Pediatric Ureteral Anomalies

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Cover Illustration by Christine Schaar

NOTE FROM THE PUBLISHER:
This publication has been developed without involvement of or review by the American Board of Urology.

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Clinically significant ureteral anomalies often are identified in childhood in association with urinary tract infection (UTI), incontinence, renal stones, or renal insufficiency. Before birth, ureteral anomalies may be detected on fetal ultrasonography in association with hydronephrosis. A basic understanding of the normal embryology of the genitourinary tract, the pathophysiology of ureteral anomalies, the standard evaluation techniques, and the risks and benefits of various surgical options helps the urologist to determine the treatment approach that will likely yield the best results for a particular patient.

This manual begins with a discussion of the terminology used to describe ureteral anomalies and the normal embryology of the genitourinary tract. Issues relating to pathophysiology, patient evaluation, and surgical options are then highlighted through case-based discussions. The specific anomalies discussed in this review are ureteroceles, ureteral duplications, ureteral ectopy, and ureteral reflux.

**TERMINOLOGY**

A standard set of definitions for ureteral anomalies has been established. A **duplex (duplicated) system** refers to one kidney with two separate pelvicaliceal systems. If such a kidney (also called a **duplex kidney**) has one ureter extending from each of the two pelvicaliceal systems and the two ureters empty separately into the bladder, it is considered a **complete duplication**. However, a duplex system may also be **incomplete** (or **partial**). A **bifid system** is a form of incomplete duplication in which two ureters from a duplex kidney are joined at the ureteropelvic junction (**bifid pelvis**) or before emptying into the bladder (**bifid ureters**), forming a Y shape.

An **upper pole ureter** or a **lower pole ureter** refers to a ureter draining the upper or lower pole of a duplex kidney and draining into the bladder via the **upper pole orifice** or the...
lower pole orifice, respectively. A laterally ectopic ureter inserts lateral to the normal position on the trigone, and a medially or caudally ectopic ureter inserts medial or distal to the normal position on the trigone. An ectopic ureter drains to an abnormal site and generally refers to an orifice located medially or caudally.

A ureterocele is a cystic dilation or ballooning of the intravesical submucosal ureter. An intravesical ureterocele is contained entirely within the bladder, whereas an ectopic ureterocele has a part of its wall arising from the bladder neck or urethra. A single-system ureterocele refers to a ureterocele occurring in relation to a kidney with only one ureter; a duplex-system ureterocele refers to a ureterocele that drains the upper pole of a kidney with a complete ureteral duplication.

**EMBRYOLOGY**

An understanding of genitourinary embryology is facilitated by viewing the mesonephric (wolffian) duct as the central piece to the embryologic puzzle. The ureter forms from the ureteric (metanephric) bud, which arises from the mesonephric duct. The mesonephric duct drains into the cloaca, which divides into the urogenital sinus and rectum. The ureteric bud grows and penetrates the metanephric blastema (Figure 1). The interaction between the ureteric bud and metanephric blastema results in the formation of the kidney and ureter. The ureteric bud forms the ureter, renal pelvis, calyces, papillary ducts, and collecting tubules.

The common excretory duct consists of the portion of the mesonephric duct between the origin of the ureteric bud and cloaca. This segment, along with a short segment above the ureteric bud, expands and inverts into the urogenital sinus and rectum. The ureteric bud grows and penetrates the metanephric blastema (Figure 1). The interaction between the ureteric bud and metanephric blastema results in the formation of the kidney and ureter. The ureteric bud forms the ureter, renal pelvis, calyces, papillary ducts, and collecting tubules.

The common excretory duct consists of the portion of the mesonephric duct between the origin of the ureteric bud and cloaca. This segment, along with a short segment above the ureteric bud, expands and inverts into the urogenital sinus, forming half of the trigone. It is unclear by what mechanism the ureteric bud expands and is incorporated into the urogenital sinus, separate from the mesonephric duct. As the developmental process proceeds, the ureteral oriﬁce migrates cephalad and laterally, while the mesonephric duct moves distally and medially. By the 12th week of gestation, the mesonephric duct reaches its ﬁnal position in the posterior urethra (at the level of the verumontanum in the male). The mesonephric duct becomes part of the epididymis, seminal vesicle, and vas deferens in the male and Gartner’s duct in the female.

Incomplete ureteral duplication results from a ureteric bud that bifurcates before it grows into the metanephric blastema. If the bifurcation occurs after the ureteric bud grows into the metanephric blastema, a bifid pelvis results.

When the ureteric bud originates at a signiﬁcantly higher-than-normal location, it may not be incorporated into the bladder and may end in the urethra or mesonephric remnants. In the male, this would result in an ectopic ureter to the epididymis, vas deferens, or seminal vesicle (Figure 2). In the female, this would result in an ectopic ureter to Gartner’s duct. This duct runs from the broad ligament along the lateral wall of the vagina to end at the hymen. An ectopic ureter to Gartner’s duct may drain into the vagina following secondary rupture of the duct into the vagina. In the female, ectopic ureters commonly exit below the level of sphincteric control and may present as constant wetting from the ectopic ureter. In the male, all ectopic ureters terminate above the level of the external urethral sphincter such that urinary incontinence is uncommon.

A complete duplication with double ureters requires two ureteric buds arising from the mesonephric duct. A ureteric bud originating at a lower-than-normal position on the mesonephric duct is incorporated into the urogenital sinus earlier than normal, allowing the ureteric bud to migrate more laterally and cranially. In this position, there is likely to be less trigonal support for the ureter, permitting ureteral reﬂux. If one of the ureteric buds is located normally and the other is in a lower-than-normal position, the result is ureteral reﬂux into the
ureter that, during migration, was carried most cranially and laterally (ie, the lower pole ureter).7 A ureteric bud originating at a higher-than-normal position on the mesonephric duct is incorporated into the urogenital sinus later than normal. This results in a shorter cranial and lateral migration, resulting in displacement of the ureteral orifice toward the bladder neck. The upper pole ureter would thus insert ectopically (Figure 3). The Meyer-Weigert law states that when complete ureteral duplication exists, the medial and distal orifice is that of the ureter draining the upper pole of the kidney.8,9 The more lateral orifice drains the lower pole of the kidney.

