Ureteric Duplication Anomalies

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Thambipillai Sri Paran and Prem Puri

92.1 Introduction

Incomplete and complete duplication anomalies of the upper urinary tract are estimated to be present in up to 0.8% of the normal population. The majority of these anomalies have normally developed renal moieties and cause no functional problems. When detected during antenatal scans, it is of paramount importance that the clinician keeps in mind that the vast majority of these anomalies are benign, incidental findings.

Overall, ureteric duplications are more common in females than in males. In 15% of patients, the ureteric duplication is bilateral (Privett et al. 1976). There is an increased incidence of up to 12% within some families, suggestive of an autosomal dominant inheritance (Atwell et al. 1974).

However, a percentage of ureteric duplications are associated with ureterocoele, ectopic ureter, VUR, and pelviureteric junction obstruction (PUJO). One or more of these abnormalities are frequently found in the presence of a complicated complete duplication (Fig. 92.2), where two separate ureters are seen to insert separately into the bladder or an ectopic opening. In incomplete duplication (Fig. 92.1), the two bifid ureters may join at any level from the ureteropelvic junction to the bladder and open as a single entity into the bladder; though rare, these can also be associated with VUR and PUJO as much as with a non-duplicated ureter.

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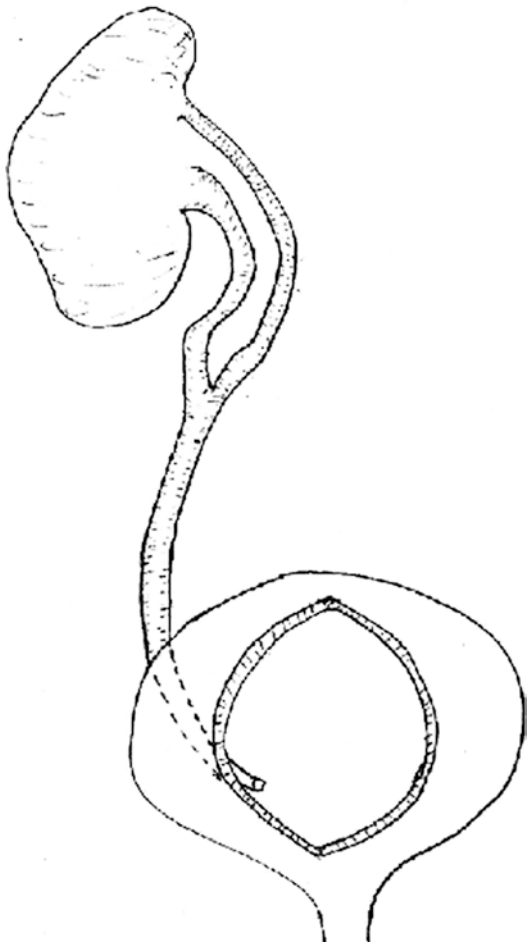

Fig. 92.1 Incomplete duplication. The two ureters unite well above the bladder and open into the bladder with a single opening

92.2 Incomplete Duplication

At 5 weeks of gestation, the ureteric bud appears from the Wolffian duct. Premature division of a single ureteral bud before it reaches the metanephric blastema is thought to result in incomplete duplication of the ureter (Kozlov and Schedl 2020). The premature division may occur at variable distance from the kidney, but there is only one ureteric orifice present on the affected side (Fig. 92.1). Incomplete duplication is three times more common than complete duplication. However, cadaveric studies have estimated the incidence to be even higher than current estimates (Arumugam 2020). The commonest complications seen with this type of anomaly are VUR and PUJO. The management of these complications will be discussed later in this chapter.

