. Complete duplication with reflux to lower pole of right kidney and chronic pyelonephritic scarring. Upper-pole ureter of left kidney is ectopic, and its associated renal parenchyma is often dysplastic.

Although many patients with duplication of the ureter are asymptomatic, a common presentation is persistent or recurrent infections. In females, the ureter to the upper pole may be ectopic, with an opening distal to the external sphincter or even outside the urinary tract. Such patients have classic symptoms: incontinence characterized by constant dribbling and at the same time, a normal pattern of voiding. In males, because the mesonephric duct becomes the vas and seminal vesicles, the ectopic ureter is always proximal to the external sphincter, and associated incontinence does not occur. In recent years, prenatal ultrasonography has led to the diagnosis in many asymptomatic neonates.

At the present time, ultrasound is the study of choice in these children. Generally, a hydronephrotic upper pole and a dilated distal ureter are seen, and in addition, one can readily evaluate parenchymal thickness and the presence of a ureterocele or other bladder anomalies. A voiding cystourethrogram is important to determine the presence of vesicoureteral reflux and confirm the presence of a ureterocele. Renal scanning (especially with ^{99m}Tc-dimercaptosuccinic acid) is helpful for estimating the degree of renal function in each renal segment (Figure 37–3).

Figure 37–3.



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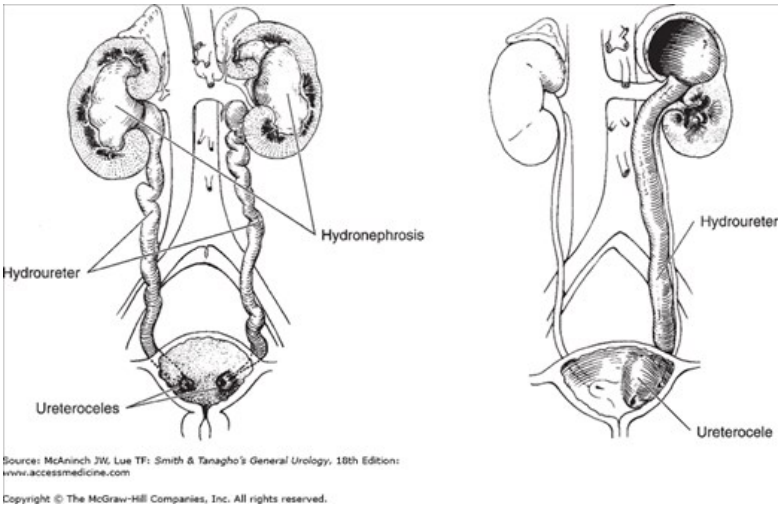
Duplicated left kidney. ***A:*** Ultrasound showing marked hydronephrosis of the left upper pole (*large arrow*) in continuity with a large tortuous ureter. The lower pole of the kidney is well preserved (*small arrows*). ***B:*** 99mTc-DMSA scan showing the relative function of the different renal segments.

The treatment of reflux is controversial (Chapter XXXX), but the treatment should not be influenced by the presence of ureteral duplication. Lower grades of reflux are generally treated medically and higher grades of reflux are more likely to be treated surgically. Because of anatomic variations, many surgical options are available. If upper-pole obstruction or ectopy is present, surgery is almost always required. Numerous operative approaches have been recommended (Belman et al, 1974). If renal function in one segment is very poor, heminephrectomy is the most appropriate procedure. In an effort to preserve renal parenchyma, treatments by pyeloureterostomy, ureteroureterostomy, and ureteral reimplantation are all appropriate (Amar, 1970, 1978). This can be done laparoscopically or open (Lowe et al, 2008; Prieto et al, 2009).

Ureterocele

A ureterocele is a sacculation of the terminal portion of the ureter (Figure 37–4). It may be either intravesical or ectopic; in the latter case, some portion is located at the bladder neck or in the urethra. Intravesical ureteroceles are associated most often with single ureters, whereas ectopic ureteroceles nearly always involve the upper pole of duplicated ureters. Ectopic ureteroceles are four times more common than those that are intravesical (Snyder and Johnston, 1978). Ureterocele occurs seven times more often in girls than in boys, and about 10% of cases are bilateral. Mild forms of ureterocele are found occasionally in adults examined for unrelated reasons.

Figure 37–4.

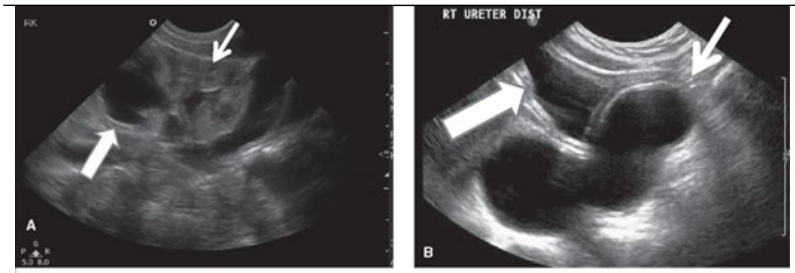


Ureterocele. ***Left:*** Orthotopic ureterocele associated with a single ureter. ***Right:*** Ureterocele associated with ureteral duplication and poor function of upper pole of kidney.

