Hydronephrosis and Pyonephrosis

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18.1 Introduction

Urinary tract infections (UTIs) are a frequent clinical problem in infants and children, and they may develop into serious complications with long-term sequelae. Their prevalence varies according to age and sex: males are more inter- ested in the neonatal period and in the first year of age, while females are more involved after that time. The clinical outcome and the severity of UTIs depend on many risk factors including uri- nary tract malformations and dysfunctions, viru- lence and properties of the pathogen, host response to the infection, and promptness of diagnosis and management.

Febrile UTIs usually are the result of a pyelo- nephritis, but more serious and uncommon pre- sentation such as renal abscess and a pyonephrosis may occur and they are clinically indistinguish- able from pyelonephritis. Various manifestations

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sometimes coexist and represent different expressions of the same infectious process that may evolve from a mild infection to dangerous complications, like emphysematous pyelone- phritis. The pyonephrosis is included in this inflammatory spectrum especially in the pres- ence of obstructed and/or dilated collecting sys- tems (Bitsori et al. 2015).

18.2 Hydronephrosis and Pyonephrosis

A pyonephrosis is associated with dilation and obstruction of the collecting system and is defined as an infected hydronephrotic kidney, which results in the accumulation of pus in the upper excretory pathways.

It is a severe and aggressive infection origi- nated in the enlarged pelvicalyceal system that causes destructive changes of renal parenchyma and inflammatory reactions in the perirenal tis- sues; in cases of delayed diagnosis, it may deter- mine sepsis and irreversible renal function deterioration. The purulent exudate is composed of sloughed urothelium, bacteria, debris, and inflammatory cells; the amount of pus varies depending on the type of infection and severity of the inflammatory process and can become obstructive or worsen a prior obstructive condition with consequent hydronephrosis. The pathogenesis of pyonephrosis implies infection

(almost always ascending dissemination), obstruction, and stasis as trigger mechanisms. The bacterial spectrum responsible for this path- ological entity does not differ from that of other forms of UTIs; the vast majority of

etiological agents derives from the intestinal flora, including *E. coli*, *Klebsiella*, *Proteus*, and *Enterobacter* species between the most frequent. Pyonephrosis may occur as a complicated evolution of pyelo- nephritis or represent the clinical manifestation of a superimposed infection developed on a pre- existing hydronephrosis; the latter needs further investigations to diagnose the underlying cause, once the acute phase has been solved and the child has recovered. Pediatric hydronephrosis is classified as obstructive and nonobstructive; the most common causes are vesicoureteral reflux (VUR) and pelviureteric junction obstruction, while less frequent are ureteral duplication, ure- terocele, ectopic ureter, and posterior urethral valves (Sharma et al. 2004; Patel et al. 2013).

Obstructive uropathy secondary to lithiasis has already been treated in Chap. 16.

18.2.1 Diagnosis

The clinical manifestations of pyonephrosis may range from asymptomatic bacteriuria to frank sepsis. In the older children, the presentation is similar to that of the adults: a history of high- grade fever, chills, flank pain, and tenderness is very suspicious for a patient affected by upper UTIs. Neonates and younger children are not able to describe their clinical condition and may present nonspecific symptoms like fever, lack of appetite, lethargy, irritability, vomiting, or diar- rhea. At physical examination, the child mani- fests pallor, dehydration, abdominal distension, and loose of bowel motion; rarely, a palpable abdominal mass is appreciable in the site of the hydronephrotic kidney. Laboratory studies include blood and urine exams. Hematology results show raised white cell count, C-reactive protein, urea, and creatinine; blood culture is needed in case of suspected sepsis. Urine analy- sis reveals bacteriuria with leukocytes and nitrites; pyuria may be present; urine culture

must be obtained in order to guide antibiotic therapy (Bitsori et al. 2015; St Lezin et al. 1992). All these clinical and laboratoristic features are not specific for pyonephrosis and have to be associated with the evidence of obstruction, hydronephrosis, and suppurative infection in order to support the diagnosis; these findings are shown by imaging investigations, especially ultrasound (US) that represents the first-level

examination.

18.2.2 Imaging Findings

US provides a sensitivity of 90 % and specificity of 97% in the diagnosis of pyonephrosis versus simple hydronephrosis, by showing in addition to pelvicalyceal dilation the following findings: lack of a completely anechoic collecting system due to echogenic debris, representing the most reliable signs of pyonephrosis, internal echoes dispersed or stratified with evidence of fluid- debris levels, and echogenic foci with acoustic shadowing corresponding to gas within the pel- vis, secondary to gas-forming organisms (Fig. 18.1). These findings are specific and their absence excludes pyonephrosis with a high degree of accuracy (Craig et al. 2008; Subramanyam et al. 1983; Jeffrey et al. 1985).

The associated dilation of upper excretory system is commonly evaluated basing on the hydronephrosis grading system represented in Table 18.1 (Figs. 18.2, 18.3, and 18.4) (Riccabona et al. 2008; Schlomer et al. 2014).

Depending on the variable nature of the puru- lent exudate, occasionally pyonephrosis may be totally anechoic simulating uncomplicated hydronephrosis, and US may only demonstrate nonspecific findings like nephromegaly and/or increased parenchymal echogenicity. In these cases, clinical manifestation and laboratory anal- ysis support the diagnosis when upper UTI is strongly suspected (Schneider et al. 1989).

Computed tomography (CT) is quite rarely performed in pediatric patients, usually in cases of emergency and complicated form of pyelonephritis. Considering that infants have higher radiation sensitivity, it obviously requires

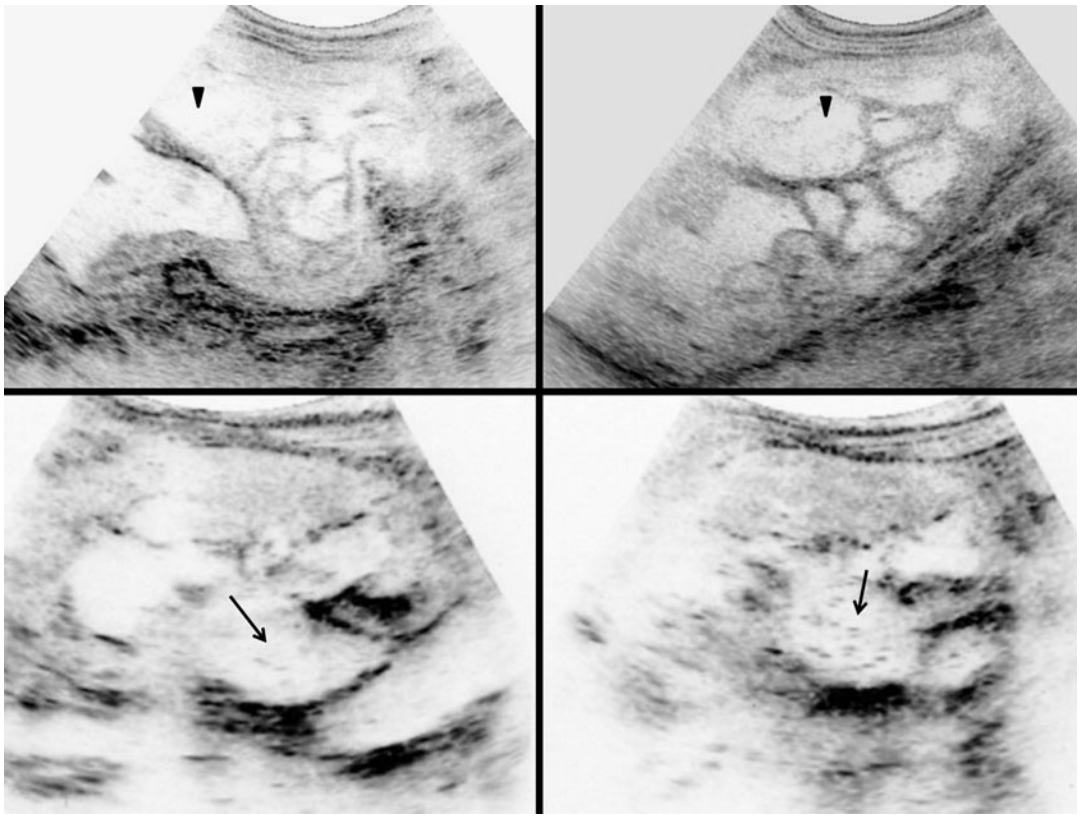
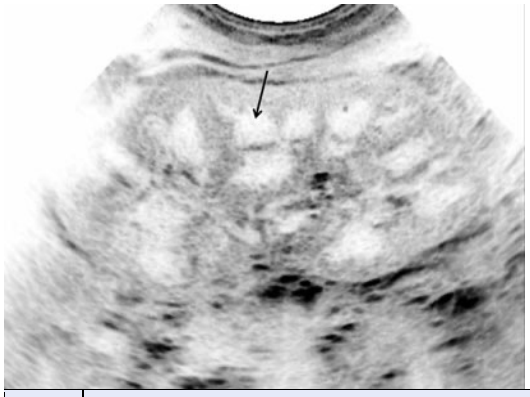