92.3 Complete Duplication

This anomaly is thought to result when two ureteral buds arise from the Wolffian duct instead of one (Campbell and Walsh 1992). The ureteral bud that gives rise to the upper pole ureter is more closely associated with the Wolffian duct and is carried medially and caudally along with the Wolffian duct (Rasouly and Lu 2013). This is thought to result in the upper pole ureter opening more medially and inferiorly than the lower pole ureter into the bladder (Weigert-Mayer law; Fig. 92.2). Sometimes, this upper pole ureter has

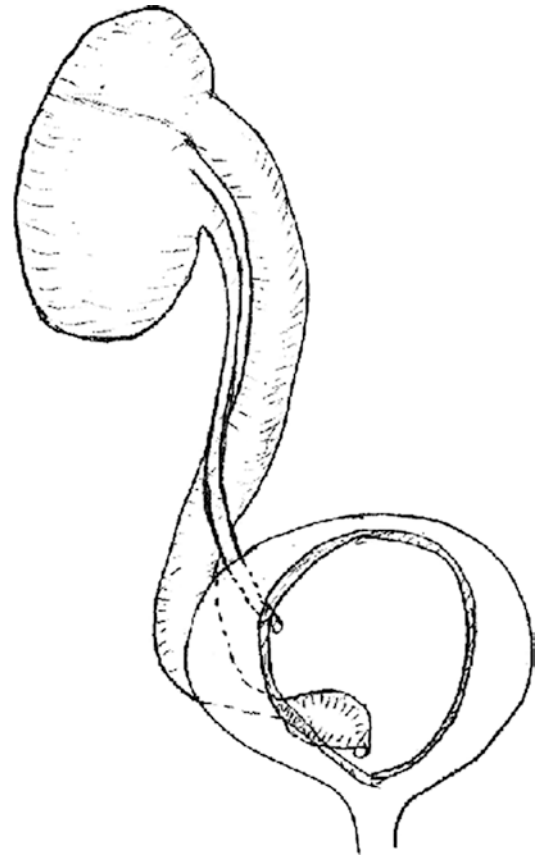

Fig. 92.2 Complete duplication with intravesical ureterocoele associated with the upper pole ureter. The lower pole normal ureter inserts superiorly and laterally with a short intramural tunnel

an abnormally prolonged or close attachment to the Wolffian duct, which may result in it opening at an ectopic location such as the urethra, seminal vesicles, vas deferens, or even epididymis.

The upper pole ureters are associated with ureterocoeles and ectopic insertions, while the lower pole ureters are associated with VUR and PUJO. These pathologies and their clinical manifestations, together with their management, are discussed below.

92.4 Investigations

92.4.1 Renal Ultrasound

Following on from either antenatal scan or clinical history suggestive of renal pathology, a renal ultrasound is the first line of investigation. It may identify ureteric duplication and associated renal pelvic dilatation or ureteric dilatation clearly. Usually, the kidney in ureteric duplication is larger in size than a normal kidney, and the upper pole has fewer numbers of calyces (usually one third of the total). When VUR is present, lower pole hydronephrosis and ureteric dilatation will be seen. When the renal parenchyma is hypoechoic, then renal dysplasia should be suspected. Absence of ureteric jet on ultrasound in conjunction with VUR on voiding cystourethrogram prompts to the diagnosis of refluxing type of obstructed megaureter (Santhalia et al. 2018). Ultrasound can also identify a ureterocoele well. Ectopic dilated ureters may mimic a ureterocoele during ultrasound study.

92.4.2 Voiding Cystourethrogram (VCUG)

This study is best performed with a feeding tube rather than with a foley catheter, which has a balloon that may confuse the diagnosis of an ureterocoele. The bladder should not be overfilled, as it may compress the ureterocoele. It is also important to take an oblique view to demonstrate the ureterocoele, which is posteriorly placed.

