# CHAPTER 90
# Ureteric Duplications and Ureterocoeles

**V. T. Joseph**

## Introduction

Duplications of the ureter represent one of the most common anomalies of the urinary tract. Duplications may be complete or incomplete and can be associated with functioning renal moieties with orifices that open into the bladder. Duplications are often completely asymptomatic and often come to light only in the course of investigations for other reasons. Clinical problems are the result of obstruction, reflux, or ectopic openings, giving rise to hydronephrosis, infection, and incontinence. Antenatal diagnosis provides useful information for postnatal evaluation with a view to preserving renal function, when appropriate, or removing dysplastic components that potentially may cause infection and compromise the other elements of the renal tract.

## Embryology

The ureteric bud develops from the wolffian (mesonephric) duct between 4 to 8 weeks in utero from a point just proximal to the junction with the cloaca. It elongates in a cranial direction to reach and penetrate the metanephric intermediate mesoderm to induce the formation of the kidney. The portion of the wolffian duct distal to the ureteric bud is called the common excretory duct. The common excretory duct is absorbed into the urogenital sinus to form the trigone, fusing with its component of the opposite side. The opening of the wolffian duct ends at the utricle, and the duct gives rise to the prostate, seminal vesicles, vas deferens, and epididymis. In females, the wolffian duct eventually undergoes involution but may persist as a Gartner's duct.

## Abnormalities of Development

A large number of different types of anomalies of the ureteric bud exist. The ureteric bud may be atretic and fail to reach the metanephric mesoderm, giving rise to a multicystic dysplastic kidney. Or the ureteric bud may originate from a site other than its normal position on the mesonephric duct, giving rise to ectopia. The orifice may lie in the bladder but lateral or caudal to the normal site. The orifice may also lie outside the bladder, in the urethral, vestibular, vaginal, or vassal positions.

In males, ectopic ureters drain proximal to the urinary sphincter and hence wetting is not evident. In females, ectopic ureters discharge into the genital tract where they bypass the urinary sphincter, thus causing the classic pattern of wetting associated with normal voiding. Three circumstances associate ectopic ureters with wetting:

- The ectopic orifice is distal to the midpoint of the urethra (females only).
- The ectopic orifice is in the vagina (rare) or vestibule (common).
- The ectopic orifice is in the urethra but the hiatus where the ureter enters into the bladder wall is at the bladder neck and disrupts the sphincter, causing incontinence.

### Ureteric Duplications

Ureteric duplications occur either due to bifurcation of a single ureter or when two ureteric buds arise from the wolffian duct. Single ureteric bifurcation may result in a whole spectrum of anomalies, ranging from bifid pelvis to almost complete duplication down to the intramural part of the ureter. Complete duplication occurs when two ureteric buds arise from the mesonephric duct and meet the metanephric mesoderm at separate points, thus giving rise to duplex kidneys.

In complete duplication, the cranially positioned bud reaches the upper portion of the mesoderm and the caudally positioned bud reaches the lower portion. However, at the bladder end, the relationships become more complex, owing to the tissue migration and incorporation into the outflow tract. When the buds are close to each other, the ureteric orifices are in the bladder in the normal position. When the buds are widely separated, the orifices may be ectopic. In such situations, the lower pole ureter (LPU) inserts normally and the upper pole ureter (UPU) crosses it anteriorly in the lower third and inserts ectopically (Weigert-Meyer law). The UPU is normally dilated and tortuous and is connected to dysplastic upper moiety. The lower end of the ureter is usually stenotic, causing obstruction, or may be associated with a ureterocoele. The LPU is usually of normal calibre but may be associated with vesicoureteric reflux.

