Congenital and Acquired Pathologies of the Pediatric Urogenital Tract

21

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Learning Objects

- Challenges and rational use of imaging modalities in congenital and acquired urinary tract pathologies.
- Imaging algorithms for most important clinical queries including incontinence.
- Get familiar with tumors of the urogenital tract.
- Learn about cardiovascular consequences of kidney diseases.

Key Point

Children are mirrors of the environment. Therefore for a successful investigation, a child friendly environment as well as staff is mandatory. Therefore, children should be imaged in dedicated Pediatric Radiology units and not mixed up with adults.

21.1 Introduction

Imaging of the urinary tract (UT) contributes considerably to the workload in Pediatric Radiology due to numerous diseases.

The purpose of this contribution is to present a short overview about most important CAKUT and acquired UT diseases.

21.2 Imaging Modalities

Due the many, physiologic, differences between children and adults it can be stated that "smallest children need the biggest machines" [1]. In order to avoid fear and uncooperative patients, Pediatric Radiology has to ensure an adequate environment for them as well as an appropriate set up of imaging modalities.

Ultrasound (US): US is the most important and starting modality of choice. US enables not only UT morphological assessment but quantitative parameters like bladder volume, residual void, and renal volumes [2]. Using the ellipsoid formula for bladder volumetry, it has to be considered that if the 3D baldder shape deviates from an ellipsoid the volume estimation getting less reliable.

Moreover, Doppler ultrasound and all its variants allow noninvasive assessment of blood flow, for example, thrombosis and vessel stenosis. Doppler tracings depict information about peripheral vessel resistance, and much more. Intravasal US contrast injection enables to visualize organ perfusion at almost no risk. Contrast-enhanced Sono Voiding Cysto-Urethrography (ceVUS) has already an established place in the diagnostic workup of urinary tract infection and suspected vesicoureteral reflux (VUR).

Due to the huge variation of body size in Pediatric Radiology, several transducers must be available including high resolution linear transducers, in order to ensure appropriate scanning an image quality for all children—regardless of age and size. It should not be forgotten that it can be performed bed-side.

Voiding Cysto-Urethrography (VCU): For decades VCU was the imaging modality of choice for VUR detection. Urine testing should be done before the procedure in order to avoid catheterization during urinary tract infection. In boys, appropriate imaging of the urethra during voiding is a must in order not to overlook posterior urethral valves

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(PUV). Modifications of the standard technique enable to assess also lower urinary tract dysfunction [3].

Intravenous Pyelography: It does not play a role anymore since ultrasound and Doppler ultrasound can deliver almost the same information. One exception of the rule might be ureteral stones in low resource countries.

Magnetic Resonance Imaging (MRI): MRI is second-line diagnostics and delivers high resolution anatomic details for almost all referrals. Using dedicated imaging sequences as well as free available post-processing techniques functional data like in Nuclear Medicine can be obtained including split renal function [4, 5].

Computed Tomography (CT): CT is rarely used, and there is almost no indication in pediatrics except for emergency situations like septic patients due to renal abscess, therapeutic interventions if ultrasound cannot be used as a navigational modality as well as in some cases of UT stone formation. There should be sized adapted protocols available on the CT machine in order to keep radiation exposure "as low as reasonable achievable (ALARA)." Furthermore, dual energy CT can help to characterize urinary tract stones for further patient management [6].

Nuclear Medicine: It enables to assess side-related functional renal parenchyma as well as in isotope renography the quantitative study of urine flow and in particular differentiation between obstructive and non-obstructive situation in PCD. It should be noted, that response to Furosemid can be missing due to kidney immaturity within the first 2–3 months of life and therefore can be misinterpretated as obstructed urinary flow.

Key Point
US and VCU represent the "workhorses" of UT imaging in children.

21.3 Normal Variations

It is difficult to make a distinct border line between normal variants and variants predisposing to illness. Persistent fetal lobulation, hypertrophied column of Bertin, and dromedary hump belong to the first group. It is already more complicated, for example, kidney hypoplasia (normal, configured but smaller kidney, hypertrophy of the contralateral one) since smaller kidneys are vascularized by smaller arteries thus increasing the risk of later hypertension. Double kidneys are frequently associated with double ureters and can predispose to hydronephrosis and VUR—see section antenatal hydronephrosis. Same applies to ectopic kidneys or crossed or fused kidneys (Fig. 21.1). The malposition itself is not the problem but associations with VUR or obstructed urinary flow. It is noteworthy to memorize that dysplastic or displaced kidneys usually have abnormal shaped calyces too.

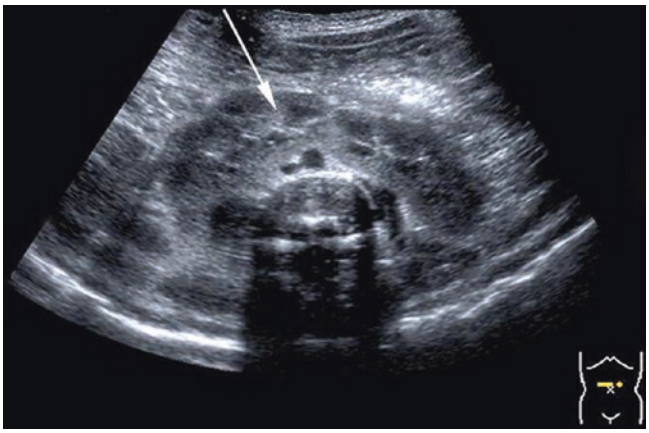

Fig. 21.1 US image from a horseshoe kidney, transverse cut of the abdomen. White arrow marks the kidney parenchymal bridge ventral to spine

21.4 Antenatal Hydronephrosis and CAKUT

Fetal antenatal hydronephrosis is a frequent finding on prenatal imaging in the order of 1.0–2.0% [7]. In order to ensure an adequate diagnostic algorithm, grading of the pelvicalyceal dilatation (PCD) can be graded according to a modified score of the "Society for Fetal Urology"(Fig. 21.2). According to literature, this is 50–70% transient/physiologic, due to ureteropelvic junction obstruction in 10–30%, vesicoureteral reflux 10–40%, ureterovesical junction obstruction/megaureter in 5–15%, multicystic dysplastic kidney disease 2–5%, posterior urethral valves 1–5%, and more uncommon ureterocele, ectopic ureter, duplex system, urethral atresia, Prune belly syndrome, and polycystic kidney diseases [7].

Furthermore, normal values according to gestational age were defined as listed in Table 21.1 [7]. Based on those ultrasounds, three patients' groups with different risk levels were defined as well as the appropriate imaging follow-up on these patients [8]. An isolated finding of an ampulla-shaped renal pelvis without PCD does not require further diagnostic workup.

