Renal Duplication Anomalies

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Introduction

Complete renal duplication is a condition in which there are two discrete renal moieties each with its own renal pelvis and ureter. The incidence of ureteral duplication in unselected autopsy series is 0.8 %. Most duplication anomalies are uncomplicated, do not result in clinical problems, and do not merit urological consultation. However, if the duplication anomaly is associated with an upper pole moiety (UPM) ectopic ureter or ureterocele or lower pole moiety (LPM) vesicoureteric reflux or ureteropelvic junction obstruction, urological consultation is recommended. Clinically relevant duplication anomalies are seen twice as commonly in females, with no side predilection. There is a genetic predisposition to renal duplication anomalies, as 1/8 of parents or siblings of an affected child are similarly afflicted.

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Embryology of the Kidney and Ureter

During embryologic development, three different fetal kidneys are formed: pronephros, mesonephros, and metanephros. The former two completely regress and disappear, while the latter forms the kidney.

The mesonephric (Wolffian) duct appears at 24 days of gestation. Shortly thereafter its distal end joins the primitive cloaca, and it becomes a hollow tubular structure. At 28 days of gestation, a sprout from the distal portion of the Wolffian duct, called the ureteric bud, interacts with the metanephric blastema and triggers a mutual stimulus for the development of the kidney and ureter. Abnormalities with this interaction are regarded as the cause for renal and ureteral anomalies, and renal duplication is one of them. Complete renal duplication occurs when two distinct ureteric buds emanate from the Wolffian duct. The most caudal of these buds is associated with the LPM of the kidney, while the cranial bud is associated with the UPM. The ureteric buds are then absorbed into the developing bladder trigone and migrate cranially and laterally. Since the most caudal bud is absorbed first, it has more time to migrate cranially and laterally, resulting in the LPM orifice being more cranial and lateral on the trigone than the UPM orifice, which is more caudal and medial. This relationship is known as the Weigert-Meyer law, and there are very rare

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exceptions to this rule. The ectopic ureteric bud may interact defectively with the metanephric blastema resulting in a moiety which is dysplastic and poorly functional. This is particularly common with upper pole moieties associated with ectopic ureters and ureteroceles.

Clinical Presentation and Investigation

As mentioned previously, many patients with uncomplicated renal duplication anomalies do not present with urological problems and may come to light as an incidental finding on abdominal ultrasound performed for an unrelated reason. Clinically relevant duplication anomalies may present in varied ways. Antenatally detected hydronephrosis in the fetus on ultrasound assessment is a common mode of presentation. Patients may also present with urinary tract infection/urosepsis, recurrent flank pain with nausea and vomiting (ureteropelvic junction obstruction—UPJO), urinary incontinence or purulent vaginal discharge (ectopic ureter in a female), or recurrent epididymitis (ectopic ureter in a male).

Investigation usually starts with a renal ultrasound and may also require the performance of a voiding cystourethrogram (VCUG) or renal scan depending on the pathology detected. Newer imaging modalities, such as MRI, have been found useful in selected cases.

Anomalies Associated with Upper Pole Moiety

There are two major anomalies associated with the UPM of a duplex system: ectopic ureter and ureterocele.