### Ureterocoeles

Ureterocoeles represent a cystic dilatation of the lower end of the ureter. The aetiology of ureterocoeles is not clear; it has been proposed that they result from the persistence of Chwalla's membrane or as a result of deficiency of the muscle in the distal ureter. They may be classified in several ways; the simplest system is to regard them as either intravesical or extravesical. In each category, additional features can be described as follows:

- *Stenotic:* The orifice is very tiny and difficult to identify.
- *Sphincteric:* The orifice is situated within the urethral sphincter zone.
- *Caecoureterocoele:* There is a caudal extension in the submucosal plane of the urethra.
- *Blind ureterocoele:* There is atrophy of the ureter distal to the ureterocoele.

## Clinical Presentation

Routine antenatal maternal ultrasonography (US) can identify most urinary tract abnormalities. Currently, two-dimensional US is employed in routine screening with detection rates as high as 88%. The 20-week anomaly scan is designed to detect major foetal abnormalities as part of the foetal anatomical survey. US is highly observer-dependent, so the success of the screening program depends as much on the training of the professionals as on the quality of the equipment.

In ureteric duplications, the most common US findings are those of hydronephrosis and hydroureter. These findings are not specific and require postnatal evaluation to define the anomaly. Bladder outlet obstruction due to ureterocoeles presents with signs similar to posterior urethral valves and requires postnatal investigation with micturating cystourethrography to establish the cause.

Postnatal presentations are related to obstruction, infection, and urinary incontinence. Specifically:

- Acute obstruction occurs at the bladder outflow tract and is caused by ectopic ureterocoeles extending into the urethra. This represents the most common cause of bladder outlet obstruction in girls and is the second most common cause in boys after posterior urethral valves. The clinical presentation is that of a distended, palpable bladder with failure to pass urine.
- A prolapsing ureterocoele presents as a purplish mass protruding from the urethral orifice, showing congestion and oedema. The urethral opening can usually be identified and catheterised.
- Obstruction can also lead to hydronephrosis, which in severe cases presents with an abdominal mass.
- Urinary tract infection (UTI) occurs in 50% of cases. Patients present with flank pain and fever, indicating a pyelonephritis. This may also be associated with failure to thrive and nonspecific abdominal pain.
- Acute epididymo-orchitis in male infants may be the presentation of an ectopic ureter. Clinically, the child presents with an acute scrotum which is indistinguishable from torsion, and the diagnosis is made only on exploration.
- Incontinence in girls may be a manifestation of ectopic ureters opening outside the bladder. The typical history is that of voiding normally but wetting between voids. The degree of wetting may vary from slight dampness to significant leaks. Many cases are diagnosed as bladder dysfunction, and the true condition may remain undetected until later years, particularly after pregnancy.

## Diagnosis

### Ultrasound

The investigation of choice in the initial evaluation of ureteric duplications and ureterocoeles is the US scan, which includes complete evaluation of the kidneys, ureters, and bladder. The postnatal scan should be delayed for up to a week in the newborn because the relative state of dehydration at birth may mask some urinary tract abnormalities. However, in urgent situations, the scans should be conducted as soon as possible. Further investigations then can be used to follow up the findings. The kidney may show hydronephrosis of varying severity; in a duplex kidney, this may be confined to one moiety, thus making the diagnosis easy. In nondilated duplex systems, US scans do not show a clear demarcation of the two moieties, and thus other imaging techniques may be required.

Massive dilatation of the ureter is easily identified on US; it may be an obstructed ureter usually draining the upper moiety or a refluxing ureter to the lower moiety. Ectopic ureters can sometimes be identified passing behind and below the bladder, but it is not possible to determine the location of its opening with ultrasound.

US is the best modality for identifying ureterocoeles. These are visualised as cystic protrusions into the bladder; sometimes, urinary jets may be seen from their orifices.