Fig. 18.1 (**a**–**d**) Hydropyonephrosis at US. Dilated and enlarged pelvicalyceal system with lack of normal anechoic aspect due to accumulation of inflammatory material in the excretory pathway: the exudate is well

Table 18.1 Grading systems of hydronephrosis

detected by revealing the presence of internal echoes that may appear stratified with evidence of fluid-debris levels (*arrowhead* in **a** and **b**) or dispersed as echogenic foci with acoustic shadowing (*arrow* in **c** and **d**)

Fig. 18.2 Normal pediatric kidney at US. The aspect of a healthy kidney especially in neonates is characterized by a physiological increased echogenicity of the renal paren- chyma with hypo-/anechoic pyramids that are therefore more evident and may resemble dilated calyces, being wrongly diagnosed as hydronephrosis



Grade 0	Normal examination with no dilation of the renal pelvis
Grade 1	Mild dilation of renal pelvis only (axial diameter less than 5–7 mm), without dilation of the calyces
Grade 2	Moderate dilation of the renal pelvis (axial diameter between 7 and 10 mm) including a few calyces with normal forniceal shape
Grade 3	Marked dilatation of the renal pelvis (larger than 10 mm) and all the calyces with reduced forniceal and papillary differentiation, normal renal parenchyma
Grade 4	Gross dilatation of the collecting system, parenchymal atrophy seen as thinning of the renal cortex

Adapted from the Society of Fetal Ultrasound classifica- tion for postnatal use Riccabona et al. (2008)

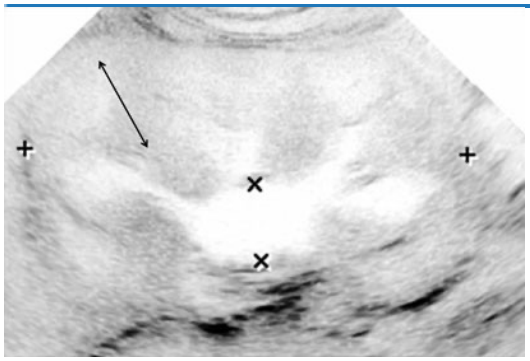


Fig. 18.3 Hydronephrosis: grade 3. Marked dilation of the renal pelvis and calyces with normal thickness of renal parenchyma that is still preserved (*doublehead arrow*)

Fig. 18.4 Hydronephrosis: grade 4. Marked dilation of the collecting system with reduced thickness of the renal parenchyma seen as thinning of the renal cortex (*double- head arrow*)

dedicated age- and weight-adapted protocols for exposure parameters and contrast medium, and it may be performed in case of correct indications and after accurate evaluation of the diagnostic advantages (Riccabona 2009).

Actually CT cannot always reliably differen- tiate an uninfected hydronephrotic kidney from pyonephrosis, especially only on the base of fluid attenuation measurements within the pel- vis that should result superior to the water den- sity value (>10–20 Hounsfield unit). In addition to obstruction and dilatation of the collecting system, CT findings suggesting pyonephrosis include thickening of the renal pelvic wall (>2 mm), renal and perirenal inflammatory changes, and layering of contrast material above the purulent debris on excretory study (Craig et al. 2008; Fultz et al. 1993).

The added value of CT consists of showing perinephric findings (such as fat stranding and thickening of the renal fascia), evaluating the nephrogram appearance to reveal the parenchy- mal involvement, confirming the presence of gas in the collecting system (the strongest indicator for pyonephrosis on CT), and ruling out abscesses or stone disease. Furthermore, CT is relevant when a complicated evolution is suspected espe- cially because a different and more aggressive treatment may be required.

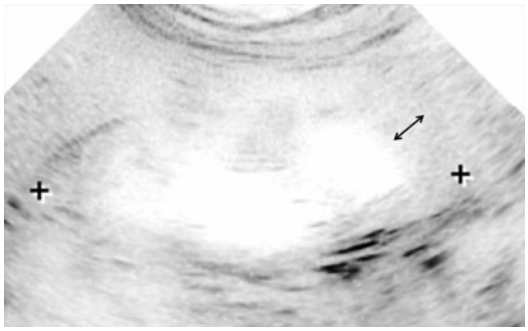
18.2.3 Treatment and Prognosis

The treatment of choice is represented by percu- taneous nephrostomy in combination with appro- priate antibiotics and other supportive therapy. Nephrectomy is exceptional and limited to com- plicated cases (Roebuck 2011; Ng et al. 2002).

The prognosis is good in most patients who receive prompt diagnosis and treatment. Pyonephrosis should be diagnosed as soon as possible in order to avoid a rapid decline in renal function risking permanent loss of kidney paren- chyma and development of septic shock with severe systemic complications.

UTIs, especially when recurrent, remain a fre- quent indication for further imaging evaluation of the pediatric urinary tract, and once the acute phase of the infectious disease is solved, poten- tial predisposing factors must be investigated. The goal is early recognition of urinary tract mal- formations and VUR, in order to improve out- come and prevent end-stage renal failure due to late or inappropriate treatment. Indeed, in spite of prenatal US screening, a number of urinary tract malformations are only detected after symptom- atic clinical presentation. On the other hand, in a child with known normal anatomy and a clini- cally evident diagnosis, additional imaging may be unnecessary.

Second-level investigations include cystoure- throgra phy for VUR assessment and morphologi- cal evaluation, bladder function studies, dimercaptosuccinic acid (DMSA) scintigraphy, and magnetic resonance imaging (MRI) to evalu- ate renal parenchymal involvement and scars.



18 Hydronephrosis and Pyonephrosis

18.3 Vesicoureteral Reflux

The VUR is one of the most important risk fac- tors for recurrent UTIs in children, although VUR itself does not represent the direct cause of infections or renal damage and it may be associ- ated with bladder dysfunction. VUR refers to the retrograde flow of urine from the urinary bladder into the ureter and often to the calyces (Bates and Riccabona 2013). It is an anatomic and func- tional disorder with potentially serious conse- quences, such as recurrent febrile UTI and renal parenchymal scars (Altobelli et al. 2014).

VUR is detected in 0, 4–1, and 8 % of normal population without complaints (Eccles and Jacobs 2000; Mak and Kuo 2003; Lama et al. 2000). Its prevalence is probably underestimated because reflux is diagnosed in those patients who are symptomatic. In newborns, VUR is more prevalent in males, but female-to-male ratio of reflux in children over 3 years old seems to be of 2:1 (Altobelli et al. 2014; Chand et al. 2003). White children are affected ten times more than black ones, in particular red-hair children seems to have an increased risk of VUR.