Ureterocele has been attributed to delayed or incomplete canalization of the ureteral bud leading to an early prenatal obstruction and expansion of the ureteral bud prior to its absorption into the urogenital sinus (Tanagho, 1976). The cystic dilation forms between the superficial and deep muscle layers of the trigone. Large ureteroceles may displace the other orifices, interfere with the muscular backing of the bladder, or even obstruct the bladder outlet. There is nearly always significant hydroureteronephrosis, and a dysplastic segment of the upper pole of the kidney may be found in association with a ureterocele.

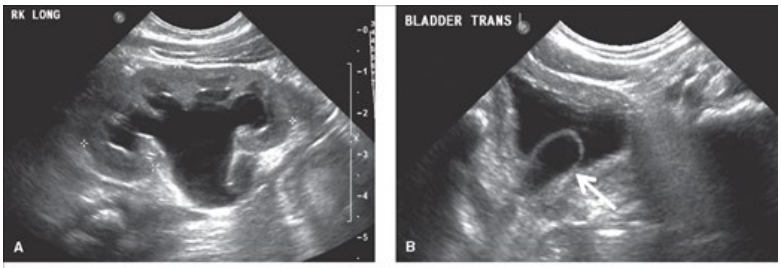
Clinical findings vary considerably. In the past, patients presented with infection, bladder outlet obstruction, or incontinence (and rarely a ureterocele may prolapse through the female urethra). However, most of the current cases are diagnosed by antenatal maternal ultrasound. After birth, sonography and voiding cystourethrography should be performed. The former confirms the diagnosis and defines the renal anatomy and the latter demonstrates whether there is reflux into the lower pole or contralateral ureter (Figures 37–5 and 37–6). Renal scanning is helpful for estimating renal function and the combination of findings is critical in planning therapy.

Figure 37–5.



Ureterocele in a girl with a duplication. ***A:*** Ultrasound showing marked hydronephrosis of the right upper pole (*large arrow*). The lower pole of the kidney is well preserved (*small arrow*). ***B:*** In contrast to an ectopic ureter, the dilated distal right ureter ends in a large ureterocele (*small arrow*) within the bladder (*large arrow*).

Figure 37–6.



Ureterocele in a boy with a single system. ***A:*** Ultrasound showing moderate right hydronephrosis with a dilated proximal ureter. ***B:*** The right distal ureter ends in a small orthotopic ureterocele within the bladder (*arrow*).

Treatment must be individualized. Transurethral incision has been recognized as the definitive procedure in many instances, particularly in patients with intravesical ureteroceles and may be the initial therapy in neonates. When an open operation is needed, the procedure must be chosen on the basis of the anatomic location of the ureteral meatus, the position of the ureterocele, and the degree of hydroureteronephrosis and impairment of renal function. In general, choices range from heminephrectomy and ureterectomy to excision of the ureterocele, vesical reconstruction, and ureteral reimplantation. When the ureterocele is ectopic, definitive treatment often entails excision of the ureterocele and reconstruction (Wang, 2008). Often, more than one procedure is necessary.

Ectopic Ureteral Orifice

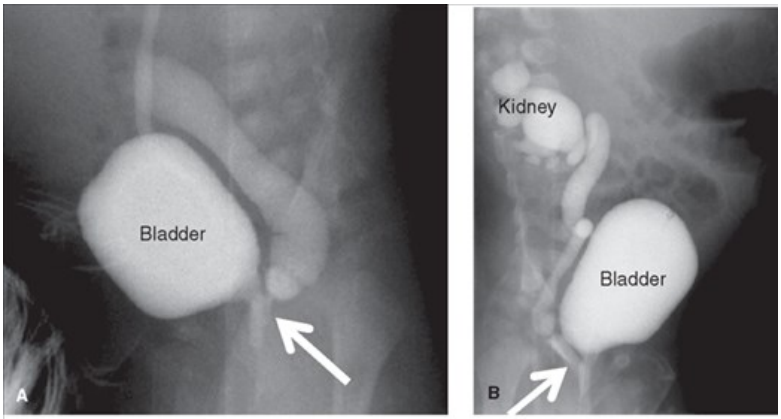
Although an ectopic ureteral orifice most commonly occurs in association with duplication of the ureter (see preceding sections), single ectopic ureters do occur. They are caused by a delay or failure of separation of the ureteral bud from the mesonephric duct during embryologic development. Again, the genetic determinants of these ureteral bud abnormalities are currently being determined, but at least GDNF signaling via the RET receptor is needed and apoptosis of the common nephric duct is critical (Michos et al, 2010). In anatomic terms, the primary anomaly may be an abnormally located ureteral bud; that also explains the high incidence of dysplastic kidneys associated with single ectopic ureters.

The clinical picture varies according to the sex of the patient and the position of the ureteral opening. Boys are seen because of urinary tract infection or epididymitis. In these cases, the ureter may drain directly into the vas deferens or seminal vesicle. In girls, the ureteral orifice may be in the urethra, vagina, or perineum. Although infection may be present, incontinence is the rule. Continual dribbling despite normal voiding is pathognomonic, but urgency and urge incontinence may confound the diagnosis.