### Micturating Cystourethrogram

A micturating cystourethrogram (MCUG) involves filling the bladder with x-ray contrast media via a catheter under fluoroscopy. It is carried out in all cases where US has demonstrated a dilated upper tract or the presence of a ureterocoele (Figures 90.1 and 90.2). The MCUG may show reflux into the lower moiety of a duplex system and thus delineate the dilated ureter. However, where US shows a dilated ureter and MCUG shows either no reflux or reflux with a normal calibre ureter, the interpretation is that the dilated ureter is associated with the upper moiety. In this situation, further investigations are necessary.

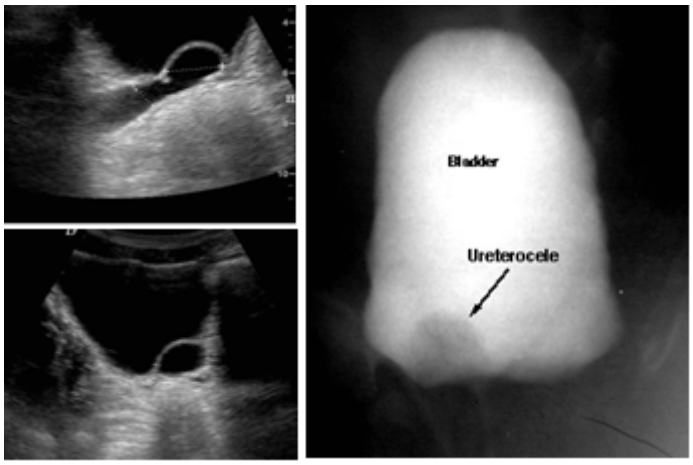


*Figure 90.1: US scan (left) and MCUG (right), showing ureterocoele.*

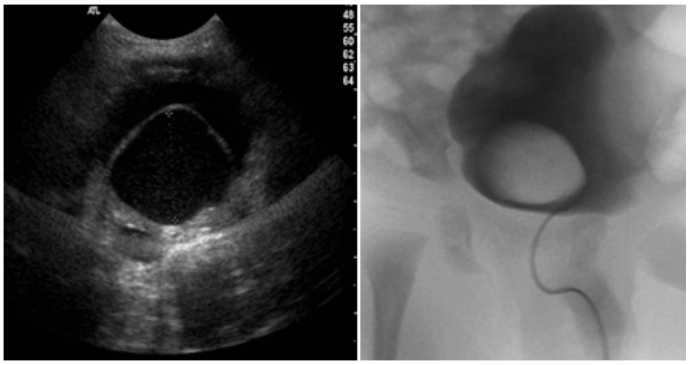

*Figure 90.2: Large ureterocoele causing bladder outlet obstruction.*

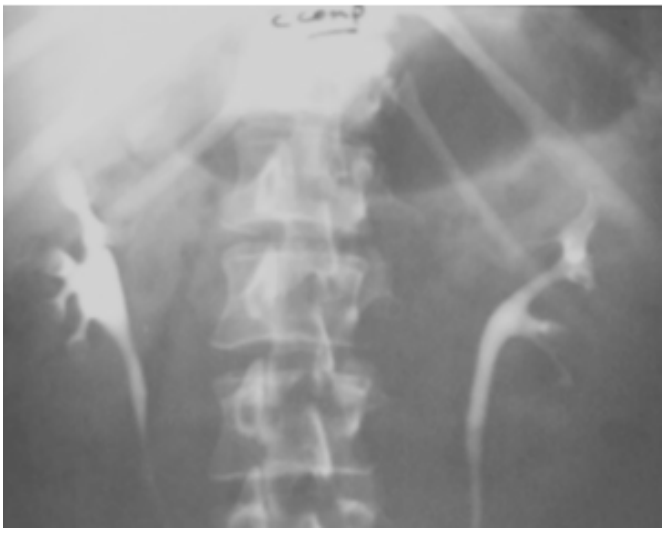

*Figure 90.3: Duplex system shown by IVU.*

### Intravenous Urography and Magnetic Resonance Urography

Until recently, intravenous urography (IVU) was the only radiological method of demonstrating the renal elements in duplication anomalies (Figure 90.3). Because the upper moiety is frequently dysplastic with poor function it was not possible to visualise it with contrast. Instead, radiological signs such as the absent upper calyx, displacement of the lower moiety, and indentation of the lower moiety ureter were used to infer the presence of a dilated upper moiety.