The embryologic origin of ureteroceles is not completely understood. Chwalla10 identified a transient two-cell–layer ureteral membrane that is present at the time the ureteric bud arises from the mesonephric duct. In theory, if this membrane were abnormally persistent, an obstructed ureteral-meatal orifice could result and lead to a ureterocele. Alternatively, it has been suggested that the distal ureteral segment may be affected by the same force that causes the expansion of the urogenital sinus to form the bladder, thus leading to the development of a ureterocele.11–13

**Figure 2.** (A) Ureteral ectopia in the male. Possible sites of an ectopic ureter are above the external sphincter. (B) Ureteral ectopia in the female. An ectopic ureter may be located beyond the continence mechanism and produce incontinence. (Adapted with permission from Johnson JH. Problems in the diagnosis and management of ectopic ureters and ureteroceles. In: Johnston JH, Scholtmeijer RJ, editors. Problems in pediatric urology. Amsterdam (NL): Excerpta Medica; 1972:57.)

**Figure 3.** (A) Complete ureteral duplication formed by two ureteric buds originating from a normal site on the mesonephric duct. (B) One of the two ureteric buds has a lower-than-normal origin, resulting in complete duplication with ureteral reflux into the ureter that, during migration, was carried most cranially and laterally (ie, the lower pole ureter). (C) One of the two ureteric buds has a higher-than-normal origin, resulting in complete duplication with an ectopic upper pole ureter. (Adapted with permission from Snyder HM 3rd. Anomalies of the ureter. In: Gillenwater JY, Howards SS, Grayhack JT, Duckett JW, editors. Adult and pediatric urology. 3rd ed. St. Louis: Mosby; 1996:2201.)

**INTRAVESICAL URETEROCELE**

**CASE PRESENTATION**

A 30-year-old pregnant woman is referred to a pediatric urologist by her obstetrician for evaluation of her 26-week-old fetus after routine fetal ultrasonography reveals dilation of the renal pelvis and a “circle in the bladder.” The woman has no significant past medical, family, or surgical history. Evaluation begins with repeat fetal ultrasonography.

- **What information is most important to obtain on repeat fetal ultrasonography?**

The amount of amniotic fluid is a critical piece of information at this time. If it is within the normal range (normal amniotic fluid index, 5 to 24 cm; normal single deepest pocket of amniotic fluid, 2 to 8 cm), no fetal
intervention is considered necessary. However, if the fetus has oligohydramnios or anhydramnios, the physician may want to consider an intervention to improve urinary drainage. Fetal interventions may involve placement of a vesicoamniotic shunt if there is bladder outlet obstruction (ie, posterior urethral valves) or puncture of a ureterocele draining a solitary kidney or one that obstructs the bladder outlet in a fetus with two kidneys. Alternative interventions might also be chosen and, depending on the week of gestation and presence of other anomalies, would include amnioinfusions, early delivery, or doing nothing. Prior to intervention, consideration should also be given to any other fetal abnormalities that may have been detected.

**DIAGNOSIS OF CASE PATIENT**

Repeat fetal ultrasonography reveals a duplication of the right kidney with dilation of the right upper pole and right upper pole ureter. The left kidney is within normal limits. A ureterocele is clearly identified as an echogenic ring or “circle” within the bladder (Figures 4–6). The amount of amniotic fluid is estimated to be normal, as is the rest of the fetal anatomy.

- How common are ureteroceles, and how are they typically detected?

**INCIDENCE AND ASSOCIATED ANOMALIES**

The incidence of ureteroceles ranges from 1 in 500 to 1 in 4000 live births per year.\textsuperscript{14,15} Ureteroceles are seen most commonly in whites and occur four to seven times more frequently in females than in males.\textsuperscript{16} Some series suggest a slight left-sided predominance. Approximately 10% of ureteroceles are bilateral,\textsuperscript{17} 60% to 80% are ectopic,\textsuperscript{3,18,19} and approximately 80% are associated with the upper pole ureter of a duplex kidney.\textsuperscript{20} The upper pole frequently displays renal dysplasia.\textsuperscript{21} Single-system ectopic ureteroceles are unusual and occur most frequently in males; they may be associated with cardiac and genital anomalies.\textsuperscript{22} Associated renal anomalies of fusion and ectopia are frequently detected.

Renal tissue associated with ectopic ureters or ureters joining ureteroceles often is dysplastic or hypoplastic. One theory suggests that the metanephric ridge is made up of blastema with variable potential for the formation of normal renal tissue.\textsuperscript{23} On either side of the center of the metanephric ridge, the blastema may have a decreased

![Figure 4](image-url) Fetal sonogram showing an echogenic “circle” from a ureterocele wall within the fetal bladder.

![Figure 5](image-url) Sonogram of a kidney associated with an upper pole ureterocele showing upper pole hydronephrosis.

![Figure 6](image-url) Pelvic sonogram showing hydroureter extending to a ureterocele within the bladder.
potential to form normal renal tissue. If a ureteric bud is located above or below the normal point of origin, it may induce the formation of renal tissue with an increased propensity for dysplasia or hypoplasia. Alternatively, a poorly draining ureter may result in dysplastic or hypoplastic renal tissue secondary to the effects of obstruction during nephrogenesis.

**CLINICAL PRESENTATION**

Currently, the most frequent means by which ureteroceles are detected is by identification of hydronephrosis on fetal ultrasonography. The sensitivity of prenatal ultrasonography for the detection of hydronephrosis increases with gestation. After birth, ureteroceles are most commonly identified in association with UTI.24–27 Also, an infant may present with a palpable abdominal mass from a distended bladder or obstructed kidney and ureter. Prolapse of a ureterocele may occasion ally cause bladder outlet obstruction, which is the most common urethral obstruction in girls.28–30 A large ureterocele associated with an abnormally lax bladder neck may lead to incontinence.31,32

• What postnatal studies should be recommended?

**EVALUATION OF THE PATIENT WITH URETEROCELE**

The standard radiographic evaluation for fetal hydronephrosis requires ultrasonography, a voiding cystourethrogram (VCUG), and a nuclear renal scan. Ultrasonography should be performed or repeated after several days of life, because the transient oliguria after birth may result in an artificial decrease in hydronephrosis. Ultrasonography may show a well-defined cystic intravesical mass along the posterior bladder wall. Because many ureteroceles are compressible with bladder filling, observing the bladder when it is very full may cause one to miss the mucosal irregularity of the bladder base. The dilated ureter identified on ultrasonography behind the bladder may also be confused with an ectopic ureter or primary obstructive megaureter.

