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PICTORIAL ESSAY

The duplicated collecting system of the urinary tract: embryology, imaging appearances and clinical considerations

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Abstract Duplication anomalies of the urinary collecting system are common and can be discovered and characterized with multiple imaging modalities. The embryology, imaging manifestations and clinical ramifications of duplicated ureters and renal collecting systems vary from a normal anatomical variant to urological pathology and are discussed and illustrated in this review.

Keywords Children · Duplex kidney · Fetal · Ureteral duplication · Ureterocele · Uroradiology

Introduction

A duplicated ureter and urinary collecting system is a common anatomical anomaly that can be an asymptomatic normal variant or, when abnormal, can be associated with vesicoureteral reflux (VUR), incontinence, ureterocele or obstructive uropathy as well as renal parenchymal scarring or dysplasia and decreased renal function. The incidence and prevalence of urinary tract duplication has been reported to be 0.7–4% of the population and affects females more than males [1]. The diagnosis and evaluation of duplication can be performed with multiple imaging modalities including ultrasonography, voiding cystourethrography (VCUG), intravenous pyelography (IVP), computed tomography (CT), dimercaptosuccinic acid (DMSA) renal cortical scintigraphy, ^{99m}technetium-mercaptoacetyltriglycine (^{99m}Tc-MAG-3) renography and magnetic resonance urography (MRU). The purpose of this manuscript is to review the embryology, highlight the imaging manifestations and discuss the clinical ramifications of duplicated ureters and renal collecting systems in the fetus, infant and child.

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Embryology

During normal fetal development, a single ureteric bud arises from the primitive mesonephric (Wolffian) duct and migrates to meet the metanephros, the embryological precursor of the kidney (Fig. 1). The ureteric bud induces the metanephros to form nephrons, the functional units of the renal parenchyma. In turn, the metanephros induces the ureteric bud to bifurcate sequentially to form the renal collecting system including the calyces and renal pelvis. When there is failure in the reciprocal signaling between the ureteric bud and the metanephros, urinary tract anomalies result. Although the embryological details have not been completely elucidated, researchers have shown that overexpression of the glial cell-derived neurotrophic factor (GDNF)-RET. signaling pathway induces the formation of multiple ureteral buds, and anomalous spatial expression influences the site of insertion in the cloaca/primitive bladder [2]. Multiple genes have been implicated in the GDNF-RET. signaling pathway. For example, mice without angiotensin type II receptors have been shown to develop urinary tract anomalies, of which ureteral duplication is the most common [3]. Temporally, the rostral ureteral bud (upper pole) inserts after the caudal ureteral bud (lower pole),

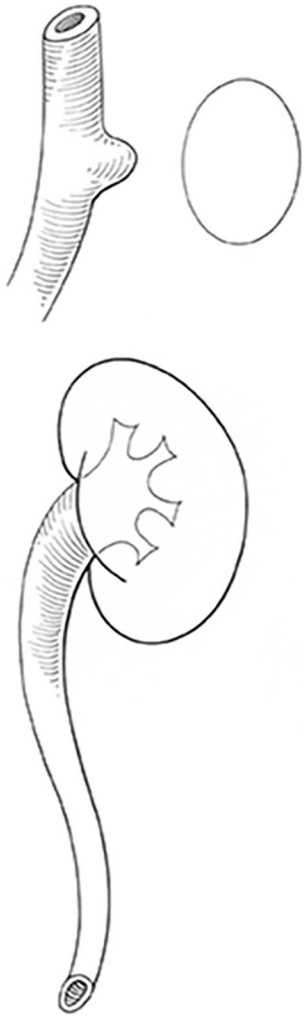

Fig. 1 Single system. Illustration depicts a single ureteric bud arising from the mesonephric (Wolffian) duct, which meets with the metanephros to form a single ureter and renal collecting system

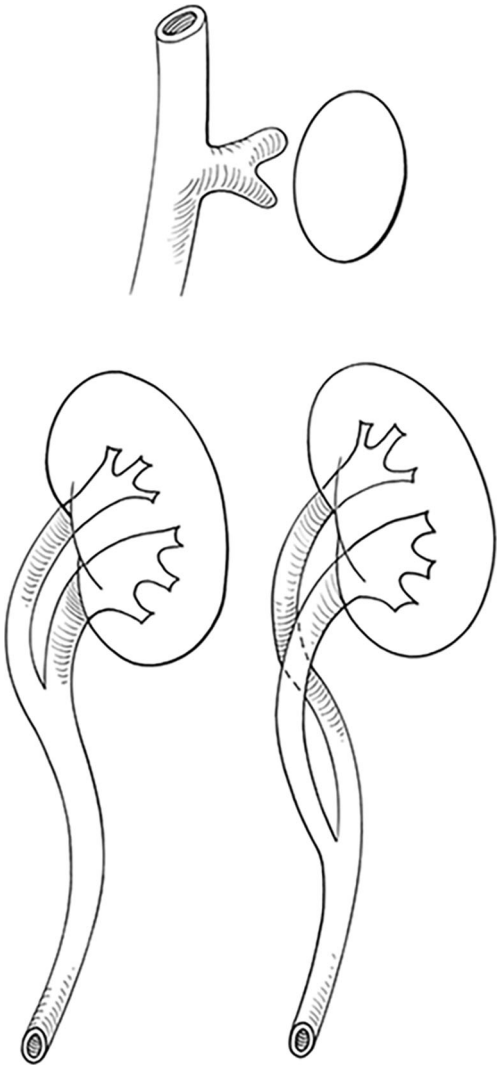

Fig. 2 Incomplete ureteral duplication. When the ureteric bud bifurcates prior to meeting the metanephros, the collecting system is partially split in two, with a common distal ureteral orifice. The level of bifurcation can occur along the ureter from the renal pelvis to just above the ureteral insertion

which allows the rostral ureteral bud to migrate inferiorly before contacting the cloaca/primitive bladder. Furthermore, for distal ureteral maturation to occur a series of events including distal apoptosis (programmed cell death) and a 180° rotation of the ureter result in correct positioning of the ureter in the cloaca/primitive bladder. This process is under the guidance of leukocyte common-antigen-related family receptor protein tyrosine phosphatases, involving retinoic acid, which interacts with RET. proteins [4].

Incomplete duplication/bifid ureter

If a single ureteric bud bifurcates prior to meeting the metanephros, an incomplete duplication, or bifid ureter/renal collecting system, results (Fig. 2). This bifurcation can occur anywhere along the ureter including high (at the level of the renal pelvis), low (near the bladder), or anywhere in between. Any pathology affecting the ureter below the bifurcation influences the upper and lower renal poles similarly, presenting identically to abnormalities affecting single systems. For example, distal obstruction or an incompetent vesicoureteral orifice can result in symmetrical upper- and lower-pole collecting system dilation (Fig. 3).

Complete duplication

Alternatively, two separate ureteric buds can arise from the mesonephric duct and can migrate to meet the metanephros (Fig. 4). Signaling pathways induce the two ureteric buds to bifurcate into two separate collecting systems, i.e. complete duplication. Many duplicated collecting systems are asymptomatic and in this scenario can be considered a normal anatomical variant. However, when either the upper pole or lower

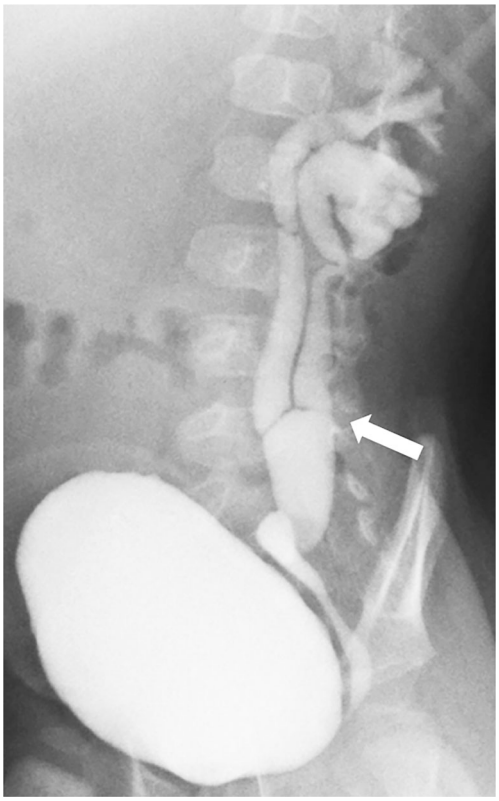

Fig. 3 Single anteroposterior fluoroscopic image from a voiding cystourethrography in a 2-year-old boy demonstrates vesicoureteral reflux into a bifid left ureter (*arrow* at bifurcation) with symmetrical upper- and lower-pole renal collecting system dilation