In the population with UTIs, the incidence varies from 30 % to 50 % depending on age and gender. The reported prevalence of VUR in sib- lings (27 %) and in offspring (36 %) suggested that there is a genetic component, still undefined, but supposed to be dominant with variable pene- trance (Skoog et al. 2010; Kelly et al. 2005).

Approximately 20–30 % of children with VUR present with renal lesions: despite neonatal VUR, there is a high percentage of spontaneous resolution (30–40% of grade 4–5 VUR can resolve within 2–6 years of age); nevertheless this population have high risk of acute pyelone- phritis, hypertension, and chronic renal failure during the first months of life (Lama et al. 2000; Caione et al. 2004). In children and young adults who need substitutive renal therapy, VUR inci- dence is about 6 % and it represents the fifth most common cause of chronic renal insufficiency (CRI) (Ardissino et al. 1995). However, the severity of VUR greatly varies and thus may affect patients differently, and some patients seem to have a genetic predisposition to renal

injury (Bates and Riccabona 2013). Hypertension develops in 10 % of children with unilateral scars and in 18.5 % with bilateral scars. Approximately 4 % of children with VUR progress to end-stage renal failure (Ardissino et al. 2004).

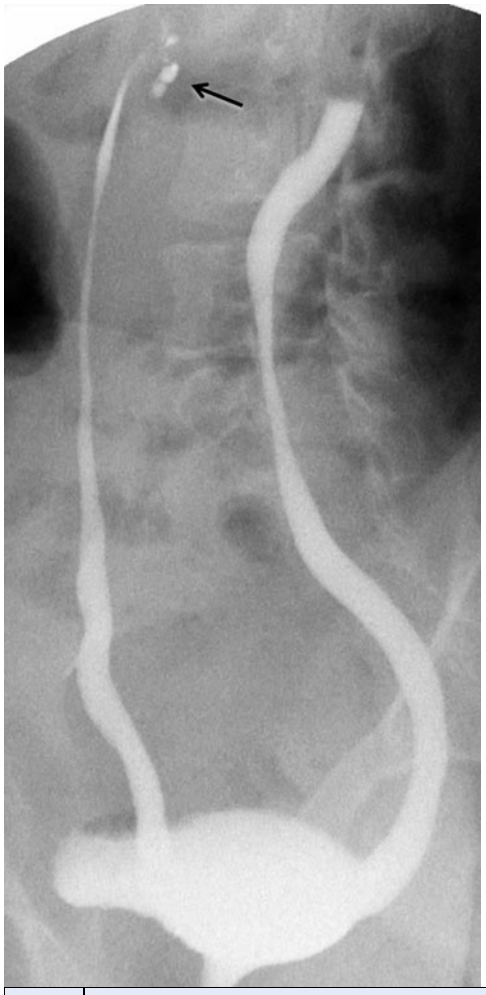
VUR is associated with anomalies of the ure- terovesical junction (UVJ), and it is often seen in patients with other urinary tract anomalies (Knudson et al. 2007a). UVJ anomalies consist in excessive lateralization, larger dimensions, shorter length of submucosal ureteral tract, or deficiency in longitudinal muscular fibers of the terminal ureter (Bates and Riccabona 2013).

Congenital or primary VUR may be caused by a sort of immaturity of UVJ, which spontane- ously solves in large percentage within the first year of life. Secondary VUR is a functional disor- der found in patients with bladder outlet obstruc- tion or neurogenic bladder disease, which lead to thinning and weakening of the UVJ musculature and increased intravesical pressure (Bates and Riccabona 2013).

Two types of urine may flow back to the caly- ces: infected or sterile urine. Intrarenal reflux of infected urine seems to be the cause of renal damage, whereas intrarenal reflux of sterile urine (under normal intrapelvic pressure) has not been identified as responsible for clinical significant renal scars. Thus, renal lesions seem to develop only in the setting of intrarenal reflux in combi- nation with UTIs, except in patients with neuro- genic bladder disease (Knudson et al. 2007b). In this condition also reflux of sterile urine may lead to renal damage in association with the highly pressurized system, especially in high grade of reflux (Knudson et al. 2007b). In children, high intravesical pressure may be sustained by overac- tive bladder (e.g., detrusor hyperreflexia, detrusor instability), aggravating a preexisting primary VUR or causing secondary VUR.

Historically, VUR is classified by the International Reflux Committee into five grades (Table 18.2), based on the degree of retrograde filling and dilatation of the renal collecting sys- tem seen on voiding cystourethrogram (VCUG) (Figs. 18.5, 18.6, 18.7, 18.8, and 18.9) (Lebowitz et al. 1985). Higher grades of reflux are associ- ated with decreased self-resolution rate, which is

Table 18.2 VUR classification into 5° on VCUG by the International Reflux Committee (Knudson et al. 2007b)



Grade 1	Urine backs up into the ureter only, and the renal pelvis appears healthy, with sharp calyces
Grade 2	Urine backs up into the ureter, renal pelvis, and calyces; the renal pelvis appears healthy and has sharp calyces
Grade 3	Urine backs up into the ureter and collecting system; the ureter and pelvis appear mildly dilated, and the calyces are mildly blunted
Grade 4	Urine backs up into the ureter and collecting system; the ureter and pelvis appear moderately dilated, and the calyces are moderately blunted
Grade 5	Urine backs up into the ureter and collecting system; the pelvis is severely dilated, the ureter appears tortuous, and the calyces are severely blunted

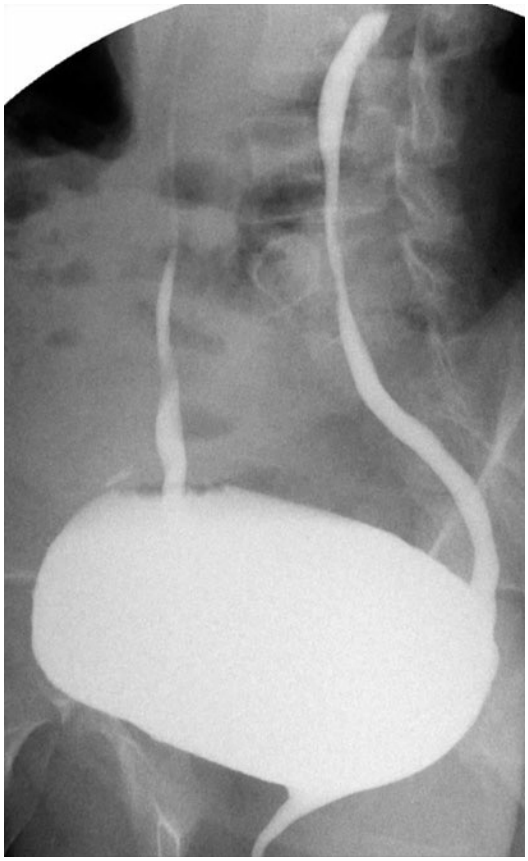


Fig. 18.5 VCUG: vesicoureteral reflux, grade 1. Retrograde filling of contrast medium into the ureters only that are not dilated

Fig. 18.6 VCUG: vesicoureteral reflux, grade 2. Retrograde filling of contrast medium into the ureters bilaterally and until the pelvis and calyces on the right side (*arrow*), without dilation

also related to some other factors such as age pre- sentation, gender, laterality, bladder volume, and pressure at the onset of reflux, voiding dysfunc- tion, history of UTI, ureteral anatomy, and preva- lence of renal scars (Bates and Riccabona 2013).