Sonography and voiding cystourethrography help delineate the problem. If the ureter is ectopic to the urethra, the ectopic ureter may be seen on the voiding film from the voiding cystourethrography (Figure 37–7). However, because an ectopic kidney may be both tiny and in an abnormal location, it may be difficult to find by ultrasound, and magnetic resonance imaging, cystoscopy, or laparoscopy may be necessary to confirm the diagnosis (Lipson et al, 2008). During cystoscopy, a hemitrigone may be seen and the ectopic orifice may be visualized directly or demonstrated by retrograde

catheterization. Renal scanning is also helpful in estimating relative renal function. As in ureteroceles and duplication of the ureter, the clinical picture and the degree of renal function dictate the therapeutic approach.

Figure 37–7.

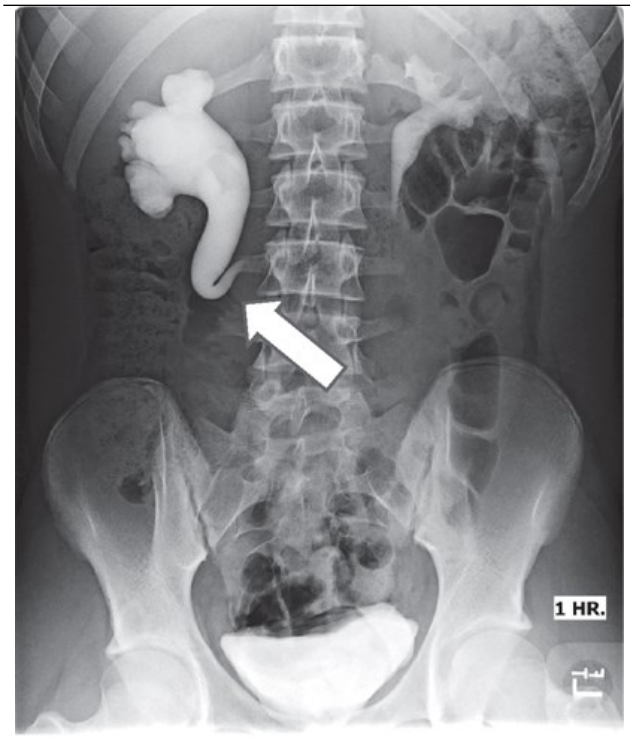


Ectopic ureter. ***A:*** Voiding cystourethrogram in a boy showing voiding into the posterior urethra with reflux into a very dilated ureter, seen entering the prostatic urethra (*arrow*). ***B:*** A voiding cystourethrogram in a girl showing voiding with reflux into a very dilated ureter and continuing all the way up to a dilated lower pole of the right kidney. Note that the ureter is ectopic, entering the proximal urethra (*arrow*).

Abnormalities of Ureteral Position

Retrocaval ureter (also called circumcaval ureter and postcaval ureter) is a rare condition in which an embryologically normal ureter becomes entrapped behind the vena cava because of abnormal persistence of the right subcardinal (as opposed to the supracardinal) vein. This forces the right ureter to encircle the vena cava from behind. The ureter descends normally to approximately the level of L3, where it curves back upward in the shape of a reverse J to pass behind and around the vena cava (Figure 37–8). Obstruction generally results.

Figure 37–8.



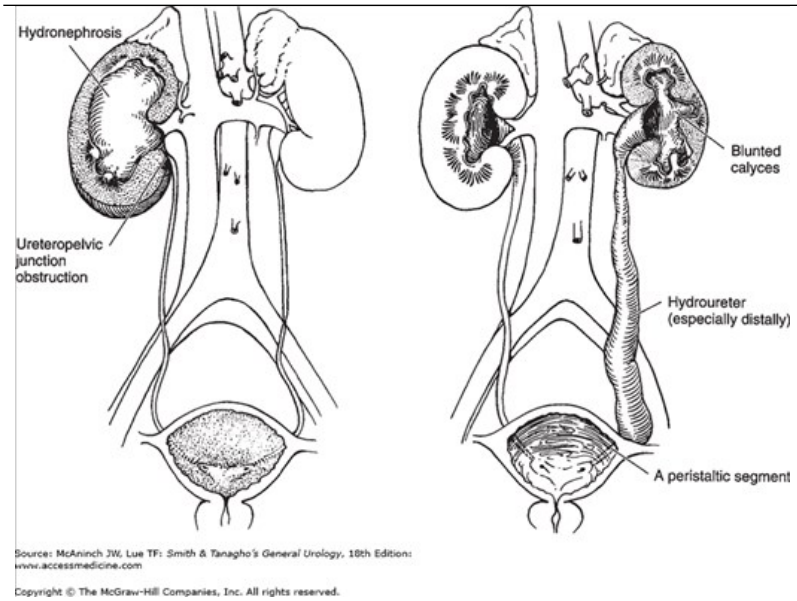
Retrocaval ureter. An intravenous urogram showing the classic appearance of a J-hooking of the ureter behind the inferior vena cava (*arrow*).