An important point to remember is, that due to neonatal,physiologic oliguria PCD can be missed in US scans in the first days of life (Fig. 21.3).

In double kidneys with double ureters, both moieties can show PCD—due to the Weigert-Meyer law the ureter belonging to the upper moiety enters the bladder within the urinary bladder basal plate and thus leading to obstruction of urinary

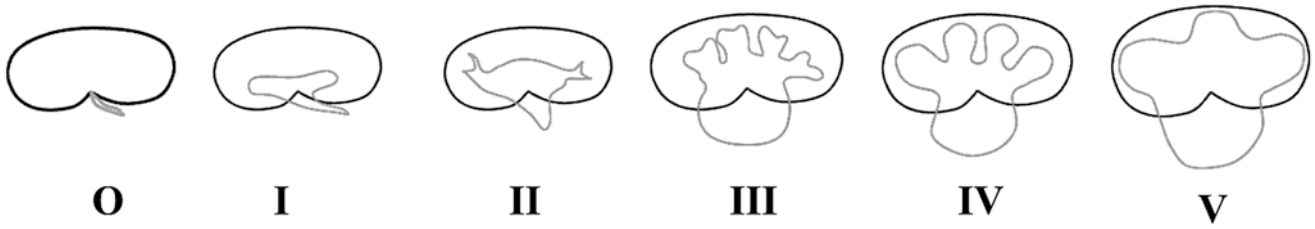

Fig. 21.2 schema of PCD dilatation. Numbers below the individual parts of the schema indicates type: O = no dilatation, I: only renal pelvis visible, II: renal pelvis and normaly shaped calyces are recognizeable, III: marked dilatation of renal pelvis (>10.0 mm), calyx fornix angles are rounded and papillary impression just reduced, no parenchymal narrowing, IV: same as III but parenchymal thickness is reduced, V: used in some instutions for the situation where parenchyma ist only a rim - modified after [2]

Table 21.1 normal sonography values for fetal UT [7]

Ultrasound findings		Time at presentation		
		16–27 weeks	>28 weeks	Postnatal (>48 h)
Renal pelvis anterior—Posterior diameter		<4 mm	<7 mm	<10 mm
Calyceal dilatation	Central	No	No	No
	Peripheral	No	No	No
Parenchymal thickness	Thickness	Normal	Normal	Normal
	Appearance	Normal	Normal	Normal
Ureter(s)		Normal	Normal	Normal
Bladder		Normal	Normal	Normal
Unexplained oligohydramnions		No	No	NA

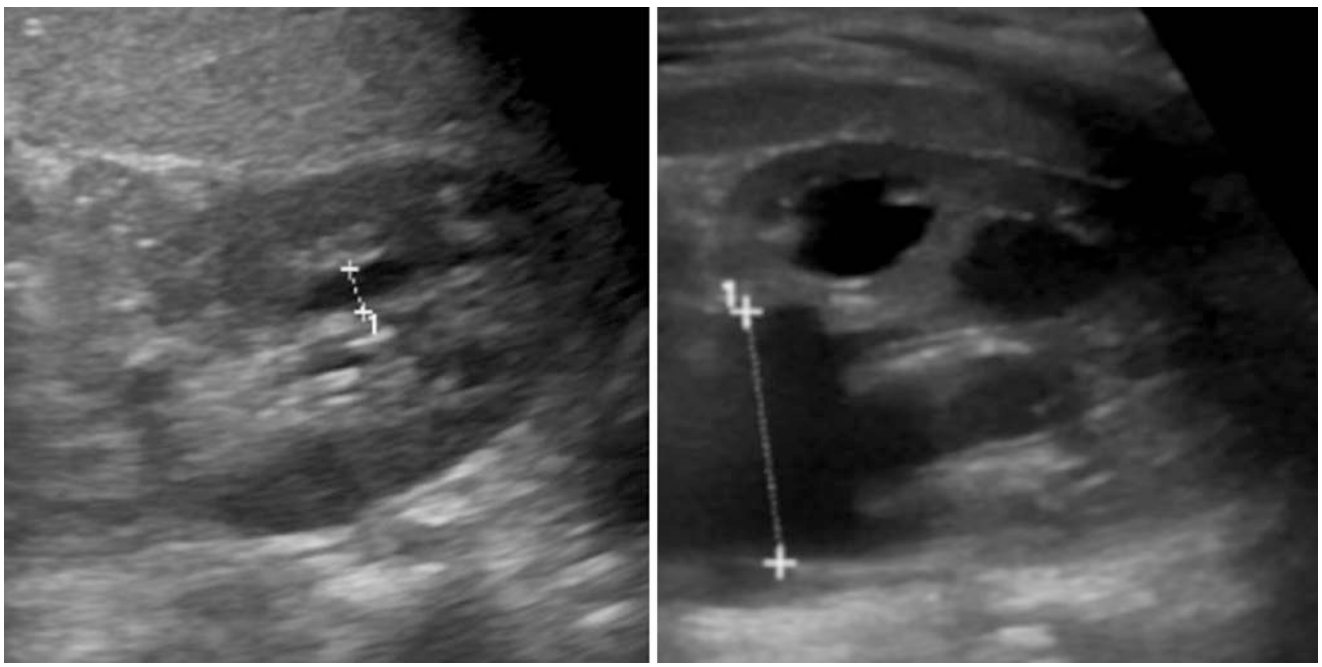

Fig. 21.3 influence on PCD by hydration. Neonate with known antenatal PCD. Left part: US on second day of life, no dilatation due to physiologic oliguria during first days of life, right part: US after a week PCD type IV

flow whereas the ureter draining the lower moiety enters the bladder in abnormal high position and therefore VUR is common [9].

Key Point
US scans in babies with antenatal diagnosed hydronephrosis should be taken after the third day of life in order to avoid false normal findings.

In double kidneys, remember the Weigert-Meyer law.

Congenital anomalies of kidneys and urinary tract (CAKUT) are defined as "any structural and functional abnormalities of kidney, collecting system, bladder, and urethra" are frequent findings in 20–50% of fetal congenital abnormalities imaging [10] and up to 1 in 500 live births [11]. A list of those anomalies is given in Table 21.2. CAKUT pathogenesis is based on the disturbance of normal nephrogenesis, secondary to environmental or genetic causes (Capone et al. 2017). As environmental causes maternal diabetes as well as intrauterine exposure to ACE inhibitors were detected. In non-syndromic cases, mutations on HNF1B order PAX2 genes may be responsible [12].