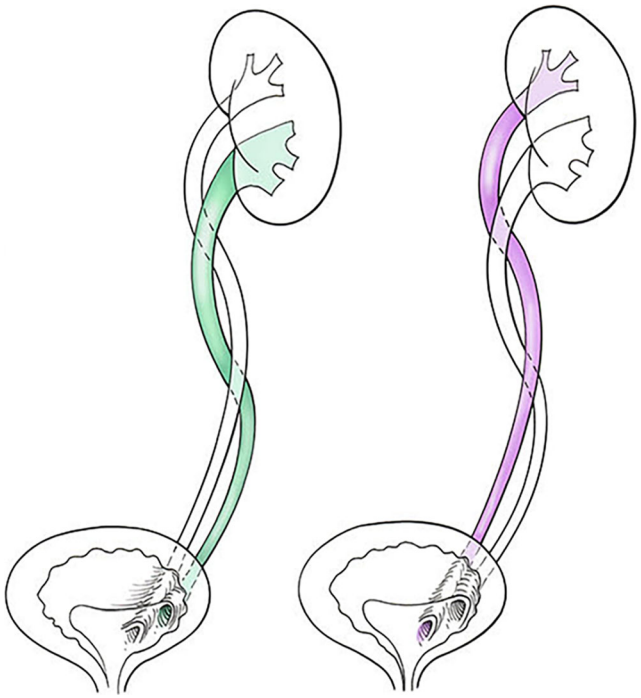

Fig. 4 Complete ureteral duplication. When two distinct ureteric buds, which arise from the mesonephric (Wolffian) duct, each meet the metanephros separately, complete duplication of the ureter and renal collecting system occurs. The lower pole ureter is the analogue of the single-system ureter and is therefore orthotopic (*green*). The upper pole ureter is ectopic (Weigert–Meyer rule), inserting inferior and medial to the orthotopic ureteral orifice (*purple*)

pole (or both) is abnormal, the depiction of the abnormal anatomy usually requires further uroradiologic imaging.

Duplicated ureter and renal collecting system

Lower pole

The lower pole of the duplicated ureter and renal collecting system is the analog of the normal single collecting system with one normal orthotopic ureter. Therefore the abnormalities that affect a single renal collecting system have been shown to similarly affect the lower pole of the duplex kidney. For example, obstruction affects the lower pole at the ureteropelvic junction (UPJ) and, less commonly, the lower pole ureterovesical junction (UVJ). Vesicoureteral reflux into the lower pole collecting system and ureter is common, just as in a single (non-duplicated) ureter.

Upper pole

Based upon observations described independently by Weigert and Meyer, the upper moiety drains via a ureter that inserts ectopically, inferior and medial to the orthotopic UVJ. This is the origin of the Weigert–Meyer rule [5, 6]. The rule states that the ectopic ureter can insert anywhere along an "ectopic pathway" (Fig. 5), which includes the structures arising from the urogenital canal and mesonephric (Wolffian) duct. The ectopic pathway is different in boys and girls. In boys, the ureter can insert into the bladder, the prostatic urethra or the genital ducts that arise from the mesonephric (Wolffian) ducts (including seminal vesicles, ejaculatory ducts, epididymis and vas deferens), all of which are above the external urethral sphincter (Fig. 6). In girls, the ectopic ureter can insert into the structures that arise from the urogenital canal including the bladder, the urethra or the distal vagina. In girls, the ureter can insert into the paramesonephric duct structures adjacent to the regressed mesonephric (Wolffian) duct, and these include the upper vagina, the uterus and the fallopian tube (Fig. 7). Unlike boys, the site of ureteral insertion in girls can be either above or below the external urethral sphincter. While uncommon, if the insertion is below the external sphincter, this can lead to urinary incontinence and the classic clinical presentation of a young girl who has "always been wet, never dry," i.e. she has always had continuous urinary dribbling. This girl should be easily distinguishable from the clinical presentation of either urinary frequency or enuresis, neither of which is associated with duplication anomalies.

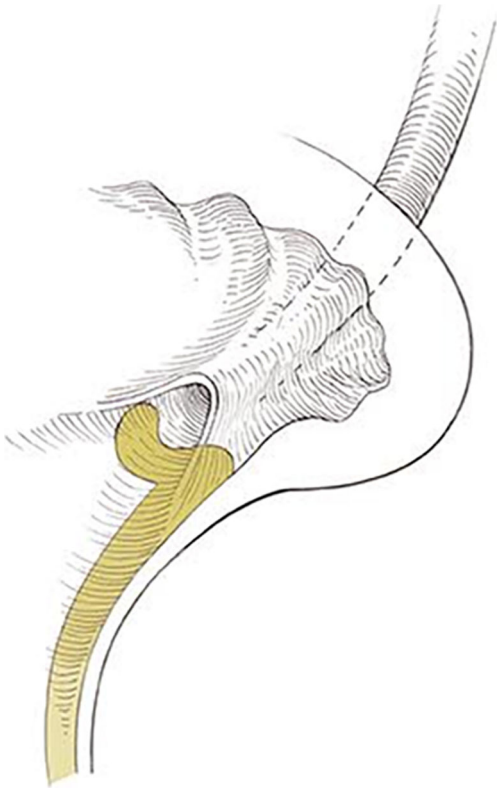

Fig. 5 Ectopic pathway in the Weigert–Meyer rule. In complete ureteral duplication the upper pole ureter is ectopic. This illustration shows the bladder, the orthotropic ureteral orifice and the pathway of potential sites of the ectopic upper pole ureter (*yellow*)

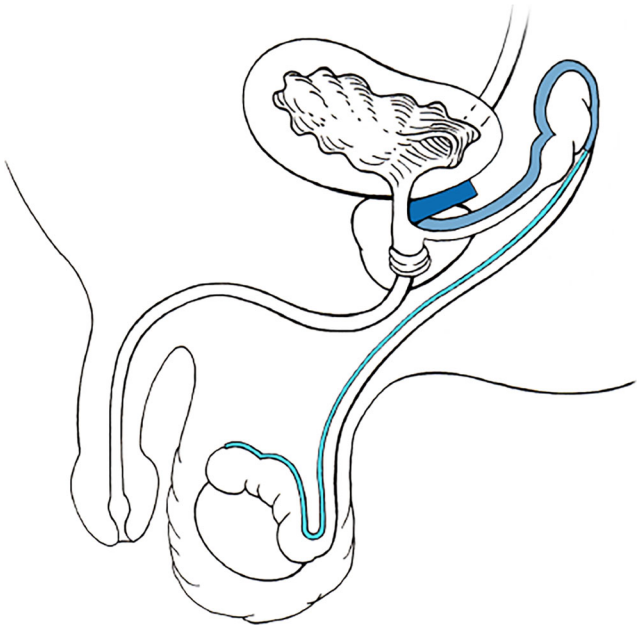

Fig. 6 Ectopic pathway in boys. Midline sagittal illustration of the male pelvis with complete ureteral duplication shows the pathway of the upper pole ectopic ureter in boys. The pathway of the upper pole ectopic ureter is outlined in blue. The most common ectopic locations are from the trigone of the bladder to the bladder base and external sphincter (*thick dark blue line*). Less commonly, an ectopic ureter inserts into the ejaculatory duct or seminal vesicle (*medium blue line*). Rarely, the ectopic ureter inserts into the vas deferens (*thin light blue line*). Note that the ectopic ureter always inserts proximal to the external urethral sphincter. Thus a boy with an ectopic ureter never presents with incontinence. This is in contrast to girls (see Fig. 7)

If the upper pole ureter inserts sufficiently ectopically, renal parenchymal dysplasia can result. The further the ectopic ureteral orifice is from the orthotopic insertion site, the more dysplasia is seen in the upper pole parenchyma. This is thought to be related to abnormal signaling with the metanephros and subsequent abnormal renal development. In addition, parenchymal injury or scarring from VUR or obstruction can contribute to decreased upper pole function. The upper pole ureter might be obstructed or have VUR, either of which can manifest as renal collecting system dilation.