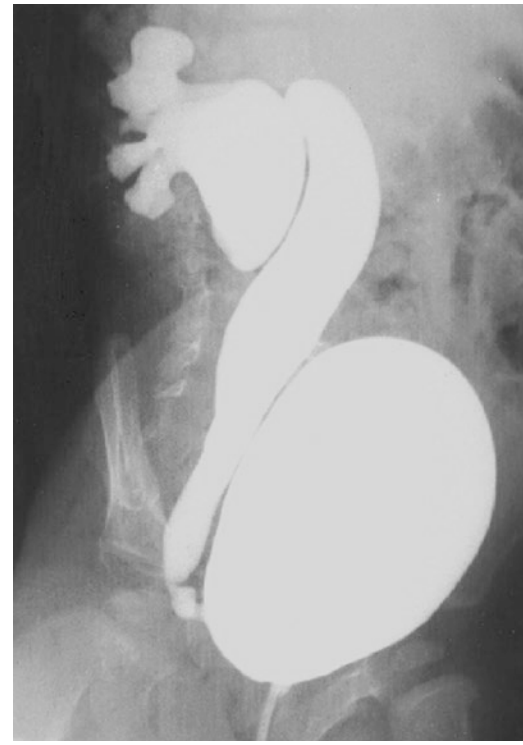

Fig. 92.3 Voiding cystourethrogram showing gross reflux into the lower pole ureter (Drooping Lilly sign)

This is the study of choice to demonstrate VUR and its severity (Fig. 92.3). It is important to appreciate that an ectopic ureter, opening at the bladder neck, can show reflux and have features suggestive of obstruction as well. It may be necessary to repeat the study to demonstrate this complex phenomenon (Santhalia et al. 2018).

92.4.3 Intravenous Pyelogram (IVP)

Though the use of this investigation has become very infrequent in pediatric urology with the introduction of CT and MR urograms, when confronted with a dilemma of demonstrating difficult ectopic ureters, IVP still has a role to play. During the latter stages of this study, when the bladder is filled with contrast, an ureterocoele could also be clearly demonstrated. A "drooping lily sign," which is the inferior and lateral displacement of lower renal moiety by the nonfunc-

tioning upper renal moiety, is a good sign of occult duplex kidney with an ectopic ureter (Figs. 92.4 and 92.5).

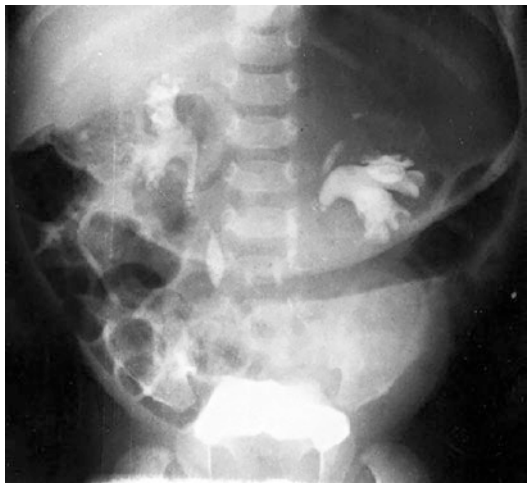

Fig. 92.4 Intravenous pyelogram demonstrating PUJ obstruction in the lower pole ureter

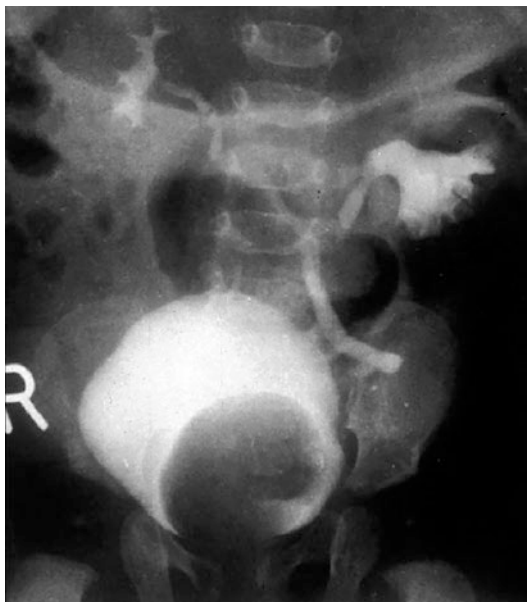

Fig. 92.5 Intravenous pyelogram demonstrating a large ureterocoele on the left side with no uptake of contrast seen within the upper renal moiety (dysplastic moiety)

92.4.4 DMSA (^{99m}Tc Dimercpatosuccinic Acid) Scan

Unlike the MAG3 scans, DMSA scans result in renal tubular labeling and are unaffected by obstruction to drainage. Therefore, they provide more accurate assessment of function and also highlight any scarring within the parenchyma. Since the newborn kidneys may not uptake the radionuclides properly, all such studies should be deferred until at least 6 weeks after delivery to allow for maturation of renal function.