A VCUG is required in the evaluation of all ureteroceles. With duplex system ureteroceles, reflux occurs in the ipsilateral lower pole ureter in approximately 50% of cases.20,26,33 In approximately 25% of cases of ureterocele there is contralateral reflux, and in approximately 10% there is reflux into the ureterocele itself.33,34 Reflux into a single system ureterocele is less frequent but can occur.33,35 Evaluation of the early filling films is critical to identify the filling defect caused by the ureterocele, because some ureteroceles will be compressed with filling, making detection difficult. If detrusor support behind the ureterocele is poor, the ureterocele may prolapse through the detrusor with voiding and mimic a bladder diverticulum (Figure 7).36,37 Prolapse may occur either into the dilated ureter associated with the ureterocele or through the hiatus paraureterally.38

A nuclear renal scan also is performed in the postnatal evaluation of fetal hydronephrosis. This test is useful in determining renal function and in detecting a urinary obstruction. In the case of a duplicated system with a ureterocele, the upper ureter subserves approximately one third of the function of the affected kidney. This suggests that, at best, the upper pole unit contributes only approximately 15% of overall renal function. In reality, this contribution often is much less and, therefore, is not often a major consideration when determining the best treatment approach.

Serum creatinine concentration often is measured to evaluate overall renal function in an infant with a ureterocele. It must be remembered, however, that the creatinine in the first day of life will reflect the maternal level; a more accurate assessment of the infant’s renal function will be obtained by waiting several days before checking the serum creatinine concentration. If UTI is suspected, urinalysis and urine culture should be performed. An infant with hydronephrosis, and any child

![Figure 7. Cystograms showing a large intravesical ureterocele (A), which prolapses through the ureteral hiatus (B, C), with subsequent reflux into the more proximal ureter (D). (Reprinted with permission from Cooper CS, Snyder HM 3rd. Ureteral duplication, ectopy, and ureteroceles. In: Gearhart JP, Rink RC, Mouriquand PD, editors. Pediatric urology. Philadelphia: WB Saunders; 2001:437.)](image-url)
with reflux, may benefit from antibiotic prophylaxis. Typically, amoxicillin is safe for use in the first several months of life.

- What treatment options are available for ureteroceles?
- What factors should be considered when choosing a treatment?

**TREATMENT GOALS AND OPTIONS**

The goals of ureterocele treatment are control of infection, protection of normal ipsilateral and contralateral units, preservation of renal function, facilitation of subsequent reconstructive procedures, and maintenance of continence. Several factors help determine which treatment approach is optimal for the child with a ureterocele; no one approach is appropriate in all cases. Because the natural history of asymptomatic ureteroceles is unknown, the effect of any treatment options on asymptomatic neonatal ureteroceles remains difficult to determine.

Treatment options include cystoscopy with endoscopic puncture or incision of the ureterocele and open surgical techniques. The open techniques include heminephrectomy with or without a distal ureterectomy, ureteropyelostomy, ureteroureterostomy, and ureteral reimplantation. In some patients, a combination of treatments is required. A bladder neck reconstruction may also be required to prevent stress incontinence in a patient with a dilated, poorly developed bladder neck.

**Cystoscopy with Ureterocele Puncture**

Cystoscopic detection of ureteroceles can be confusing. A small ureterocele may not be apparent until a peristaltic wave or flank compression causes it to fill (Figure 8). With very large ureteroceles, identification of any ureteral orifice in the bladder may be impossible. A compressible ureterocele may resemble only a minor mucosal fold with bladder filling. A ureterocele that prolapses may be misdiagnosed as a bladder diverticulum.

Monfort et al and Blyth et al were the first to suggest ureterocele decompression via a small puncture placed low on the ureterocele to preserve a flap valve of the collapsed ureterocele and, thus, prevent reflux. Because many ureteroceles are compressible with bladder filling, it often helps to keep the bladder relatively empty and massage the flank to distend the ureterocele. A 3 French Bugbee electrode with the cutting current at a high enough level to ensure a clean puncture is used. With a very large ureterocele, it may be necessary to look anteriorly and laterally with the cystoscope to identify the bladder neck. The bladder neck can then be followed around from this point to identify a location on the ureterocele that is within the bladder.

A low incision or puncture on the front wall of the ureterocele is made just above the bladder neck; however, if the puncture is too low, it may potentially be below the level of the ureterocele floor and may not decompress the ureterocele. A very small 3 French hole is adequate in most cases, as the thermal injury of the incision will lead to a further enlargement in the hole. If the ureterocele appears to be very thick walled, a slightly larger incision may be appropriate.
Heminephrectomy

For an upper pole partial nephrectomy, a transverse flank incision below the tip of the 12th rib directed posteriorly to the edge of the erector spinae muscles permits mobilization of the kidney. The ureter to the upper pole usually runs posterior to the renal vessels, and no attempt is made to dissect out the hilar vessels. The upper pole of the kidney usually can be mobilized and rotated up into the incision. After identification and division of the ureter to the upper pole, gentle traction guides dissection along the upper pole ureter to the upper pole parenchyma. If dissection is kept close to the upper pole collecting system, vessels running to this unit can be clearly identified and divided.

As the vessels are tied, a line demarcating the devitalized upper pole occurs. Often the division between the upper and lower poles is apparent prior to devascularization by a cleft in the parenchyma. The upper pole capsule is incised relatively high on the upper pole and may be stripped back for later use in closure. The parenchymal incision is made just on the upper pole side of the line of demarcation from the lower renal unit following the division of the upper pole vessels. The upper pole can then be excised. The upper pole ureter can then be dissected down to below the level of the iliac crest, where it enters a common sheath with the lower pole ureter. The upper pole ureter usually is tortuous and frequently appears to wrap around the lower pole ureter. Dissection kept immediately on the wall of the upper pole ureter prevents injury to the lower pole ureter. A large amount of the tortuous ureter usually can be excised with this technique through a small incision (Figure 9). If no reflux into the ureteroceles is noted, the stump of the excised upper pole ureter is left open. If reflux into the ureteroceles is noted, the upper pole ureter should be ligated following aspiration of urine from the system. Drains are left in the area of the ureteral stump and in the area of the removed upper pole.