The ectopic ureter can also end in a ureterocele, i.e. saccular dilation of the submucosal distal ureter secondary to congenital narrowing of the ureteral orifice at its insertion can produce a ureterocele. It is important to note that a ureterocele is a dynamic structure; as the bladder fills, the ureterocele might be effaced and decompress into its ureter. A large ureterocele might also affect the ipsilateral orthotopic or the contralateral ureter and cause displacement or obstruction.

Rare anomalies

Additional rare anomalies can occur. When three ureteric buds meet the metanephros, urinary tract triplication can occur. Sometimes a ureteric bud can fail to meet the metanephros, which might result in a blind-ending ureter, which can be shown on VCUG if there is VUR into the ureter (Fig. 8).

What a radiologist needs to know

A duplex kidney should have two collecting systems, and when the upper and lower poles are normal, it is typically longer than a single-system kidney. This is considered a normal variant. When the upper or lower pole or both are abnormal, complete ureteral duplication and its associated anomalies should be considered. In complete ureteral duplication, it is the upper pole ureteral orifice that is ectopic, and is often obstructed, resulting in upper pole hydroureteronephrosis. The upper pole ureter might terminate in a ureterocele. Often, greater degrees of ectopia result in greater associated dysplasia of the upper pole renal parenchyma. When the upper pole of a duplex kidney is severely dysplastic, the duplex kidney might be shorter than the contralateral normal kidney. Since the lower pole ureter is the analogue of the single-system kidney it might be normal, reflux or obstruct, and the lower pole can appear normal, dilated or scarred/dysplastic.

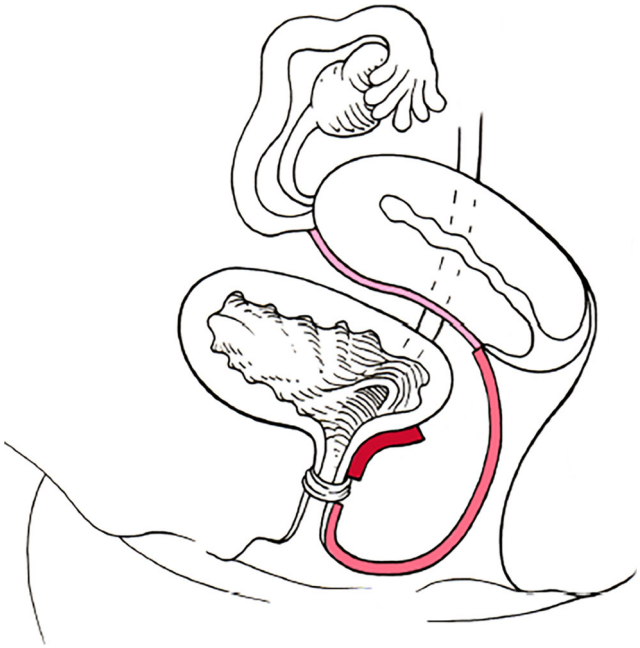

Fig. 7 Ectopic pathway in girls. Midline sagittal illustration of the female pelvis with complete ureteral duplication shows the pathway of the upper pole ectopic ureter in girls. The pathway of the upper pole ectopic ureter is outlined in pink. The most common ectopic locations are from the trigone of the bladder to the bladder base and external sphincter (*thick dark pink line*). Less commonly, the ectopic ureter inserts into the urethra below the external sphincter, perineum or vagina (*medium pink line*). Rarely, the ectopic ureter inserts into the uterus or fallopian tube (*thin light pink line*). Unlike boys, the ureter can insert proximal or distal to the external urethral sphincter. Thus an ectopic ureter in a girl inserting below the external sphincter creates the classic clinical presentation of constant wetness, day and night, because of the relentless dripping from the ectopic ureter

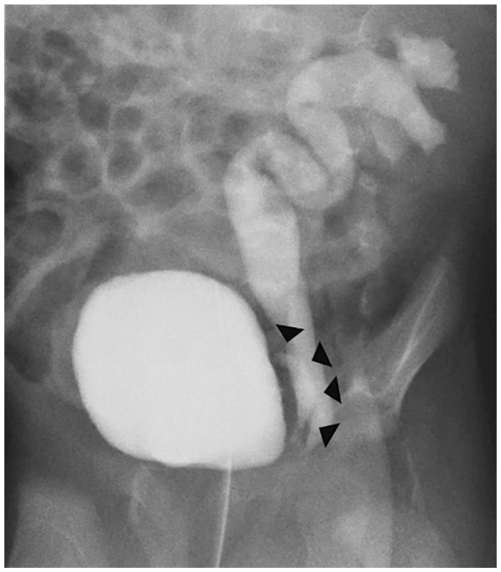

Fig. 8 Duplicated system. Lateral oblique fluoroscopic image in a 2-year-old boy demonstrates grade V vesicoureteral reflux into the lower pole of the left kidney and a blind-ending rudimentary ureter (*arrowheads*) related to a dysplastic left upper moiety in a duplicated system, seen on the boy's preceding ultrasound (not shown)

Thus the following should be considered when evaluating a duplex kidney. First, the radiologist should have an awareness and suspicion that a duplex kidney might exist. Thereafter the radiologist should consider the renal length, number of collecting systems separated by a bar of renal parenchyma, renal parenchymal thickness and appearance, dilated collecting system(s) and presence or absence of a ureterocele. The combination of imaging studies needed to fully evaluate the child is dependent on the child's symptoms and the initial imaging modality that detected the duplex kidney. These observations can be made on any imaging modality, and the appearances are described in the subsequent sections.

Imaging of ureteral and renal collecting system duplication

Ultrasonography

Ultrasonography offers a noninvasive, relatively inexpensive imaging modality without ionizing radiation in the evaluation of suspected urinary tract duplication. Additionally, because this examination is frequently performed in the fetus and the child for a variety of clinical indications, it is the imaging study through which a duplex kidney is often discovered. In the fetus, this diagnosis can be made on a routine anatomical survey that is typically performed at about 20 weeks' gestational age or in the evaluation of fetal hydronephrosis. In the infant or the child, ultrasonography is commonly performed to follow up prenatal hydronephrosis or during the evaluation of an illness, particularly a febrile urinary tract infection, and duplication anomalies are sometimes found.