92.4.5 MAG3 (Mercaptoacetyltryglycerine) Scan

This study combines information regarding the relative renal function with that of any degree of obstruction to drainage. As mentioned above, in the presence of obstruction, the relative function values may be somewhat overestimated. As with DMSA scans, MAG3 should also be postponed to allow maturation of neonatal kidneys.

92.4.6 Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) Scans with or Without Urogram

Over the last few years, CT and MR urograms have slowly replaced the need for IVP. These provide clear images of renal parenchyma, any mass lesions and that of urothelium (Kawashima et al. 2004a, b). In addition, they also provide detailed images of the intra-abdominal organs in relation to the urinary tract, highlighting any external compression on the pelvis and ureters. MRI is particularly useful in cases where occult dysplastic renal moieties, ectopic ureters, and ureterocoeles are suspected and not clearly defined by all other modalities of investigation. MR urogra-

phy is highly accurate in the assessment of ectopic ureters. In incontinent girls, MR urography should be the method of choice for depicting or ruling out ectopic ureter (Figueroa et al. 2014).

92.5 Vesicoureteric Reflux (VUR)

VUR is the commonest problem associated with both complete and incomplete ureteric duplications. It is more common in girls than in boys. In complete duplications, it is mostly seen in the lower pole ureter, which inserts superiorly and laterally, resulting in a short intramural tunnel. However, following ureterocoele puncture or excision and reimplantation, VUR may also be seen in the upper pole ureter.

The management of VUR in ureteric duplication is the same as in normal nonduplicated systems. Studies have shown that the rate of resolution of minor grades of reflux is similar to that seen in nonduplicated systems. In the newborn period, it is essential to start chemoprophylaxis, until all the problems associated with ureteric duplication are identified. High grades of reflux, associated breakthrough infections in spite of adequate chemoprophylaxis, and progressive renal scarring are all indications for an antireflux surgical intervention.

Surgical options include endoscopic subureteric Deflux injection, reimplantation of the ureter(s), ureteroureterostomy (Fig. 92.6), or heminephrectomy (Fig. 92.7), when the function within the associated renal moiety is poor. Reimplantation of the refluxing ureter could only be undertaken in isolation, if it can be safely isolated from the normal ureter without causing ischemic injury. If this is not possible or when there is documented reflux into both ureters on the same side, then common sheath reimplantation (where both ureters are reimplanted without being separated) could be done (Fig. 92.8). Recurrence of reflux is well reported within these reimplanted ureters, and may necessitate further surgery.

Endoscopic correction of VUR (with submucosal injection of Deflux) has been shown to be highly effective for high-grade reflux in single