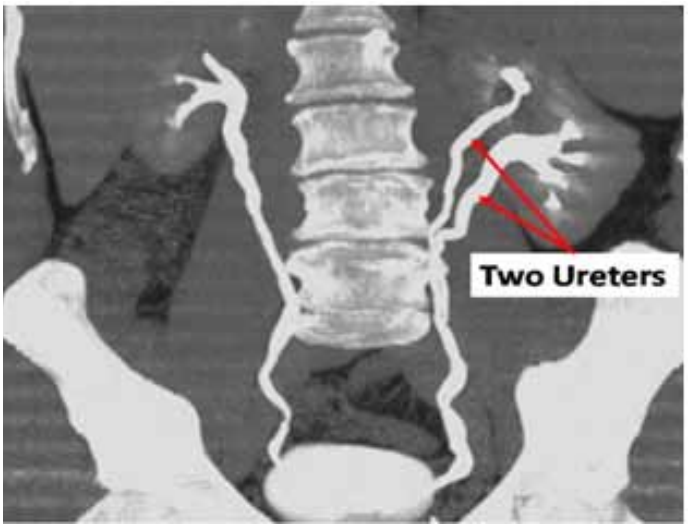


*Figure 90.4: Bifid system on MRU.*

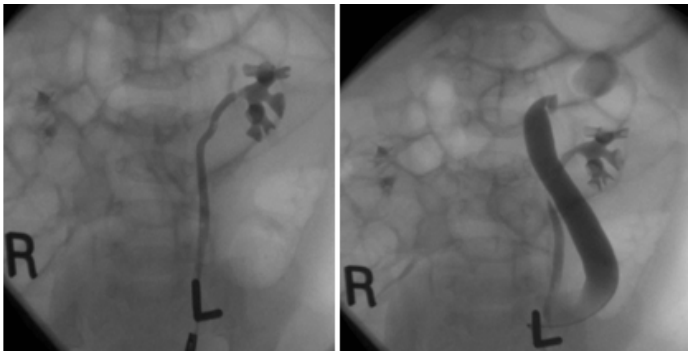


*Figure 90.5: Retrograde pyelogram showing duplex system.*

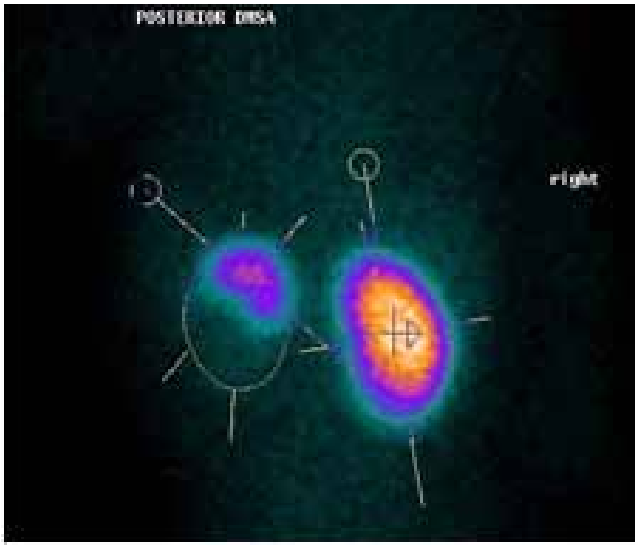


*Figure 90.6: DMSA scan showing nonfunctioning lower moeity on the left.*

The introduction of magnetic resonance urography (MRU; Figure 90.4) has made a dramatic difference in imaging the renal tract. It is now possible to sequentially image the entire upper tract and follow the ureter all the way down to the bladder and beyond. Even ectopic ureters can be traced down almost to where they actually open, thus providing vital information for the surgeon.