The diagnosis of retrocaval ureter is still made by excretory urography in some cases. However, since sonography is now usually the first test performed, the radiologist must be suspicious of the anomaly based on a dilated proximal (but not distal) ureter. Currently, magnetic resonance imaging may be the best single study to delineate the anatomy clearly and noninvasively. Surgical repair for retrocaval ureter, when indicated, consists of dividing the ureter (preferably across the dilated portion), bringing the distal ureter from behind the vena cava, and reanastomosing it to the proximal end. The procedure has been performed laparoscopically to reduce morbidity (Bagheri, 2009).

Obstruction of the Ureteropelvic Junction

In children, primary obstruction of the ureter usually occurs at the ureteropelvic junction or the ureterovesical junction (Figure 37–9). Obstruction of the ureteropelvic junction is probably the most common congenital abnormality of the ureter. In children, it is seen more often in boys than in girls (5:2 ratio), but it has recently been discovered that in adults, it is more common in women than in men (Capello, 2005). In unilateral cases, it is more often on the left than on the right side (5:2 ratio). Bilateral obstruction occurs in 10–15% of cases and is especially common in infants (Johnston et al, 1977). The abnormality may occur in several members of the same family, but it shows no clear genetic pattern.

Figure 37–9.

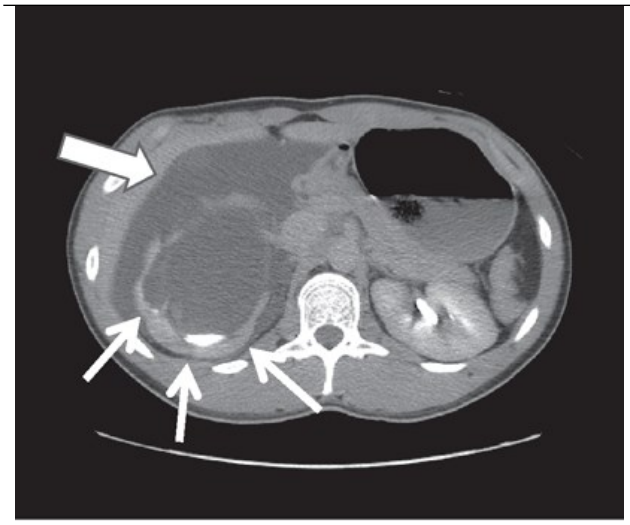


Congenital ureteral obstruction. ***Left:*** Right ureteropelvic junction obstruction with hydronephrosis. ***Right:*** Left ureterovesical junction obstruction (obstructed megaureter) with hydroureteronephrosis.

The exact cause of obstruction of the ureteropelvic junction often is not clear. Ureteral polyps and valves are seen but are rare. There is almost always an angulation and kink at the junction of the dilated renal pelvis and ureter. This by itself can cause obstruction, but it is unclear whether this is primary or merely secondary to another obstructive lesion. True stenosis is found rarely; however, a thin-walled, hypoplastic proximal ureter is observed frequently. Characteristic histologic and ultrastructural changes are observed in this area and could account for abnormal peristalsis through the ureteropelvic junction and consequent interference with pelvic emptying (Hanna et al, 1976). Current basic research suggests that decreased BMP4 signaling leads to disruption of smooth muscle investment of the ureter and may be involved (Wang, 2009). In addition, there seems to be an overexpression of extracellular matrix and depleted numbers of nerves (Kaya et al, 2010). Two other findings sometimes seen at operation are a high origin of the ureter from the renal pelvis and an abnormal relationship of the proximal ureter to a lower-pole renal artery. It is debatable whether these findings are the cause or the result of pelvic dilatation, but Stephens (1982) has suggested that abnormal rotation of the renal pelvis allows the ureter to become entrapped in the blood vessels of the lower pole of the kidney, ultimately leading to obstruction. Using careful studies at the time of operation, it is possible to define whether the principal lesion is intrinsic or extrinsic (Johnston, 1969; Koff et al, 1986).

Clinical findings vary depending on the patient's age at diagnosis. Many cases are now diagnosed in utero. These tend to be primarily intrinsic lesions and many resolve spontaneously. Later, pain and vomiting are the most common symptoms; however, hematuria and urinary infection also may be seen. A few patients have complications such as calculi, trauma to the enlarged kidney (Figure 37–10), or (rarely) hypertension. Most of the latter are extrinsic lesions but it is unclear whether they originated from intrinsic problems earlier in development.

Figure 37–10.