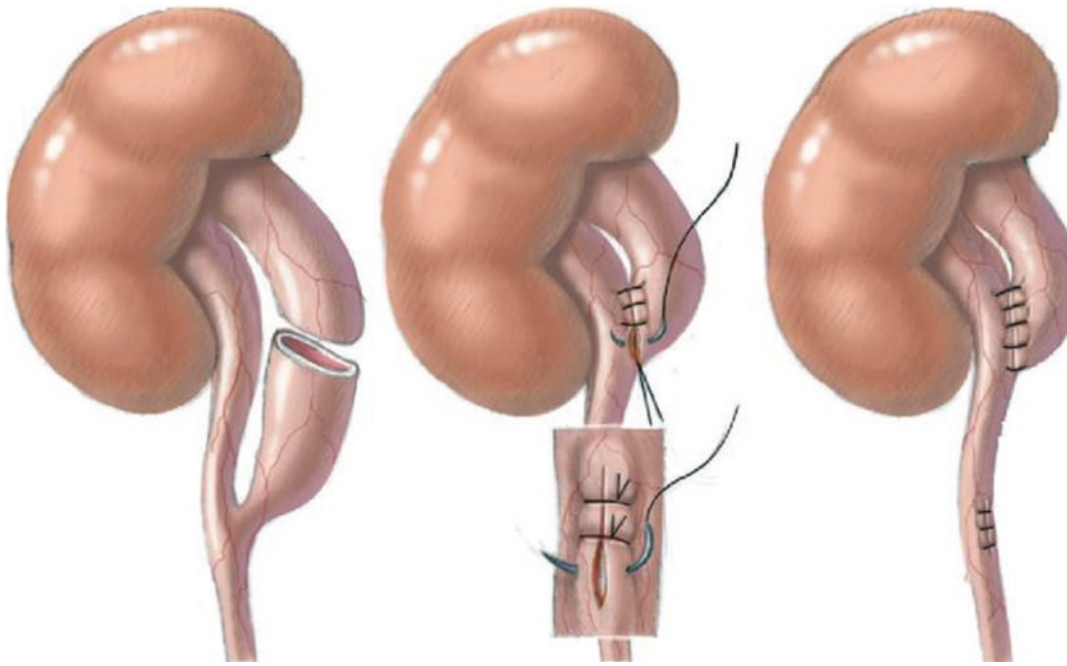

Fig. 92.6 The dilated upper pole ureter should be cut at its insertion with the lower pole ureter. Following repair of the lower pole ureteric defect, the upper pole ureter is anastamosed end-to-side to lower pole renal pelvis

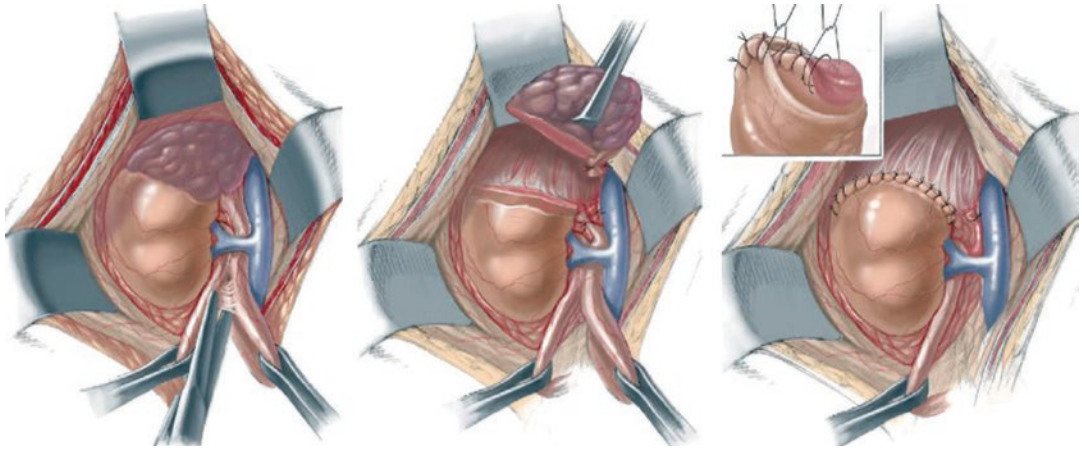

Fig. 92.7 Through a subcostal loin incision and retroperitoneal approach, the duplex kidney is exposed. The entire hilar vessels, including that of the lower pole should be clearly identified, before the upper pole vessels could be tied off. The upper pole is usually dysplastic with a clear demarcation from the healthy lower pole. The defect following the heminephrectomy is closed to achieve hemostasis