### Cystoscopy

Cystoscopy is carried out in all cases of ueterocoeles and duplications presenting clinically with symptoms relating to obstruction, infection, and incontinence. A single ureteric orifice on the side of the duplication indicates the presence of an ectopic ureter. This can be further confirmed by doing a retrograde study (Figure 90.5) via the single opening to demonstrate that it is associated with one ureter going only to the lower pole. A careful search of the bladder neck area will usually reveal the ectopic ureteric opening. Careful examination of the vulva/vestibule area in the female may occasionally show the ectopic opening and allow contrast examination to be performed.

### Dimercaptosuccinic Acid Scan

The dimercaptosuccinic acid (DMSA) scan (Figure 90.6) is useful because the isotope is taken up only by functioning renal tissue. Thus, even a small dysplastic upper moiety may be visualised and the presence of renal scarring can also be assessed. This information is useful when a decision has to be made regarding nephroureterectomy in duplex systems.

## Management

The principles of management in duplications of the upper urinary tract are based on a thorough evaluation of the symptoms and pathophysiological changes in the systems.

Each case has to be considered individually. Most duplications of the upper urinary tract are incomplete, and the majority are asymptomatic, being discovered only in the course of investigation for some other complaint. These cases do not need any further intervention. Complete duplications are often associated with clinical problems that require active intervention. The following guidelines are just a framework for detailed assessments.

### Vesicoureteric Reflux

Urinary tract infections are often associated with vesicoureteric reflux into the lower moiety of a duplex system. (Vesicoureteric reflux is described in more detail in Chapter 91.) Rarely, reflux can also occur into the upper or both moieties. The clinical presentation is that of pyelonephritis with fever, loin pain and tenderness, dysuria, and a culture positive urine. Children with these symptoms can be very ill and need hospitalisation for administration of intravenous antibiotics and fluid resuscitation. A broad-spectrum antibiotic such as coamoxiclav is combined with one providing gram-negative cover, such as gentamycin, and treatment must be given until there is complete resolution of the UTI. The reflux needs to be treated, which can be done by cystoscopy—an on-table cystogram with endoscopic submucosal injection of Deflux®. If this technique fails to stop reflux, then reimplantation of the ureter will have to be done. This often means reimplantation of both ureters because they can be very close and even share a common sheath. In this situation, careful evaluation of the upper moiety function should be made preoperatively to decide whether preservation of this renal element is justifiable.

### Obstruction

In some cases where infection occurs in an obstructed system, it is necessary to provide urgent drainage in addition to antibiotic therapy. This can be done by US-assisted percutaneous drainage if facilities are available, or by open nephrostomy. Following this, the anatomy and functional state of the affected moiety must be clearly assessed and a decision made as to the further surgical procedure that would be required. If sufficient renal function is present, the obstruction is dealt with by the appropriate method. This usually involves reimplantation of the lower end of the ureter into the bladder because most often that is the site of the obstruction. Difficulty may arise when a dilated, nonobstructed system is present. In this case, conservative treatment

with stenting by using a double-J stent and prophylactic antibiotics would be an option. The stent is left in for a prolonged period of 3 to 6 months, and periodic assessments need to be made during this time, usually with US. This method has yielded remarkable results in a number of cases with significant reduction in upper tract dilatation. If conservative management fails and recurrent infections are a problem, the only definitive treatment is nephroureterectomy of the affected moiety.

Obstruction is usually associated with ureterocoeles and is considered next in that context.

## Ureterocoeles

Single-system ureterocoeles are more common in boys than girls and are confined to within the bladder. They may obstruct the bladder outlet and can be confused with posterior urethral valves. They may also cause obstruction to the upper tract, although this is not common, and are known to be associated with recurrent UTIs. The treatment of choice is endoscopic incision, which is done with a fine electrode introduced through the cystoscope. The ureterocoele collapses and may not even be visible on subsequent imaging. Excision with reimplantation of the ureter is rarely indicated for this lesion.