Moreover, CAKUTs are responsible for 30–60% of chronic kidney disease starting already in childhood thus leading to renal replacement therapy already with 31 years as compared to others (61 years) [10].

Table 21.2 CAKUT spectrum of anomalies

Kidney anomalies	Renal agenesis Renal hypoplasia Duplication anomalies (duplex kidneys—only one collecting system Fusion anomalies (fused kidneys—One or more collecting systems, horseshoe kidney) Ectopic kidney (pelvic kidney, crossed ectopis) Renal dysplasia and multicystic kidney Cystic kidney disease (simply renal cyst, adult polycystic kidney disease, infantile polycystic kidney disease, medullary sponge kidney (MSK)
Abnormalities of the ureter and ureteropelvic junction	Ureteral atresia Obstruction of the ureteric junction obstructionDuplication of the ureter Ureterocele Ectopic ureter Prune belly syndrome (PBS) Obstructed mega-ureter
Abnormalities of the bladder	Bladder exstrophy Persistent urachus Vesicoureteral reflux
Anomalies of the penis and urethra in males	Males: Posterior urethral valves, double urethra
Anomalies of the testis	
Female genital anomalies	
Gonadal dysgenesis	
Disorders of sex development	

21.5 Urinary Tract Infection (UTI)

UTI affects during first year of life about 0.7% of girls and 2.7% of uncircumcised boys [13]. There is bimodal age distribution with a peak within the first year of life and another between 2 and 4 years [13]. Diagnosis seems easy with urine testing but since there the used urine bag collection leads quite often to false-positive results. Differentiation between cystitis and pyelonephritis is not possible clinically. Any kidney parenchymal scar will increase the likelihood of hypertension later in life.

An imaging algorithm was published by the Taskforce Abdomen [14]. US is used as imaging modality of choice and should always include urinary bladder. In pyelonephritis, the nephritis part can be diagnosed swelling of kidney, loss of cortical differentiation, areas of reduced echogenicity (representing edema) or increased one due to hemorrhage. Pyelitis causes wall thickening as well as pus, free flowing particles, or sedimentation levels (Fig. 21.4)—but the latter needs time to happen, so patience is needed before scanning the child.

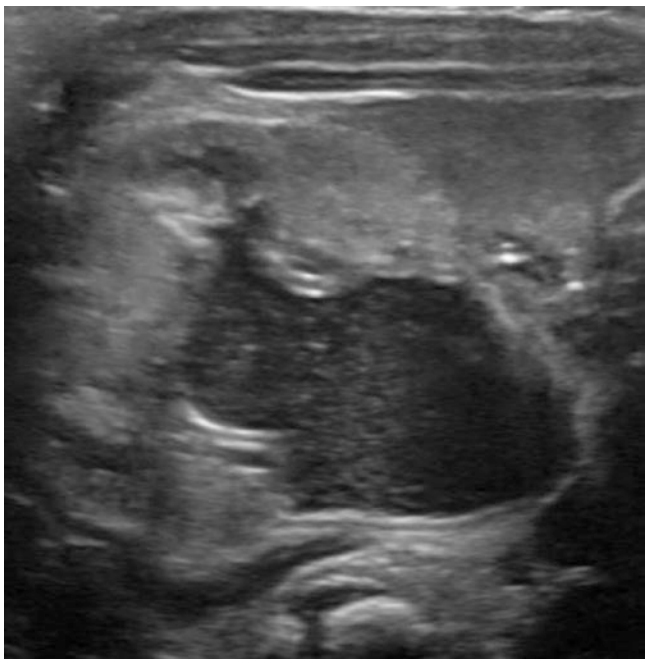

Fig. 21.4 child with urosepsis, kidney US, transverse section. There is massive dilated renal pelvis with free, flowing, echogenic particles corresponding to pus

Pyelonephritis can be diagnosed with equal accuracy by CT, MRI, and DMSA scan and ultrasound was reported to be less performant [15] Recently, it was published that contrast-enhanced US (ceUS) proves to be a valuable tool with almost comparable performance in regard to CT and DMSA scan but avoiding radiation exposure and sedation in small children [16].

Key Point
As in UTI, US is the starting modality of choice. ceUS enables to diagnose pyelonephritis with high confidence in doubtful cases.

21.6 Incontinence/Enuresis

Incontinence must be separated from enuresis. Incontinence represents an incomplete micturition at the wrong time point (e.g., urge) whereas enuresis is a complete micturition at the wrong time, being further divided in enuresis during night (enuresis nocturna) and/or during daytime (enuresis diurna),. Unfortunately, many cases of enuresis are labeled incorrectly as incontinence, especially many cases enuresis diurna are belonging to the group of incontinence (urge incontinence). Due to incorrect use of both terms, the problem is a wetting child. This could be due to anatomy (e.g., an ectopic ureter entering perineum or vagina in girls from a double system) or functional (e.g., urge incontinence) or a combination (enuresis nocturna due to small bladder volume, high fluid intake in evenings together with late wake up in nights), and there is also a family factor, where all relatives, for example, were suffering from enuresis nocturna. An overview about these entities can be found in [17].

Key Point
Do not mix terms incontinence and enuresis—these are different entities.

As usual starting imaging with US represents a good choice. Double systems can be ruled out and during full bladder scanning opening and closure the bladder sphincter can be observed—thus indicating urge incontinence. Moreover, an open bladder neck also points to bladder instability [18].

As mentioned already in the imaging modality section Fotter's VCU modification (only the procedure and no additional hardware needed) allows to analyze lower urinary tract dysfunctions with a performance almost comparable to bladder urodynamics.

21.7 Renal Masses

Kidney angiomyolipomas (fat content) are known to be in associated in 20% with tuberous sclerosis complex and pulmonary lymphangioleiomyomatosis [19]. Cystic renal masses include simply renal cysts, multicystic dysplastic kidneys, hereditary cystic renal diseases (autosomal dominant or recessive polycystic kidney disease) to cysts in renal dysplasia [20].

Cystic nephroma and cystic partially differentiated nephroblastoma cannot be distinguished by imaging, and it is believed that they represent the benign end of tumors originating from metamorphose, whereas Wilms tumor being on the malignant end of the spectrum. Another tumor originating from nephrogenic rests is the "Ossifying renal tumor of infancy (ORTI)." It appears like a staghorn calculus but enhances after contrast injection. Mesoblastic nephroma is the most common renal tumors in neonates and descend from mesenchyma. "Clear Cell Carcinoma of the kidney (CCSK)" is a solid tumor with cystic components and metastasizes in bones, which would be uncommon in Wilms tumor. Renal rhabdoid tumor is a solid tumor of toddlers which also shows subcapsular hemorrhage. Furthermore in 15%, it is associated with primary or secondary brain tumors.