Pretreatment Considerations

Patient age is an important consideration. If the ureterocele is detected antenatally, endoscopic puncture has the advantage in the newborn of providing a simple and direct decompression of the obstructive uropathy. An infant may tolerate a short endoscopic procedure better than a more complex upper pole partial nephrectomy. If open excision of the ureterocele is later required, it is facilitated by previous endoscopic decompression. Because bladder neck surgery causes significant postoperative discomfort after the toilet-training stage of childhood development, treatment is best accomplished before this stage. For ureterocele treatment after toilet training, a simplified approach with an upper pole partial nephrectomy avoids the need for bladder surgery if there is no associated reflux.

Another consideration is whether the kidney is a single or duplex system. In a single-system ureterocele, a primary endoscopic approach usually is appropriate, after which future ureteral reimplantation is facilitated by having a smaller decompressed ureter. When the renal unit is a duplex system, the decision is more complex, as more treatment options are available.

The degree of ureteral dilation also is important. If the ureter subserving the ureterocele is massively dilated, attempts at reimplantation have a greater complication rate. Occasionally, the associated ureter is too dilated to permit reimplantation with a 5:1 submucosal tunnel length to ureteral diameter width ratio. In such cases, the ureter requires plication or tailoring. If there is no reflux driving surgery at the bladder level and if upper pole function justifies salvage, a renal level ureteroureterostomy or ureteropyelostomy may be used effectively.

Both the endoscopic and open surgical techniques vary according to whether the ureterocele is intravesical or extravasical. In one study of extended follow-up of patients who underwent endoscopic incision, a second operation was required in 61% of patients with an extravasical ureterocele and only 18% of patients with an intravesical ureterocele. Reflux is the major factor driving the second operation and occurs more frequently with extravasical ureteroceles. The high percentage of second operations required in children with extravasical ureteroceles managed with initial endoscopic treatment is well recognized. Prior endoscopic decompression, however, should permit definitive treatment of the ureterocele via one open incision made at the bladder level. The decompressed ureter serving a ureterocele can be reimplanted with results approaching those seen for reimplantation of ureters without a ureterocele.

Associated ureteral reflux appears to best predict the need for open surgery and emphasizes the importance

Figure 9. The redundant excised ectopic upper pole ureter is shown alongside the infant, following an upper pole heminephrectomy through a 2.5-cm incision.
If the ureterocele is associated with high-grade reflux (see page 16 of this manual for discussion of grading of ureteral reflux), a primary endoscopic incision facilitates subsequent surgery at the bladder level, if necessary, by decompression of the ureterocele.\(^2\) If ureteral reflux is absent, a reasonable treatment option for an ectopic ureterocele would be a simplified approach via an upper pole partial nephrectomy. In a review by Husmann et al,\(^3\) if less than grade III reflux was present in only one ureter, 60% of patients did not require further surgery; in contrast, a higher grade reflux into one or more renal moieties almost invariably led to further surgery (96% of patients).

- What follow-up studies are needed after ureterocele treatment?

**FOLLOW-UP STUDIES**

Follow-up after an endoscopic incision or puncture involves renal and bladder ultrasonography at 1 month after the procedure. Although residual hydrourerteronephrosis often persists, a diminution of some degree indicates that decompression has been achieved. The mucosal folds of the ureterocele often remain detectable on a postpuncture sonogram. Subsequent imaging of the upper tracts and a VCUG 6 months following the procedure help direct further treatment.

Follow-up after a heminephrectomy also requires renal ultrasonography or a nuclear renal scan to assess the remaining lower pole to ensure that no injury or obstruction has occurred. A VCUG may also be warranted to check for reflux, which may occur following the heminephrectomy.

**TREATMENT AND OUTCOME OF CASE PATIENT**

Following birth at term, the affected infant undergoes renal ultrasonography, which reveals hydrourerteronephrosis of the right upper pole draining into a ureterocele. A VCUG is obtained, which confirms an intravesical ureterocele (Figure 10). Cystoscopy and endoscopic puncture of the ureterocele are performed on the seventh day of life (Figure 11). Repeat ultrasonography 4 weeks later reveals decompression of the upper pole, and a VCUG reveals no new onset of reflux.

**EXTRAVESICAL URETEROCELE**

**CASE PRESENTATION**

A 22-month-old girl presents with left flank pain, fever, and a UTI consistent with left pyelonephritis. Subsequent ultrasonographic evaluation reveals a duplication of the left kidney with dilation of the upper pole ureter associated with a ureterocele.

- How common are duplicated ureters?

**URETERAL DUPLICATION**

**Incidence and Associated Anomalies**

Many people with ureteral duplication are asymptomatic. In an autopsy population, ureteral duplication occurs in 1 in 125 patients, or 0.8%, and constitutes the most frequent ureteral anomaly.\(^1\) In clinical series of
patients with urinary symptoms, the incidence of duplication is higher (ie, 2% to 4%). Duplication affects the right and left kidneys equally. Bilateral duplication occurs in about 40% of cases. Twice as many females as males are affected by ureteral duplication. UTI is the most common associated finding.

It is thought that ureteral duplication may be transmitted as an autosomal dominant trait with incomplete penetrance. When an index child with a duplication is identified in a family, the frequency of a sibling being similarly affected rises to 1 in 8 or 9. The incidence of childhood UTIs is increased with duplications, as might be expected with the associated increased incidence of reflux or obstruction. Other urinary tract anomalies associated with ureteral duplication include scarring, hydronephrosis, or both. Histologically, renal hypoplasia or dysplasia and pyelonephritic scarring have an increased incidence in patients with ureteral duplications.

**What further evaluation is required for this patient?**

A VCUG is a critical component in this child’s evaluation. It will be important to note if the ureterocele is intravesical or extravesical, because this affects the chance of an endoscopic approach serving as definitive therapy. As previously noted, 18% of patients with an intravesical ureterocele require a second operation following endoscopic incision versus 64% of patients with an extravesical ureterocele. Reflux constitutes the major factor driving these second operations and occurs much more frequently with extravesical ureteroceles.

**DIAGNOSIS OF CASE PATIENT**

A VCUG is obtained and reveals reflux into the lower pole ureter on the left and an extravesical ureterocele (Figure 12). The patient is placed on antibiotic prophylaxis. A subsequent nuclear renal scan is obtained and demonstrates a relative function equal to 42% on the left and 58% on the right. No function is noted in the left upper pole (Figure 13).