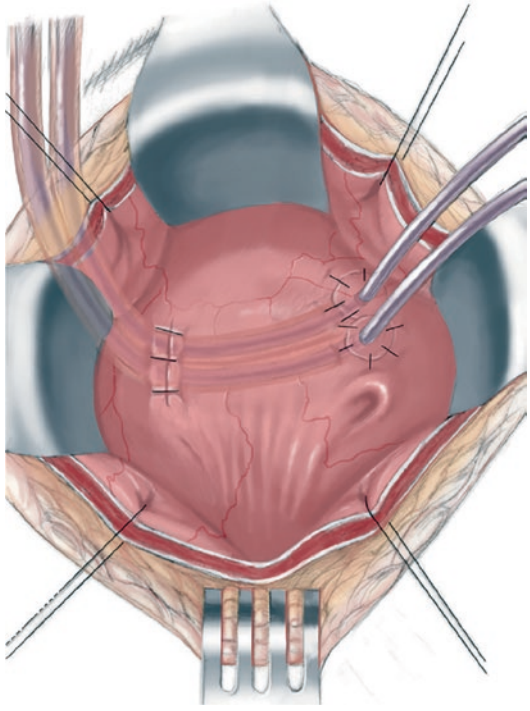

Fig. 92.8 The two ureters, which are bound closely, are dissected out together and reimplanted en bloc to avoid ischemic injury to the lower segments of the ureters

nonduplicated systems by several authors. Puri et al. have shown that this technique could be safely and effectively adopted in ureteric duplication as well. The injection is made under the bladder mucosa, 2–3 mm below the refluxing ureteric opening (which is usually the one belonging to the lower renal moiety that opens superiorly and laterally in the bladder) at the 6 o'clock position. The needle needs to be advanced fully and enough material is injected until a "volcanic" bulge of the mucosa is seen around both ureteric orifices, resulting in a slit-like appearance of both orifices.

Ureteroureterostomy (Fig. 92.6), where the markedly dilated ureter is anastamosed to the side of the normal ureter, was initially used to overcome obstructed ureters secondary to ureterocoeles. However, this approach has been successfully used in dilated ureters secondary to severe VUR as well. But, de novo ipsilateral VUR of the normal ureter has been noted following ureteroureterostomy. The advantages of this approach are that it could be performed laparoscopically with minimal morbidity, shorter hospital stay, and better cosmetic results.

When the refluxing ureter is associated with a poorly functioning or nonfunctioning dysplastic renal moiety, then heminephrectomy would be the best option (Fig. 92.7). At times, both upper and lower renal moieties may have very poor function and nephroureterectomy will become the option of choice. Most reports show that a second incision to remove the residual ureteric stump is not necessary in most children and could

be safely undertaken in the minority of children who develop problems related to residual ureteric stump (De Caluwe et al. 2002). However, when heminephrectomy or nephrectomy is undertaken laparoscopically, complete removal of the ureter(s) could safely be carried out at the same time.

92.6 Ureterocoele

Ureterocoele is a cystic dilatation of the terminal intramural segment of the distal ureter. It is usually associated with the dilatation of the ureter and calyces with a dysplastic poorly functioning renal moiety. Ureterocoeles can be classified into intravesical and extravesical based on the position of their opening. In the intravesical ureterocoele, the opening of the ureterocoele is located between the normal position of the ureteric orifice and the bladder neck. The extravesical ureterocoele opens ectopically at the bladder neck or urethra, and the opening is usually proximal to the external sphincter. These ectopic ureterocoeles are associated with significant obstruction and dysplastic upper renal moieties. A large ureterocoele could be seen prolapsing through the vestibule (Fig. 92.9).

Ureterocoeles have been reported to be 4–8 times more common in females than in males. In females, 95% of the ureterocoeles are associated with complete duplication of ureters, while in males only two thirds are associated with complete duplication; the remainder is associated with single system normal kidneys. The cystic swelling of the ureterocoele and the dilated ureters associated with them are easily identified in the antenatal scans, after 20 weeks of gestation. Most intravesical ureterocoeles are treated with endoscopic puncture. In neonates with an uninfected ureterocoele discovered by antenatal scans, endoscopic puncture of the ureterocoele alone may be sufficient. If a dysplastic renal moiety is identified in these asymptomatic children following endoscopic puncture or deroofing, on functional imaging studies, one could adopt a wait-and-see policy or elect to carry out heminephrectomy at a convenient time. Of late, the consensus has shifted to minimal intervention in these antenatally diagnosed asymptomatic children during the 1st year of life and subsequent evaluation as to what surgical reconstruction, if any, is required.

