4 Diseases of the Ureter

4.1 Congenital Diseases

4.1.1 Ureteropelvic Junction Obstruction

Definition

The ureteropelvic junction (UPJ) is between the proximal ureter and renal pelvis. Congenital malformations with obstruction of the ureteropelvic junction are a common cause of hydronephrosis (Tan et al. 2004). Guidelines of the EAU (Radmayr et al. 2022).

Epidemiology

Neonatal period: Ureteropelvic obstruction causes up to 48 % of fetal hydronephrosis.

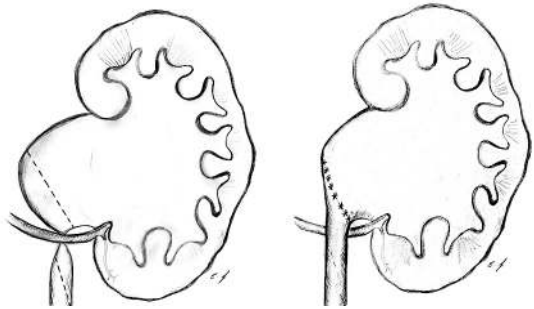


Figure 4.1: Schematic drawing of a UPJ obstruction caused by a lower pole renal artery (left). After surgical correction (technique by Anderson-Hynes), the vessel is transposed dorsally of the ureteral anastomosis (right).

Postnatal period: Incidence 1:1500. 60 % left, 10–40 % bilateral manifestation. Male to female = 2:1 in the neonatal age group.

Etiology

Ureteral causes: Insufficient tubularization of the proximal ureteral segment in the 10th to 12th week of pregnancy obstructs the UPJ, probably caused by a lack of innervation or imbalance of growth factors. This leads to a disturbed structure of the smooth muscles within the ureteral wall and insufficient peristalsis of the proximal ureteral segment. Rare intrinsic causes are ureteral valves or polyps.

Extrinsic causes: Compression of the ureter by a lower pole renal artery (15–40 %) or fibrotic strands [fig. 4.1 and fig. 4.2]. Commonly associated malformations are pelvic kidney, horseshoe kidney, renal malrotation, or duplex kidney.

Secondary UPJ obstruction: In 10 % of patients with severe vesicoureteral reflux, UPJ obstruction exists (or develops).

Pathophysiology

Increased resistance of the urinary flow: The obstruction leads to a chronic increase of pressure in the renal pelvis, which leads to smooth muscle hypertrophy, dilatation of the renal pelvis, and increased renal intratubular pressure.

Loss of renal function: The increased renal pressure decreases renal blood flow (RBF) and glomerular filtration rate (GFR). The reduced urine production normalizes the pressure within the renal pelvis, which remains dilated. An uncorrected UPJ obstruction may lead to functionless hydronephrosis. An activated RAAS system seems to be the central mediator of the pathophysiological effects (reduction of RBF and GFR). The administration of ACE inhibitors has a protective effect on renal function.

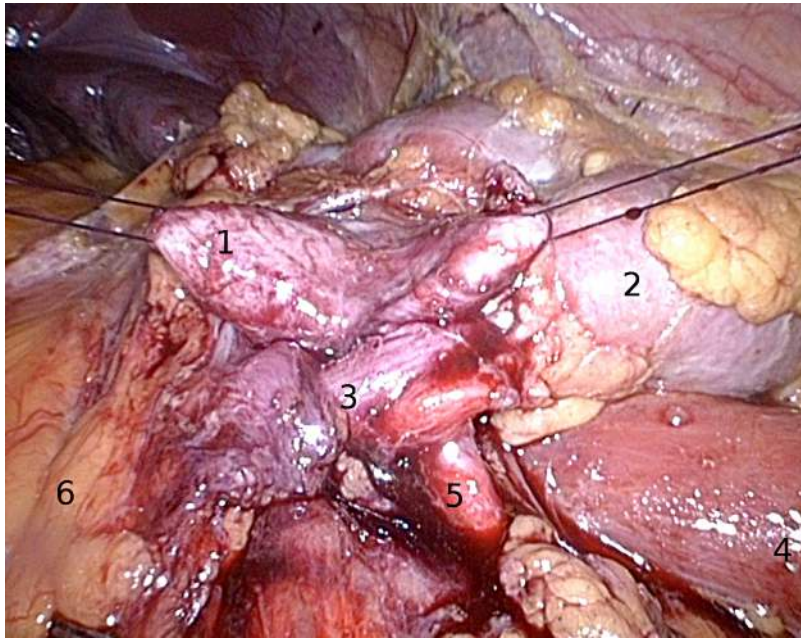


Figure 4.2: Intraoperative laparoscopic findings of a left-sided UPJ obstruction: renal pelvis (1), left kidney (2), crossing lower pole artery (3), psoas muscle (4), ureter (5), descending colon (6). Please see retrograde pyelography fig. 4.4 of the same patient.