**What treatment options are available for extravesical ureterocele?**

**TREATMENT OPTIONS**

At this time three standard treatment options exist, each with advantages and disadvantages that must be presented to the parents while obtaining their informed consent. Although other reconstructive options may be considered, the three basic options include an upper pole heminephrectomy, a common sheath ureteral reimplant, or an endoscopic puncture of the ureterocele. Both heminephrectomy and endoscopic puncture of the ureterocele avoid a relatively symptomatic operation on the bladder, but neither operation will correct the lower pole ureteral reflux. If one of these operations is performed, the child could be maintained on prophylactic antibiotics, with the hope that the reflux would resolve as the child grows. The chance of reflux resolution is diminished in this situation because of the poor detrusor support in the region of the ureterocele, and at some point the child will likely require a ureteral reimplant. Endoscopic puncture has the advantage of being a relatively simple outpatient procedure compared with heminephrectomy.
A common sheath reimplant has the advantage of correcting both the reflux and the obstructive ureterocele in one operation through one incision. At the same time, a bladder neck reconstruction may be required if the ureterocele has distorted the internal sphincter at the bladder neck. The reimplant may also require ureteral tapering of the dilated upper pole ureter to achieve an adequate submucosal tunnel length to diameter of ureter ratio to prevent reflux.

**TREATMENT OF CASE PATIENT**

The parents elect for their child to undergo an endoscopic puncture, which is performed and confirms an extravesical ureterocele. The ureterocele is noted to be thick walled at the time of the puncture. Follow-up ultrasonography reveals persistence of the hydronephrosis and ureterocele. The parents are presented again with the three main treatment options, and this time they choose a common sheath reimplant with excision of the ectopic ureterocele. At the time of surgery, a bladder neck reconstruction is also performed.

- How is an extravesical ureterocele completely excised?

The anterior mucosa of the ureterocele within the bladder is easily unroofed; however, this becomes more difficult with an extravesical ureterocele that extends down into the urethra. The surgeon must ensure that a lip of the distal anterior wall of the ureterocele does not persist and create an obstructive flap. One simple way to address this is to take a lacrimal duct probe and bend the end of it into a hook. By passing a catheter antegrade through the bladder and urethra, the hooked end of the probe may be inserted into the eye of the catheter. The lacrimal duct probe can then safely be drawn into the bladder through the urethra by pulling back on the catheter. After disengaging the lacrimal duct probe from the catheter, it is oriented with the hook facing in a posterior direction. By withdrawing the probe through the urethra, the distal lip of the anterior wall of the ureterocele may be engaged by the hook and drawn to the external urethral meatus, where it can simply be cut, helping to ensure that no obstruction within the urethra has been created.

**OUTCOME OF CASE PATIENT**

The patient undergoes follow-up ultrasonography 6 weeks after the operation, which reveals a decrease in the amount of upper pole hydronephrosis. A VCUG obtained 6 weeks later reveals no reflux and a patent bladder neck. Antibiotic prophylaxis is discontinued at this time.

**ECTOPIC URETER**

**CASE PRESENTATION**

A 3-day-old boy is referred for a VCUG to evaluate right upper pole hydronephrosis that had been identified on prenatal ultrasonography. The amniotic fluid volume had been normal. The child was born at term and is otherwise healthy.

A urologist is called for a consultation. The urologist’s physical examination of the patient is unremarkable. Ultrasonography performed on the third day of life reveals a dilated upper pole of the right kidney and a severely dilated ureter behind the bladder; no evidence of a ureterocele is noted within the bladder (Figure 14). The patient’s serum creatinine level and results on urinalysis are normal. He is placed on prophylactic antibiotics and sent for a VCUG.

- What instructions should be given to the radiologist when requesting a VCUG?

The radiologist should be instructed to perform a cyclic VCUG. This requires repeated filling and voiding. The ectopic ureter frequently traverses a portion of the musculature of the bladder neck and is obstructed when the bladder neck is closed during filling. During voiding, this obstruction is relieved and the retained ureteral urine can drain. Ureteral reflux occurs in many ectopic ureters, producing the paradox of both reflux and obstruction. By having the bladder neck repeatedly open, the cyclic VCUG of Lebowitz and Wyly provides an opportunity for the obstructed ectopic ureter to drain before contrast is voided and thus increases the likelihood that the contrast will reflux into the ectopic system. This method frequently identifies reflux that would not be detected with a single VCUG.

**EVALUATION OF CASE PATIENT**

The patient’s cyclic VCUG reveals reflux into a severely dilated right upper pole ureter entering distal to the bladder neck (Figure 15). A nuclear renal scan is obtained, which reveals a rim of functioning parenchyma in the upper pole (Figure 16).

- How common are ectopic ureters, and how do they present?

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INCIDENCE AND ASSOCIATED ANOMALIES

The incidence of ectopic ureters is estimated at 1 in 1900 live births per year. Ectopic ureters are much more common in females, with only approximately 15% of ectopic ureters occurring in males. In 80% of cases, the ectopic ureter is from the upper pole of a duplicated renal system. When the ectopic ureter is part of a duplex system, the contralateral system is duplicated in approximately 80% of cases, and 21% of these will have contralateral ectopy as well. The incidence of an ectopic ureter draining a single system is more common in males than females. Ten percent of ectopic ureters are bilateral.

Although renal hypoplasia or dysplasia is the most frequently encountered anomaly associated with an ectopic ureter, the degree of ectopia often correlates with the degree of renal abnormality. This correlation is more apparent for duplex systems with ectopy than for single systems with ectopy. Severe ectopia with an orifice in the genital system is almost always associated with poorly functioning renal tissue.

Ureteral Ectopia in Females

In females, approximately one third of ectopic ureters open at the level of the bladder neck or slightly more distally in the upper urethra. The higher the orifice, the less likely the chance for associated urinary incontinence; however, obstruction is more common with higher insertions. One third of ectopic ureters terminate in the area of the vaginal vestibule, adjacent to the urethral orifice. In approximately 25% of cases, the orifice opens into the proximal vagina. More rarely (less
than 5% of cases), an ectopic ureter ends at a higher site on Gartner’s duct, with an opening at the level of the cervix or even the uterus.

**Ureteral Ectopia in Males**

In the male, an ectopic ureter may end in the bladder neck, prostatic urethra (down to the level of the verumontanum), epididymis, seminal vesicle, or vas deferens. Half end in the posterior urethra, and more than one third join the seminal vesicle.

**CLINICAL PRESENTATION**

Ectopic ureters often are identified subsequent to the detection of hydronephrosis on prenatal ultrasound. They also may present after birth in association with UTI.