Findings of a kidney with a duplicated collecting system include unequal renal lengths, abnormal parenchymal contour and asymmetrical upper- and lower-pole pelvicalyceal dilation. The duplex kidney can be larger than the non-duplicated kidney on ultrasonography, although when there is underlying renal scarring or dysplasia, it might be smaller than expected. If renal parenchymal tissue is seen extending from the cortex to the renal hilum separating the upper and lower poles, underlying duplication is suggested. Typically, this parenchymal tissue does not bisect the kidney. Instead it is most commonly seen in the upper third of the duplex kidney. Additionally, the upper and lower poles in duplicated renal collecting systems might demonstrate asymmetrical pelvicalyceal dilation, which is unlikely to occur in incomplete duplication or single ureteral systems. These findings can be recognized on prenatal or postnatal ultrasound (Fig. 9). The findings of asymmetrical renal

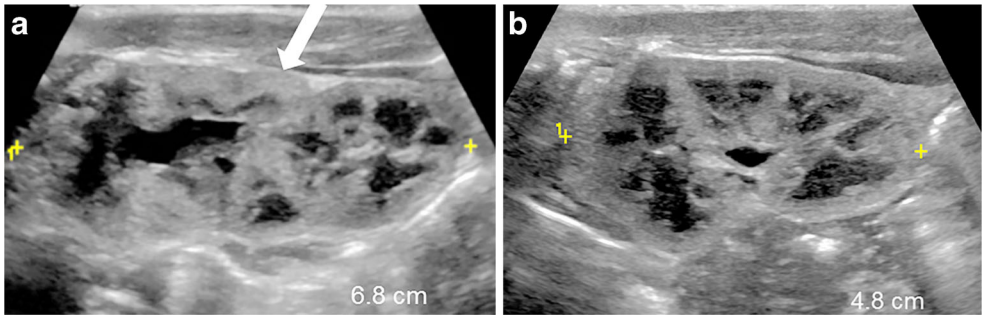

Fig. 9 Renal asymmetry on ultrasonography. **a, b** Two sagittal US images in a 31-day-old girl depict (**a**) the right kidney and (**b**) the left kidney with asymmetrical renal lengths, right longer than left. A parenchymal column separates the right upper and lower poles, with continuity of the cortex between the poles (*arrow*) and with asymmetrical dilation of the two pelves

lengths, dividing parenchymal tissue, or disproportionate pelvicalyceal dilation of the upper and lower poles are very suggestive of an abnormal duplicated renal collecting system.

A ureterocele can be diagnosed by identifying a well-circumscribed fluid-filled structure within the bladder on prenatal or postnatal ultrasound (Fig. 10). Ureteroceles are dynamic and might only be seen when the bladder is incompletely distended. As the bladder fills, the ureterocele might be effaced as the urine within it empties into its ureter. The ureterocele can even evert. With bladder emptying, urine within the ureter refills the ureterocele. Ureteroceles can be seen with single collecting systems, almost always in boys. However, if a ureterocele is identified, close examination to evaluate for an associated duplication anomaly is important, especially in girls, where most are associated with duplicated collecting systems. Ureteroceles have also been associated with obstruction at the contralateral or ipsilateral lower pole ureterovesical junction.

If urinary tract duplication is suspected or diagnosed by ultrasonography, additional imaging to further evaluate the urological anatomy and to assess for potential complications might be needed. Referral to a pediatric urologist is usually warranted to discuss workup, management and treatment options. For example, ureteroceles might require surgical incision or resection.

Voiding cystourethrography (VCUG)

VCUG is often performed to further evaluate the infant or child when an abnormal duplex kidney is discovered to determine whether vesicoureteral reflux is present or to confirm the presence of a ureterocele. In girls presenting with a febrile urinary tract infection, the most common indication for VCUG, 5% have a completely duplicated urinary collecting system [7]. The appearance of VUR on VCUG can suggest or diagnose either incomplete (VUR into a bifid ureter) or complete (VUR only into the lower pole) urinary tract duplication. The axis of a non-duplicated renal collecting system is oriented toward the child's contralateral shoulder (Fig. 11). However if VUR is seen into a pelvicalyceal system and its axis is oriented toward the child's ipsilateral shoulder, urinary tract duplication with VUR into the lower pole is likely (Fig. 11). In the absence of VUR, by exclusion, urinary collecting system dilation is caused by obstruction. For example, ureteropelvic junction obstruction typically affects the lower pole, which is the analogue of the single collecting system, and might coexist with lower moiety VUR (Fig. 12).

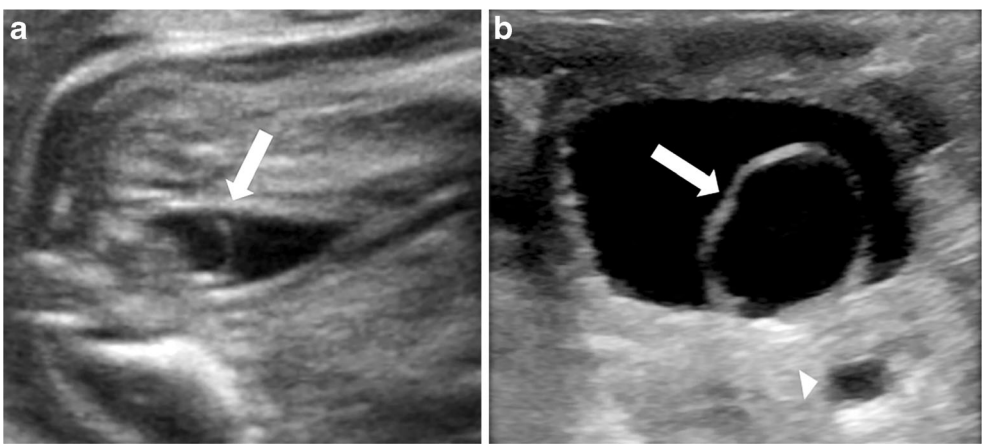

Fig. 10 Ureterocele on ultrasonography. **a** Prenatal US image of the bladder in a male fetus at 24 weeks 4 days of gestational age demonstrates a ureterocele (*arrow*). **b** Postnatal sonogram in a 3-day-old girl demonstrates a left-side ureterocele within the bladder and a dilated distal left upper pole ureter (*arrowhead*)

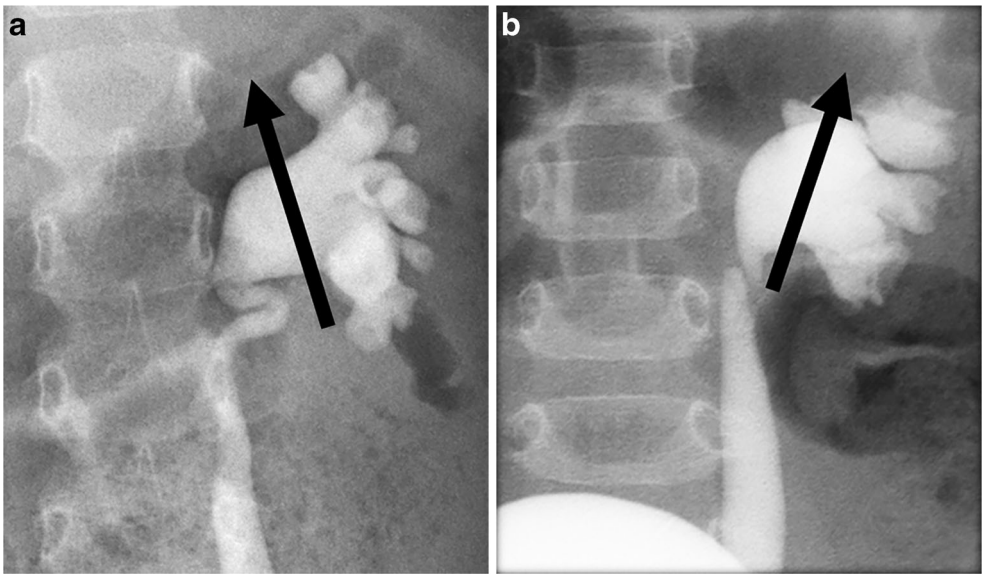

Fig. 11 Calyceal axis on voiding cystourethrography (VCUG). Anteroposterior fluoroscopic images from VCUG in two children. **a** VCUG shows vesicoureteral reflux into a non-duplicated left renal collecting system in a 3-year-old boy in whom the calyceal axis is oriented toward the boy's contralateral shoulder. **b** Contrast agent opacifies the lower pole of a duplicated left renal collecting system in a 9-month-old girl in whom the calyceal axis is oriented toward the girl's ipsilateral shoulder

A ureterocele can be identified as a filling defect within the bladder on early filling images (Fig. 13). Later during bladder filling, the ureterocele is effaced and can become invisible, underscoring the importance of awareness and identification during early bladder filling. The ureterocele can be dynamic, sometimes even everting during voiding in up to 3% of patients with a duplicated renal collecting system and a ureterocele (Fig. 13) [8]. In this situation the ureterocele can mimic a bladder diverticulum. Therefore, if a bladder filling defect disappears as the bladder fills and begins to look like a "bladder diverticulum" on late filling images, an everting ureterocele should be suspected [8]. As with ultrasonography, ureteroceles can be seen with single-system urinary tracts, but suspicion for a duplicated renal collecting system should be heightened when a ureterocele is identified [9]. Rarely, the ureterocele prolapses into the urethra (Fig. 14). When the ureterocele is large and the associated ureter, collecting system and renal parenchyma are relatively small in size, the descriptive term "ureterocele disproportion" has been used and should suggest that (a) duplication is present and (b) the associated upper pole renal parenchyma is dysplastic and poorly functioning (Fig. 15) [10].

