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Magnetic resonance urography of congenital abnormalities — what the radiologist needs to know

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Abstract

Congenital abnormalities of the kidney and urinary tract include a wide range of malformations ranging from asymptomatic to life-threatening conditions. Although pediatric urogenital system imaging is based on the use of US (pre- and postnatal), voiding cystourethrography and scintigraphic study, magnetic resonance (MR) urography plays a fundamental role in the classification and management of congenital abnormalities of the kidney and urinary tract, giving an overview of the different clinical pictures, thanks to its panoramicity and high anatomical detail. In fact the anomalies of the urinary tract are phenotypically variable because they can affect simultaneously several segments of different embryonic derivation, with complex clinical pictures; they can appear both as isolated phenotypes or as complex malformative conditions, involving renal parenchyma, collecting system and bladder. A deep knowledge of this complex embryogenesis and its possible phenotypic patterns allows a correct interpretation of MR urography images. We describe the embryology and pathophysiology of congenital abnormalities of the kidney and urinary tract as well as MR urography technique and findings. Congenital abnormalities of the kidney and urinary tract are classified into four groups: (1) obstruction (proximal, middle and distal), (2) budding with respect to the Wolffian duct (site and number of ureter), (3) ascent and rotation (ectopia, malrotation and fusion of kidney) and (4) anomaly of metanephric differentiation (dysplasia, megapolicalycosis).

Keywords Children · Congenital abnormality · Embryology · Imaging technique · Kidney · Magnetic resonance urography · Urogenital system

Introduction

Congenital anomalies of the kidney and urinary tract are a spectrum of malformations resulting from either an abnormal development of the ureteric bud from the Wolffian duct or from an abnormal mutual interaction between the ureteric bud and the metanephric blastema during organogenesis. Congenital abnormalities of the kidney and urinary tract affect about 1 in 500 newborns and account for 20–30% of all abnormalities identified in the prenatal period. Some anomalies are asymptomatic, but many malformations are relevant causes of infant mortality as well as morbidity in older children and adults, including the progression to renal failure [1]. Although imaging of the pediatric urogenital system relies on the use of US, it is not sufficient; MRI is currently considered the cross-sectional examination of choice to define abnormalities. In fact, MRI is a useful technique to recognize and accurately describe the various phenotypes of congenital anomalies of the kidney and urinary tract,

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including complex cases. Several papers have described the congenital anomalies of the kidney and urinary tract in detail [2–6]; in this paper, however, we give a comprehensive overview and a simple and quick classification that is based on the link between the embryonic developmental abnormalities and clinical phenotypes (Table 1). During embryogenesis, three phases of development of the urogenital system can be distinguished (Fig. 1). Initially, kidneys are located in the pelvic area; thereafter, they gradually migrate cranially (Fig. 2). Nephrogenesis ends at approximately 34 weeks of gestation [7].

Magnetic resonance urography protocol

Magnetic resonance (MR) urography has an increasing role in congenital abnormalities of the kidney and urinary tract thanks to its high spatial and contrast resolution and the lack

Table 1 Summary of the physiopathological and phenotypic correlation of congenital abnormalities of the kidney and urinary tract

Physiopathology	Phenotype
Obstruction: proximal, middle and distal	Ureteropelvic junction obstruction Congenital midureteral obstruction Megaureter Ureterocele
Budding with respect to the Wolffian duct: site and number of ureters	Duplex system escretory (bifid pelvis, complete and incomplete double ureters) Zinner syndrome
Ascent and rotation: ectopia, malrotation and fusion of kidney	Simple ectopia Cross ectopia Horseshoe kidney Sigmoid kidney L-shape kidney Discoid kidney Cake kidney
Anomaly of metanefric differentiation	Hypoplasia Dysplasia Multicystic dysplastic kidney Megapolicalycosis

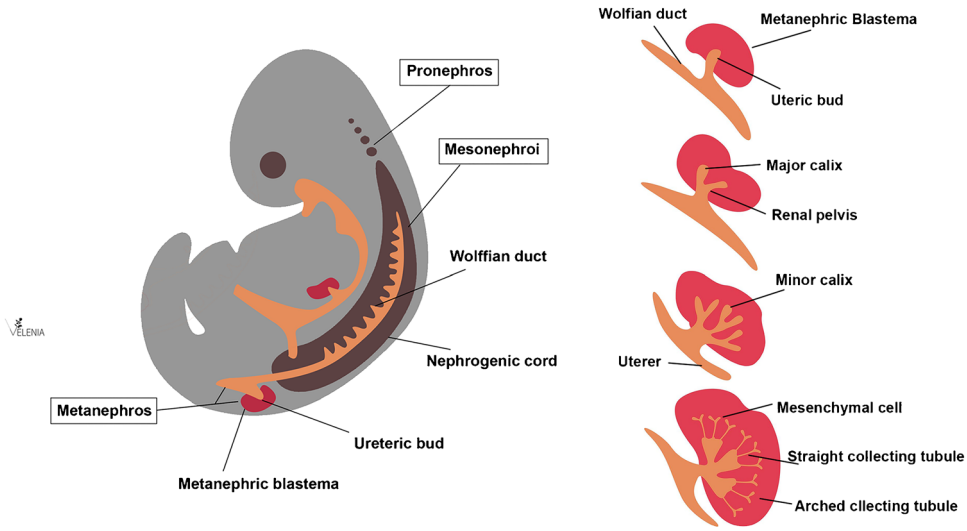

Fig. 1 Urogenital embryogenesis illustration. **a** Three stages of the urogenital system development: rudimentary pronephros (before the 4th week); mesonephroi (middle of the 4th week) composed of the nephrogenic cord and the Wolffian duct; metanephros, definitive excretory system (5th week), composed of the ureteric bud and metanephic blastema. **b** Development of renal parenchyma and excretory system: the mutual interactions between the ureteric bud and the metanephric blastema induce the development of the kidney and the excretory system. The ureteric bud forms a peduncle that develops into the ureter. Its cranial expansion becomes the renal pelvis while the mesenchymal cells of the metanephric blastema gradually evolve into the primordial nephrons