Duplex-system ureterocoeles are associated with the upper moiety and can cause obstruction to the bladder outlet, the lower moiety ureter, and prolapse through the urethra in girls, sometimes appearing as a reddish-purple swelling at the urethral orifice. The initial management in all these cases is cystoscopy and incision of the ureterocoele. Subsequently, full investigations are carried out to determine renal tract anatomy and function.

A duplex-system ureterocoele associated with very poorly functioning upper moiety is best treated by upper pole nephroureterectomy, leaving the distal stump of ureter and ureterocoele behind. This usually does not cause any further problems.

When both upper and lower moieties have satisfactory renal function, the procedure of choice is excision of the ureterocoele and reimplantation of the ureters. With large ureterocoeles, the underlying bladder wall may be deficient and needs to be repaired to prevent diverticulum formation. Excision of the urethral lip of the ureterocoele has to be complete to avoid leaving a rim of tissue that can cause obstruction to outflow. At the same time, great care has to be taken to ensure that the sphincter musculature is not damaged. If both moieties have poor renal function, then nephrectomy and removal of as much of the ureters as possible is performed.

In some cases, it may be necessary to carry out complete excision of the upper moiety, its ureter, and the ureterocoele to achieve a satisfactory result. This operation also often requires reimplantation of the lower moiety ureter. Previously, this kind of surgery required the use of separate incisions for the kidney and bladder parts of the procedure. However, with technological advances in minimally invasive surgery, it is now possible to do this operation with suitably placed ports by using an intraperitoneal or retroperitoneal approach.

## Evidence-Based Research

Table 90.1 presents a retrospective study of the long-term effectiveness of endoscopic puncture of a ureterocoele in children.

*Table 90.1: Evidence-based research.*

| | |
|---|---|
| **Title** | Endoscopic puncture of ureterocele as a minimally invasive and effective long-term procedure in children |
| **Authors** | Chertin B, Fridmans A, Hadas-Halpren I, Farkas A |
| **Institution** | Department of Urology, Shaare Zedek Medical Center, Jerusalem, Israel |
| **Reference** | Eu Urol 2001; 39(3):332–336 |
| **Problem** | Over a period of years, the surgical approach to ureterocele has evolved from complicated major surgery to minimally invasive endoscopic treatment. Because of the high rate of secondary surgery in some recently reported series, an upper pole partial nephrectomy is again recommended as the procedure of choice. |
| **Intervention** | A retrospective evaluation of the long-term results of endoscopic puncture of a ureterocele and its long-term effectiveness and applicability in children. |
| **Comparison/ control (quality of evidence)** | Over the past 8 years, 34 patients (20 female, 14 male) were treated with primary endoscopic puncture of a ureterocele. The mean age of the patients was 1.1 ± 4.3 (mean ± SD) years. Mean follow-up was 6.1 ± 2.4 years. Antenatal ultrasound detected the ureterocele in 5 (14%) patients, foetal hydronephrosis leading to the postnatal diagnosis in 13 (38%); 16 (48%) children presented with symptoms of urinary tract infection (UTI). The ureteroceles presented as part of renal duplication in 31 (91%) patients, 3 (9%) in a single system, and 1 had bilateral ureteroceles of a duplex system. Twenty (58%) children had intravesical ureteroceles, and the remaining 14 (42%) had ectopic ureteroceles. Very poorly functioning upper pole moiety presented in 26 (75%) of the cases, and nonfunctioning upper poles in 5 (14%). Twenty of 34 (58%) children had initial vesicoureteric reflux (VUR) to the lower moiety, either to the ipsi- (60%) or contralateral kidney (40%). A cold knife incision was carried out in 4 (11.7%), puncture by a 3-Fr Bugbee electrode in 20 (58%), and the stylet of a 3-Fr ureteral catheter was utilized to puncture the ureterocele in the remaining 10 patients (30.3%). |
| **Outcome/ effect** | Complete decompression of the ureterocele was observed in 32 of 34 (94%) children. Two patients required secondary puncture two years following the primary procedure and are doing well. Upper pole moiety function improved postoperatively in 2 infants and remained stable in all 32 patients; no patient presented with deterioration of the renal function. Six of 20 (30%) patients who had initial VUR to the lower pole, accompanied with recurrent UTI, required surgery. Three underwent ureteric reimplantation and another 3 submucosal polytetrafluoroethylene paste (Teflon®) injection. Eight (40%) patients presented with spontaneous resolution of VUR to the lower moiety following puncture of the ureterocele. An additional 6 (17.6%) patients developed VUR to the upper moiety following the puncture of the ureterocele, 3 after cold knife incision and 3 after simple puncture. In 2, submucosal Teflon injection solved the VUR; the remaining 4 patients were maintained on prophylactic antibiotics. In 1 child, the reflux resolved spontaneously, and no patient presented with UTI. In 2 cases with nonfunctional upper poles, partial nephrectomy was performed due to symptomatic UTI one and two years following the initial puncture, respectively, in spite of complete collapse of the ureterocele. No difference was observed in the reoperation rate between the patients with ectopic versus intravesical ureteroceles ($p < 0.05$). |
| **Historical significance/ comments** | Endoscopic puncture of a ureterocele presents an easily performed procedure that allows the release of obstructive ureters and avoids major surgery in the majority of cases, even after a long follow-up. |