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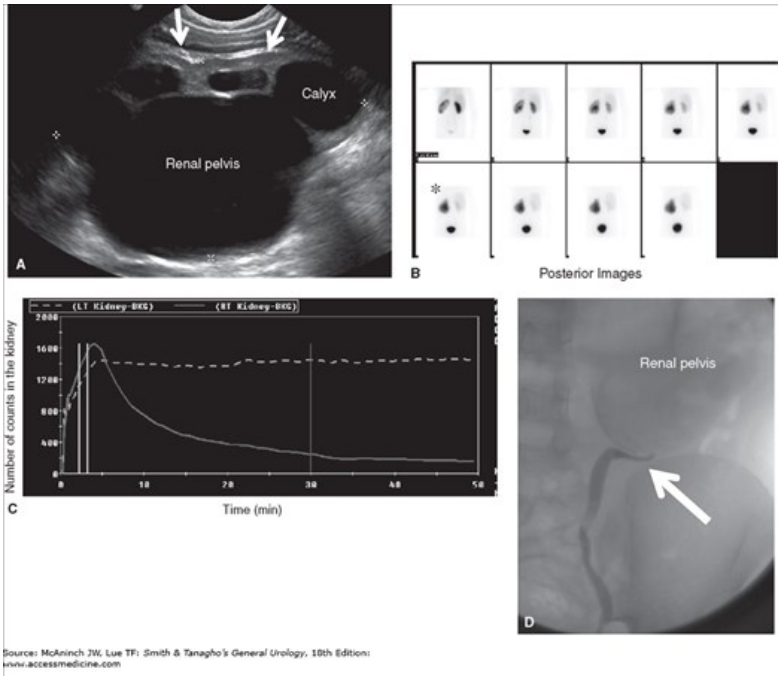
Ureteropelvic junction obstruction discovered after trauma. CT scan in an 18-year-old boy performed after abdominal trauma shows a markedly hydronephrotic right kidney (*small arrows*) and a urinoma due to urinary extravasation from a rupture of the renal pelvis (*large arrow*).

The diagnosis is made most often by sonography. In equivocal cases, diuretic renography or (rarely) antegrade urography with pressure-flow studies is helpful (Thrall et al, 1981; Whitaker, 1973) (Figure 37–11). Many surgeons consider a voiding cystourethrogram a routine part of the preoperative workup, since radiographic findings in vesicoureteral reflux may be similar to those in ureteropelvic junction obstruction. This fact is especially relevant when the ureter is well seen or dilated (or both) below the ureteropelvic junction.

Figure 37–11.



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Ureteropelvic junction obstruction after prenatal hydronephrosis. ***A:*** An ultrasound shows marked hydronephrosis with a large renal pelvis and dilated calyces. There is some preservation of renal parenchyma (*arrows*). ***B:*** Images taken every 5 minutes from the 99mTc-MAG-3 diuretic renogram showing good function on the left kidney, but hydronephrosis and slow drainage from the kidney. The *asterisk* indicates when furosemide is given. Of

note, there is no increase in drainage from the left kidney after furosemide. ***C:*** The computer calculated time/activity curves from the renogram pictures. Both kidneys have good function (high counts in the first couple of minutes). The right kidney (*solid line*) excretes the radioisotope rapidly. There is poor drainage of the left kidney (*dotted line*). ***D:*** A retrograde pyelogram demonstrates a normal ureter with obstruction at the ureteropelvic junction (*arrow*) and a large renal pelvis above the obstruction.

Symptomatic obstruction of the ureteropelvic junction should be treated surgically. Because most cases are now detected by hydronephrosis on prenatal ultrasonography and the infants are asymptomatic, it becomes important to assess the significance of the hydronephrosis. On the one hand, early surgery may prevent future urinary tract infections, stones, or other complications; on the other hand, many of the patients could live their whole lives without experiencing a consequence of the hydronephrosis. This remains an area of considerable controversy. Early surgery is recommended for patients who have kidneys with diminished function, massive hydronephrosis, infection, or stones. Nonoperative surveillance with good follow-up is thought to be safe, although about 25% of patients will ultimately require an operative repair for pain, urinary infection, or reduced renal function on repeat nuclear scan. Clinicians and scientists are searching for biomarkers of obstruction, and urinary proteome analysis seems promising (Drube et al, 2010). This subject remains particularly controversial, but there are some data suggesting that early childhood treatment reduces the risk of ureteropelvic junction obstruction in adults (Capello, 2005).

Because of anatomic variations, no single procedure is sufficient for all situations (Smart, 1979). Regardless of the technique used, all successful repairs have in common the creation of a dependent and funnel-shaped ureteropelvic junction of adequate caliber. Although preservation of the intact ureteropelvic junction is feasible in some circumstances, when the obstruction appears to be caused by a dyskinetic segment of proximal ureter, the most popular operation is a dismembered pyeloureteroplasty (Anderson, 1963). Dismembered pyeloureteroplasty is also favored when the proximal ureter is hooked over a lower-pole blood vessel. When there is a dilated extrarenal renal pelvis, dismembered pyeloureteroplasty can be combined with a Foley Y-V plasty to create a more funnel-shaped ureteropelvic junction (Foley, 1937). Pelvic flap procedures (Culp and DeWeerd, 1951; Scardino and Prince, 1953) are suited ideally to cases in which the ureteropelvic junction has remained in a dependent position despite significant pelvic dilatation. They also have the advantage of interfering less with the ureteral blood supply; this is particularly relevant when distal ureteral surgery (eg, ureteral reimplantation) is contemplated in the future. In most centers, the dismembered pyeloureteroplasty is the mainstay of repairs.

Both the Y-V plasty and the flap techniques are useful in managing ureteropelvic junction obstructions in horseshoe or pelvic kidneys, in which the anatomy may prevent creation of a dependent ureteropelvic junction if a dismembered technique is attempted. The use of stenting catheters and proximal diversion at the time of pyeloplasty has been the subject of debate, and the issue has not been resolved. Excellent results have been reported both with and without stents and diversions (Smith et al, 2002).