VUR may be diagnosed in the prenatal period at sonographic examination when transient dila- tation of the upper urinary tract is noted in con- junction with the bladder emptying in late gestation (>28 weeks). Approximately 10% of neonates diagnosed prenatally with dilatation of the upper urinary tract will be found to have reflux postnatally (Ylinen et al. 2003). In neonates

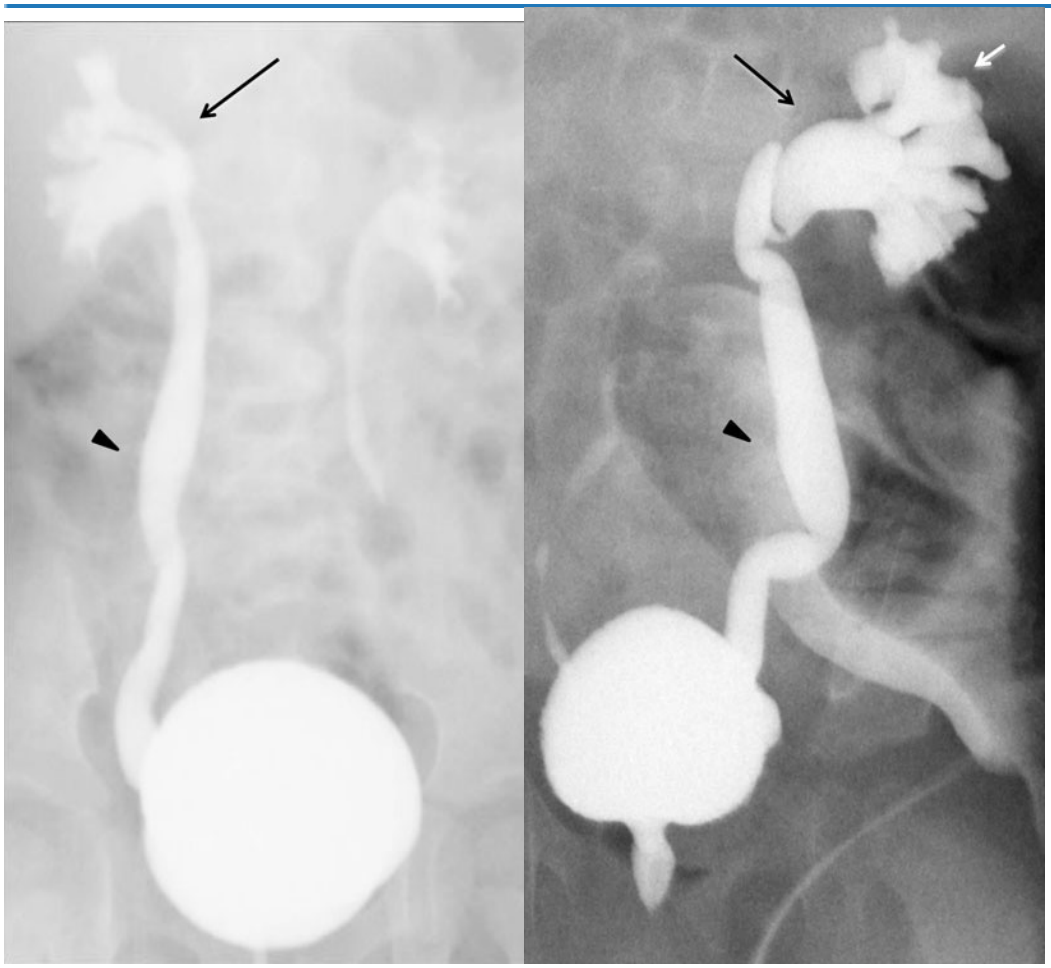


Fig. 18.7 VCUG: vesicoureteral reflux, grade 3. Retrograde filling of contrast medium until the pelvis and calyces bilaterally, with mild pelvis (*arrow*) and ureter (*arrowhead*) dilation on the right side

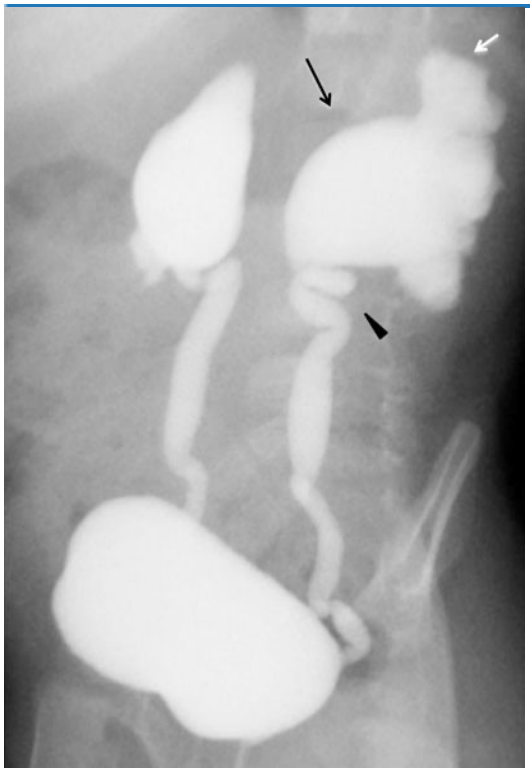
clinical presentation is characterized by respira- tory distress, persistent vomiting, failure to thrive, renal failure, flank masses, and urinary ascites, and VUR may subsequently be diagnosed with severe UTI. Recurrent febrile UTIs are the most important and common presentation.

In the past, Academy of Pediatrics guidelines recommended renal-bladder US and VCUG for any child from 2 to 24 months of age who pre- sented with their first febrile UTI. After a revision in 2011, only US is recommended after first epi- sode of UTI (Roberts 2011). To evaluate second- ary VUR caused by bladder/bowel dysfunction, recent studies have identified specific voiding

Fig. 18.8 VCUG: vesicoureteral reflux, grade 4. Retrograde filling of contrast medium until the pelvis and calyces on the left side, with dilation of the ureter (*arrow- head*) and pelvis (*white arrow*) and moderate blunted aspect of the calyces (*black arrow*)

patterns during urodynamic study in children with voiding dysfunction without anatomical and/or neurological abnormalities (Altobelli et al. 2014).

US is necessary to examine the upper urinary tract morphology, dilatation, parenchymal thick- ness, and echogenicity (Altobelli et al. 2014). US can identify indirect signs of VUR, such as uro- epithelial thickening of the ureter or renal pelvis, changing diameter of the pelvicalyceal system and ureter, quick refilling of the bladder after voiding, asymmetric ureteral inflow jets, and a lateralized position or unusual shape of the ure-



VCUG: vesicoureteral reflux, grade 5. Bilateral retrograde filling of contrast medium until the pelvis and calyces, with severe dilation of the pelvis (*white arrow*) and ureter that is tortuous (*arrowhead*); severe blunted aspect of the calyces (*black arrow*)

teral ostium (Bates and Riccabona 2013). Great dilatation of the whole collective system, with clubbing of the calyces and elongated and tortu- ous ureters, is typical of severe VUR. These signs are often associated with poor ureteral peristalsis, especially in correlation with high-grade VUR and UTI.

Moreover, VUR may be directly observed by contrast-enhanced voiding ultrasonography (ce- VUS) that consists in a US examination with injection of shaken saline solution with air or commercially available contrast agents via supra- pubic or transurethral catheterization till bladder filling (Bates and Riccabona 2013).

Ultrasound techniques such as harmonic imaging, stimulated acoustic emission, or other contrast-specific techniques have further enhanced ce-VUS potential for VUR depiction and grading, resulting in a reported sensitivity

and specificity equal to VCUG (Bates and Riccabona 2013; Riccabona 2008).

Nevertheless, VCUG remains the basic imag- ing technique to detect VUR and to determine its degree (Figs. 18.5, 18.6, 18.7, 18.8, and 18.9). This exam is performed with iodinated contrast medium instilled into the catheterized and emp- tied urinary bladder, which is filled to near capac- ity. Bladder capacity is estimated on the basis of weight, multiplied by 7 (mL = kg × 7). Over 1 year of age, it equals age in years plus 2, multi- plied by 30 [mL = (age + 2) × 30].