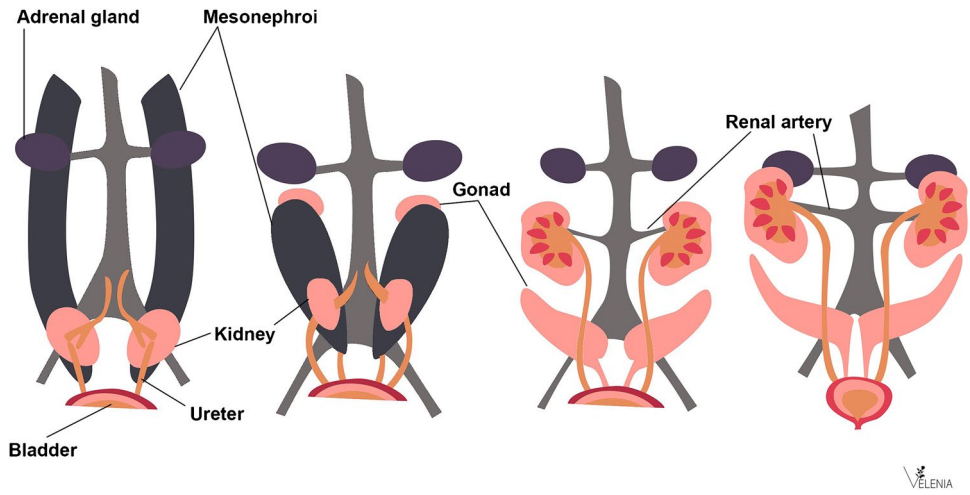

Fig. 2 Renal embryogenesis illustration. During the development of abdomen and pelvis (6th–9th weeks), kidneys migrate cranially, reaching the adrenal gland in the retroperitoneum, while the mesonephros disappears. At the same time, hilum rotates 90°, from ventral to medial position, crossing vessels and nerves; at the 9th week, the hilum is placed anteromedially. The vascularity of the kidneys originates from branches of the common iliac arteries. During the ascent they receive new branches from the aorta while the rudimentary arteries gradually disappear; this is why it is common to find anatomical variants of the renal blood supply

of ionizing radiation. Therefore, MR urography can combine an anatomical–morphological study of the kidney and the excretory system, with functional information obtained from the quantitative analysis of a three-dimensional (3-D) sequence performed after contrast medium injection ("one-stop shop" technique) [8].

Commonly, MR urography plays a crucial role in different clinical scenarios. It is in fact a complementary imaging tool to US in the diagnosis of complex anatomical malformations

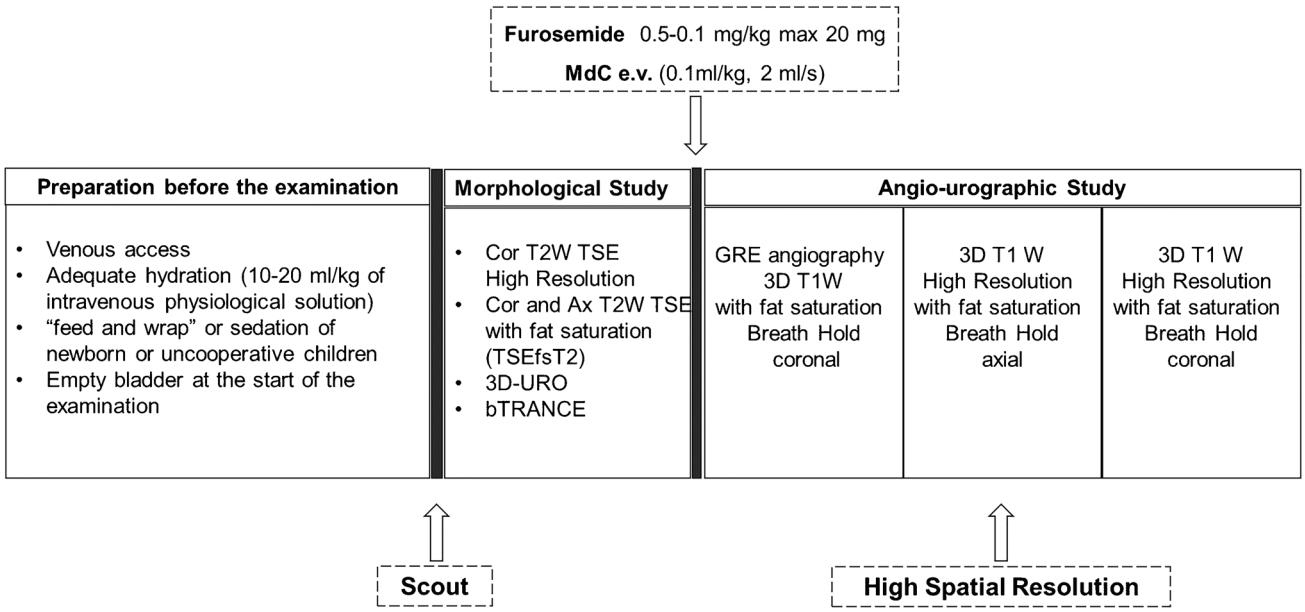

Fig. 3 Magnetic resonance urographic protocol. Intravenous hydration and bladder emptying are required before the exam. After the morphological study with T2-weighted sequences, furosemide is administered. The functional part of the study is based on dynamic acquisitions after injection of contrast medium. In our clinical practice, angiographic (bolus tracked), nephrographic (80–140 s) and urographic (8–12 min) phases are acquired. *3D* three-dimensional, *Ax* axial, *bTRANCE* balanced triggered angiography non-contrast enhanced, *Cor* coronal, *GRE* gradient echo, *T1W* T1-weighted, *T2W* T2-weighted, *TSE* turbo spin echo, *URO* volumetric hydrographic sequence

Table 2 Tips and tricks: possible solutions to common magnetic resonance (MR) urography problems

Problems	Solutions
Poor-quality image: over-distension of bladder, uncooperative children	Foley catheter Feed-and-wrap approach with free-breathing or sedation
Child with glomerular filtration rates less than 30 mL/min/1.73	Use an MR hydrography technique without contrast medium, possibly repeating the sequence until good image quality is achieved
Loss of signal in the excretory tract in post-contrast T1-weighted images due to T2*	The use of furosemide, intravenous hydration and administration of half a dose of contrast medium reduces the concentration of the contrast medium and the T2* effect
Partial or intermittent obstruction without clear hydronephrosis	Furosemide treatment and intravenous hydration can make hydronephrosis more evident
Unrecognizable distal ureter, ureteral insertion or suspected ectopic ureter	Acquire a smaller field of view, preferably 3-D T2-weighted fast spin echo and post-contrast 3-D T1-weighted gradient echo

of the kidneys and urinary tract, and it establishes the possible etiology of the obstruction (intrinsic or extrinsic causes) in cases where the type of urinary obstruction is difficult to classify with first-line imaging methods.

Functional MR urography helps in therapeutic choices (clinical follow-up or surgical therapy) and in the selection of the best surgical approach (minimally invasive surgery or pyeloplasty). Furthermore, it is useful for a study of the surgical anastomoses of the urinary tract already treated (i.e. postoperative controls in obstruction of the ureteropelvic junction or of the reimplanted ureters) [2].

Patient hydration before the MR urography investigation is needed (10–20 mL/kg of intravenous physiological solution). Venous access must be placed in all children before the examination. Collaborating children are asked to empty the bladder before MR urography and a bladder catheter is placed in those who undergo MR under sedation. For non-collaborating children younger than 6 months, the feed-and-wrap technique can be used [9], while for older non-collaborating children, sedation is performed according to institute protocols. Children are positioned supine on the imaging table with arms placed over the head.

At our hospital MR images are acquired using a 1.5-tesla (T) MR system (Achieva Intera; Philips Healthcare, Best, The Netherlands) with a body or cardiac coil according to patient size. Before contrast administration, two sequences are performed: a rapid free-breathing morphological T2-weighted sequence (with and without fat saturation) and a volumetric hydrographic sequence, which allows a strong contrast of fluids within the excretory system.