In females, ectopic ureters can terminate at a level distal to the external sphincter and cause incontinence. Approximately half of females with ectopic ureters present with a classic history of continuous dribbling incontinence despite what appears to be a normal voiding pattern. Occasionally, an affected female presents with a persistent foul-smelling vaginal discharge. If the ureter ends in a Gartner’s duct cyst, the patient may present with a mass on the anterior vaginal wall. Any infant may present with an abdominal mass from a severely obstructed ectopic ureter.

In males, ectopic ureters may present as epididymitis and should be considered in the prepubertal male with such a diagnosis. In some males, a genitally ectopic ureter does not become symptomatic until the onset of sexual activity. At that time, the male may present with epididymitis, prostatitis, seminal vesiculitis, or occasionally an infected seminal vesicle cyst. A dilated single-system ectopic ureter that inserts into the prostatic urethra may elevate the bladder neck, causing outlet obstruction.

- **How are ectopic ureters diagnosed?**

**DIAGNOSIS**

Without reflux, the diagnosis of an ectopic ureter in the female may be very difficult. Physical examination with close observation of the area around the urethral meatus and distal vagina may reveal a recurring drop of liquid over a very small opening that can be probed and retrogradely injected to confirm the presence of an ectopic ureter. With ectopy into the external genitalia, there is likely to be poor function and nonvisualization of the associated renal unit by intravenous pyelography (IVP) or nuclear renal scan. Ultrasonography may be useful if a dilated ectopic ureter exists behind the bladder. If hydroureteronephrosis is minimal, the diagnosis may depend on recognizing the absence of an upper pole calyx. Computed tomography of the kidney or magnetic resonance imaging delineating the fluid-filled ureter may be useful. Vaginograms may show reflux into a vaginal ectopic ureter. Cystoscopy and vaginoscopy may also show the ectopic orifice.

In the male, an ectopic ureter entering the genital tract often is single and drains a nonfunctioning renal unit. Ultrasonography may show hydroureteronephrosis. The greatest diagnostic difficulties occur in duplex kidneys with a tiny upper pole draining into a minimally obstructed ectopic ureter with little dilation. Most ureters ectopic to the urethra or bladder neck will reflux on cyclic VCUG. Cystoscopy and examination under anesthesia may establish the diagnosis. A mass may be felt in the area of the seminal vesicle, or elevation of the floor of the bladder (“pseudoureterocele”) may be noted at cystoscopy. The ectopic ureteral orifice may be seen at the bladder neck or urethra, or there may be an enlarged ejaculatory duct, permitting a retrograde study.

- **What treatment options are available for this patient?**

**TREATMENT OPTIONS**

Reflux into an ectopic ureter is unlikely to resolve with time, in contradistinction to primary reflux into a ureter with an orifice located at the bladder level. Treatment options include an upper pole heminephrectomy with distal ureterectomy, an upper pole ureter to lower pole renal pelvis ureteropyeloplasty, a
distal ureteroureterostomy, or a temporary cutaneous ureterostomy with subsequent ureteral reimplant once the child’s bladder has become larger. There is no consensus on which treatment option is best nor the optimal timing of surgery, and these decisions should be made on an individual basis.

The problem in the case patient is one of reflux into a relatively obstructed system. A reimplant at this stage might be considered but would be technically very difficult and require ureteral tailoring. Also, this patient is at high risk for complications given the large size of the ureter and the small size of his infant bladder. In an older patient with a single-system ectopic ureter and significant renal function, a reimplant would be a reasonable option.

In females, if an ectopic ureter enters into the introitus or vagina, the entire distal ureter may not need to be removed. In these patients, the distal ureteral segment is a rare source of later problems. Removal of the ureteral stump is more likely to be needed for urinary ectopic ureters. An ectopic ureter ending in the bladder neck or urethra may have reflux of voided urine into the residual ureteral stump, causing a small amount of dribbling incontinence after micturition or infections.

• What follow-up studies are needed after treatment?

A nuclear renal scan or renal ultrasonography should be performed following the surgical treatment of an ectopic ureter. The vasculature to the lower pole of the kidney could be injured during the upper pole nephrectomy. The upper pole ureter is frequently entwined with the lower pole ureter. Unrecognized injury to the lower pole ureter may also occur and result in a ureteral stricture.

TREATMENT AND OUTCOME OF CASE PATIENT

The patient undergoes an uncomplicated right upper pole heminephroureterectomy at 3 months of age. The pathologic examination reveals severe hydronephrosis with tubular atrophy and interstitial fibrosis. At a 2-year follow-up, the child is doing well without any urologic complications.

URETERAL REFUX

CASE PRESENTATION

A 22-month-old girl is referred to a urologist by her primary pediatrician for evaluation and treatment of a second UTI. Her first infection occurred 6 months ago and was associated with urinary frequency and dysuria. The child’s parents report that she is currently running a low-grade fever and complaining of pain in her left side. The referring physician noted bacteria in her urine. The child has no other significant past medical history.

• What additional information should be obtained in the history and physical examination of this child?

CLINICAL EVALUATION OF UTI

History

In children, UTIs are strongly linked to constipation. Because most UTIs are caused by ascending infections, it is important to take a history of the child’s bowel movements. Often this information is not known by the parent or child and requires a diary. Constipation in children presents in a variety of manners, such as very large or hard bowel movements, encopresis, episodic diarrhea, or stool streaks on the child’s underpants. It is easy to understand how residual stool in undergarments—which are kept in a moist place at body temperature just outside the urethra—could predispose a child to UTI. Successful treatment of constipation has been shown to prevent UTI.

A voiding history also is critical in the evaluation of a child with a UTI. Dysfunctional voiding may predispose to UTI by stasis of urine due to incomplete bladder emptying. Also, infrequent voiding during the day may give any bacteria that enter the bladder more time to multiply and adhere to the bladder surface. Again, information regarding a child’s voiding habits may not be known initially, and the use of a voiding diary is often helpful. The parent or child should be asked questions regarding dribbling, incontinence, double voiding, urgency, “holding” maneuvers (ie, curtseying or squatting), and difficulty initiating a voluntary void.

Physical Examination

On physical examination, the blood pressure should be noted, as hypertension may reflect renal damage. Attention also should be given to the child’s spine and gluteal cleft, looking for any evidence of spina bifida occulta (eg, a sacral dimple, fat pad, hairy patch, or skin tag; asymmetry). Along this line, a neurologic examination should also be performed. The abdomen may be palpated and assessed for evidence of constipation as well as kidney enlargement. The flanks are checked for costovertebral angle tenderness and evidence of kidney enlargement. The external genitalia are inspected for any congenital abnormality including phimosis or labial adhesions.
• How is the diagnosis of a UTI made?