(a) Ectopic Ureter

The UPM ureteric bud may branch more craniad than normal from the Wolffian duct and thus enter the trigone later than it usually would or even continue to be attached to the Wolffian duct. This results in a ureter that inserts into the bladder neck or urethra in either sex. In boys it may also insert to the ejaculatory duct, vas deferens, or seminal vesicle. In girls, the ureter may enter the Gartner's duct and rupture into the vagina or introitus. It may also rarely insert into the cervix or uterus. Presentation may include antenatal hydronephrosis, urinary tract infection, or urinary incontinence in a girl with an otherwise normal voiding pattern. Rarely a girl may present with recurrent purulent vaginal discharge. Males do not present with incontinence, as the ectopic ureter inserts to the urinary tract proximal to the external sphincter complex. However, males may present with acute epididymitis as the ectopic ureter may insert into the reproductive tract. The physical examination may be normal, especially in infants detected as having an UPM ectopic ureter by antenatal hydronephrosis. In such patients a flank mass may occasionally be palpable. If presenting with a urinary tract infection, there may be concurrent flank tenderness to palpation or percussion. An enlarged, erythematous, and tender hemiscrotum would be consistent with epididymitis in a male. Careful examination of the introitus in the female may reveal the slow continual dribbling of urine from an ectopic ureter inserting to the urethra or vagina. Investigation should include a renal ultrasound in all children. The UPM is usually hydronephrotic and associated with a tortuous hydroureter. Those presenting with urinary tract infection also require a VCUG to rule out reflux, as in some cases UPM ureters ectopic to the bladder neck or proximal urethra reflux. A renal scan is usually obtained to assess the function of the UPM, which is often minimal due to underlying dysplasia. In most cases an UPM heminephrectomy is performed to remove the poorly functioning UPM and as much of the associated ectopic ureter as possible. A stump of ectopic ureter is left behind and generally does not result in problems. In the rare instance the UPM has good function,

its ureter may be joined to the LPM ureter or reimplanted into the bladder to reintegrate it into the urinary tract.

(b) Ureterocele

Ureterocele is a cystic dilatation of the distal ureter, which may be contained entirely within the bladder (intravesical) or may extend into the bladder neck or urethra (ectopic). Duplex systems are more commonly associated with ectopic ureteroceles and single systems with intravesical ureteroceles. Duplex system ectopic ureteroceles are more common in girls and more commonly left sided and may be bilateral in 10 % of cases. Patients may present with antenatally detected hydronephrosis, urinary tract infection/urosepsis, bladder outlet obstruction, or prolapse through the urethral orifice (females only). Physical findings mimic those discussed for ectopic ureter, with the exception of scrotal findings. Additionally, in a female infant, the ureterocele can rarely prolapse through the urethra resulting in a mass at the introitus. Investigation comprises a renal ultrasound and VCUG. The ultrasound demonstrates a hydronephrotic UPM associated with a dilated tortuous hydroureter, which culminates in a bubble-like appearance in the bladder (Fig. 4.1). There may also be ipsilateral LPM hydronephrosis, and indeed contralateral hydronephrosis, particularly in the setting of bladder outlet obstruction. Vesicoureteric reflux (VUR) is seen in 50 % of LPM ipsilateral to the ureterocele and in 20 % of contralateral ureters. Renal scans are often obtained to assess the degree of UPM function and/or obstruction by the ureterocele. Like ectopic ureters, the UPM function is often poor when associated with a ureterocele. Management depends on the acuity of patient presentation, presence of VUR, and UPM function. If a patient presents with urosepsis not responding to antibiotics or prolapse of the ureterocele through the urethral orifice, emergency drainage is required. In more elective circumstances, management varies greatly from endoscopic incision through upper pole heminephrectomy to complete reconstruction (upper pole heminephrectomy with ureterocele excision and LPM ureteric reimplantation).

Anomalies Associated with Lower Pole Moiety

VUR

Vesicoureteric reflux is the most common urinary tract abnormality associated with duplex systems and complete ureteral duplication. As

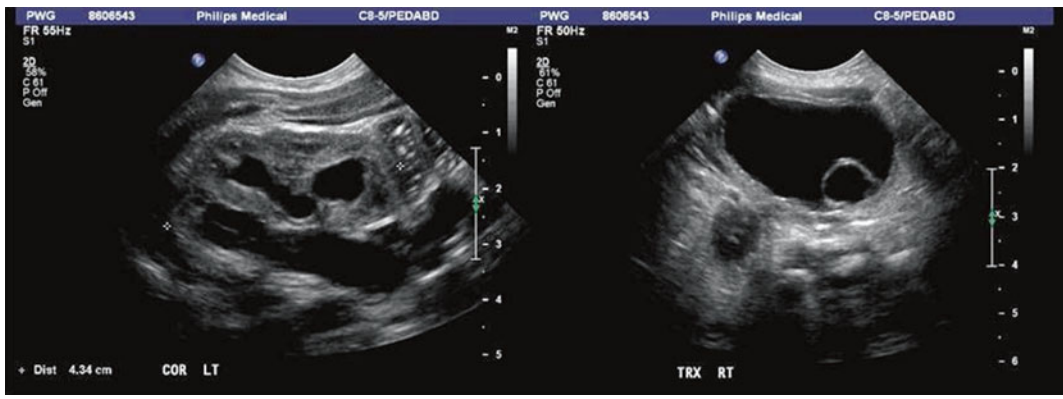

Fig. 4.1 One-month-old boy with left renal duplication, hydroureteronephrosis of the upper unit, and ureterocele inside the bladder