A balanced free-breathing cardiac-triggered 3-D sequence combined with a slab-selective inversion prepulse sequence without contrast enhancement is acquired (balanced triggered angiography non-contrast enhanced MRI, or bTRANCE-MRI) for non-contrast angiographic mapping. Furosemide is used to improve visualization of the urine flow and to promote distribution and dilution of contrast medium. It is administered before contrast (F0), with dose of 1 mg/kg in infants and 0.5 mg/kg in older children at a maximum dose of 20 mg, as is explained in detail in the work of Vivier et al. [10]. It is not recommended to

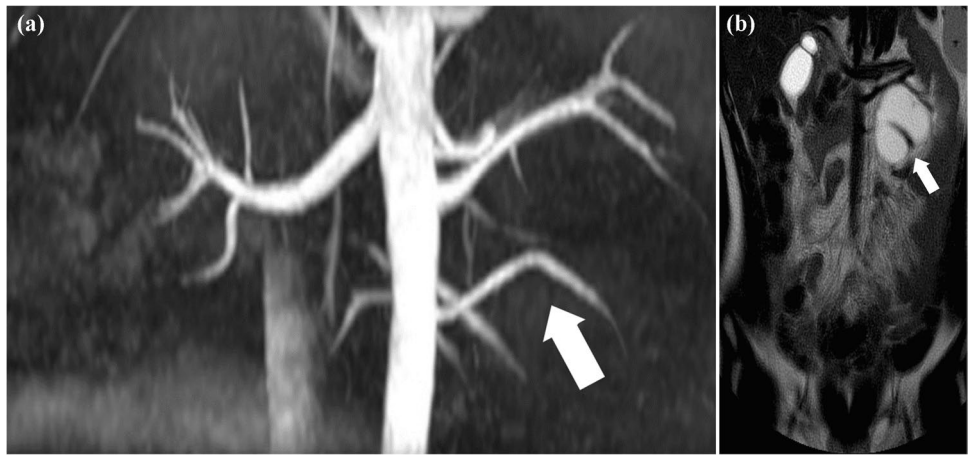

Fig. 4 Ureteropelvic junction obstruction in a 13-year-old girl. **a, b** Coronal gradient echo three-dimensional T1-weighted maximum-intensity projection reconstruction **a** and coronal T2-weighted image **b** show left inferior polar artery (*arrow*), which is the cause of ureteropelvic junction obstruction

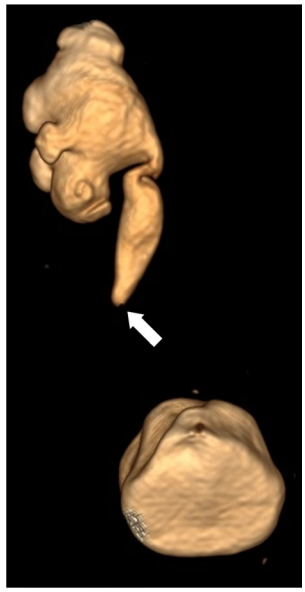

Fig. 5 Congenital midureteral obstruction in a 6-year-old girl. Coronal volume-rendered three-dimensional MRI reconstruction shows midureteral stenosis (*arrow*), with associated dilation of the proximal ureter and calico-pyelic system

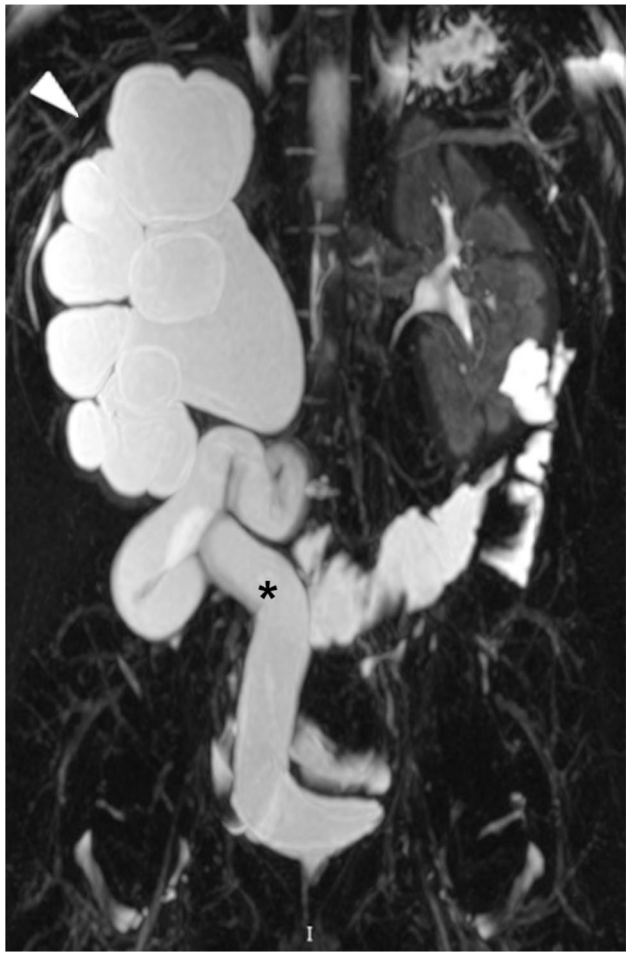

Fig. 6 Primary megaureter in a 5-year-old girl. Coronal three-dimensional urographic maximum-intensity projection reconstruction shows the whole dilated excretory system with convoluted ureter (*asterisk*) and thinning of the right renal parenchyma (*arrowhead*)

administer the contrast medium to infants younger than 1–2 months because of their renal immaturity. In older children a low dose of gadolinium (0.05–0.1 mmol/kg) is injected to limit the T2* effects of high concentrations in the aorta, the medulla and the collection system [10]. After injection of contrast medium, angiographic (bolus tracked), nephrographic (80–140 s) and urographic (8–12 min) phases are acquired. Figure 3 summarizes the MR urographic protocol recommended by the uroradiological task force of the European Society of Paediatric Radiology and the European Society of Urogenital Radiology Paediatric Working Group [8]. Table 2 shows technical problems and solutions in MR urography [2].

Anomalies associated with obstruction

Congenital obstructions in childhood usually occur as a proximal, middle or distal form.

Ureteropelvic junction obstruction

Ureteropelvic junction obstruction is the most frequent cause of obstructive uropathy and pelvicalyceal dilation in children, with an incidence of 1 in 1,500 newborns [11]. It is characterized by a complete or partial obstacle to the normal outflow of urine from the pelvis to the proximal tract of the ureter.

The obstruction can be intrinsic or extrinsic (Fig. 4). The vascular obstructive form is the most frequent extrinsic cause, usually determined by an accessory artery in conflict with the ureteropelvic junction [12]. The coexistence of an aberrant artery and an instrinsic obstruction has also been described as a mixed form [13]. Rarer causes are adhesions, retrocaval ureter, lymphadenopathy, retroperitoneal neoplasms and retroperitoneal fibrosis. The intrinsic ureteropelvic junction obstruction is characterized by an abnormal development of smooth muscle and innervation during the fetal age with the consequent absence of peristalsis and propulsive wave propagation from the pelvis to the ureter [13]. It has been described that the spiral musculature is replaced by longitudinal muscle or fibrous tissue [14].