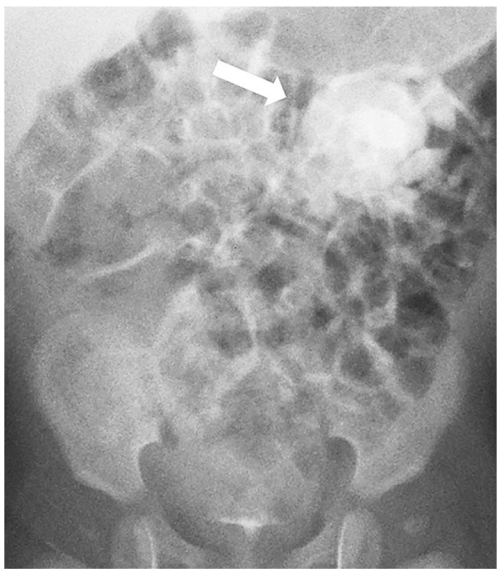

Fig. 12 Ureteropelvic junction (UPJ) obstruction on voiding cystourethrography (VCUG). Anteroposterior fluoroscopic image from VCUG in a 3-month-old girl demonstrates reflux into a dilated left lower moiety and possible concomitant UPJ obstruction. Subsequent ^{99m}Tc-MAG-3 diuresis renography demonstrated very minimal drainage of the tracer from the dilated left lower moiety after administration of furosemide with obstructive post-diuresis parameters, thus confirming high-grade UPJ obstruction (not shown)

Careful evaluation of the expected location of the UVJ should be performed in the steep oblique position. Because the abnormal ureteral insertion can be distal to the external urethral sphincter in girls, special attention should be paid by the radiologist during voiding because when the ectopic ureter inserts at or below the urethral sphincter, VUR can only be detected during voiding (Fig. 16).

Although VCUG is the most common imaging procedure performed in the evaluation for vesicoureteral reflux, radionuclide cystography and contrast-enhanced voiding urosonography are also available imaging modalities.

VUR might require antibiotic therapy, follow-up imaging or a surgical procedure. VUR into an ectopic ureter rarely resolves without intervention and sometimes requires surgical resection or reimplantation. As discussed, ureteroceles might need surgical incision or excision. Referral to a pediatric urologist should be considered.

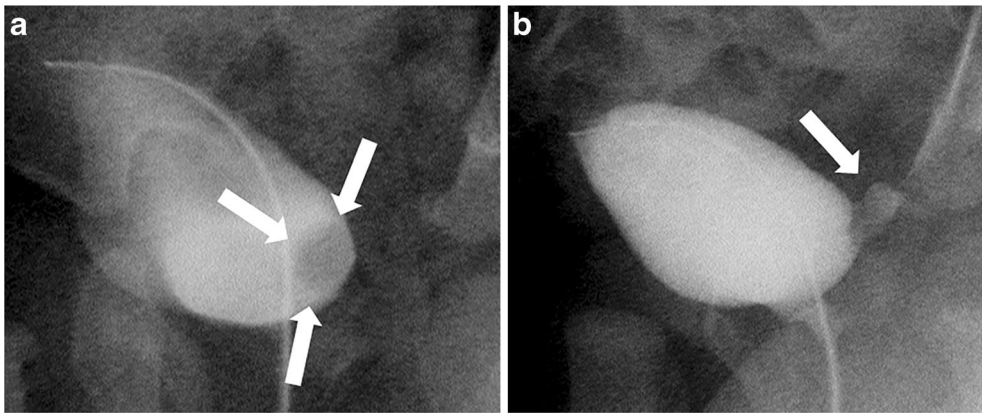

Fig. 13 Ureterocele on voiding cystourethrography (VCUG). **a, b** Two oblique lateral fluoroscopic images from a VCUG in a 10-month-old boy demonstrate (**a**) a filling defect (*arrows*) in the bladder representing a ureterocele, which (**b**) everts with bladder filling and mimics a bladder diverticulum (*arrow*)

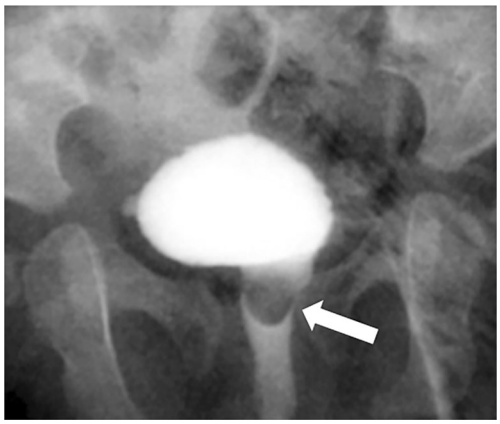

Fig. 14 Prolapsed ureterocele on voiding cystourethrography (VCUG). Anteroposterior fluoroscopic image from a VCUG in a 6-month-old girl with a ureterocele (identified as a bladder filling defect on early filling images, not shown) demonstrates prolapse of the ureterocele into the urethra with voiding (*arrow*)

Other imaging modalities to diagnose vesicoureteral reflux (VUR)

Radionuclide cystography (RNC) can also be used to evaluate VUR. Typically, RNC is used to follow known VUR because its sensitivity for detection of VUR is similar to that of VCUG but the radiation exposure is less [11]. However RNC is limited in its ability to delineate the anatomy of the ureters and kidneys and is insensitive in detecting duplex kidneys and associated anomalies. Voiding urosonography with contrast agents offers an additional method for detecting VUR in children and provides the benefit of no radiation exposure and

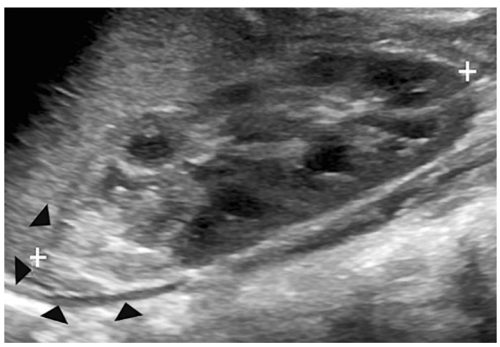

Fig. 15 Ureterocele disproportion. Sagittal US image of the left kidney in a 2-month-old girl with a ureterocele within the bladder (not shown) demonstrates a small echogenic residual upper pole parenchyma (*arrowheads*) and no calyceal dilation. This constellation of findings with a ureterocele and a small dysplastic upper moiety without calyceal dilation is described as "ureterocele disproportion"

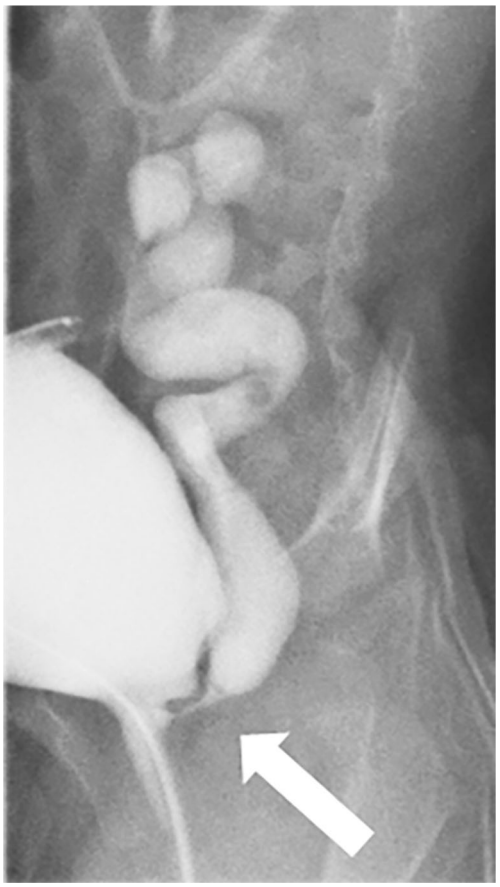

Fig. 16 Vesicoureteral reflux (VUR) on voiding cystourethrography (VCUG). Steep oblique fluoroscopic image from VCUG in a 2-month-old girl demonstrates vesicoureteral reflux into an ectopic left upper pole ureter inserting into the urethral sphincter (*arrow*), seen only during voiding

similar performance characteristics when compared to VCUG [12, 13]. In addition to detecting and characterizing voiding urosonography, the gray-scale US images obtained during the voiding urosonography can provide anatomical information, including that of duplex kidneys (Fig. 17).