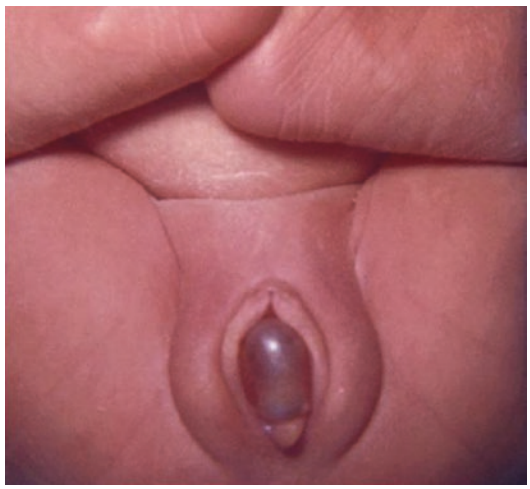

Fig. 92.9 Large ureterocoele seen to prolapse through the vestibule

In those infants with infected ureterocoele, early endoscopic puncture of the ureterocoele must be undertaken. Imaging studies, such as DMSA scans, should be delayed until 6–8 weeks, following the endoscopic deroofing, to attain accurate values. The overall incidence of VUR following endoscopic puncturing of the ureterocoele is much less than with open surgical procedures. Studies have shown that almost 90% of intravesical and 50% of extravesical ureterocoele renal moieties show useful function, if drained in early life. Therefore, a full anatomic and functional assessment should be delayed till 1 year of age, and appropriate reconstructive surgery must then be undertaken.

In almost 50% of extravesical and 15% of intravesical ureterocoeles, further reconstructive surgery will be required. A dysplastic nonfunctioning upper moiety, prone to recurrent infection is better removed (heminephroureterectomy; Fig. 92.7). When the upper moieties have reasonable function, then ureteroureterostomy (Fig. 92.6) or excision of ureterocoele followed by ureteric reimplantation can be carried out (Fig. 92.10). The final management must be tailored according to the functional and anatomical findings in each child, and demands a flexible approach.