Congenital midureteral obstruction (congenital midureteral stricture, or ureteral valve)

Congenital midureteral obstruction is a rare cause of pelvicalyceal dilation that is often mistaken for ureteropelvic junction obstruction. Second-level imaging like MR urography retrograde pyelography are often necessary for an accurate diagnosis (Fig. 5). The abnormality results from a failed recanalization of the ureter caused by a prenatal vascular insult or persistence of a ureteral valve [15].

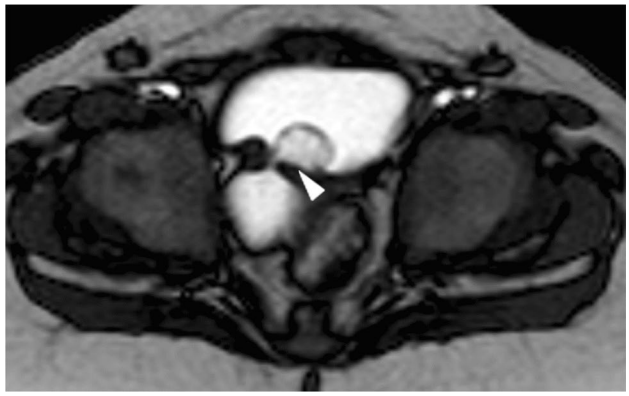

Fig. 7 Ureterocele in a 5-month-old boy. Axial balanced fast field-echo MRI shows a ureterocele, which appears as a cystic lesion projecting into the bladder (*arrowhead*)

Megaureter

A megaureter is a markedly dilated ureter (>7 mm) (Fig. 6) [16]. It is more common in males (4:1) and on the left side; it can be bilateral in 15–25% of cases. It results either from an idiopathic congenital immaturity of the vesicoureteral junction (primary) or bladder and urethral disorders (secondary) [17]. The primary form is characterized by the last 3–4 cm of an adynamic segment, which does not allow the passage of the peristaltic wave. Parietal muscle tissue is disorganized or absent, largely replaced by collagen tissue [18].

Ureterocele

The term "ureterocele" refers to the congenital cystic dilation of the intravesical tract of the ureter (Fig. 7). It can be very small but also so large that it fills the entire bladder lumen and prolapses into the urethra; it can be simple when localized in the intravesical area with the ureter opening in the trigone of the bladder, or ectopic when the outlet, with associated cystic dilation, is found medially and below the bladder neck. It often drains the upper pole of a kidney with a duplex complete collection system, acting, therefore, as an obstructive factor [19].

Budding with respect to the Wolffian duct

Defects in ureteral budding from the Wolffian duct often manifest themselves as anomalies affecting both the renal parenchyma and the collecting system, as shown in Fig. 8 (the most frequent phenotypes). The absence of development of the ureteric bud from the Wolffian duct leads to the failure of the kidney and excretory system to develop. On the other hand, the development of the number of ureters and kidneys depends, respectively, on the number and timing of duplication of the ureteric bud and the metanephric blastema at the end of the 4th week of gestation. When there is premature division of the ureteric bud or a double ureteric bud, two separate collecting systems might be formed within the kidney with development of two separate renal pelves. There could be a complete double ureter, or the ureters might join anywhere along their course before entrance into the bladder [20–25].

In addition, a supernumerary kidney develops when duplication of the metanephric blastema occurs simultaneously with the ureteric bud. MR urography might be needed to fully understand upper urinary tract duplication abnormalities (Table 3) [26].

Duplex collecting system

Duplex renal collecting systems are one of the most common congenital anomalies of the urinary tract (Fig. 9). They occurs when two separate ureteric buds arise from a single Wolffian duct [27]; they have been described as unilateral or bilateral and complete or incomplete [26].

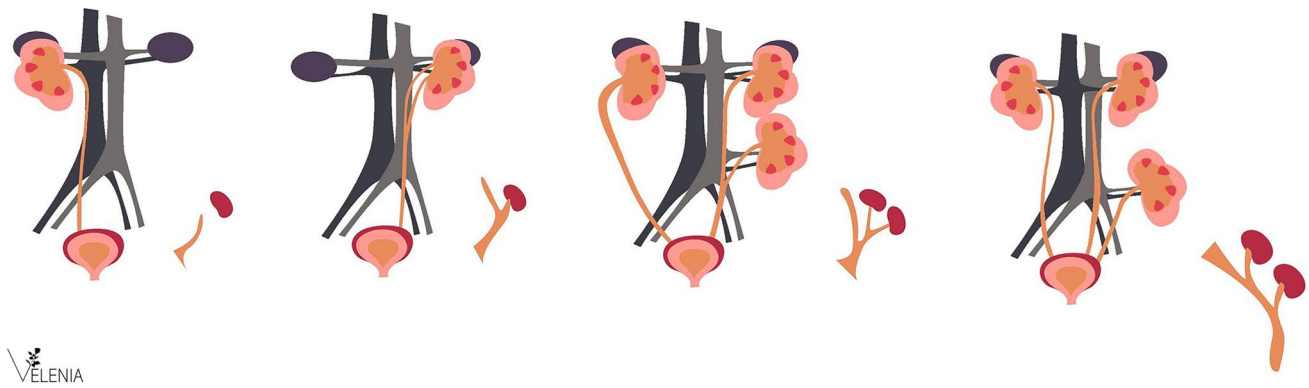

Fig. 8 Illustration of the most frequent phenotypes of possible defects in the interaction between the ureteral bud and Wolffian duct

Table 3 Indications for magnetic resonance (MR) urography in upper urinary tract duplication abnormalities

Indications for MR urography
To confirm the presence of a duplex kidney
To determine whether the duplication abnormality is incomplete or complete
To establish the exact path of the ureters and their insertions
To evaluate the differential function
To evaluate other coexisting congenital anomalies such as pelvi-ureteric junction obstruction, ectopic ureter, ureterocele
To identify any scar of the renal parenchyma, diffuse thinning or dysplasia of the same

Incomplete ureteral duplication originates from a ureteral bud that bifurcates shortly after its origin from the Wolffian duct. If the division occurs after the ureteric bud penetrates into the metanephric blastema, a bifid pelvis results; if it occurs before the 5th week of gestation, varying degrees of ureteral duplication result with fusion distally as a single ureter entering the bladder [25].

A duplex kidney with complete double ureters requires two ureteric buds arising from the Wolffian duct (Fig. 10). The bud that arises more cranially on the mesonephric duct extends to the upper pole of the kidney, and the caudally positioned bud drains the lower pole (Weigert–Meyer rule) [25].