"Renal Cell Carcinoma ("RCC" is similar to adults but occurs in children with Hippel Lindau disease.

Wilms tumor (nephroblastoma) represents the most common renal neoplasm in infancy (90%) and arises from nephrogenic rests (or nephroblastomatosis) [21]. Peak incidence is about 2–3 years of age and appears sold but can also be heterogenous due to hemorrhage. Calcifications can be seen up to 15% in CT [21]. In addition, renal vein invasion can be found. In 2%, there is a familial predisposition and several associations due to mutations of WT1 (WAGR syndrome, Denys-Drash syndrome, Frasier syndrome, Bloom syndrome) and WT2 (Beckwith-Wiedemann syndrome, Perlman syndrome, Simpson-Golabi-Behmel syndrome, Sotos syndrome) genes. Moreover, isolated abnormalities can be found like isolated abnormalities: cryptorchidism in 3%, hemihypertrophy in 3%, hypospadias in 2%, sporadic aniridia, and renal fusion (https://radiopaedia.org/articles/wilms-tumour).

Key Point
Wilms tumor most frequent renal tumor in childhood. Renal tumors in children may have several important associations.

21.8 Hematuria and Renal Calculi

Hematuria is a relatively common finding in children. It may be found incidentally by urine analysis (microscopic hematuria) or when gross hematuria is evident. Ultrasound is the primary imaging modality looking for structural abnormalities such as renal anomalies, ureteric calculi, and renal or bladder masses. The most common causes of gross hematuria are inflammatory processes in the bladder and glomerulonephritis. Other considerations include renal calculi and bladder rhabdomyosarcoma.

Adolescents and school aged children with renal colic present in the typical way, but infants may present with irritability and inconsolable crying. Ultrasound and radiography are the initial imaging modalities of choice with low dose CT being the most definitive. The goal is to identify the presence, position, number, and size of the renal calculi. Stones can be seen in the pelvicalyceal systems, ureters, or bladder. On ultrasound, signs of renal calculi include shadowing echogenic foci, dilatation of the urinary tract, and increased parenchymal echogenicity. Color Doppler ultrasound can be used to elicit the twinkle artifact. Low dose CT is used if the ultrasound is normal or if further anatomic details are needed for surgical planning. CT is complementary to US and is used for problem solving. When properly performed radiation doses are minimized and optimally adapted to the child's size.

Renal stones are unusual in children and their presence may indicate an underlying metabolic abnormality. In those with a metabolic abnormality, there can be repeated episodes over the years so judicious use of imaging is important.

Key Point
Hematuria is common in childhood. The common causes are renal calculi as well as inflammatory and neoplastic conditions.

21.9 Trauma

The kidney is the most commonly injured organ of the urinary tract in children and can occur in up to 20% of all blunt injury cases [21]. Most children are treated conservatively, but if they are hemodynamically unstable operative management may be required. Injuries to the ureter, bladder or urethra are usually seen in the setting of polytrauma [22]. The pediatric kidney is relatively mobile within Gerota's fascia so lacerations and contusions are caused by crushing of the kidneys against the spine or ribs. Undiagnosed pre-existing renal abnormalities are found incidentally in up to 20% of children who are imaged in the setting of acute trauma [23].

Imaging has a pivotal role in managing blunt or penetrating trauma to the genitourinary tract. In many places, ultrasound is first modality used especially if the patient has minimal symptoms. In cases of urinary tract injury, multiphase post-contrast CT of the urinary tract is recommended including delayed post-contrast scans (Fig. 21.5) [22].

In children with renal trauma, imaging is used to classify any injury to the kidney, to identify underlying congenital abnormalities and demonstrate the extent of any other injury. Initial ultrasound helps to identify patients needing more extensive investigation and is particularly useful for follow-up of renal injuries, hematomas, and urinomas. It must be remembered that ultrasound is insensitive for detecting renal lacerations. Most urinomas are asymptomatic and will resolve spontaneously.

CECT with delayed urographic phase is the gold standard for grading renal injuries. It allows accurate evaluation of injuries to the renal parenchyma, the renal vessels, and collecting systems. CT is recommended in children with high energy or penetrating trauma and/or when there is a drop in hematocrit associated with any degree of hematuria [24].

Renal injuries are graded based on CT findings using the AAST Organ Injury Scale [25]. It describes a scale of progressively more severe injury with Grade I representing parenchymal contusion and subcapsular hematoma to Grade V which represents a completely shattered kidney. Most renal injuries in children are Grade I–III while only about 20% are Grade IV or V. Most children are treated conservatively and surgical intervention is required only in clinically unstable patients. Ureteral injuries are uncommon in children [26]. Bladder rupture can be either intra-peritoneal or extra-peritoneal and is usually associated with fractures of the pelvis. Urethral injuries are rare and most often seen in boys with blunt perineal trauma. Retrograde urethrogram is performed to evaluate urethral trauma. In children being evaluated for trauma, if they have normal genitourinary examination, normal voiding and without gross hematuria, no imaging of the lower GU tract is required.

Contrast-enhanced US has emerged as a promising tool to assess renal injuries [27].

Key Point
The kidney is the most commonly injured organ of the urinary tract. Most children are treated conservatively. Underlying congenital abnormalities are commonly found.