**DIAGNOSIS OF A UTI**

Many children are misdiagnosed with a UTI based on symptoms and a voided urine sample. It is important to determine how the diagnosis of a UTI was made. Dysuria and frequency are nonspecific symptoms of a UTI. A voided specimen or a bag collection is easily contaminated and is not reliable to make a true diagnosis of UTI.

The American Academy of Pediatrics recently developed a practice guideline for the diagnosis, treatment, and evaluation of UTI in infants and young children from 2 months to 2 years of age. The guideline recommends that any young child with an unexplained fever should be evaluated for a UTI. Infants with a UTI often present with nonspecific signs and symptoms, such as irritability, vomiting, diarrhea, and failure to thrive. Therefore, the physician must maintain a high index of suspicion to diagnose the UTI. Urine for culture should be obtained prior to starting antibiotics.

**Urine Collection and Analysis**

A seemingly reasonable way to collect urine from a child with a suspected UTI is by applying a bag to the perineum. The bag-collected urine, however, frequently is contaminated; therefore, a bag-collected urine specimen is insufficient to document the presence of a UTI. Of course, if the urinalysis results are normal, it is unlikely that the child has a UTI. A bag-collected urinalysis suggestive of a UTI requires the performance of a more invasive method of obtaining urine, with the possible exception of circumcised boys older than 1 year. Urine obtained by suprapubic bladder aspirate is the least likely to be contaminated, and that obtained by transurethral bladder catheterization is next best.

A urine culture is required for the diagnosis of a UTI. For a child who appears sufficiently ill to warrant immediate antimicrobial therapy, one of the invasive methods of obtaining urine for culture should be performed prior to starting antibiotics.

**Imaging Studies**

Imaging of the urinary tract is recommended for every febrile infant or young child following the first UTI. Imaging includes renal and bladder ultrasonography and a VCUG. Approximately 40% of children may have an abnormality detected by these studies following a febrile UTI. Renal ultrasonography may detect hydronephrosis, duplication anomalies, stones, or abnormalities of the bladder wall and should be performed at the earliest convenient time. A cystogram may be obtained by instillation of contrast with fluoroscopy or by instillation of a radionuclide. Radionuclide cystography has the advantage of decreased radiation. The contrast-voiding cystourethrogram, however, has the advantage of providing better anatomic detail, which may help detect bladder or urethral abnormalities, and is often the initial study of choice. Either method should include a voiding phase, because reflux is the most likely abnormality to be detected and may only occur with voiding. The cystogram should be obtained once the child is free of infection.

• What are the risks of a missed diagnosis of UTI in a child?
• What are the goals of initial treatment?

**RISKS OF MISSED UTI DIAGNOSIS**

Children can suffer renal damage from UTIs that are not promptly diagnosed or subsequently evaluated. The sequelae of a UTI include renal damage and hypertension. Young children appear more susceptible to renal injury from UTI than older children or adults. The highest incidence of underlying urinary tract abnormalities (eg, ureteral reflux) also occurs in this young age-group. Scars are less likely to develop in the older child and rarely occur in the adult. Prompt treatment of pyelonephritis prevents renal scar formation. Many episodes of UTI in infants may go undiagnosed or are misdiagnosed. By prompt recognition and appropriate treatment of the UTI, the physician minimizes the risk of injury to the kidneys.

**UTI TREATMENT GOALS**

The goals of UTI treatment include eliminating the infection, preventing urosepsis, and minimizing renal damage. Initial treatment with parenteral antibiotics is recommended for those children who appear toxic, dehydrated, or unable to retain oral intake. For children who do not appear as ill, initial oral antibiotics usually include amoxicillin, a sulfonamide-containing antimicrobial, or a cephalosporin. If the expected clinical response is not obtained after 2 days of antimicrobial therapy, the child should be reevaluated with another urine culture. Adequate treatment requires a 7- to 14-day course of antibiotics. However, antibiotics in prophylactic dosages should continue until the child undergoes imaging studies.

**DIAGNOSIS OF CASE PATIENT**

Urine obtained by catheterization is sent for urinalysis and culture, which confirm the presence of a UTI. Renal and bladder ultrasonography reveal some mild
left hydronephrosis without evidence of duplication anomalies, stones, or intravesical lesions. A VCUG is performed and reveals grade III left-sided ureteral reflux (Figure 18). The patient has a smooth-walled bladder that empties completely after voiding.

• What causes ureteral reflux?
• How is reflux clinically graded and managed?

URETERAL REFLUX

Reflux has both primary and secondary causes. Primary reflux is thought to be due to an abnormal ureterovesical junction, which results from a defect during embryologic development of the genitourinary tract (see discussion in “Embryology” section on page 3). Secondary reflux may occur due to elevated bladder pressures from a noncompliant neurogenic bladder or urethral obstruction (valves or voiding dysfunction). Identification and correction of the underlying cause of secondary reflux may result in the resolution of reflux. Distortion of the trigone from ureteroceles or ectopic ureters may also be associated with reflux, and these conditions must be recognized so that the child is managed appropriately.

Grades of Primary Reflux

Primary reflux is graded based on the VCUG. The more severe grades are associated with an increased incidence of renal injury. Although many grading systems have been used, the American Urological Association (AUA) developed guidelines using the International Reflux Study classification (Figure 19). Grade I reflux occurs if contrast enters a nondilated ureter. Grade II occurs if contrast makes it up to the level of the kidney. With grade III reflux, there is mild to moderate dilation of the ureter and pelvis with minimal blunting of the fornices. Grade IV occurs when the ureter is noted to be tortuous and the calyces are dilated. Grade V reflux consists of gross dilation of the ureter, pelvis, and calyces.

According to the meta-analysis performed by the AUA guidelines panel on primary reflux in children, the spontaneous resolution rates for grades I and II reflux were generally the same, regardless of age at presentation or laterality (unilateral versus bilateral). Resolution rates after 5 years were approximately 90% in children with grade I reflux and 80% in children with grade II reflux. In children with grade III reflux, an increased age at presentation and bilateral reflux decreased the probability of resolution. With unilateral grade III reflux, the resolution rates at 5 years were approximately 70% for those younger than 2, 50% for those between ages 2 and 5, and 45% for those older than 5. The resolution rates for bilateral grade III reflux in these age-groups decreased to 50%, 30%, and 12%, respectively. With unilateral grade IV reflux, the resolution rates at 5 years were approximately 60% versus resolution rates of approximately 10% with bilateral grade IV reflux.