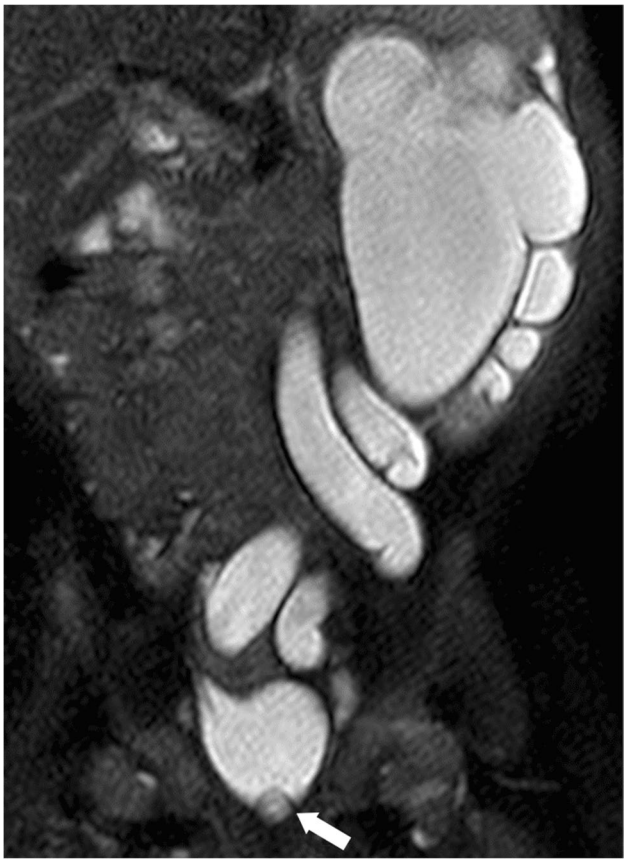

Fig. 9 Duplex collecting system in a 3-month-old boy. Coronal turbo spin-echo fat-saturated T2-weighted MR image shows severe calico-pyelo ectasia, ureteral dilation with thin renal parenchyma, and loss of corticomedullary differentiation. Moreover, it shows the two ureters as tortuous and convoluted, crossing several times with an ectopic insertion of the upper ureter in the bladder neck (*arrow*)

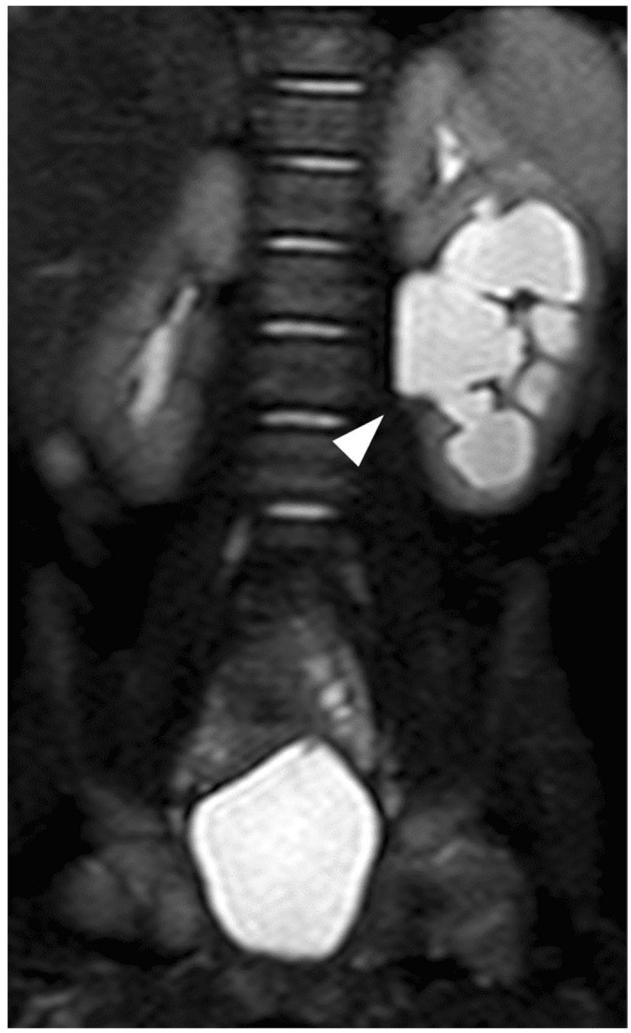

Fig. 10 Duplex collection system and ureteropelvic junction obstruction in a 5-month-old boy. Coronal turbo spin-echo fat-saturated T2-weighted MR image shows the upper pole composed of a single calyx and the lower pole with pelvicalyceal dilatation caused by an intrinsic ureteropelvic junction obstruction (*arrowhead*)

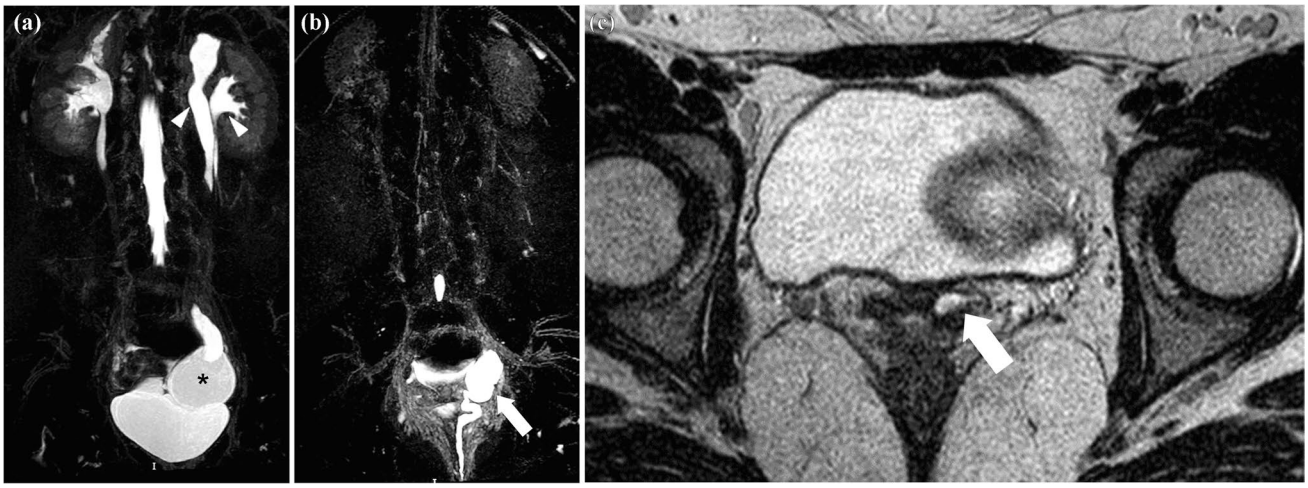

Fig. 11 Duplex collection system in an 11-year-old girl. **a**, **b** Coronal turbo spin-echo fat saturated T2-weighted images show a complete duplex collection system (*arrowheads*) of the left kidney. The ureter of the upper pole ends as ureterocele (*asterisk*) in the dome of the bladder while the lower ureter pole ends in the vaginal vault (*arrow*). **c** Axial turbo spin-echo T2-weighted image shows the ectopic site of the ureter of the lower pole which inserts at the level of the vaginal vault (*arrow*)

The trigone of the bladder arises from the interaction between the Wolffian duct and the cloaca. The ventral cloaca develops into the urogenital sinus and thereafter into bladder and urethra, while the common excretory duct develops from the Wolffian duct. This duct gradually expands to be incorporated into the urogenital sinus and forms a portion of the trigone and the underlying detrusor musculature.