Signs and Symptoms

Newborns and infants: Abdominal tumor, failure to thrive, and urosepsis are the classic presentation. Nowadays, many (more) children are diagnosed via prenatal ultrasound imaging, and many of them are asymptomatic.

Children and adults: Flank or upper abdominal pain, often with nausea and vomiting, particularly after fluid intake (Dietl crisis). Further symptoms are pyelonephritis, nephrolithiasis, and hematuria.

Diagnosis

Renal ultrasound: With renal ultrasound, differentiation between physiologic dilatation and significant hydronephrosis is impossible. The anteroposterior diameter of the renal pelvis is used as a parameter for the extent of obstruction of the UPJ obstruction.

With an anteroposterior diameter of the renal pelvis of 12 mm three months after birth, renal scintigraphy should be initiated. The ap diameter three months after birth is also a risk factor for future necessary operation: 75 % of children with an anteroposterior diameter of the renal pelvis of more than 21 mm will need an operation. Doppler ultrasound may reveal an increased resistive index (RI, usually $< 0{,}7$) in the affected kidney.

Intravenous urography: Dilatation of the renal pelvis and calyceal system with a stenotic ureteropelvic segment. Delayed visualization and drainage of contrast medium from the renal pelvis. Intravenous urography is often replaced by better alternatives (e. g. MR urography or CT).

Renal scintigraphy: After administration of a radionuclide (MAG3), renal scintigraphy assesses renal function separately for each kidney and measures the drainage from the renal pelvis to the bladder. If drainage from the renal pelvis is poor, stimulation with furosemide enhances urinary flow. Poor drainage from the renal pelvis and, therefore, significant hydronephrosis means a tracer washout of less than 50 % within 20 minutes after furosemide stimulation. A tracer washout half-time of fewer than 10 minutes after furosemide stimulation is considered normal. A half time between 10–20 min is considered uncertain to relevant obstruction. Indications for surgery are relevant obstruction, especially with symptoms or decreasing fractional renal function (< 40 %).

Voiding cystourethrogram: Vesicoureteral reflux is associated in 10 % of cases with UPJ obstruction. A voiding cystourethrogram is indicated for dilated ureters.

Retrograde pyelography: And ureteroscopy are indicated for unclear proximal ureteral stenoses, before ureteral stenting in patients with symptomatic obstruction, and before surgery to confirm the diagnosis depending on available imaging [Abb. 4.3 und 4.4].

CT or MRI angiography and urography: Imaging technique for differential diagnosis of hydronephrosis, identification of a lower pole renal artery, and to identify associated kidney malformations.

Figure 4.3: Retrograde pyelography of a duplex renal system with UPJ obstruction due to a lower pole renal artery. With kind permission, Dr. R. Gumpinger, Kempten.

Whitaker test: Invasive measurement of the renal pelvis pressure during infusion through a percutaneous nephrostomy with 10 ml/min. The bladder pressure is subtracted from the renal pelvis pressure; the difference should be lower than 20 cm water column. Due to the invasiveness of the test, it is seldom used in practice.

Figure 4.4: Retrograde pyelography of a UPJ obstruction of the left kidney with a lower pole renal artery; see also fig. 4.2 of the same patient.

Differential Diagnosis

See section hydronephrosis on p. 484, in children especially megacalicosis, higher grade VUR and megaureter

Therapy

Regular monitoring of renal function is sufficient in the absence of symptoms and even fractional renal function.

Indications for invasive therapy:

- Significant obstruction in renal scintigraphy
- Decreasing fractional renal function ($< 40\,\%$)
- Equivocal obstruction in renal scintigraphy with recurrent flank pain, pyelonephritis, or nephrolithiasis.

Pyeloplasty: The most popular pyeloplasty is the dismembered Anderson-Hynes technique, which can be done via an open lumbar, open subcostal, laparoscopic, retroperitoneoscopic or robotic-assisted approach (Munver et al. 2004). The Anderson-Hynes technique consists of the transection and reduction of the renal pelvis, excision of the narrow ureteropelvic junction, spatulation of the proximal ureter, and reanastomosis of the proximal ureter

to the renal pelvis (end-to-side). If a lower pole renal vessel is present, the ureter is repositioned after transection, and anastomosis is done anterior to the vessel. For surgical technique and complications see p. 1151.