Intravenous pyelography (IVP)

IVP was the traditional method to evaluate the appearance and function of the urinary tract. After intravenous injection of contrast agent, the appearance of the kidneys, ureters and bladder were evaluated by radiographs and fluoroscopy over time to assess for the appearance and function of the renal parenchyma and collecting system, to diagnose obstruction and scarring and other abnormalities. IVP provided the first radiologic method to show duplex kidneys and clearly show abnormal orientation of the renal collecting system, multiple ureters, ureteroceles and ectopic ureteral insertion sites, but now this has been supplanted by other imaging modalities.

Computed tomography (CT)

CT is not the primary imaging modality used in the evaluation of suspected urinary tract duplication, but duplication anomalies are often found incidentally. CT provides excellent spatial resolution and the ability to delineate complex anatomy. However this technique is typically used less frequently in children because of to its use of ionizing radiation. In contrast to adult imaging, where multiphase CT protocols are used, single-phase imaging is often adequate in the diagnosis of complete and incomplete urinary tract duplication in children. The findings of duplication anomalies include renal size discrepancy, a dividing cleft of renal parenchymal tissue, asymmetrical dilation of the upper and lower pole collecting systems, or two ipsilateral ureters (Fig. 18). The insertion site of the ectopic ureter can be delineated on the excretory phase images. CT might be most useful in children with complex anatomy that is not well-defined with VCUG, i.e. in absence of VUR. While information regarding renal function can be obtained with CT, other modalities such as nuclear medicine studies and MRU are used more frequently, and are more precise.

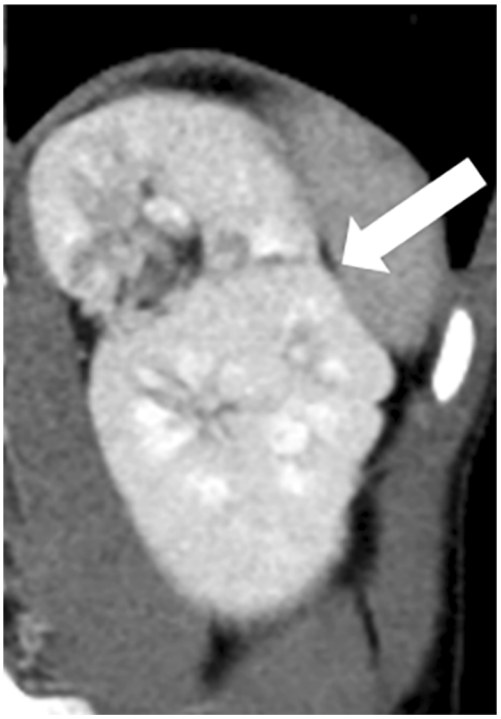

Fig. 18 Duplex kidney on CT. Coronal contrast-enhanced CT image in a 13-year-old boy demonstrates a parenchymal column (*arrow*) separating the upper and lower moieties in a duplex left kidney

Dimercaptosuccinic acid (DMSA) renal cortical scintigraphy

DMSA scans are typically performed to assess cortical function of the upper and lower moieties of the duplex kidney and can confirm suspected dysplasia and scarring of one or both moieties on anatomical imaging. The appearance of a duplex system on DMSA scans varies depending on the size and

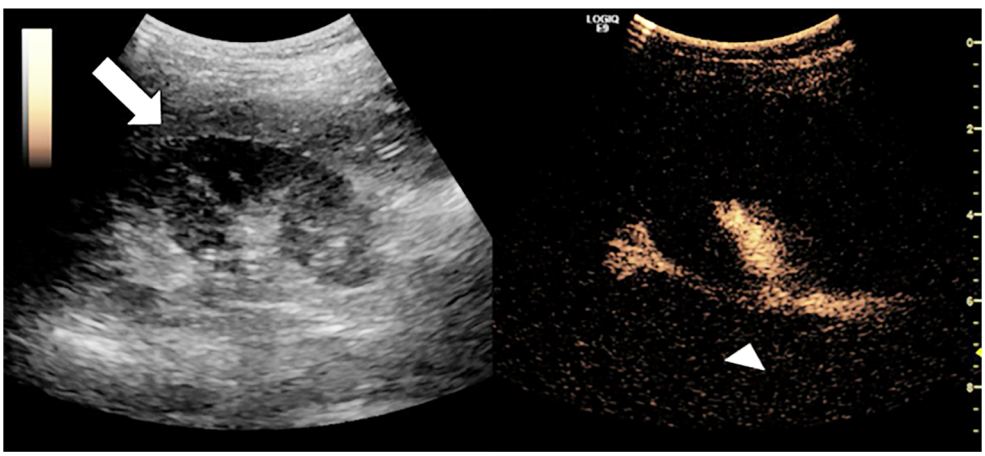

Fig. 17 Urosonography. Sagittal gray-scale image of the right kidney in a 9-year-old girl (*left image*) demonstrates a duplex kidney with a parenchymal bar of tissue separating the upper and lower poles (*arrow*). Simultaneous contrast-enhanced voiding urosonogram (*right image*) demonstrates reflux into a bifid ureter (*arrowhead* bifurcation)

function of each moiety and the nature and severity of any associated complications [14]. When there is no underlying parenchymal abnormality, renal cortical uptake is normal and duplication anomalies might not be recognized. Incidentally noted asymmetry of renal sizes on a DMSA scan sometimes provides a clue to underlying unilateral renal collecting system duplication, and this can be confirmed by morphological imaging such as ultrasonography. A duplex kidney frequently shows a cortical band with radiotracer uptake separating the upper and lower poles (Fig. 19). A dysplastic upper moiety of a duplex kidney might be seen as a subtle focal indentation along the medial aspect of the upper pole when small and nonfunctioning, or as focally decreased uptake when poorly functioning, and might mimic a focal cortical scar or pyelonephritis (Fig. 20). Hydronephrosis of one of the moieties, when present, appears as eccentric photopenia within the renal contour instead of the typical central photopenia that is seen with single-system hydronephrosis (Fig. 21). A duplex system might also appear as a small kidney located at the level of either the upper or the lower pole of the contralateral normal kidney, indicating nonfunctioning lower or upper moieties, respectively [8].

Functional information from a DMSA scan is often complementary to anatomical imaging. Assessment of differential function is very useful in clinical decision-making, particularly for surgical planning in children in whom partial nephrectomy might be a treatment option to surgically resect nonfunctional renal tissue.

Dynamic renal scintigraphy and diuresis renography

^{99m}Tc-MAG-3 (99 m–technetium mercaptoacetyltriglycine) renography is useful in the evaluation of differential renal function and differential drainage of the upper and lower moieties of the duplex kidney. It can show asymmetrical radiotracer uptake or unequal clearance between the moieties (Fig. 22). Diuresis renography can be used to assess suspected obstruction of the upper or lower moieties in a duplicated collecting system (Fig. 22). Following furosemide administration delayed or absent clearance of radiotracer is seen with high-grade obstruction at the level of either the UVJ (typically upper moiety) or the UPJ (uncommon, typically lower moiety). As with a DMSA scan, the examination might provide important functional information to complement anatomical imaging because nonfunctional renal tissue can be surgically resected.