Typically, the ureteric orifice draining the upper pole develops caudal to the normal location and it might be ectopic. In girls, the ectopic ureter can be located in the vagina or perineum, causing incontinence (Fig. 11) [28]. In boys, the ectopic ureter is always cranial to the sphincteric plane and incontinence is caused by a bilateral ectopia that distorts the bladder neck. This is frequently associated with ureterocele, often causing obstructive hydronephrosis, while the ureter of the lower pole is more prone to reflux caused by a defect in its intramural path, leaving it without the anti-reflux mechanism [29].

Zinner syndrome

Zinner syndrome is a triad of Wolffian duct anomalies characterized by unilateral renal agenesis, ipsilateral ejaculatory duct obstruction and seminal vesicle cyst on the side of the obstruction (Fig. 12). It is rare, with only 200 cases reported in the available literature [30]. The ipsilateral ureter can be absent or incomplete [31–33]. Generally, the dilated ureteral remnant arises from a supernumerary ureteric bud or from the inability of a normal ureteric bud to connect with the metanephric blastema, thus causing renal agenesis [34].

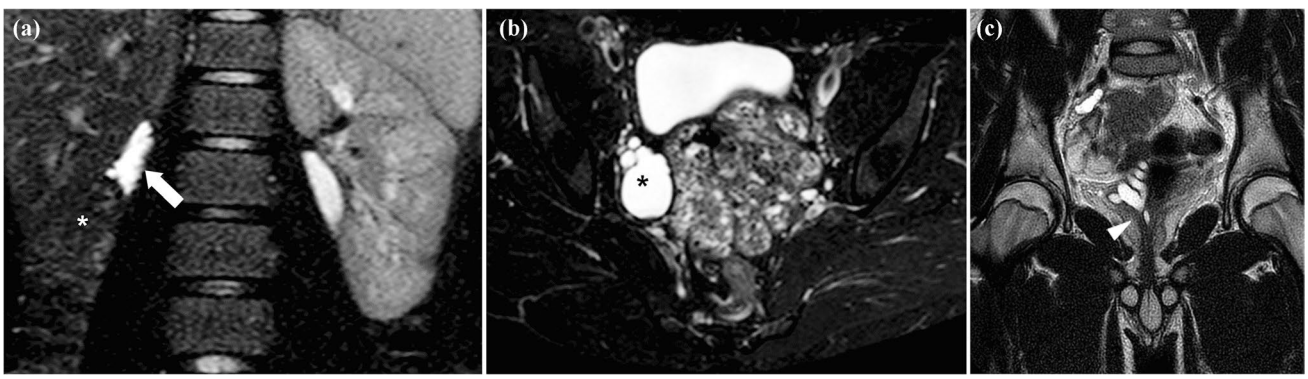

Fig. 12 Zinner syndrome in a 13-year-old boy. **a** Coronal turbo spin-echo fat-saturated T2-weighted MR image shows right renal agenesis (*asterisk*) and residual ureteric bud (*arrow*). **b** Axial turbo spin-echo fat-saturated T2-weighted MR image shows right seminal vesicle cyst (*asterisk*). **c** Coronal turbo spin-echo T2-weighted MR image shows ejaculatory duct obstruction (*arrowhead*)

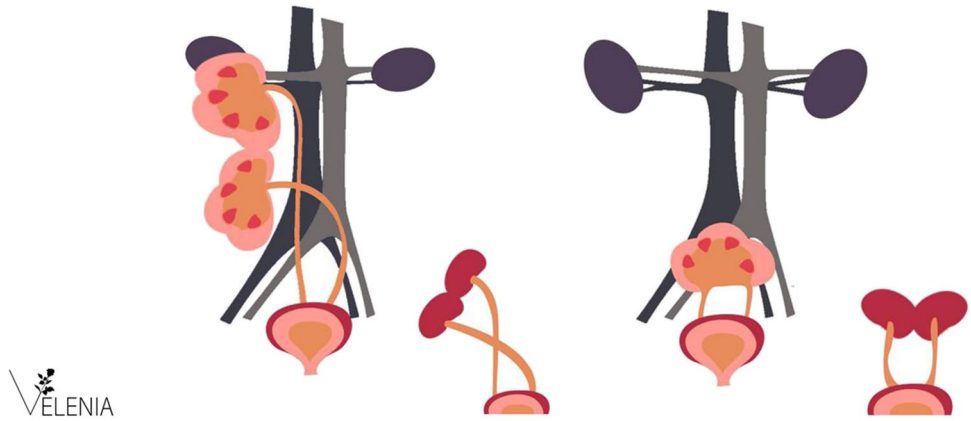

Fig. 13 Illustration of phenotypes of anomalies of ascent and fusion of the kidneys. Ascent arrest of the kidney can take place at any level and results in abnormal location in the abdomen. Crossed renal ectopia is defined as the presence of the kidney in the opposite retroperitoneal space, which can often lead to fusion of the kidneys, resulting in crossed fused renal ectopia. In horseshoe kidney there is a fusion of two distinct functioning kidneys on the midline. They are connected by an isthmus that consists of either functioning renal parenchyma or fibrous tissue

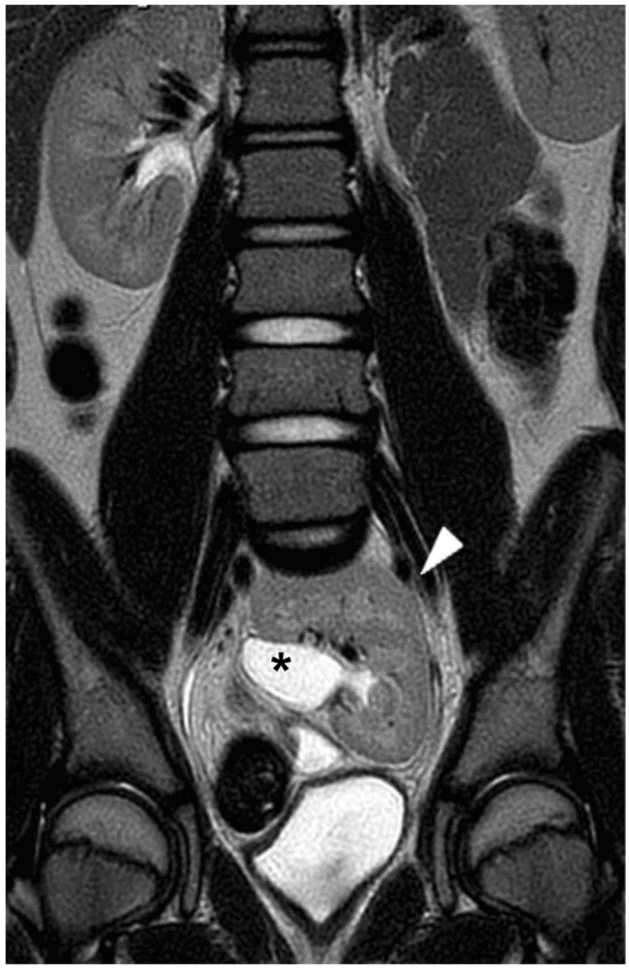

Fig. 14 Simple ectopic left kidney in a 10-year-old boy. Coronal turbo spin-echo T2-weighted MR image shows an ectopic left kidney, located in hypogastric region, malrotated (*arrowhead*) and with associated ureteropelvic junction obstruction (*asterisk*)

The dilated ureteral residue can be orthotopic (trigone of the bladder) or ectopic.