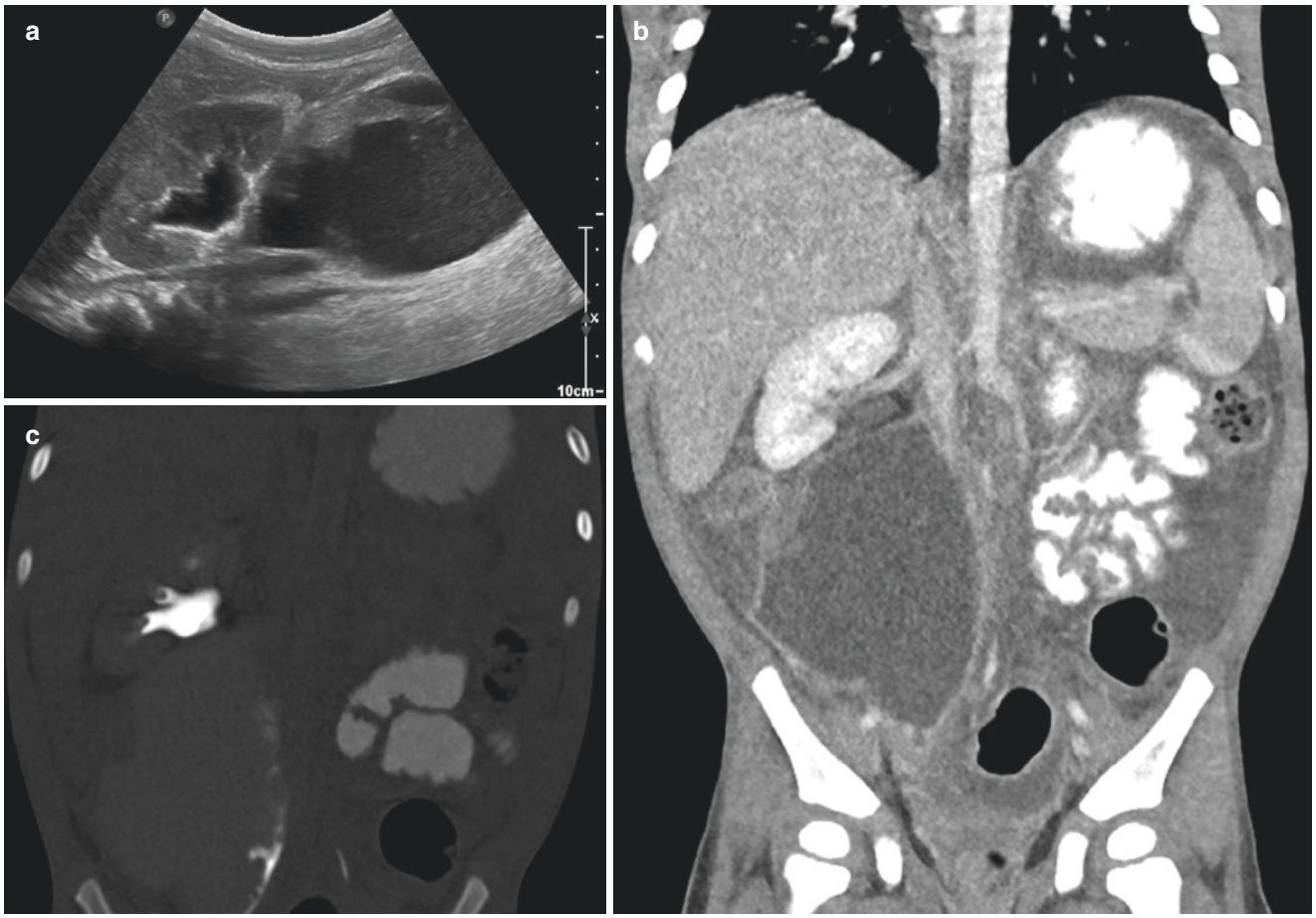

Fig. 21.5 6 year old boy involved in motor vehicle accident (**a**) Sagittal ultrasound image through the right kidney shows mild right-sided hydronephrosis with an inferior fluid collection with moderate low level echos (**b**) nephrogenic phase CT demonstrates homogeneous enhancement of the right kidney with inferior fluid collection. (**c**) delayed phase CT with wide windowing showing contrast collecting in the inferior fluid collection indicating a urinoma

21.10 Acute Kidney Injury (AKI) and Chronic Renal Failure (CRF)

Renal failure in infants and children may be acute or chronic, reversible, or irreversible and may lead to dialysis or renal transplantation. Pathophysiologically, there are three major causes:

- Intrinsic renal disease
- Obstructive uropathy
- Pre-renal disease secondary to a systemic or extra-renal disease

Acute kidney injury is characterized by an abrupt deterioration of kidney function and is commonly seen in critically ill children. It can be seen in 30% of children in intensive care units. Clinically, there is usually increased blood pressure with oliguria or anuria. The diagnosis is based on laboratory findings with elevated serum creatine, electrolyte disturbances, low protein, and often metabolic acidosis. US plays a central role in evaluating the etiologies of renal failure and helping to differentiate acute from chronic failure. The imaging findings must be correlated with biological and clinical data.

Neonatal AKI may be suggested prenatally, but the diagnosis is often only established after birth. The most common renal causes are ARPKD, congenital nephrotic syndromes (CNS), or neonatal glomerulonephritis (GN). In children with ARPCKD, the kidneys are large, echogenic and have a salt and pepper appearance of the parenchyma. Neonatal CNS and GN present with large kidneys and non-specific echo pattern, often with loss of the normal corticomedullary differentiation. Colour Doppler findings are also non-specific. Corticomedullary differentiation depends on whether the cortex, medulla, or both are affected. Medullary and cortical necrosis in the neonate results from lack of renal perfusion. On US, the cortex in cortical necrosis first appears hyperechoic, then shrinks and finally calcifies. In medullary

necrosis, calcifications develop within the medulla. The value of ultrasound is not to be specific, but to rule out other causes of AKI.

Renal vein thrombosis (RVT) is most often seen in neonates with adrenal gland hemorrhage, dehydration, or thrombotic syndromes. RVT can occur in utero and unilateral RVT usually presents with hypertension and hematuria. In the acute phase on US, the kidney is enlarged, echogenic and with loss of the CMD. On CDS, the color signals from the affected renal veins are missing. The kidney may atrophy with calcification in the vessels.

The three most common causes of ARF in children in developing countries are hemolytic uremic syndrome (HUS), glomerulonephritis, and postoperative sepsis/pre-renal ischemia. In industrialized countries, the three commonest causes are intrinsic renal disease, postoperative septic shock, and organ/bone marrow transplantation [28]. Hemolytic uremic syndrome is comprised of hemolytic microangiopathic anemia, thrombocytopenia, and AKI and is caused by toxins released from certain strains of *E coli*. The patients often have a history of hemorrhagic enterocolitis. In the acute phase, the renal cortex becomes markedly hyperechoic bilaterally with increase corticomedullary differentiation. On Doppler analysis, the RI is markedly elevated with diffuse decreased cortical perfusion on CDS. Proximal tubular necrosis may follow toxic ingestions or medication. Tubular and vascular obstruction causing prolonged renal ischemia can follow renal parenchymal uric acid accumulation, sickle cell crisis, myoglobulinemia, and renal vein thrombosis. Cortical and tubular necrosis may occur following hemorrhagic shock, severe dehydration, crush injuries, thermal burns, and septic shock.

The incidence of ESRD has been stable over the past 30 years worldwide, but prevalence has increased [29]. The most common cause of pediatric CRF is CAKUT accounting for up to 50% of cases. The next most common causes are the hereditary nephropathies and glomerulonephritis. Infants of low birth weight and have an increased risk of developing ESRD in adolescence. Chronic renal failure is defined as a GFR <50 ml/min per 1.73 m/kidney. On ultrasound, the kidneys are small with loss of CMD and small cysts.