Fig. 19 Posterior DMSA SPECT images of the kidneys in a 4-year-old boy demonstrate asymmetrical renal lengths, right longer than left. The duplex right kidney demonstrates asymmetrical uptake in the upper and lower moieties with diffusely decreased uptake of the right upper moiety, consistent with dysplasia. The left kidney is also duplex, with a prominent cortical column (*arrow*) separating the upper and lower poles. *DMSA SPECT* 99mTc-dimercaptosuccinic acid single-photon emission computed tomography

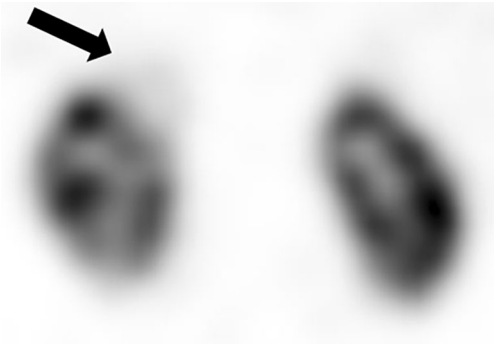

Fig. 20 Posterior DMSA SPECT image of the kidneys in a 76-day-old girl demonstrates a focal defect in the upper pole of the left duplex kidney. There is preserved cortex underneath the defect (*arrow*), suggesting a non-functioning/dysplastic upper moiety of a duplex kidney rather than pyelonephritis in a single-system kidney. *DMSA SPECT* 99mTc-dimercaptosuccinic acid single-photon emission computed tomography

Magnetic resonance urography (MRU)

MRU is a useful modality gaining popularity in the evaluation of urinary tract anomalies in children and provides important anatomical information, especially if complex anatomy is inadequately characterized by ultrasonography or VCUG [15]. Additionally, MRU can provide functional information, similar to that achieved with DMSA and ^{99m}Tc-MAG-3 scans, which might obviate the need for the nuclear medicine studies. The percentage of functional tissue in each kidney and each moiety can be calculated and the drainage parameters of each collecting system can be determined. While no ionizing radiation is employed, the risks of sedation should be considered in children.

The spatial resolution of MRU is less than than that of CT; however the contrast resolution is superior because MRU exploits the inherent contrast difference between the urine (fluid signal) and adjacent renal parenchyma and pelvic viscera (soft-tissue characteristics). Therefore the urine in the collecting system provides an ideal contrast agent in MRU. Findings of duplicated renal collecting systems are the same as on CT, including renal size discrepancy, a dividing cleft of renal

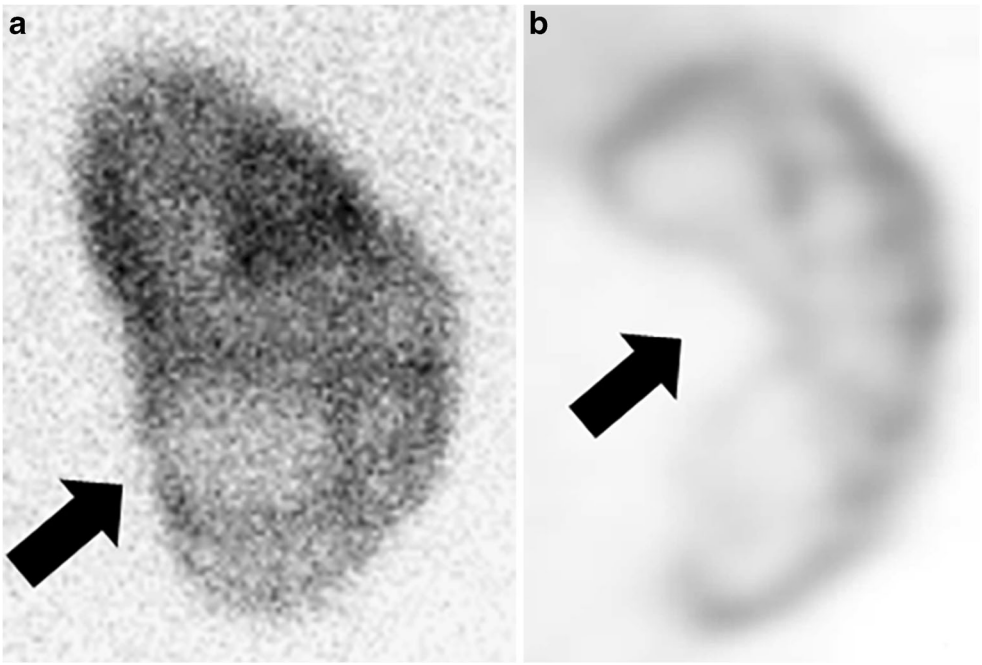

Fig. 21 Single-system hydronephrosis. **a** Posterior pinhole image of the right kidney on a DMSA scan in a 3-month-old boy demonstrates eccentric photopenia in the lower pole region of the right kidney (*arrow*) with preserved cortex in the upper pole, suggesting the presence of a duplex right kidney with hydronephrosis of the lower moiety and a normal upper moiety. **b** In comparison, a summed coronal SPECT image of a hydronephrotic single-system right kidney on a DMSA scan in a 1-year-old boy demonstrates more central photopenia (*arrow*), corresponding to pelvicalyceal dilation. *DMSA* 99mTc-dimercaptosuccinic acid, *SPECT* single-photon emission computed tomography

parenchymal tissue, asymmetrical dilation of the upper- and lower-pole collecting systems and multiple ureters (Fig. 23). Again, careful evaluation and description of the insertion site of the ectopic ureter is essential. MRU is valuable in the evaluation of ectopic ureters because they are frequently filled with T2-hyperintense fluid representing excreted urine (Fig. 24); therefore one needs very little renal function to opacify the collecting system, so delineation of the ureter can be made even in the setting of renal dysplasia. Additionally, excreted contrast material might opacify the ectopic ureter and the place where the ureter drains, e.g., the perineum or the vagina in girls with continuous urinary dribbling, or the ejaculatory ducts in boys (Fig. 25) [16]. With excellent anatomical resolution and inherent contrast (T2-hyperintense urine), MRU also provides an opportunity to visualize dysplastic renal parenchyma [17]. Furthermore, dynamic contrast-enhanced MRU can be used to obtain functional information that could show asymmetrical uptake or unequal clearance between moieties or kidneys

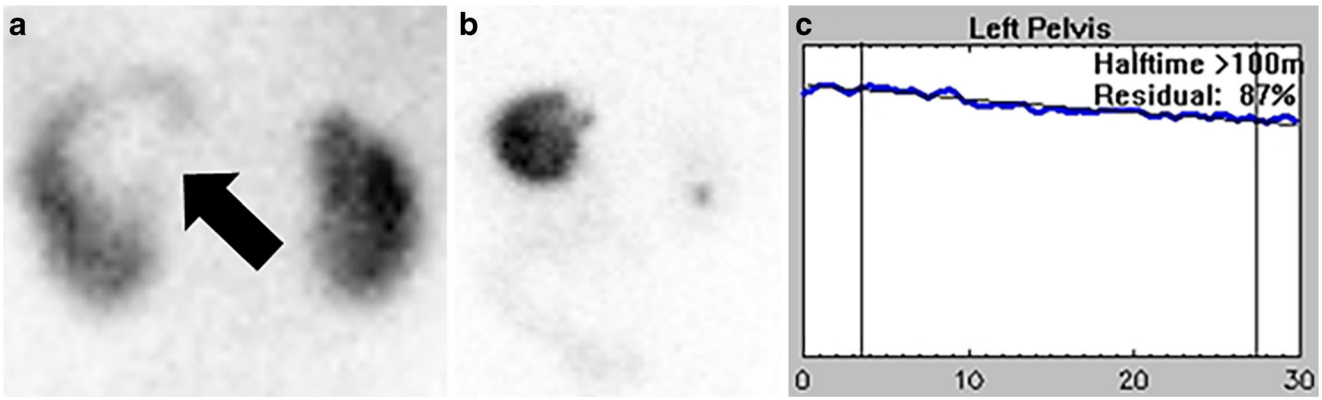

Fig. 22 ^{99m}Tc-MAG-3 (99 m–technetium mercaptoacetyltriglycine) scan in duplex kidneys. **a** Posterior planar image of the kidneys on a ^{99m}Tc-MAG-3 scan in the parenchymal phase demonstrates a duplex left kidney with hydronephrosis of the upper moiety. **b, c** Post-diuretic 30-min posterior planar image of the kidneys (**b**) and post-diuretic washout curve of the left upper moiety (**c**) depict no significant drainage of the tracer from the left upper moiety after administration of furosemide, indicating a high-grade obstruction in this 5-year-old boy with left upper pole hydronephrosis and ureterectasis seen on US (not shown)