mentioned in the Embryology section, the Weigert-Meyer law explains the differences observed in the ureteral orifice location and submucosal tunnel length of the distal ureters in complete renal duplication. As the LPM ureter inserts more proximally and laterally on the bladder trigone, it has a shorter submucosal tunnel length and consequently a suboptimal anti-reflux mechanism. The UPM ureter rarely refluxes, and this is attributed to its distal location on the trigone, which confers a longer submucosal tunnel and a more effective flap-valve mechanism. VUR is diagnosed by VCUG or nuclear cystogram. The typical VCUG image of VUR to the LPM in a complete duplication will show the "drooping lily" appearance of the collecting system: fewer calices than expected in a single system with the upper calices in a more horizontal axis. Reflux to the LPM in a duplex kidney may be associated with hydronephrosis, recurrent urinary infection, and in severe cases thinning of the LPM parenchyma. Physical examination is nonspecific for the presence of VUR and would be expected to be normal in a child who is asymptomatic. Management for reflux in a duplex collecting system is similar to that for VUR in a single collecting system, with antibiotic prophylaxis being the first choice in most cases. Surgery is reserved for more severe grades of reflux associated with breakthrough urinary infections and/or progressive renal scarring with loss of ipsilateral renal function. Reported data shows the same rate of resolution of reflux to LPM of duplex systems when compared with VUR to a single system. When surgical treatment is elected, reimplantation of both ipsilateral ureters and heminephrectomy of the LPM are options. The choice is mainly dictated by the amount of functioning parenchyma in the LPM and the severity of dilation of the ureters. More recently, endoscopic sub-ureteral injection of bulking substances for correction of VUR has shown acceptable rates of cure in duplex systems.

UPJO

UPJO is the most common congenital obstruction in a single system; however the incidence of UPJO in duplex collecting systems is less frequent (2 %) and most commonly affects the LPM. Obstruction of the UPJ is rarely seen in the UPM. It may present with antenatal hydronephrosis, flank pain, recurrent urinary infection, or kidney stones. Physical findings are nonspecific and may range from a normal examination to a patient with severe upper quadrant and flank tenderness ipsilateral to the obstruction. UPJO may be an intrinsic primary congenital malformation of the ureter, but in duplex collecting systems, it is often associated with crossing vessels of the renal pedicle or high-grade VUR. Severe VUR to the LPM may cause significant ureteral dilation with secondary kinking of the UPJ, resulting in obstruction. UPJO of the lower moiety may be associated with different degrees of dilation of the collecting system and impairment of the urinary drainage. Usually the LPM ureter is not dilated, unless there is a concomitant severe grade of VUR, which would be documented by VCUG. UPJO of the LPM is suspected by an US that shows a dilated collecting system in the lower portion of a duplex kidney. Dilation of the urinary system is not always associated with obstruction of the kidney, whereas renal scarring and diffuse thinning of the parenchyma on the US may represent an indirect sign of kidney damage. The presence of obstruction is assessed using a diuretic renal scan, which provides the differential renal function of both kidneys, the differential function of the UPM and LPM of the duplex system, and the drainage curves of both kidneys and ipsilateral moieties to allow for assessment of obstruction. A significant obstruction with compromise of the relative LPM function is an indication for surgical correction, which may be accomplished by an open or laparoscopic pyeloplasty. Severe UPJO of a nonfunctioning LPM is best managed with heminephrectomy of the LPM and excision of as much ureteral length as possible if VUR is present.

Recommended Reading

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