Congenital nephrotic syndromes (CNS) encompass diseases in which there is massive proteinuria occurring after birth. The most common form of CNS is the Finnish type. On US, at birth, the kidneys are large and hyperechoic. The CMD is present but the pyramids are irregular and within weeks will no longer be visible. Other causes of CNS include diffuse mesangial sclerosis which can be part of Denys-Drash syndrome.

Renal diseases that include primary and secondary tubulopathy are numerous. Hypercalciuria is a constant finding and may lead to nephrocalcinosis which is easily seen on US (Fig. 21.6). Secondary hyperparathyroidism may develop leading to renal osteodystrophy which can result in abnormalities affecting the growth plates, epiphyseal displacement, and fractures.

Key Point

US is the key imaging examination in children with AKI or CRF. Ultrasound is key to differentiating pre-, post-, and intrarenal causes. Most nephropathies have a similar appearance with large kidneys usually indicating acute disease and small kidneys in chronic diseases.

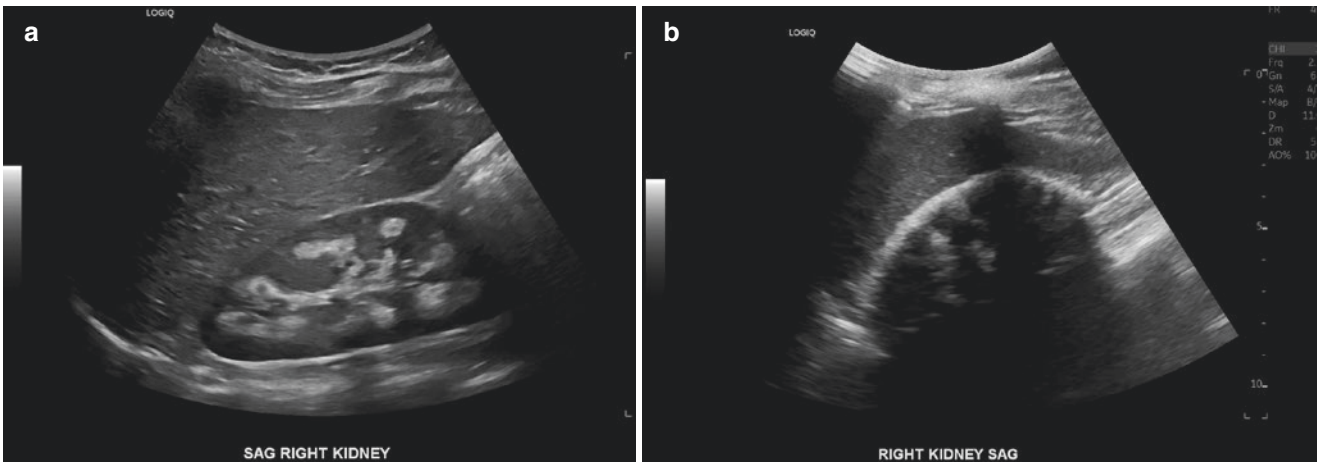

Fig. 21.6 Nephrocalcinosis in two different children (**a**) Medullary nephrocalcinosis: sagittal ultrasound image through the right kidney in an 11 year old girl demonstrates increased echogenicity in the medulla of the right kidney consistent with medullary nephrocalcinosis (**b**) Cortical nephrocalcinosis: sagittal image through the right kidney in a 7 year old boy demonstrates increased echogenicity and shadowing from the renal cortex

21.11 Renal Causes of Hypertension

A renal cause for hypertension is suspected when hypertension is severe or refractory to multiple drugs. Renovascular hypertension is responsible for 5–25% of hypertension in children [30]. There are numerous causes of aortic and renal artery narrowing leading to renal hypertension. These are often syndromic and include idiopathic/fibromuscular dysplasia, NF1, Williams syndrome, mid-aortic syndrome, inflammatory arteritis as well as extrinsic compression.

Ultrasound is the initial modality to assess for renal anomalies or scarring as well as non-renal lesions such as pheochromocytoma. Doppler evaluation can be used to assess for renal artery stenosis by showing a tardus parvus pattern of the spectral waveform with slow systolic acceleration. Pathologic flow parameters include peak systolic flow >180 cm/s, acceleration time > 80 ms, renal artery to aortic flow velocity ratio >3 and difference in RI more than 0.05 [30]. Renal Doppler ultrasound is reasonably specific but not sensitive enough to exclude renal vascular abnormalities [31].

CT angiography or MR angiography can verify a renovascular cause for hypertension by demonstrating one or more areas of stenosis or if there are collateral vessels present. CTA and MRA are excellent for evaluating the aorta and main renal arteries, but for smaller intraparenchymal branches visualization is limited [32]. Children are referred for catheter renal angiography if no abnormality has been identified on noninvasive techniques and there is persistent failure of medical therapy. Angiography is considered the gold standard in establishing the diagnosis of renovascular disease as no noninvasive technique can exclude renovascular disease [32].

Key Point
Renal causes of hypertension are common in children. Ultrasound is the initial modality used to identify underlying renal abnormalities. However, only catheter angiography can exclude renovascular disease.

21.12 Disorders of Sexual Differentiation

Disorders of sexual development (DSD) are defined as conditions in which chromosomal sex is not consistent with phenotypic sex or in which the phenotype is not classifiable as either male or female [33]. DSD can be divided into three categories: those with 46 XX karyotype, those with a 46XY karyotype, and those relating to sex chromosomes [34]. Disorders of sex development occur when the male hormone (androgens and anti-Mullerian hormone) secretion or action is insufficient in the 46 XY fetus or when there is androgen excess in the 46 XX fetus. DSD with ambiguous genitalia are typically diagnosed clinically in the newborn period, whereas those associated with male and female phenotypes may not present until adolescence. Patients with pure gonadal dysgenesis or complete androgen insensitivity usually are phenotypic females who present at puberty with primary amenorrhea.

Diagnosis and classification of these disorders is complex and the role of imaging in infants is to identify a uterus and or cervix, to locate the gonads, and to define the anatomy of cloacal malformations, Mullerian duct anomalies, urinary tract anomalies as well as anorectal and spine malformations. Ultrasound has been the primary modality to identify the internal organs, and occasionally fluoroscopic genitography and VCUG are used to assess the vagina, urethra, and any fistulas or complex tracts. Contrast-enhanced ultrasound and MR genitography are being used more often in the evaluation of these anomalies especially in defining the anatomy of the urogenital tract, anorectal malformations, and to identify otherwise occult gonad [35, 36]. Both techniques are similar to traditional fluoroscopic genitography in that contrast material is used to demonstrate the anatomy of the various cavities.