Endopyelotomy: After retrograde access (ureteroscopy) or antegrade access to the ureteropelvic junction, an endoscopic incision of the narrow segment (posterior position to avoid any lower pole vessel) is done with a cold knife, laser fiber or special cutting devices. Endopyelotomy is an option if a lower pole renal vessel has been excluded and after failed pyeloplasty. Larger series have reported success rates around 75–90 %. Endopyelotomy is seldom used in children.

Nephrectomy: For UPJ-obstruction with a low fractional renal function ($<$ 10–20 % of total renal function). Nephrectomy is usually possible with a minimally invasive technique (laparoscopic or retroperitoneoscopic).

4.1.2 Ureteral Duplication

Definition

Ureteral duplication or duplex kidney is an anatomic variant of the ureter and pyelocalyceal system with a large variability [fig. 4.5]. EAU Guideline: (Radmayr et al. 2022).

Complete ureteral duplication: Duplex kidney with complete duplicated ureters, renal pelvis, and two orifices of the ureters.

Ureter fissus or bifid ureter: Duplex kidney with incomplete ureteral duplication and a common orifice of the ureter.

Bifid renal pelvis: Duplex kidney with duplication of the renal pelvis and a common ureter.

Inverted Y ureteral duplication: Two separate ureteral buds merge on the way to the metanephric blastema, resulting in a common renal unit. Very rare.

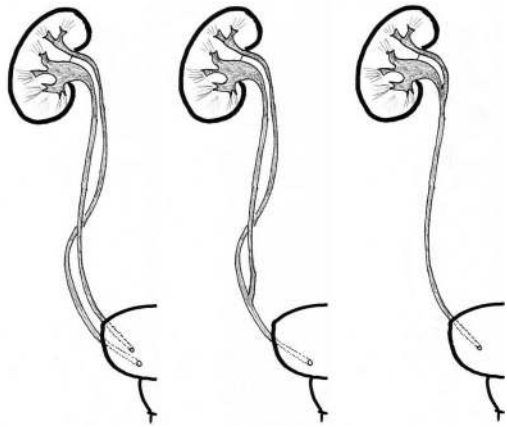


Figure 4.5: Different variants of ureteral duplication: duplicated ureter (left), ureter fissus (center) and bifid renal pelvis (right).

Ureteral triplication: Three independent ureteral orifices with three complete ureters or incomplete ureteral triplication with two orifices of the ureters (see ureter fissus).

Epidemiology

Prevalence 1:150 (0,7 %).

Etiology

Two ureteric buds: On the mesonephric duct (Wolffian duct) result in two separate ureters and a duplex kidney in a common renal capsula. The ureteric buds rotate by 180 degrees while being incorporated into the urogenital sinus: the latero-cranial orifice drains the lower pole of the kidney, and the medio-caudal orifice drains the upper pole of the kidney. This relation is called the *Weigert-Meyer rule*.

Division of the ureteric bud: Incomplete ureteral duplications (bifid ureter, bifid pelvis) are caused by a division of a single ureteric bud on its way to the metanephric blastema.

Pyelocalyceal system of the duplex kidney: Compared to a normal kidney with 8–9 calyces, the duplex kidney consists of 11–12 calyces. The upper pole system is smaller, with an average of 3–4 calyces.

Associated malformations: Vesicoureteral reflux to the lower pole system (40 %), ureteropelvic junction obstruction of the lower pole system, ectopic ureteral orifice of the upper pole system (with or without obstruction), ureterocele of the upper pole ureter, hypoplastic and dysplastic renal parenchyma most common of the upper pole system.

Pathophysiology: Duplication of the ureter is associated with vesicoureteral reflux or hydronephrosis, leading to a susceptibility to urinary tract infections or nephrolithiasis.

Signs and Symptoms

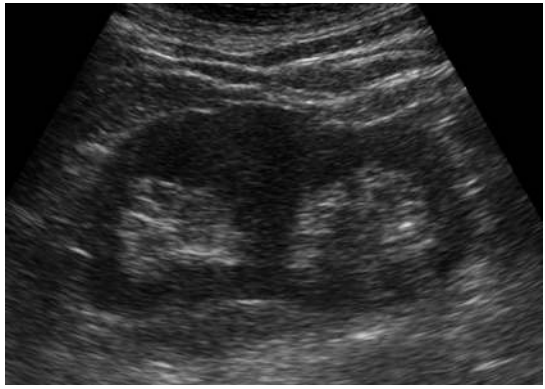
Associated malformations may lead to urinary tract infections, nephrolithiasis, fever, or abdominal tumor. Without malformations, a duplex kidney usually presents without symptoms.