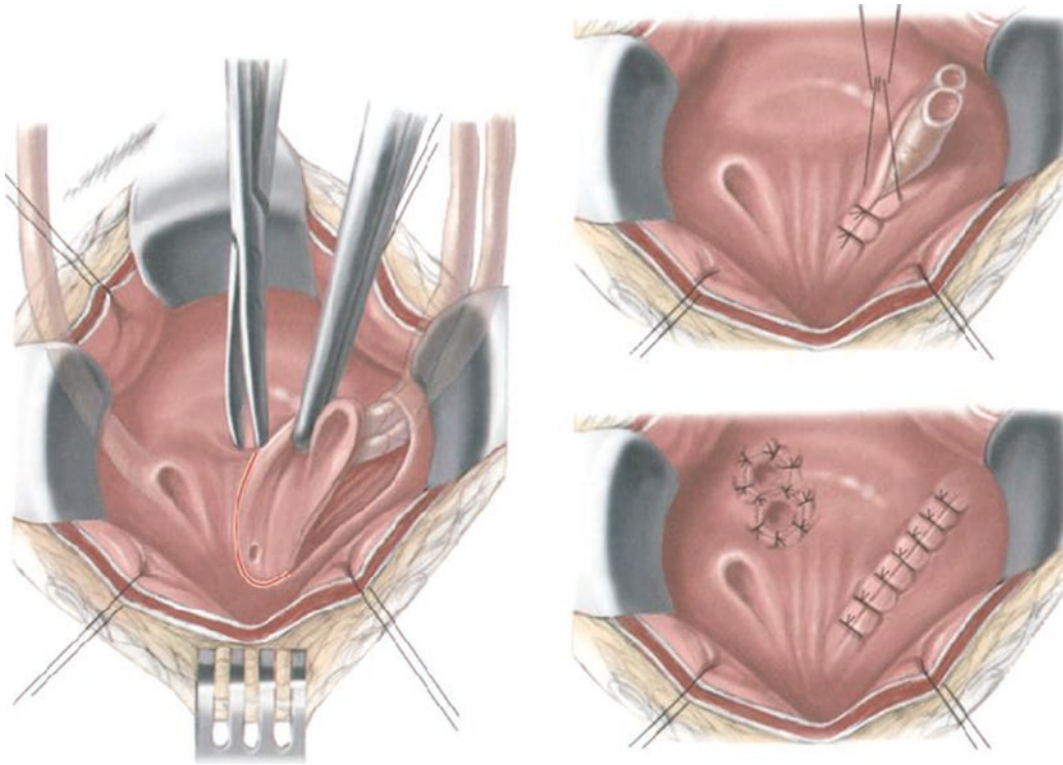

Fig. 92.10 Excision of ureterocoele and reimplantation of duplex ureters

Studies have shown that in up to 50% of those who were treated with heminephrectomy and extended ureterectomy, a second bladder surgery is necessary to deal with VUR of the lower pole ureter or rarely for bladder outlet obstruction. However, the management of VUR that only occurs following heminephrectomy could be guided by the same principles as discussed above.

92.7 Ectopic Ureters

Ectopic ureters are nearly three times more common in females than in males. As with ureterocoeles, the vast majority of the ureters are associated with duplication in females. In males, the majority of ectopic ureters are associated with single system kidneys. The com-monest sites of ectopic ureteral openings in males are posterior urethra and prostatic urethra; a small percentage is seen to be associated with seminal vesicles and epididymis. In females, the commonest sites of ectopic ureteric openings are the urethra, vestibule, and vagina.

When ureteric duplication is detected in antenatal scans, a thorough evaluation must include a search for ectopic openings. However, when present later in life, the females tend to present with a history of normal voiding pattern, with damp underwear day and night. In males, the symptoms may be one of urgency and frequency or epididymo-orchitis (Warchol et al. 2014). The most important factor in diagnosing ectopic ureters is high index of clinical suspicion.

The ectopic ureters are usually associated with the upper renal moiety, and may be seen easily with the initial ultrasound scanning. However, at times, demonstrating an ectopic ureteric opening with imaging studies can be highly challenging. Intravenous pyelography, contrast-enhanced delayed CT scan, and MRI scan have all proven useful in the diagnosis of an ectopic ureteric opening and the associated nonfunctioning, nondilated renal moiety.

When diagnosed later in life, there has already been a period of infection and dysplasia in the associated renal moiety, and heminephrectomy is the treatment of choice in these children. Of late, due to early detection following antenatal diagnosis, the upper moieties could be saved by ureteropyelostomy, ureteroureterostomy, or ureteric reimplantation, when good relative function is confirmed with imaging studies.

At times, when continued vaginal discharge or local infection is noted, a second surgery may be necessary to deal with the residual distal segment of ureter following the initial surgery.

92.8 Pelviureteric Junction Obstruction (PUJO)

Though PUJO in ureteric duplication is relatively rare, when present, it is mostly seen in association with the lower renal moiety. On renal ultrasound, a dilated lower pole renal pelvis, with no dilatation of the ureter is seen. MAG3 scintigraphy is necessary to confirm the degree of obstruction and the relative function of the affected renal moiety.

Depending on the exact anatomy, a standard pyeloplasty or a pyeloureterostomy (anastomosing the lower pole pelvis to the upper pole ureter) could be carried out. During pyeloureterostomy, the remainder of the distal ureter should be removed to avoid VUR into this remnant ureteric stump and associated infections in future. However, if the function in the associated renal moiety is poor, then heminephrectomy will become the treatment of choice.

92.9 Conclusion

With the increased referrals of antenatally diagnosed renal anomalies, including that of duplication of ureters, it is essential for a clinician to remember that only a small percentage of this group will need therapeutic interventions. Vast majority of incomplete duplications are asymptomatic and need no more than ultrasound follow-up. However, those presenting with urinary tract infections and/or hydronephrosis need appropriate investigations and interventions to preserve ipsilateral and contra lateral nephrons from further damage. When children present with persistent urinary tract infections and/or urinary leakage, high index of suspicion is necessary to look for occult duplication anomalies.

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