The prognosis is generally good. In several large series, the reported reoperation rate has been only 2–4%, but the postoperative radiographic appearance of the area may be disappointing. There can be marked improvement when a large extrarenal pelvis has prevented massive calyceal distortion; however, in most cases, considerable deformity persists despite adequate drainage of the kidney. Furthermore, it is usually many years before the radiographic appearance improves.

The progressive move to minimally invasive surgery has resulted in a number of trends in the repair of ureteropelvic junction obstruction. Endopyelotomy can be performed percutaneously or via ureteroscopy and treats the problem by incising directly through the obstruction, stenting the area and allowing healing via secondary intention. The technique is similar to that reported by Davis (1943) but is done entirely endoscopically. Unfortunately, the success rate is only 80–85% at best, and currently, it is primarily used for cases that have failed an initial repair. Another selected indication is when there are stones in the kidney with a ureteropelvic junction obstruction. Both conditions can be dealt within one endoscopic procedure. However, these procedures have largely been supplanted by laparoscopic repair. In particular, the remarkable growth of robotic urological surgery has allowed for many more surgeons to become proficient with robotic-assisted pyeloplasty and this has become a viable option. It remains unclear whether the long-term results are as good as open procedures, but there is a clear trend towards performing an increasing number of repairs this way. Although the morbidity of open surgery is decreasing, success rates with laparoscopic and robotic approaches are comparable and parents seem to prefer the robotic approach (Braga et al, 2009; Chacko et al, 2006; Freilich et al, 2010).

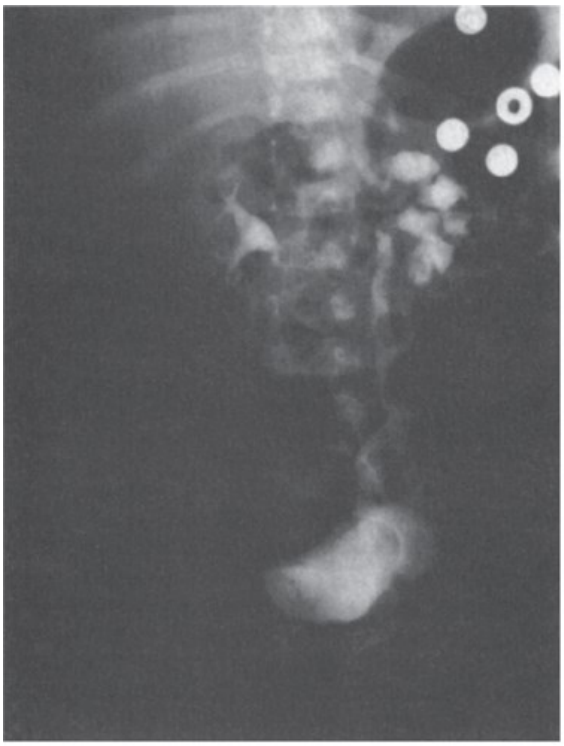
Obstructed Megaureter

Obstruction at the ureterovesical junction is four times more common in boys than in girls. It may be bilateral and is usually asymmetric. The left ureter is slightly more often involved than the right.

The embryogenesis of the lesion is uncertain. It is clear that in most cases, there is no stricture at the ureterovesical junction. At operation, a retrograde catheter or probe can usually be passed through the area of obstruction. Close observation either at operation or by fluoroscopy reveals a failure of the distal ureter to transmit the normal peristaltic wave, resulting in a functional obstruction. Moreover, on fluoroscopy, retrograde peristalsis is seen. This transmits abnormal pressures up to the kidney, resulting in calyceal dilation out of proportion with the renal pelvic dilation. Histologic findings include an excess of circular muscle fibers and collagen in the distal ureter that may account for the problem (Tanagho et al, 1970). Ultrastructural studies show that this obstruction is similar in appearance to obstruction of the ureteropelvic junction.

Currently, most of the cases are discovered on prenatal sonography. Sonography usually shows the pathognomonic configuration of a dilated distal ureter, a less dilated proximal ureter, a relatively normal-appearing renal pelvis, and calyces blunted out of proportion to the renal pelvis (Figure 37–12).

Figure 37–12.



Obstructed megaureter. Follow-up study in a 9-month-old boy with unilateral hydronephrosis detected by ultrasonography in utero. Excretory urogram shows the classic configuration of a dilated distal ureter, a less dilated proximal ureter, and blunted calyces.

It was assumed previously that surgery was indicated in most cases. Ureteral reimplantation with excision of the distal ureter is curative. Because of the excessive dilation of the ureter, ureteral tapering or folding may be necessary (Ehrlich, 1985; Hanna, 1982; Hendren, 1969). Because the ureteral muscle is generally healthy, these cases have an excellent prognosis. However, in recent years, it has become obvious that at least 50% of cases will undergo spontaneous resolution. A period of observation is nearly always appropriate when the diagnosis is made in an asymptomatic patient (Baskin et al, 1994; Chertin et al, 2008). Because of the high risk of infection, 1–2 years of prophylactic antibiotics are recommended in neonates.