Diagnosis

Ultrasound imaging: Without associated malformations, a normal finding in renal ultrasound is often diagnosed. A duplex kidney may be recognized if two separated renal pelves are separated by renal parenchyma [fig. 4.6(a)]. It is easier to detect a duplex kidney with ureterocele or hydronephrosis, most often the upper pole system is affected [fig. 4.6(b)]..

Intravenous urography: A duplex kidney is often an incidental finding during the diagnostic workup for other diseases [fig. 4.7]. Due to poor renal function, urography often lacks to contrast the upper part of the duplex kidney. Hints for a non-contrasting upper portion are obtained from the small number of calyces shown, inferior-lateral displacement of the lower pole (dropping lily sign) and the greater distance of the renal system to the spine [fig. **??**]. Sometimes, a ureterocele is delineated in the bladder [fig. 4.16].

Voiding cystourethrogram: Indicated for recurrent urinary tract infections or dilatation of the pyelocalyceal system to diagnose vesicoureteral reflux. A possible ureterocele is detectable in the early filling phase.



(a) Ultrasound imaging of a duplex kidney.

(b) Ultrasound imaging of a duplex kidney with hydronephrosis of the upper pole system due to a ureteral stone.

Figure 4.6: Ultrasound imaging of ureteral duplication.

Endoscopy: Cystoscopy, retrograde pyelography and possibly ureteroscopy depending on complaints and other findings.

Renal scintigraphy: Enables the separate determination of the renal function of the upper and lower part of the duplex kidney for further treatment planning.

Figure 4.7: Intravenous urography: left-sided duplex kidney with complete ureteral duplication. Both parts of the left-sided duplex kidney present without hydronephrosis. The urinary tract did not cause the complaints of the patient. Note the abdominal mass due to an abdominal aortic aneurysm, which displaces the left psoas shadow (*). With kind permission, Dr. G. Antes, Kempten.

Therapy

Duplex kidney or ureteral duplication is a normal variant and, thus, not subject to treatment. The associated malformations and symptoms guide the therapy (vesicoureteral reflux, ureterocele, ectopic ureter, recurrent urinary tract infections, nephrolithiasis, or urinary tract obstruction).

4.1.3 Ectopic Ureter

Definition

An ectopic ureter is a ureter with an abnormally located ureteral orifice. Instead of draining into the bladder, the ureteral orifice drains in the urethra,

Figure 4.8: CT of a left-sided ureteral duplication: visualization of the anatomy with 3D volume rendering (VR). With kind permission, Dr. H. Ruhnke, Radiologie im Zentrum, Augsburg.

vagina or mesonephric duct structures (ductus deferens or seminal vesicles).

EAU guidelines (Radmayr et al. 2022).

Epidemiology

1:2000, more girls than boys.

Etiology

See etiology of ureteral duplication p. 438.

Figure 4.9: Ectopic ureter (marked by an ureteral catheter) draining into the vestibulum vaginae. With kind permission, Prof. Dr. R. Harzmann, Augsburg.

Pathology

80 % of ectopic ureters are associated with a ureteral duplication, especially in girls. In boys, the ectopic ureter may also drain a single renal system. In girls, the ectopic ureter drains into the urethra (35 %), vulval vestibule (34 %), vagina (25 %), or uterus (5 %). In boys, the ectopic ureter drains into the prostatic urethra (47 %), seminal vesicles (33 %), prostatic utricle (10 %), or vas deferens (10 %).

The further the distance of the ureteral opening from its normal position, the higher the likelihood of renal malformation (renal dysplasia, renal hypoplasia) and dysfunction. If the ectopic ureter is associated with a duplex system, the ectopic ureter drains the upper pole of the kidney.

Signs and Symptoms

Recurrent urinary tract infections, flank pain, fever, and arterial hypertension. Additional symptoms depend on the gender and location of the ureteral orifice:

- Girls: urinary incontinence (day and night, but sometimes also intermittent), vaginal discharge.
- Boys: LUTS, epididymitis.

Diagnosis

Pelvic examination: Indicated in girls with urinary incontinence; sometimes, the orifice of the ureter can be identified in the vagina or vulval vestibule [fig. 4.9].

Cystoscopy and retrograde pyelography: To search for ureteral orifices.

Ultrasound imaging: Ureteral ectopia may cause hydronephrosis. If a duplex kidney is present, urinary obstruction of the upper kidney portion may be visible. A dilated ureter may be detectable behind the bladder.