The fluoroscopic observation of the (early) filling phase, the distal ureters (in oblique pro- jections), the renal collecting system, and the urethra during voiding (lateral projection in boys) enables focused imaging of critical areas and conditions such as intrarenal VUR and UVJ anatomy (Bates and Riccabona 2013). VUR is not always well depicted in the first fill and void; in order to avoid missing signifi- cant VUR, VCUG should be repeated cyclically.

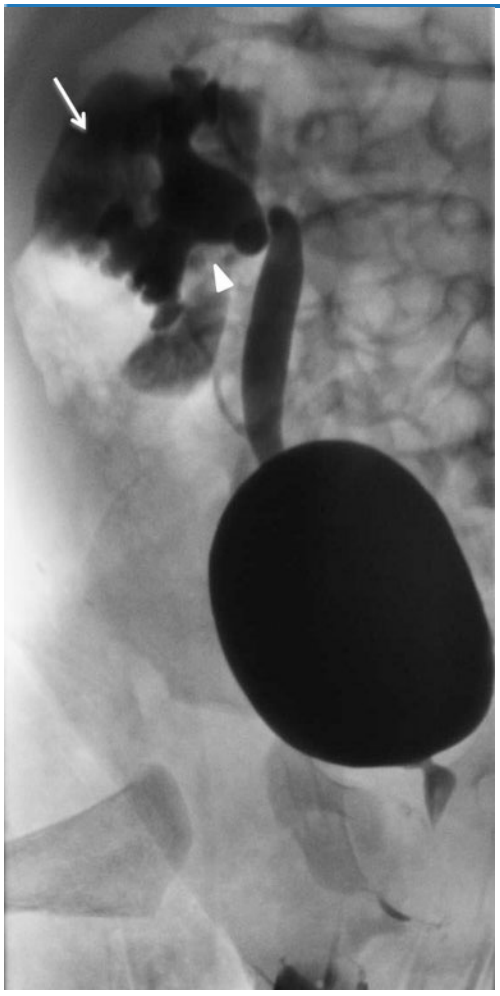
The appearance of refluxing ureters and pelvi- calyceal systems on VCUG is quite variable, ranging from normal-sized upper tracts to extreme upper tract dilatation and marked ure- teral tortuosity (Bates and Riccabona 2013). In some patients, VUR is accompanied by a tran- sient phenomenon depicted as marked ballooning of the

pelvicalyceal system in the absence of evi- dence of ureteropelvic junction (UPJ) obstruc- tion. Sometimes VUR may induce a kink at the UPJ, producing a functional valve-like UPJ obstruction.

When VUR is extended to the calyces, finding an intrarenal reflux is important because of its association with renal scarring. Intrarenal reflux may be depicted as a transient pyelotubular and interstitial reflux of contrast media extending outward in a wedge-shaped pattern from one or more papillae to the renal cortical surface (Fig. 18.10).

The physiopathological basis of intrarenal reflux is supposed to be an abnormal morphology of the opening of the collecting ducts of Bellini on the renal papilla, which appears round, more common at the poles, and therefore less resistant

Fig. 18.9



VCUG: intrarenal reflux. Image shows con- trast medium reflux until pelvis and calyces extending outward from the papillae to the interstitium of the renal cortex (*arrow*), so that the profile of the kidney results well delineated

to retrograde flow than the slitlike openings of the ducts of simple or conical papillae (Bates and Riccabona 2013).

The study of the renal parenchyma is usually performed by US and by DMSA scintigraphy. US may show parenchymal damage depicted as areas of depression in the outline of the kidney in correspondence of the

dilated and distorted calyx. Color Doppler imaging may reveal a focal reduc- tion of peripheral vascularization. Nuclear scin- tigraphy using DMSA is especially sensitive in detecting renal cortical scars and is considered

the gold standard. This technique may reveal the inflammation of acute pyelonephritis or chronic renal scars, evaluating the degree of renal dam- age, which has main importance during follow- up (Bates and Riccabona 2013).

Other methods for the evaluation of the upper tract are contrast-enhanced CT and MRI, which may become indicated for assessment of compli- cated disease (Bates and Riccabona 2013).

The European Association of Urology recom- mends a DMSA scan if VUR is diagnosed on ini- tial VCUG. On the other hand, the American Urological Association recommends the renos- cintigraphy study only if an abnormal US is reported during breakthrough UTIs, in children presenting high-grade VUR (grade III–V) or ele- vated serum creatinine (Peters et al. 2010). This diagnostic approach is "bottom-up" (Altobelli et al. 2014).

The recent National Institute for Health and Clinical Excellence (NICE) guidelines alterna- tively suggest a "top-down approach" with an initial renal ultrasound or a DMSA scan per- formed 4–6 months after the infection in order to assess renal scarring rather than pyelonephritis, in children with their first febrile UTI. This approach may avoid unnecessary VCUG in patients with normal DMSA (in 35–60% of cases), reducing children radiation exposure (Herz et al. 2010; National Collaborating Center for Women's and Children's Health (UK) 2007; Weinberg et al. 2013). This same motivation leads some authors to suggest also the use of ce- VUS with endovesical instillation (Piaggio et al. 2003). This is the main advantage of the recent "top-down approach" (Altobelli et al. 2014).

18.4 Ureteropelvic Junction Obstruction

The UPJ obstruction is the most common cause of hydronephrosis in neonatal age, and it is usu- ally due to an intrinsic cause (a short stenotic segment at the UPJ) or less commonly to an extrinsic compression secondary to bands, kinks, or aberrant vessels (Dewan et al. 1998). Obstruction can be also determined by a high

Fig. 18.10

insertion of the UPJ on the dilated renal pelvis, but the UPJ displacement could be an effect rather than a cause. Aberrant or crossing lower pole vessels are found in more than 30 % of older children and adults undergoing pyeloplasty for this cause, while they are present in less than 5 % of infants with prenatally detected UPJ obstruc- tion; they are incidental to an intrinsic obstruc- tion in the latter, but in older patients, they are the cause of the obstruction.

The male-to-female ratio is approximately equal and the left kidney is more commonly affected than the right. The condition usually occurs on a sporadic basis, but familial inheri- tance has been reported. UPJ obstruction is more common in children with other urinary tract anomalies such as multicystic dysplastic kidneys and the VACTERL spectrum of anorectal and vertebral anomalies (Thomas et al. 2008).

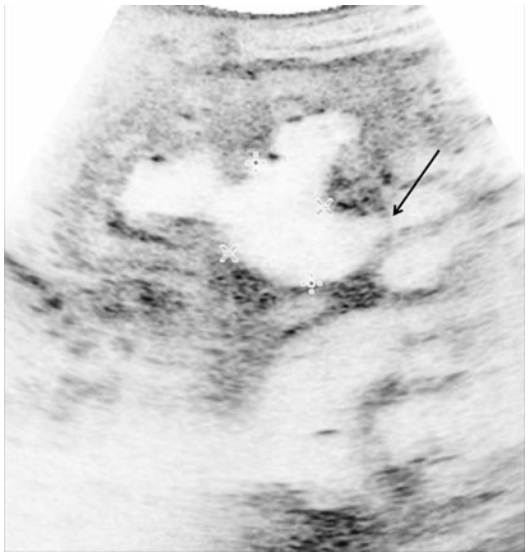
With the development of antenatal ultrasound, UPJ obstruction is now diagnosed more during the perinatal period in asymptomatic infants (Grignon et al. 1986); however, UPJ obstruction may go unnoticed antenatally and present in emergency settings with a palpable mass or abdominal distension in neonates, or with severe abdominal pain, urinary tract infection, or hema- turia in older children. Intermittent pain associ- ated with vomiting is also recognized (Marincek and Dondelinger 2007). Unlike nonspecific abdominal pain, the symptoms arising from an obstructed kidney usually last for several hours or several days. Although the pain is typically

sited in the region of the loin, it may be experi- enced predominantly in the central quadrants of the abdomen, with possible diagnostic difficul- ties (Thomas et al. 2008).