Management of Ureteral Reflux

Antibiotic prophylaxis constitutes a major component in treating the child with ureteral reflux.81 Reflux alone, without obstruction or infection, does not appear to harm the kidneys.82,83 This fact, combined with the knowledge that ureteral reflux resolves in most children...
over time, leads to the recommendation that all children with reflux should be maintained on antibiotic prophylaxis. Commonly used antibiotics include amoxicillin for children during the first 2 months of life and then either trimethoprim-sulfamethoxazole or nitrofurantoin. The use of prophylactic antibiotics is aimed at preventing pyelonephritis and renal scarring, since sterile reflux does not cause scarring.

Multiple host factors can influence whether a child with ureteral reflux will develop UTIs and pyelonephritis. Host factors such as urothelial receptors, papillae configuration, periurethral flora, voiding patterns, constipation, and immunologic resistance and reactivity determine the likelihood of developing pyelonephritis and renal scarring. In one study, 63% of children admitted for pyelonephritis did not have ureteral reflux. Bacteria, such as P-fimbriated \textit{Escherichia coli}, adhere to specific urothelial receptors and ascend the ureter with or without reflux. The adherence of bacteria to urothelial and vaginal epithelial cellular receptors varies among individuals and is increased in patients susceptible to recurrent UTI. Host factors also vary with age, since young children have an increased incidence of UTI. Early detection and treatment of reflux may explain why screened siblings with reflux have a lower incidence of renal scars than their index siblings.

Anticholinergic medications may be useful in the child with ureteral reflux and voiding dysfunction. Oxybutynin has been shown to improve resolution of reflux, most likely by decreasing uninhibited bladder contractions and intravesical pressure. Timed voiding and correction of abnormal urinary sphincter activity also improve reflux resolution rates.

**Monitoring a Patient with Ureteral Reflux**

Assessment of renal function and scarring is important and is best detected with a nuclear renal scan. After an episode of pyelonephritis, a renal scar usually appears within 6 months on a renal scan but may not be apparent on IVP or renal ultrasonography for more than a year. Knowledge of a scarred kidney may be an important factor when deciding whether to continue antibiotic prophylaxis or proceed to surgical therapy. There is no consensus on the best method and optimal frequency of periodic upper tract imaging for a child with ureteral reflux.

Most physicians routinely monitor their pediatric patients with ureteral reflux with a radionuclide cystogram performed every 12 to 18 months. Antibiotic prophylaxis is stopped once cystography shows resolution of reflux. The physician caring for a child who fails to resolve reflux frequently determines whether the child should continue on antibiotic prophylaxis or undergo surgical treatment. Some consider failure to resolve reflux over several years an indication for surgery, and others suggest that cessation of antibiotic prophylaxis in the older child with a minor history of previous UTIs is reasonable. Surgical intervention in the latter group is reserved for those who develop a UTI while not taking antibiotics.

**When is surgical intervention indicated?**

**SURGICAL INTERVENTION**

Considerations for operative intervention include breakthrough UTIs, noncompliance with antibiotics, failure of the reflux to resolve, high-grade reflux in a child with a history of febrile UTI with no voiding dysfunction, and loss of relative renal function in a refluxing kidney. In school-aged children with high-grade bilateral reflux and no voiding dysfunction, surgical therapy is often performed. Although the long-term use of antibiotics seems innocuous, the cost, inconvenience, possible side effects, and parental concerns (eg, a desire to correct the problem quickly, fear of antibiotics, or the child’s noncompliance) may prompt surgical treatment. It is important to rule out voiding dysfunction or secondary causes of reflux (eg, neurogenic bladder, urethral valves) before considering surgical correction of reflux. A urodynamic study may be valuable in assessing bladder stability, compliance, and function in children with a suspected secondary cause of reflux.

**Techniques**

Open surgical correction of reflux is highly successful regardless of the technique used. Success rates of
over 95% are anticipated for grades I through IV.\textsuperscript{98} Surgery is successful in 80% of grade V cases. New onset of contralateral reflux after unilateral ureteral reimplantation occurs in approximately 9% of children. Fortunately, this generally resolves spontaneously. Failure of reflux to resolve following surgical correction should arouse suspicion that a secondary cause of reflux may exist.

The goal of open surgical correction is to increase the length of the submucosal ureteral tunnel. A submucosal tunnel length to ureteral diameter ratio of 5:1 is thought to reliably prevent ureteral reflux. With severely dilated ureters, a ureteral tapering or plication technique may be required to obtain this ratio. The creation of the submucosal tunnel may be performed through a variety of intravesical or extravesical techniques. The primary complication following ureteral reimplantation is ureteral obstruction requiring a subsequent operation. This complication occurs in approximately 2% of cases.\textsuperscript{99}

Endoscopic injection of Teflon\textsuperscript{®} and collagen beneath the ureteral orifice has also been used for treating ureteral reflux. Teflon\textsuperscript{®} is no longer approved for use in the United States because of fear of particle migration. Short-term success rates of approximately 60% with collagen have been reported. One concern with collagen is that its volume decreases over time, and long-term success rates will be lower. Other agents for subureteral injection are currently being investigated.

Surgical intervention is warranted in an infant with high-grade reflux and breakthrough infections. With dilated ureters and a relatively small, thin-walled bladder, a ureteral reimplant is technically difficult and associated with increased complications. A reasonable surgical option in these patients consists of a cutaneous vesicostomy. This permits the bladder to fill and empty at low pressure and reduces the incidence of symptomatic UTI. With time and growth, the ureteral dilation may decrease as well as the reflux. The vesicostomy may subsequently be taken down at the time of ureteral reimplantation.

- **Should this patient’s siblings be screened for ureteral reflux?**

There appears to be a genetic component to ureteral reflux, as 34% of the siblings of children with reflux also have reflux.\textsuperscript{98} Up to two-thirds of children of women with reflux also may have reflux.\textsuperscript{100} The early detection and treatment of reflux in screened siblings is thought to result in a lower incidence of renal scars among these children, compared with their index siblings.\textsuperscript{98}