## Key Summary Points

1. Duplications of the renal tract are often diagnosed on antenatal ultrasound scans.
2. Incomplete duplications rarely cause problems and do not require intervention.
3. Complete duplications may be associated with infection, obstruction, or incontinence.
4. Recurrent urinary tract infections associated with reflux into the lower moiety require an endoscopic antireflux procedure, which involves submucosal injection of Deflux® at the ureteric orifice.
5. Obstruction with very poor renal function is best treated by upper pole heminephroureterectomy.
6. Obstruction and infection presenting acutely with pyonephrosis is treated initially with drainage and subsequently according to the renal function present.
7. Ectopic ureters in the male are single system and open above the sphincteric zone. Hence, males do not experience the symptom of wetting but can present with epididymo-orchitis. Renal function is usually preserved, and the treatment is reimplantation of the ureter into the bladder.
8. Ectopic ureters in the female may open either above or below the sphincteric zone. They are often associated with duplex systems and can present with the symptom of wetting in spite of normal voiding. In most cases, the renal element has very poor function and the treatment is excision of the upper pole with its ureter.
9. Ureterocoeles are easily diagnosed on ultrasound and initially are best managed with endoscopic incision.
10. Surgical intervention is required if further problems relating to obstruction or infection continue to be present. In such cases, if renal function is preserved, the treatment of choice is excision of the ureterocoele and reimplantation of the ureter.
11. If the renal element has poor renal function, the treatment will be nephrectomy of the upper moiety and ureter, leaving the distal stump behind. In some cases, however, it will be necessary to carry out complete excision of the upper moiety, ureter, and ureterocoele.

## Suggested Reading

Brueziere J. Ureteroceles. Ann Urol (Paris) 1992; 26:202–211.

Castagnetti M, El-Ghooeimi A. Management of duplex system ureteroceles in neonates and infants. Nat Rev Urol 2009; 6:307–315.

Chertin B, de Caluwe D, Puri P. Is primary endoscopic puncture of ureterocele a long-term effective procedure? J Pediatr Surg 2003; 38:116–119.

Merilini E, Lelli Chiesa P. Obstructive ureterocoele—an ongoing challenge. World J Urol 2004; 22:107–114.