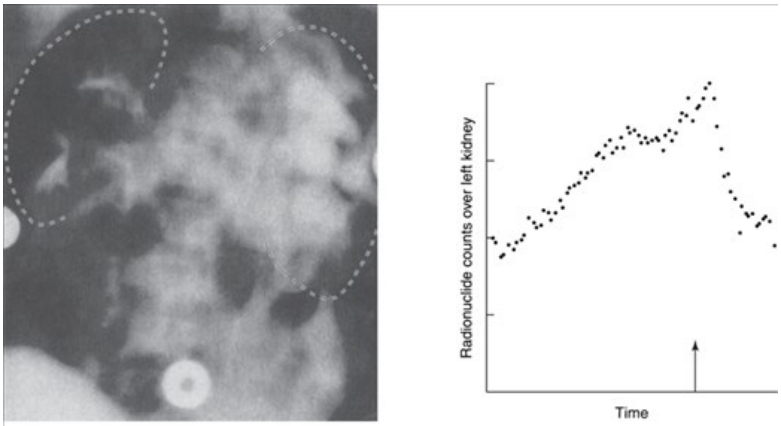
Upper Urinary Tract Dilatation Without Obstruction

It should not be assumed that every dilated upper urinary tract is obstructed. A voiding cystourethrogram is an essential part of the evaluation, not only to rule out reflux but also to ensure that no abnormality of the lower urinary tract is responsible for the upper urinary tract dilatation. Other cases in which diagnosis may be difficult include residual dilatation in a previously obstructed system, dilatation associated with bacterial infection

(presumably related to a direct effect of endotoxin on the ureteral musculature), neonatal hydronephrosis, and prolonged polyuria in patients with diabetes insipidus.

In such cases, the usual investigations may not provide sufficient information. A radionuclide diuretic renogram is especially helpful in distinguishing nonobstructive from obstructive dilation and in determining whether renal functional injury has occurred (Figure 37–13) (Thrall et al, 1981). However, the procedure must be performed carefully, as technical problems may confuse the results (Gungor et al, 2002; Nguyen et al, 1997). Use of percutaneous renal puncture is occasionally beneficial; in the dilated system, it carries minimal risk, making antegrade urography and pressure-flow studies feasible in selected cases. Measurement of the renal pelvic pressure during infusion of saline into the renal pelvis at high rates (10 mL/min) (*the Whitaker test*) may help differentiate nonobstructive from obstructive dilation (Wolk and Whitaker, 1982). Unfortunately, there is no true "gold standard," and these studies do not always agree; clinical judgment the final arbiter.

Figure 37–13.



Upper urinary tract dilatation. ***Left:*** Three months after resection of posterior urethral valves, hydronephrosis in the right kidney has completely resolved. The left collecting system remains dilated. (*Dashed lines* outline kidneys.) ***Right:*** Radionuclide diuretic renography was performed to determine if there was secondary ureteropelvic or ureterovesical obstruction. Renogram demonstrates clear-cut "washout" of radionuclide following injection of furosemide (*arrow*). There is no significant obstruction.

Acquired Diseases of the Ureter

Nearly all acquired diseases of the ureter are obstructive in nature. Although they are seen frequently, their actual incidence is unknown. Their clinical manifestations, effects on the kidney, complications, and treatment are similar to those described previously. The lesions can be broadly categorized as either intrinsic or extrinsic.

Intrinsic Ureteral Obstruction

The most common causes of intrinsic ureteral obstruction are as follows:

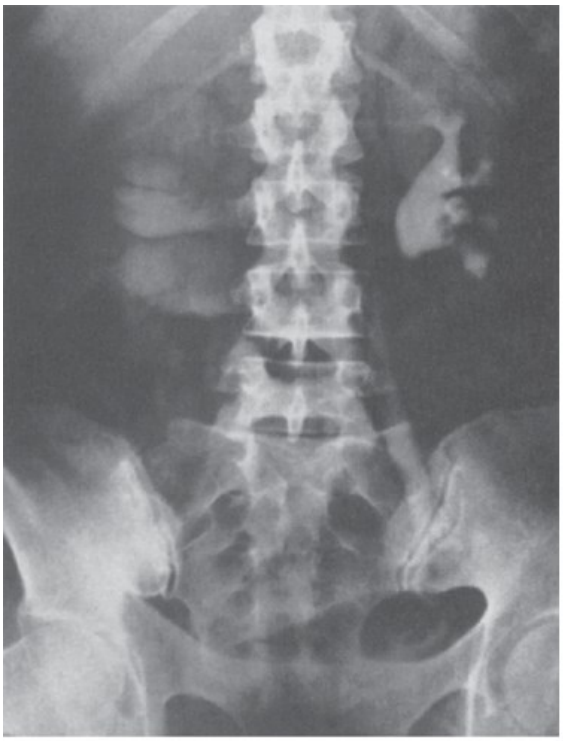
1. Ureteral stones (see Chapter 17)
2. Transitional cell tumors of the ureter (see Chapter 21)
3. Chronic inflammatory changes of the ureteral wall (usually due to tuberculosis or schistosomiasis) leading to contracture or insufficient peristalsis (see Chapter 15 and Figures 15–2 and 15–4)

Extrinsic Ureteral Obstruction

The most frequent causes of extrinsic ureteral obstruction are as follows:

1. Severe constipation, sometimes with bladder obstruction, seen primarily in children but in adult women as well.
2. Secondary obstruction due to kinks or fibrosis around redundant ureters. The primary process is either distal obstruction or massive reflux.
3. Benign gynecologic disorders such as endometriosis or right ovarian vein syndrome.
4. Local neoplastic infiltration associated with carcinoma of the cervix, bladder, or prostate.
5. Pelvic lymphadenopathy associated with metastatic tumors.
6. Iatrogenic ureteral injuries, primarily after extensive pelvic surgery (Figure 37–14) and also after extensive radiotherapy.
7. Retroperitoneal fibrosis.