Current assessment of the child with apparent UPJ obstruction rests upon a determination of structure and functional studies. Typically, the sequence comprises ultrasound followed by ^{99m}Tc mercaptoacetyltriglycine (MAG3) dynamic renography. The use of DMSA scintigraphy or ^{99m}Tc DMSA scintigraphy is reserved for poorly functioning obstructed kidneys.

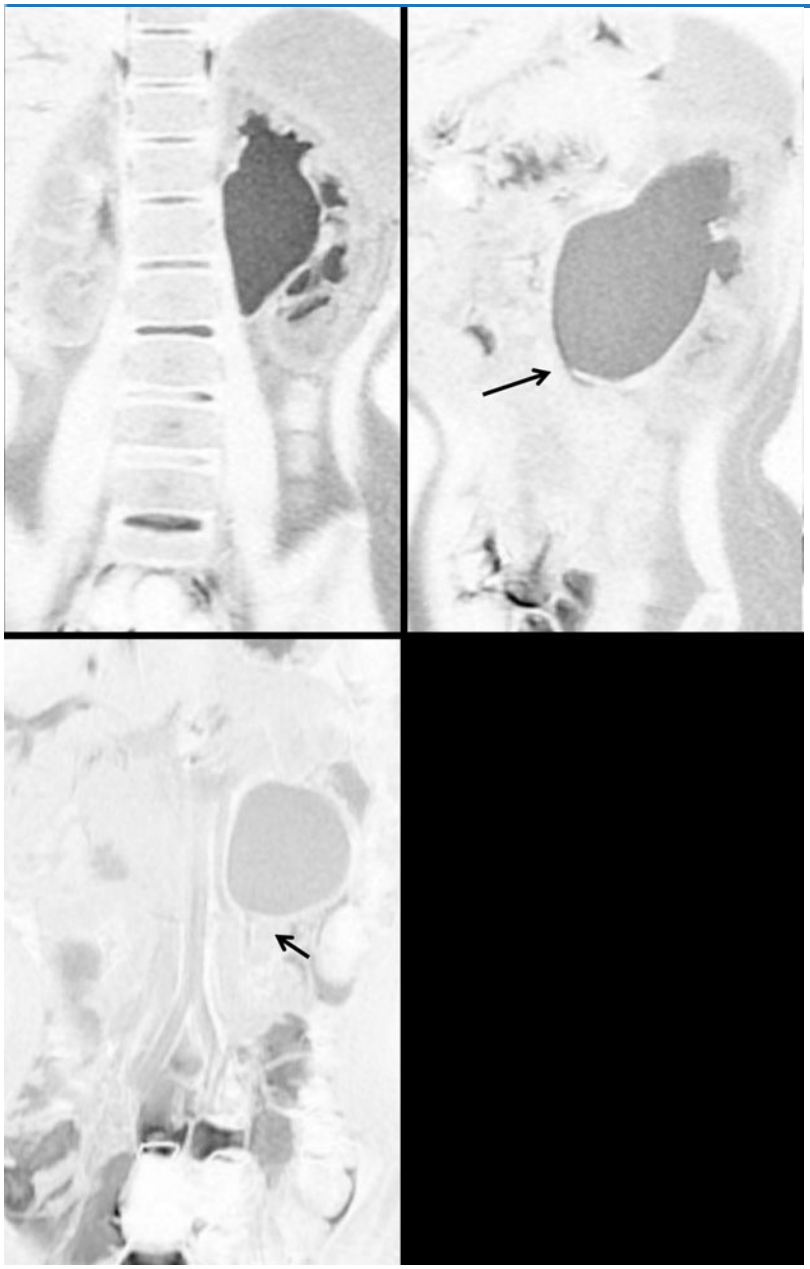
US is the initial imaging of choice used in children with suspected urinary tract obstruction. It shows dilated renal pelvis communicating with

dilated calyces in the absence of visualization of the distal ureter (Fig. 18.11). The renal cortex may appear thin, and a crossing blood vessel at the UPJ may be visualized on color Doppler ultrasound. In older children, the concern for a crossing lower pole renal vessel is higher than in infants, so occasionally CT or MR angiography is sometimes obtained to look for these vessels (Fig. 18.12). Occasionally, the distal ureter appears dilated due to associated vesicoureteric reflux or to a more distal cause of obstruction. The UPJ obstruction must be distinguished from a multicystic kidney; the latter appears as non- communicating area of cystic dilation of vari- able size (Marincek and Dondelinger 2007). A massively dilated renal pelvis without signifi- cant caliectasis suggests a lesser degree of obstruction, and one should not base clinical decisions on the degree of pelvic dilation alone (Lima and Manzoni 2015). A similar obstruc- tion can be determined by a retrocaval ureter, by a functional impairment of the peristaltic con- traction of the ureter, by secondary obstruction due to gross VUR, and by distortion of the ure- ter in a horseshoe kidney, so all this conditions must be excluded.



UPJ obstruction. Longitudinal US scan shows dilated renal pelvis communicating with dilated calyces and a sudden narrowing in the site of the ureteropelvic junction (*arrow*). The distal ureter is not visualized

Fig. 18.11



a

b

c

Fig. 18.12 UPJ obstruction at MRI: T2-weighted single- shot fast-spin-echo (SSFSE) images. Coronal SSFSE (**a**) shows severe dilation of the pelvis and calyces with asso- ciated thinning of the renal cortex. Sagittal SSFSE (**b**) and coronal fat saturated SSFSE (**c**) better highlight the site of the obstruction and the distal ureter with normal caliber (*white arrows*). In this case, a crossing blood vessel at the UPJ was not identified

The most reliable current method of assessing relative renal function is the diuretic renogram (DR), a functional study using ^{99m}Tc MAG3 per- formed to assess the renal drainage and compare the function of the kidneys (Stauss et al. 2003).

The UPJ obstruction is confirmed by showing dilatation of the upper urinary tract, failure of tracer washout after diuretic administration, and gradually rising activity on the time-activity graph. The greatest value of the DR and washout

curve is to monitor the status of the kidney over time, particularly when an observational approach is taken. This is though a burden on families and includes increased radiation. The child with more severe dilation and more delayed washout is more likely to not improve spontane- ously or to deteriorate. As long as the family is aware of this and is reliable to return for follow- up, observation is the advisable approach (Thorup et al. 2003).

If the ureter is dilated or the bladder wall is thick, a VCUG is indicated to exclude vesicoure- teric reflux and urethral valves.

The goal in diagnosing and treating UPJ obstruction is to prevent ipsilateral renal function loss and minimize associate comorbidities, like UTIs and urolithiasis.

Acute management of a UPJ obstruction is usually the placement of a percutaneous nephros- tomy, and functional imaging is performed when acute symptoms have settled. Treatment is either conservative (wait and see) or surgical, depend- ing on the severity of functional loss. It is well established that after surgery the hydronephrosis can resolve spontaneously.

18.5 Duplex Kidney

A duplex kidney is a kidney with two pelvicaly- ceal systems, generally referred to as upper and lower poles. The upper pole normally accounts for one third of the kidney, whereas the lower one for the remaining two thirds. Anatomically, duplex systems can be incomplete (bifid pelvis or ureter) or complete. The former are four to five times more common than the latter, which are most common in females. In approximately 40 % of cases, the condition is bilateral. Partial dupli- cations will have convergence of the ureters into a single system prior to its entry into the bladder.

When the separate buds incorporate into the bladder, the more cranial bud terminates in a location more caudal and medial than expected. Conversely, the most caudal bud ends more cra- nial and lateral. This phenomenon is known as the Weigert-Meyer rule and has only rare excep- tions (Mackie and Stephens 1975; Jain et al.