When performing a genitogram, it is important to ensure that all perineal orifices are examined [37]. It is also important to preserve the morphological appearance by only inserting the catheters a short distance. The goal is to define a male or female urethral configuration and identify any fistulous communication with the vagina or rectum [38]. Demonstration of the level at which the vagina opens into a urogenital sinus and its relationship to the external sphincter is important in surgical planning. The vagina is evaluated to determine its presence or absence, its relationship to the urethra and to identify the uterus. The presence of hydrocolpos associated with ambiguous genitalia and two perineal orifices confirms the presence of a urogenital sinus. In the presence of a large hydrocolpos, the bladder may be displaced anteriorly making it difficult to see so care is needed to make sure a fluid filled vagina is not confused with the urinary bladder. The uterus often is identified capping the vagina and the distended vagina often contains a fluid-debris level.

In cloacal malformations, the genital, urinary, and gastrointestinal tracts open into a single common channel classically located at the expected site of the urethra [39]. It is almost exclusively seen in girls. Cloacal malformations are divided into two groups depending on the length of the common channel. A common channel less than 3 cm is more easily repaired and has a lower incidence of associated anomalies.

Mullerian duct anomalies are a broad and complex spectrum of anomalies that often present with primary amenorrhea in adolescents. MR is the imaging method of choice in

defining these anomalies [40]. Uterus, fallopian tubes, cervix, and upper two thirds of the vagina are derived from the Mullerian ducts. The ovaries are embryologically separate and not typically involved in Mullerian duct anomalies. In patients with Mullerian duct anomalies, renal and ureteric anomalies are common. In addition to the well-known association with renal agenesis which is found in up to 30%, there is also a high incidence of ectopic, malrotated, or dysplastic kidneys. Additionally, 25% of patients with renal agenesis have distal ureteric remnants or ectopic ureteric insertion. These ureteric remnants may become distended by menstrual blood leading to abdominal pain, infection or present with urinary incontinence and recurrent UTI. All patients with Mullerian duct anomalies need assessment of the urinary tract to identify renal agenesis, ectopic ureters, or ureteric stumps.

Mayer-Rokitansky-Kuster-Hauser syndrome is a heterogeneous disorder characterized by ureterovaginal atresia in 46XX girls. Abnormalities of the genital tract may range from upper vaginal atresia to total Mullerian agenesis with associated urinary tract anomalies.

The external genitalia are normal. Cyclical abdominal pain due to endometrial tissue or even hematometra in the rudimentary uterus may be a cause of clinical confusion. The ovaries are ectopic in 40% of cases and are readily identified on pre-operative MR imaging. Herlyn-Werner-Wunderlich syndrome is characterized by uterus didelphys and unilateral hematocolpos related to an obstructed hemivagina with unilateral renal agenesis. They are often diagnosed early in infancy but may present in adolescence with hematocolpos, hematometra, or hematosalpinx. The diagnostic dilemma in these patients is that most have regular menstruation because one uterus is not obstructed.

Key Point
Disorders of sexual differentiation are complex disorders that often present at birth but may not become apparent until puberty.

21.13 Testicular and Ovarian Pathology

21.13.1 Cryptorchidism

Cryptorchidism or undescended testis is one of the most common congenital malformations on infant males seen in up to 5% of full-term and 45% of preterm neonates. In most cases, there is spontaneous descent within the first few months of life. Undescended testis is initially evaluated with ultrasound which can easily detect testes in the inguinal canal. However, ultrasound cannot reliably detect intra-abdominal testes which represent 20% non-palpable testes. MRI cannot diagnose monorchidism. Both the American Urologic Association and the European Association of Urology guidelines recommend against imaging for the routine management of patients with non-palpable testes. However, if there is associated ambiguous genitalia or hypospadias, there is a higher likelihood of an underlying disorder of sexual development. In this instance, ultrasound or MR is recommended to look for internal female pelvic organs specifically the uterus.

21.13.2 Scrotal Masses

A palpable scrotal mass should be characterized as intra- or extratesticular, solid or cystic, and characterized by its vascularity. Testicular tumors account for approximately 1–2% of all pediatric solid tumors. Most testicular tumors present as a painless scrotal mass. Hydroceles are often also present. The first line of evaluation is high resolution ultrasound (7.5–12.5 MHz) with Doppler interrogation. Testicular tumors in prepubertal boys differ in several aspects to testicular tumors after puberty: they have a lower incidence, they have a different histologic distribution (teratomas and yolk sac tumors are more common and germ cell tumors are less common), and they are more often benign. Testicular tumors can generally be classified as germ cell or stromal tumors.

Teratomas are usually benign in prepubertal children and represent about 40% of testicular tumors. They present at a median age of 13 months. Yolk sac tumors are the predominant prepubertal malignant germ cell tumor. Epidermoid cysts are of ectodermal origin and are always benign. Keratin-producing epithelium is responsible for the keratinized squamous epithelial deposits which appear hyperechogenic on US. Juvenile granulosa cell tumors usually occur in first year of life. Leydig cell tumors arising from the testosterone producing Leydig cells should be suspected in boys with premature puberty, with high testosterone and low gonadotropin levels. Patients are typically 6–10 years old. One specific tumor type is the gonadoblastoma which contains germ cell and stromal cell types and occurs almost exclusively in the setting of DSDs.

Paratesticular tumors are less common than testicular tumors and may be benign or malignant. Benign tumors include leiomyoma, fibroma, lipoma, hemangioma, and lymphangioma. The most common malignant tumor is the paratesticular rhabdomyosarcoma and the rare melanotic neuroectodermal tumor of infancy.

Testicular microlithiasis is increasingly seen in prepubertal boys and represents multiple tiny calcification in the testes. Microlithiasis appears as small non-shadowing hyperechoic foci ranging in diameter from 1–3 mm.

Microlithiasis is usually seen bilaterally. A recent metanalysis showed only 4 out of 296 boys with microlithiasis <19 developed a testicular tumor [41]. However, there is ongoing debate about the relationship to developing germ cell tumors but at present, there is no compelling evidence that regular sonographic follow-up is useful.

Up to a third of boys with congenital adrenal hyperplasia (CAH) will have testicular adrenal rest tumors (TARTS). These are thought to be ectopic adrenal cells with are growing under pathological stimulation from ACTH. They have no malignant potential but may be associated with impaired fertility.

21.13.3 Acute Scrotal Pain

The most common causes of acute scrotal pain are torsion of the testis or appendix testis, and epididymitis/epididymo-orchitis. Other causes of acute scrotal pain include mumps orchitis, varicocele, scrotal hematoma, incarcerated hernia, or appendicitis. Trauma can cause hematomas, testicular contusion, rupture, dislocation, or torsion.