Figure 37–14.



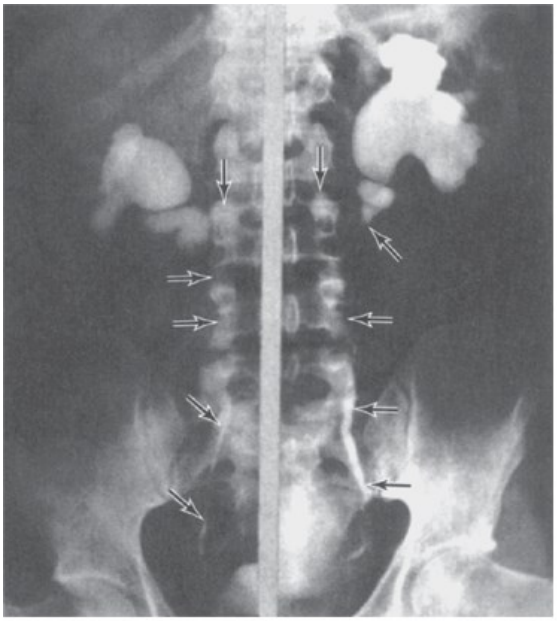
Ureteral obstruction. Excretory urogram obtained 2 weeks after a radical hysterectomy shows bilateral ureteral obstruction and marked hydronephrosis on right.

Retroperitoneal Fibrosis (Retroperitoneal Fasciitis, Chronic Retroperitoneal Fibroplasia, Ormond Disease)

One or both ureters may be compressed by a chronic inflammatory process that involves the retroperitoneal tissues over the lower lumbar vertebrae. There are numerous causes of retroperitoneal fibrosis. Malignant diseases (most commonly Hodgkin's disease, carcinoma of the breast, and carcinoma of the colon) should always be suspected and ruled out. Some medications have been implicated, most notably methysergide (Sansert), an ergot derivative used to treat migraine headaches. Rarely, membranous glomerulonephritis (Shirota et al, 2002), inflammatory bowel disease (Siminovitch and Fazio, 1980), or an aortic aneurysm (Brock and Soloway, 1980) is responsible. The remainder of cases are idiopathic, a condition sometimes referred to as Ormond disease. It is most recently thought to be autoimmune, and in men especially, IgG4 is likely involved (Zen, 2009).

The symptoms are nonspecific and include low back pain, malaise, anorexia, weight loss, and, in severe cases, uremia (Swartz, 2009). Infection is uncommon. Although the diagnosis is usually made by Computed tomography (CT) scan, an intravenous urogram (or delayed plain x-ray from the CT scan) is pathognomonic (Figure 37–15). There is medial deviation of the ureters with proximal dilation. A long segment of ureter is usually involved, and, in some cases, in the CT urogram reconstructions, there is a pipestem appearance caused by aperistalsis related to the fibrosis. Ultrasonography is useful for monitoring the response to therapy. CT scanning or magnetic resonance imaging is essential for evaluating the retroperitoneum itself, in addition to imaging the ureters (Hricak et al, 1983). Recently, positron emission tomography scanning has been advocated to diagnose metastatic cancer and to follow the severity of the inflammatory lesions (Piccoli, 2010).

Figure 37–15.



Retroperitoneal fibrosis. Right and left kidneys of same patient as shown by excretory urography. Note medial marked obstruction. (Courtesy of JA Hutch.)

Spontaneous regression has been reported (Kume and Kitamura, 2001); however, treatment is usually surgical. A course of corticosteroids may be successful, but recurrence is high if not continued (Swartz, 2009). When the response to corticosteroids is poor or the obstruction is severe, the ureter must be dissected surgically from the fibrous plaque. After it is freed, it should either be placed intraperitoneally or wrapped in omentum in an attempt to prevent recurrence; this procedure can also be performed laparoscopically (Stein, 2010). Numerous biopsies of the fibrous tissue should be obtained at the time of operation to determine whether there is a malignant tumor.

Ureteral Obstruction Secondary to Malignant Disease

Ureteral obstruction associated with widespread malignant disease was at one time a terminal event. Because therapy for malignant diseases has improved, however, urinary diversion is indicated more frequently in such cases. Diversion usually is necessary for relatively short periods of time; either the tumor is progressive or if therapy is effective, the obstruction resolves. Thus, the goal of treatment is to leave the urinary tract intact and effect as little morbidity as possible. This can be accomplished with indwelling stents passed either retrograde during cystoscopy or antegrade using percutaneous techniques. Plastic stents can easily become obstructed and metal stents may be considered for long-term use (Sountoulides, 2010).

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