2008). The lower pole ureter tends to have a shorter course within in the bladder wall due to its lateral positioning, predisposing it to VUR, whereas the upper pole ureter with a long course within the bladder wall is more often associated with obstruction or ureterocele. In addition, ure- teral ectopia is more common in the upper pole, whereas UPJ obstruction in the lower pole (Whitten and Wilcox 2001). Upper pole UPJ obstruction and primary upper pole VUR are rare for anatomical and embryological reasons.

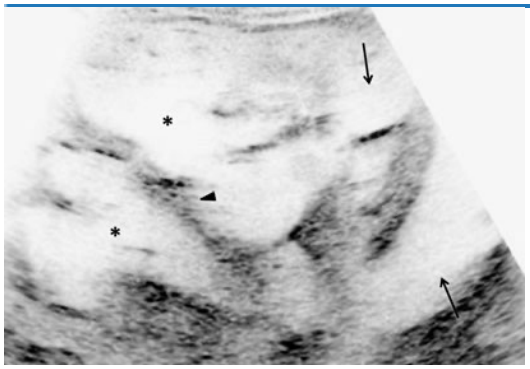
Uncomplicated duplex system anomalies often go undiagnosed for the whole life. In con- trast, complicated ureteral duplications are often associated with other urological anomalies and more easily present themselves with different symptoms.

Febrile UTIs are the most common presenting symptoms. They occur more commonly when a VUR is associated with the condition in any of the moieties or with hydroureteronephrosis (Lima and Manzoni 2015). Risk of infection might be higher in case of ureterocele, although it is still unclear whether this is due to the uretero- cele itself or to the fact that VUR is often associ- ated (Castagnetti et al. 2012).

Basic diagnostic workup in patients with a complicated duplex system should generally undergo US of the upper urinary tract and blad- der and a renal nuclear scintigraphy. A VCUG should be considered if a dilated ureter is visible on US. Additional imaging may be useful when anatomical information is necessary.

The US of the urinary tract is generally the first investigation. Sonographically, duplicated systems will have an intervening bar of renal parenchyma separating the upper and lower poles. This parenchyma is more evident in the presence of hydronephrosis (Fig. 18.13). The US should define the laterality of the condition and identify the moieties that are dilated; the degree of the dilatation, whether or not ureteral dilata- tion is associated; and the aspect of the bladder including the presence of a ureterocele.

Since duplication is often associated with VUR, VCUG should always follow but in suspi- cion of lower pole UPJ obstruction (Kim et al. 2001). VUR into a lower pole ureter may displace



Hydronephrotic duplex kidney. Longitudinal US scan shows two dilated pelvicalyceal systems (*aster- isks*), with interposed renal parenchyma (*arrowhead*) separating the upper and lower poles. Ureteral dilation of both the excretory system is associated (*arrows*)

the lower pole downward and outward, with the renal pelvis and calyces appearing like a “drooping lily.” In order to rule out the presence of a ureterocele, a picture should be taken after injection of few cc of contrast into the bladder, at low-volume bladder filling, to prevent uretero- cele collapse.

Finally, an assessment of moiety function should be achieved by nuclear scintigraphy. The dynamic ^{99m}Tc MAG3 scintigraphy allows for simultaneous assessment of cortical function and upper tract drainage: the information necessary for the decision-making is the gross presence of any function in the moiety or its complete absence. Patients with primary lower pole VUR are the only exception, in which using a static ^{99m}Tc DMSA scintigraphy can be useful to search for parenchymal scars. It is recommended, how- ever, also a MAG3 scan if any trapping of con- trast in the upper tract is seen on the VCUG, in order to rule out any concomitant obstruction that might contraindicate the injection of bulking agent as treatment of reflux, should recurrent febrile UTIs occur (Othman et al. 2012).

If additional imaging is required, i.e., for massive nonrefluxing dilatation of both upper and lower ipsilateral moieties or for incongru- ence in the basic assessment tests, uro-MRI allows for simultaneous optimal anatomical defi- nition and functional assessment (Grattan-Smith and Jones 2006).

Treatment of duplications is conservative in the majority of cases, surgical only in the pres- ence of severe complications or concomitant pathology.

18.6 Ureterocele

A cystic dilation of the distal ureter where it inserts into the bladder or urethra is called a ureterocele.

Ureteroceles are most commonly associated with the upper pole in a duplex kidney and occur more frequently in females (Shokeir and Nijman 2002a). The reported incidence of ureteroceles is 1 in every 1,000 births (Palmer and Palmer 2014). The risk of UTI in children with ureteroceles may be as high as 50% (Besson et al. 2000). Ureteroceles are classified as intravesical (or orthotopic) if they are entirely within the bladder and as ectopic if part of the ureterocele extends into the bladder neck or into the urethra. The exact embryologic explanation for ureteroceles is unknown, but one popular theory is that there is a membrane between the mesonephric duct and the ureteric bud that does not completely break down and leads to a stenotic ureteral orifice and obstruc- tion (Palmer and Palmer 2014).

Single-system ureteroceles are most common in boys and usually incidentally found during sonographic examinations, but they can present with acute signs of UTI or lithiasis. Upper tract dilatation is absent or of mild to moderate sever- ity and renal function is normally preserved. They usually lie entirely within the bladder (orthotopic ureteroceles).

Duplex system ureteroceles are associated with the upper pole ureter and can cause reflux or obstruction in the ipsilateral lower pole and, being more often ectopic, can cause bladder out- let obstruction. They can alter the function of the whole bladder, because of the trigone develop- ment already being partly abnormal due to the presence of two ipsilateral ureters (Lima and Manzoni 2015). Dysplasia of the upper pole parenchyma is the rule with duplex system ure- teroceles, usually to the extent that the upper pole has little if any useful function. Often this

Fig. 18.13

correlates with dilatation of the upper pole ureter. Clinical presentation of ureteroceles in a duplex system is typically with UTI, occasion- ally with Gram-negative septicemia, acute urinary retention, or, in females, with a vaginal mass determined by urethral prolapse of the ureterocele.

Most ureteroceles are diagnosed by ultraso- nography that demonstrates a thin-walled cystic mass within the posterior wall of the bladder and potentially hydroureteronephrosis (Fig. 18.14). Attention should be paid to the degree of bladder fullness during the exam, as ureteroceles can col- lapse and be overlooked in a full bladder. Conversely, if the bladder is too empty, the ure- terocele can easily be lost among the folds of the bladder wall. Pseudoureteroceles (the dilated dis- tal ureter pressing on the bladder wall) can be dif- ferentiated from real ureteroceles by the thickness of their walls: thin-walled for a true ureterocele and thicker bilaminar bladder wall for a pseudo- ureterocele. Upper pole dysplasia can be seen as hyperechoic parenchyma and smaller relative size with varying degrees of thickness.

A VCUG is generally performed as well because of the increased incidence of ipsilateral lower pole reflux in duplex systems. Reflux into both poles is usually indicative of incomplete duplication. Contralateral reflux can also be seen.

Fig. 18.14 Ureterocele. Axial US scan of the pelvis shows a thin-walled cystic dilation (*asterisk*) within the posterior wall of the bladder

Early images of the bladder with the VCUG will often demonstrate the ureterocele as a large, smooth, filling defect on the posterior wall/tri- gone. Once the bladder is full, the ureterocele may evert to look like a diverticulum with its location at the bladder base as the only indication of being a ureterocele. During the voiding phase, prolapse of a large ureterocele can be demon- strated with movement of it into the urethra.

Cystoscopy is routinely advisable for the assessment of duplex system ureteroceles, princi- pally in order to determine if the lesion is ortho- topic or ectopic (Shokeir and Nijman 2002b).