Ascent and rotation

Incorrect ascent of the ureteric bud and metanephric blastema can result in both renal fusion and position abnormalities (Fig. 13). Ectopia occurs whenever the kidney is

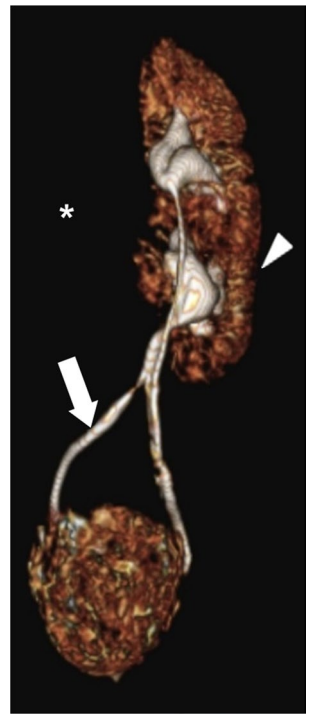

Fig. 15 Cross ectopic right kidney in a 7-year-old girl. Coronal three-dimensional volume-rendered MR reconstruction shows the empty right renal lodge (*asterisk*), the right kidney placed on the left side (*arrowhead*) and the ureteric outlet into the bladder on the right (*arrow*)

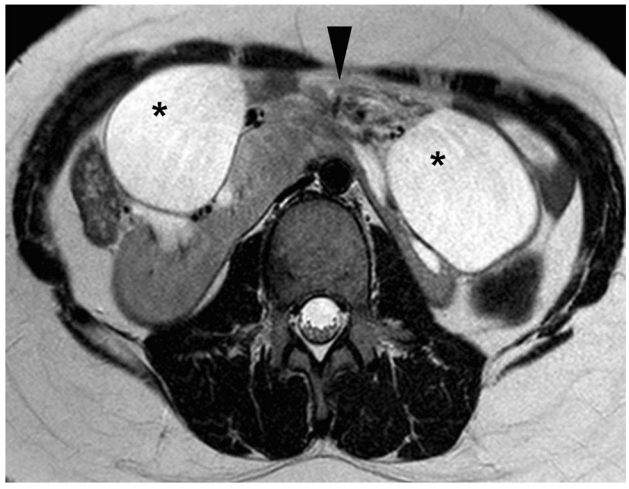

Fig. 16 Horseshoe kidney in a 10-year-old girl. Axial turbo spin-echo T2-weighted MR image shows fusion of the lower poles of the kidneys at the midline (*arrowhead*). Both renal hila are rotated anteriorly with associated bilateral ectasia of the pyelo-ureteral junction (*asterisks*), secondary to high insertion of both ureters

located outside the flank region (vertebral levels L1–L3). There are two forms: simple ectopia, when the ectopic kidney is located on the same side as its ureter (1 in 3,000 cases) [35] (Fig. 14); and cross ectopia, when the ectopic kidney is on the other side of the ureter, with an estimated incidence of 1 in 1,300–7,500 [36] (Fig. 15). Fusion anomalies arise probably between the 4th and 8th weeks of gestation, when because of abnormal flexion of the fetal spine and the mesenteric artery engaging the isthmus, there is an arrest of the ascent of the metanephric blastema [6]. The most frequent of the fusion anomalies is the horseshoe kidney, with an estimated incidence of 1 in 500 live births (Fig. 16) [37]. Less common patterns include sigmoid, L-shape, discoid and pie kidneys.

Metanephric differentiation anomaly

Dysplastic disease includes a heterogeneous group of congenital, hereditary and non-hereditary diseases characterized by the presence of cystic formations. It generally occurs sporadically or within the context of a multisystemic syndrome. Dysplastic kidneys are common: unilateral dysplastic kidneys occur in 1 of 1,000 births, and bilateral in 1 of 5,000 (Fig. 17) [38].

Embryologically, the dysplastic kidney arises either from aberrant interactions of the ureteric bud and metanephric mesenchyme or because of an obstruction [39]. Dysplastic kidneys are characterized by small size, disorganized architecture with loss of normal corticomedullary differentiation and small subcortical cysts, variable in size and number (Fig. 18), with a delayed excretion and dysmorphic calyces better appreciated in urographic sequences [40]. Multicystic dysplastic kidney is commonly diagnosed and is seen in approximately 1 of 4,300 live births. Multicystic dysplastic kidneys might persist without notable change, or increase in size or undergo spontaneous involution [40].

Megapolicalycosis is a very rare anomaly; the male/female ratio is 6:1, and most cases are asymptomatic (Fig. 19). It originates from a medullary hypoplasia with consequent dilation and multiplication (about 12 or more) of the calyces without signs of obstruction, with the renal pelvis and ureters appearing normal (Fig. 20). Embryologically, the condition is caused by an abnormality in the development of the metanephric blastema in the collecting ducts. The medullary portion is hypo-developed with a crescent shape and papillary flattening [41].

Conclusion

Magnetic resonance urography is a one-stop shop non-irradiating tool for a second-level diagnosis of a wide variety of kidney and urinary tract anomalies. The early diagnosis reduces the number of tests required for a correct

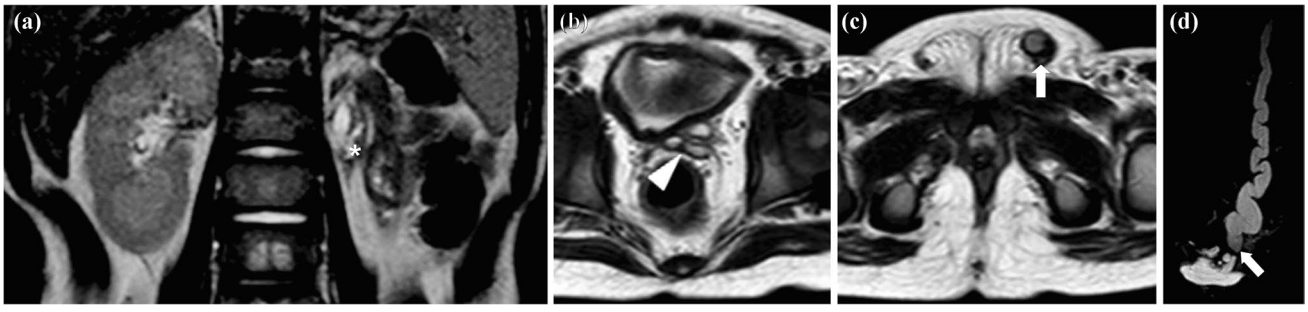

Fig. 17 Dysplastic left kidney in a 2-year-old boy. **a** Coronal Turbo spin-echo T2-weighted image shows a small dysplastic left kidney (*asterisk*). **b**, **c** Axial Turbo spin-echo T2-weighted images show distension of the left seminal vesicle with moderate ectasia of the terminal tract of the ipsilateral ductus deferent (*arrowhead*), and note the left testis retained in the inguinal canal (*arrow*). **d** Coronal volumetric hydrographic sequence demonstrates the left ureter outlet below the bladder (*arrow*)