Torsion of the testis most often occurs in the neonatal period and around puberty, whereas torsion of appendix testis occurs over a wider age range. Epididymitis affects two age groups: less than 1 year and 12–15 years. Perinatal testicular torsion most often occurs prenatally. Most cases of perinatal torsion are extravaginal, in contrast to the usual intravaginal torsion which occurs during puberty.

In general, the duration of symptoms is shorter in testicular torsion, and torsion of the appendix testis is compared to epididymitis. Prepubertal males are more likely to present with atypical symptoms and delayed diagnosis. Testicular torsion is a spectrum ranging from partial to complete. The torsed testis becomes enlarged and develops heterogeneous echogenicity. In partial torsion, there is asymmetric decreased flow to the affected testis. On Doppler ultrasound, there is absence of flow to the testis with complete torsion. With partial torsion, there may be absent or reversed diastolic flow or tardus parvus waveforms.

With torsion of the testicular appendages, there is focal pain over the superior aspect of the testis. Ultrasound demonstrates an extratesticular avascular nodule of varying echogenicity. Retrograde infection is frequently the source of epididymo-orchitis. Sexually transmitted infections are usually seen in adolescents. In acute cases, the epididymis is enlarged and hypervascular.

21.13.4 Ovarian Neoplasms

Ovarian neoplasms can be divided according to their cell of origin into three groups: germ cell, sex cord-stromal, and epithelial.

Most ovarian tumors in children are benign but 10–30% will be malignant. They usually present with pain or a palpable abdominal mass and may be associated with ovarian torsion. If the tumor secretes sex hormones, they may present with precocious puberty or virilization. The tumors are usually identified on ultrasound and more definitively evaluated with MR imaging. If the mass is malignant, FDG-PDT/CT has been shown to improve accuracy when detecting metastases [42].

Germ cell tumors are most common ovarian tumor in girls. Unlike adults, up to 30% of GCT in girls are malignant. Mature cystic teratomas are the most common benign ovarian neoplasm and are commonly known as dermoid cysts. Dysgerminomas are the most common malignant ovarian tumor. Their imaging appearance is determined by their content which is usually a mix of cyst, calcifications, fat, and sometimes hair. In contrast to epithelial neoplasms, which spread through peritoneal dissemination, GCTs usually disseminate through the lymphatic system. The prognosis for GCTs is excellent.

Sex-cord stroma tumors can be either benign or malignant. The two most common tumors in children are the granulosa cell tumor and the Sertoli-Leydig cell tumor. These tumors usually present with endocrine dysfunction and are usually confined to the ovary. The appearance is variable and includes both cystic and solid masses.

Epithelial tumors can also be benign or malignant and represent up to 15% of ovarian tumors in children. Most are benign and include serous, mucinous, and mixed cystadenomas. Carcinomas are very rare. Epithelial tumors usually appear as unilocular or multilocular cystic masses with numerous septations.

Adnexal torsion can involve the ovary and/or the fallopian tube. It occurs equally in pre- and post-menarchal girls and may be associated with a lead point such as a teratoma. The clinical presentation can be confusing with intermittent pain due to torsion/detorsion complex. On ultrasound, there is an enlarged, heterogenous pelvic mass with several peripherally dilated cysts and absent Doppler flow. Increased volume is the most common finding. It should be remembered that the presence of flow on Doppler US does not exclude torsion as the ovaries have dual arterial supply. In some cases, CT is the initial modality performed and recognition of the characteristic findings is important for prompt diagnosis (Fig. 21.7).

Key Point

The most common tumors of the testis and ovary in childhood are germ cell tumors. In prepubertal boys, most intratesticular tumors are benign, whereas after puberty they are malignant. Ovarian and testicular torsion may be intermittent leading to a confusing clinical presentation.

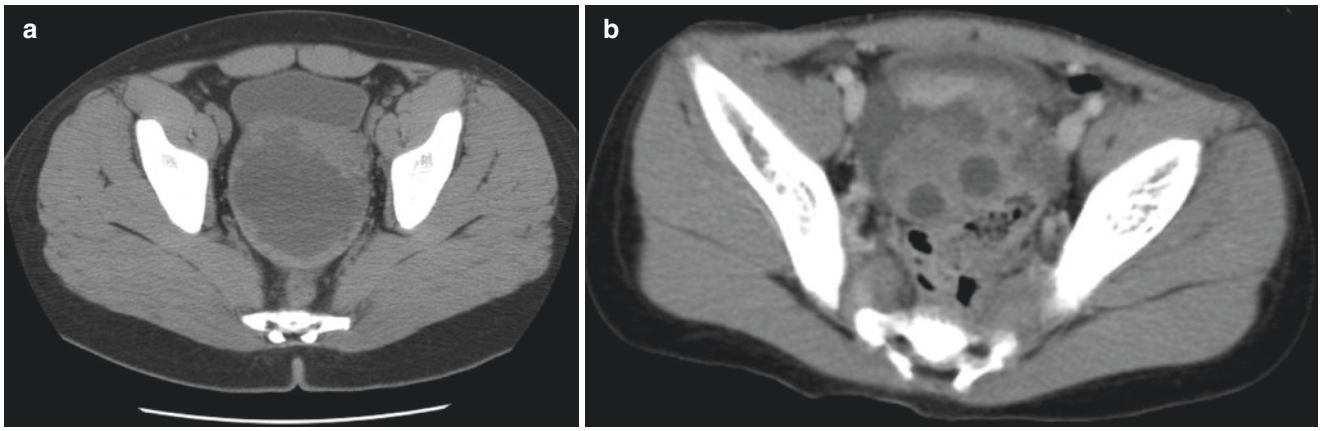

Fig. 21.7 14 year old girl with severe abdominal pain secondary to ovarian torsion (**a**) axial post contrast CT shows a large necrotic mass in the pelvis (**b**) inferior image through the pelvis demonstrates the peripheral cysts typical for ovarian torsion

21.14 Concluding Remarks

Most imaging evaluations in the infant or child begin with ultrasound. The next imaging steps are determined by the initial differential diagnosis obtained by integrating the clinical presentation with the ultrasound findings. It is important to be aware of the unique challenges involved in the imaging of children. Congenital abnormalities, urinary tract infection`, and tumors of the genitourinary tract represent a majority of indications for imaging in a pediatric radiology practice.

Take Home Points

1. Ultrasound is the workhorse of imaging the genitourinary tract.
2. Congenital abnormalities are very common.
3. Awareness of common normal variants is important.
4. Pediatric urological conditions are diverse with many different approaches to imaging and management.

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