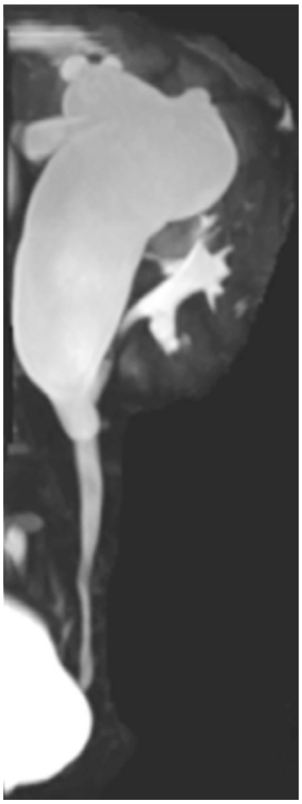

Fig. 23 MR urography findings. Coronal T2-weighted maximum-intensity projection reformation in an 8-year-old girl demonstrates upper pole pelvicalyceal dilation in a duplex left kidney

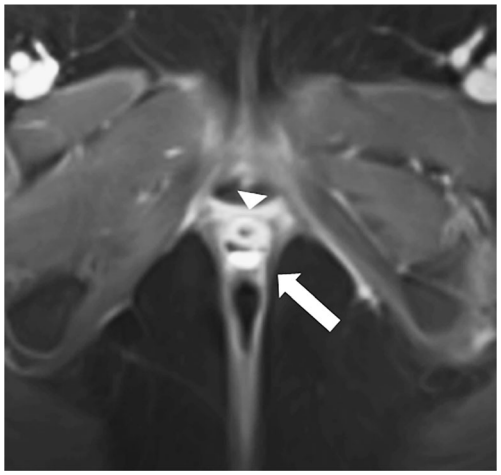

Fig. 25 MR urography in depicting ectopic ureteral insertion. Post-contrast axial T2-W fat-saturated MR image in a 9-year-old girl demonstrates the urethra (*arrowhead*) containing a bladder catheter and excreted contrast material opacifying the vagina (*arrow*), suggesting a site of ectopic ureteral insertion

in the setting of urinary tract duplication (Fig. 26). With MRU, one imaging modality can provide both anatomical and functional information in complex cases of urinary tract duplication.

Ultimately, multiple imaging modalities provide opportunities to diagnose and characterize renal collecting system duplication, and many are complementary in the evaluation of the child. The radiologist should review all of the uroradiologic imaging studies available in order to synthesize the information into a cohesive understanding of the child's anatomy and potential or existing complications. This understanding provides crucial information for treatment planning.

Conclusion

Duplication of the upper urinary tract is a relatively common anomaly that can be an asymptomatic normal variant or, when

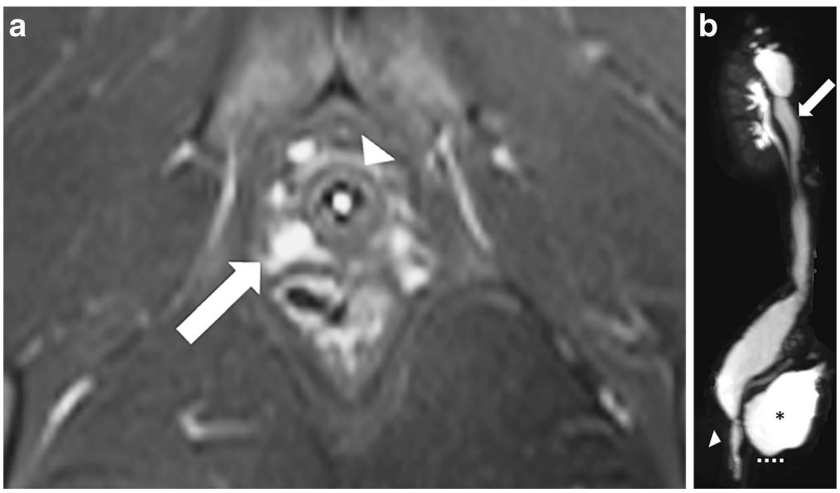

Fig. 24 MR urography in depiction of ureters. **a** Axial T2-W fat-saturated MR image in a 5-year-old girl with a duplex right kidney demonstrates the right upper pole ureter extending below the urinary sphincter (*arrow*) at the level of the urethra (*arrowhead*). **b** Oblique sagittal maximum-intensity projection MR image demonstrates a duplex right kidney with a dilated upper pole ureter (*arrow*). The distal ectopic right upper pole ureter (*arrowhead*) extends inferior to the urinary bladder (*asterisk*) and the level of the external sphincter (*dotted line*)

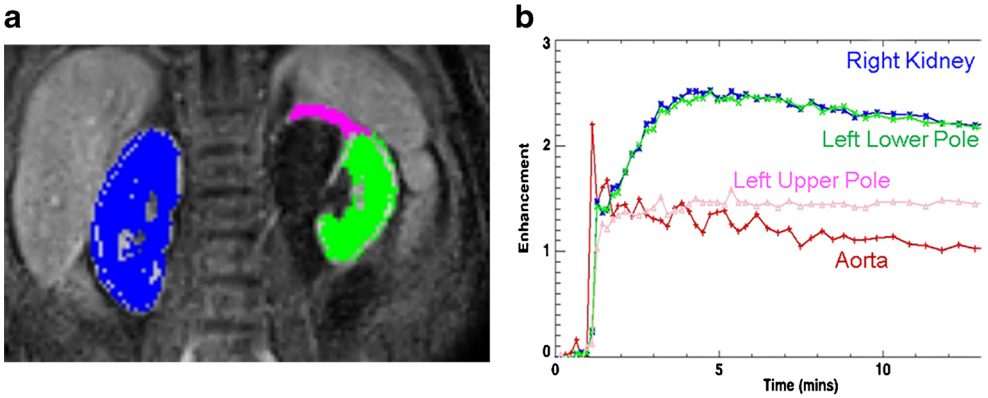

Fig. 26 MR urography for obtaining functional information. **a** Coronal image of the kidneys from a dynamic contrast-enhanced sequence in an 8-year-old girl depicts the regions of interest of the single-system right and duplex left kidneys. **b** Renal parenchymal enhancement curves demonstrate decreased enhancement of the left upper pole parenchyma when compared to both the right kidney and left lower pole

abnormal, might be associated with VUR, a ureterocele or obstruction, as well as renal dysplasia, scarring and impaired renal function. Understanding the embryology, imaging and clinical consequences of urinary collecting system duplication is essential for early detection and diagnosis. The first step is awareness of duplex anomalies and vigilance with regard to the signs seen on all imaging modalities, especially ultrasonography. The second step is understanding how the basic rules governing duplex anomalies serve to guide a thoughtful evaluation of the child. Each imaging modality can contribute to understanding the effect of the duplex anomaly in each child. With many imaging options available, uroradiologic imaging can and should be tailored to the specific clinical question.

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Compliance with ethical standards

Conflicts of interest None

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