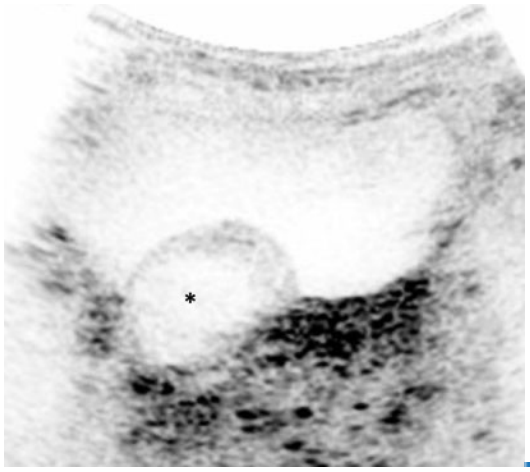
MRI has the potential to clarify anatomy and provide functional information without exposure to radiation. The major disadvantage is the requirement for general anesthesia or sedation in children to ensure they remain still while the scan is being performed.

Ureteroceles are treated to prevent or reduce UTI and to prevent renal damage. It is generally agreed upon that all children with a ureterocele should be on prophylactic antibiotics to prevent UTI and urosepsis (Husmann et al. 1995). The treatment for a child with a ureterocele and sep- sis, unresponsive to antibiotics, is urgent endo- scopic decompression via transurethral incision.

Indications to surgery are febrile UTIs and bladder outlet obstruction (Shokeir and Nijman 2002b).

18.7 Ectopic Ureter

An accessory bud that arises from the mesoneph- ric duct in a more cephalic position than the nor- mal site and which drains the upper renal pole will come to enter the urinary tract in a distally ectopic location, as said before. It may drain into either the bladder, the urethra, or the urogenital sinus. In females, such ectopic ureters may be sited above the sphincter mechanism (supra- sphincteric), close to or at the level of the striated sphincter (although usually below the bladder neck), or distal to the sphincter (infrasphincteric), either at the introitus or in the distal vagina. In males, the termination is always suprasphincteric (because mesonephric duct derivatives are all



18 Hydronephrosis and Pyonephrosis

proximal to the external urethral sphincter), con- necting with the ductus deferens, the seminal vesicle, or, most often, the ejaculatory duct.

Ureteric ectopia is usually associated with a dysplastic renal pole, while the ureter itself is fre- quently dilated, either as a consequence of reflux, obstruction, or dysmorphism. Even in the absence of dysplasia, the affected pole typically exhibits some degree of dilatation, the only common exception being in some girls with infrasphinc- teric ectopia.

Ectopic ureter is rare, affecting some 0.01% of individuals, mostly females.

Clinical presentation of suprasphincteric ecto- pic ureters is almost invariably with urinary infection, which, in males, may manifest as epididymo-orchitis. Rarely it can present with an abdominal palpable mass, represented by the hydronephrotic kidney. Urinary incontinence and hematuria can be typical of infrasphincteric ecto- pia. This incontinence is very specific, with a constant dribbling of urine superimposed upon an otherwise normal

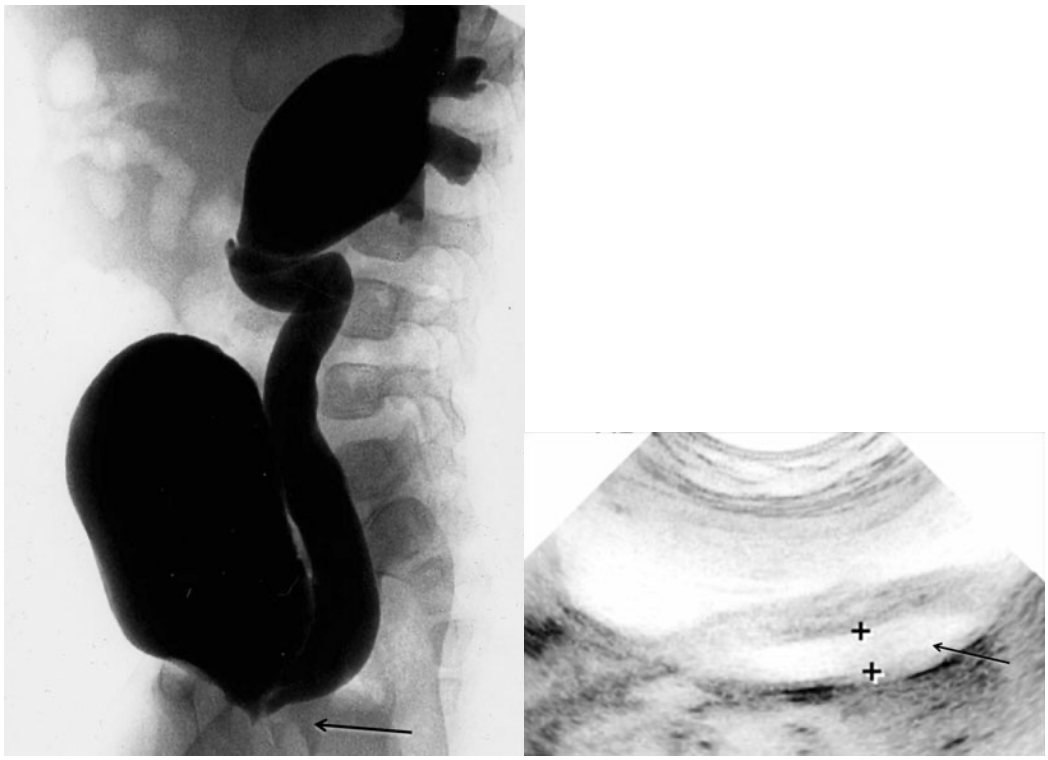
pattern of micturition, a clinical picture that distinguishes it from all other causes of incontinence (Thomas et al. 2008).

In cases where an ectopic ureter is suspected, radiological evaluation begins with a renal and bladder US. The greater the degree of ureteral dila- tation, the easier it is to determine the site of ecto- pic insertion. The location of the ectopic ureter insertion is often not determinable. The dilated dis- tal ureter can be visualized behind the bladder (Fig. 18.15). Often hydronephrosis of the upper

Fig. 18.15 Ectopic ureter. US scan of the pelvis shows a dilated distal ureter (*arrow*) behind the bladder, suggest- ing an ectopic insertion; the exact location is not easy to visualize and often not determinable on US examination

renal pole is visible. If the ectopic ureteral orifice is in the external urethral sphincter, a unique situation can occur where an obstructed, refluxing megaure- ter is found. In some girls with infrasphincteric ectopia, the upper renal pole is not hydronephrotic but is small and dysplastic (and can be ectopic) and consequently difficult or impossible to detect by ultrasonography (cryptic duplication). When visi- ble the upper pole can have hyperechoic paren- chyma and thinning as a result of the hydronephrosis and high association with renal dysplasia.

VCUG in suprasphincteric ectopia demon- strates reflux to the upper pole only during voiding, when the external sphincter is relaxed. In patients with an ectopic ureter at the bladder neck (Fig. 18.16), it is not unusual that the catheter inserted in the urethra for the VCUG instead of entering the bladder goes straight into the ectopic ureter. The injection of contrast actually results in a retrograde pyelography of the upper pole moiety.



VCUG: ectopic ureter associated with high- grade VUR. Left ectopic ureteral insertion (*arrow*) at the bladder neck with retrograde filling of contrast medium until the pelvis and calyces that appear severely dilated

Fig. 18.16

MRI is excellent in showing the location of the orifice (Krishnan and Baskin 2005; Wille et al. 2003), especially with sagittal views. No contrast is needed and ionizing radiation is absent. T2-weighted images are excellent at delineating fluid-filled structures such as an ecto- pic ureter. In most cases, utilizing MRI for young children has required sedation in some form for optimal imaging. This fact, along with the increased cost, has limited MRI to equivocal cases in need of definition of anatomy rather than as a screening modality.

DMSA radionuclide scans remain the gold standard to determine renal function and to guide surgical plan.

Treatment of suprasphincteric ectopic ureters requires reimplantation of the ureters into the bladder, plus augmentation cystoplasty when the bladder is small. The only treatment for infra- sphincteric ectopic ureters is the exeresis of the related small dysplastic kidney (Thomas et al. 2008).

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