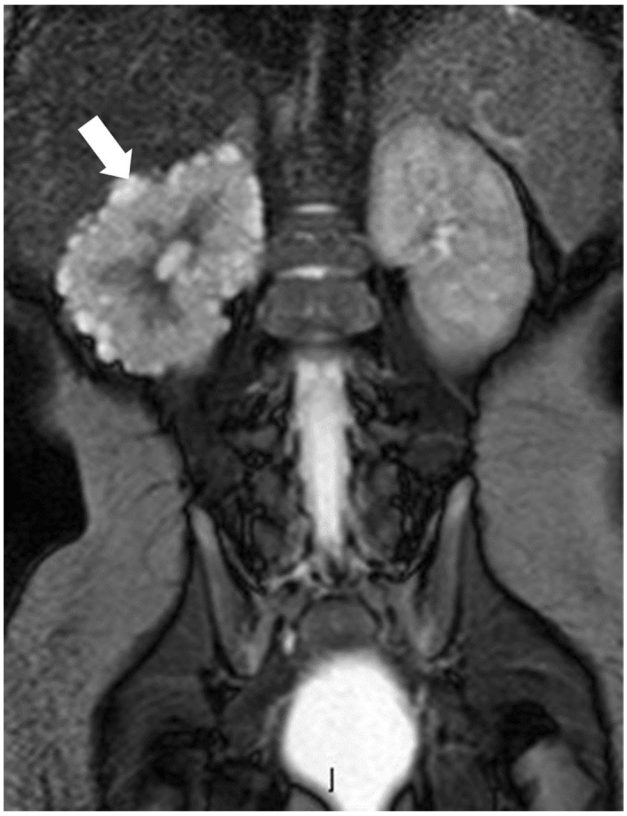

Fig. 18 Renal dysplasia in a 5-year-old girl. Coronal turbo spin-echo fat-saturated T2-weighted MR image shows a dysplastic right kidney characterized by loss of normal corticomedullary differentiation and small subcortical cysts (*arrow*)

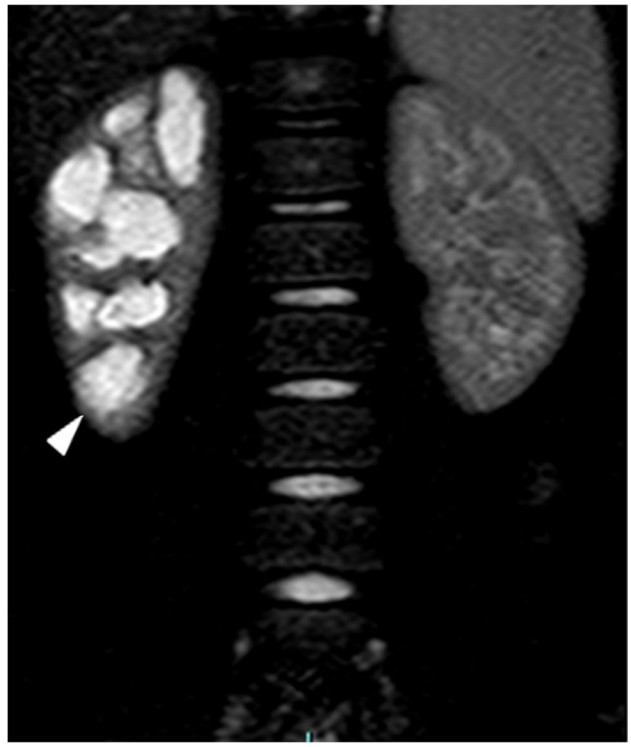

Fig. 19 Megapolicalycosis in a 10-year-old girl. Coronal turbo spin-echo fat-saturated T2-weighted MR image shows unilateral dilation and increased number of renal calyces (*arrowhead*) without obstruction

clinical–therapeutic workup. In our experience, in pre- and

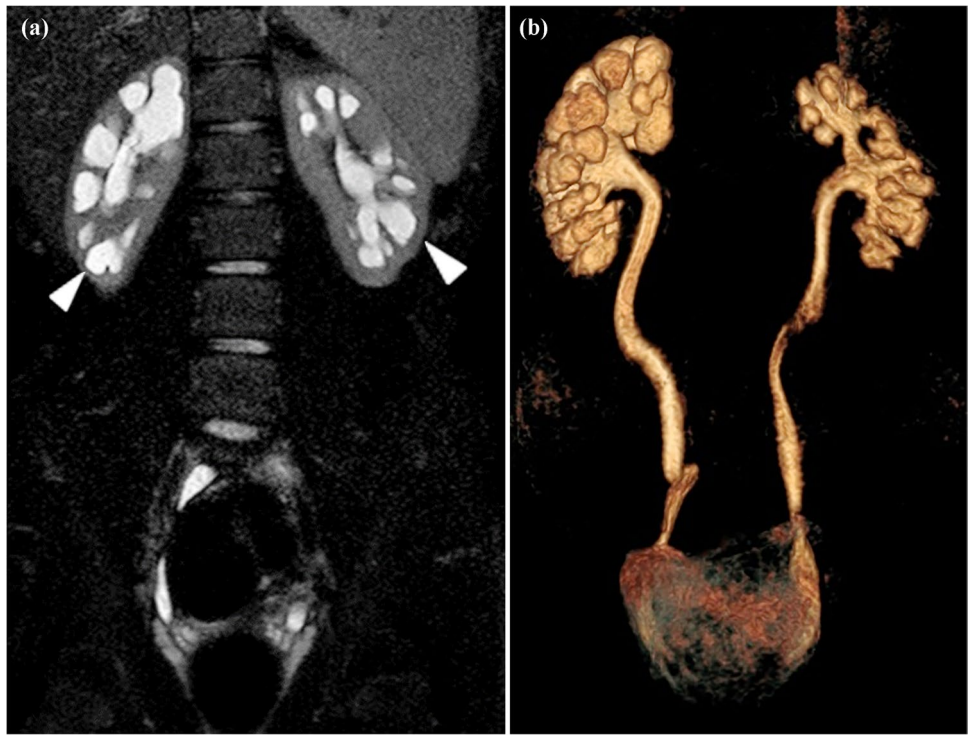

Fig. 20 Megapolicalycosis in a 15-year-old girl. **a** Volume-rendered coronal turbo spin-echo fat-saturated T2-weighted MR image shows bilateral megapolicalycosis, which is characterized by dilation and increased number of renal calyces (*arrowheads*). **b** A reduction in thickness of the parenchyma, and regular and complete excretory phase appear on volume-rendered image of the urographic phase

postnatal clinical suspicion of congenital abnormalities of the kidney and urinary tract, MR urography can be used to establish an early diagnosis, thus improving patient outcome.

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Declarations

Conflicts of interest None

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