Intravenous urography: Increasingly replaced by MR urography, or, in adults, by CT. Due to poor renal function, urography often lacks to contrast the upper part of the duplex kidney. Hints for a non-contrasting upper portion are obtained from the small number of calyces shown, inferior-

Figure 4.10: Ureterectopy with a left-sided ureteral duplication: the ureter of the upper pole system led into the prostatic urethra. The massive hydronephrosis of the upper pole system (indicated with (*) and an inserted kidney fistula) led to a caudal displacement of the lower pole calyces (dropping lily sign in intravenous urography). With kind permission, Dr. N. Dreger und Prof. Dr. S. Roth, Wuppertal.

lateral displacement of the lower pole (dropping lily sign) and the greater distance of the renal system to the spine [fig. **??**]. Late images after 1–3 hours may show contrast in the upper renal portion.

Voiding cystourethrography: VCUG may detect reflux into the lower renal pole if a duplex kidney is present.

Renal scintigraphy: Indicated to determine the renal function on the side of the ectopic ureter. If a duplex system is present, the renal function of the upper and lower pole must be analyzed separately.

MRI Urography or CT: MRI urography is the most accurate imaging tool and is indicated for imaging in children, especially if unclear findings in previous investigations are present and an ectopic ureter is suspected. CT is an imaging alternative in adults that is more sensitive than intravenous urography.

Therapy

In principle, treatment is only necessary for patients with symptoms, relevant vesicoureteral reflux, or significant obstruction.

Ectopic ureter with sufficient renal function:

- Ureteropyelostomy or ureteroureterostomy for a duplex kidney, usually the ectopic ureter of the upper pole is anastomosed with the lower pole ureter (end-to-side). If significant reflux into the ectopic ureter is present, resection of distal ectopic ureter is necessary.
- Common sheath ureteroneocystostomy (UCN) for a duplex kidney with closely spaced ureteral orifices and vesicoureteral reflux.
- Ureteroneocystostomy (UCN), if the ureter drains a single renal system.

Ectopic ureter with a nonfunctioning kidney: Standard treatment is heminephrectomy for a double system and nephrectomy for a single system; surgery is possible with a laparoscopic technique. In the case of reflux in the ectopic ureter, an additional distal ureterectomy is necessary. In the case of reflux in the lower part of a duplex kidney, ureteral reimplantation may be necessary.

Heminephrectomy is a complex operation with a small risk of organ loss due to bleeding or urinoma. Alternatively, although the partial function of the upper portion is poor, the ectopic ureter can be anastomosed (ureteroureterostomy or ureteropyelostomy) to the lower pole ureter. The procedure has fewer complications and the prognosis regarding long-term complications (UTI, hypertension) is equal (Kawal et al. 2019).

4.1.4 Ureterocele

Definition

The ureterocele is a cystic dilatation of the distal, intravesical ureter [fig. 4.12]. In 80 %, the ureterocele drains the upper pole of a duplex kidney [fig. 4.13]. A ureterocele causes a defect in the trigonum and predisposes the patient to vesicoureteral reflux into the ureter of the lower pole of a duplex kidney (A. Shokeir et al. 2002). EAU guideline: (Radmayr et al. 2022). The following classifications exist:

By Ericsson: Separates *intravesical (simple) ureterocele* from textitectopic ureteroceles, which extend to the bladder neck, and urethra.

By Stephens: *intravesical ureteroceles:* Which may be stenotic or non-obstructed.

Extravesical ureteroceles: Which may be sphincteric (non-obstructive ostium distal of the bladder neck), sphincterostenotic (stenotic ostium distal of the bladder neck), caecoureterocele (intravesical ostium, but the ureterocele extends down into the urethra and causes obstruction) or blind (no contact to the kidney).

Epidemiology

Prevalence 1:5000, more often in females (4:1).

Etiology

Different theories exist, but no theory can explain all forms of ureteroceles:

Chwalla membrane: Separates the urogenital sinus from the ureteral bud. A delayed and incomplete reabsorption of Chwalla membrane is postulated, causing a stenotic ostium which also leads to the cystic dilatation of the intravesical ureter. The theory does not explain the different sizes and forms of the ureterocele.

Disturbed fusion of Wolffian duct with the urogenital sinus: The theory aligns the ureterocele with the ectopic ureter: the more lateral the

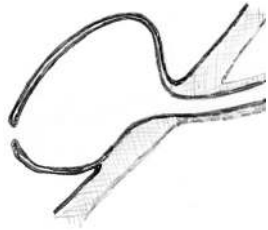


Figure 4.11: Cross-section through a ureterocele: cystic dilatation of the distal intravesical ureter.

Figure 4.12: Ureterocele in cystoscopy: bulging of the bladder mucosa with a stenotic ostium.

ureter bud, the later and more disturbed the fusion into the urogenital sinus.

Acquired ureteroceles: Ostium stenosis (caused by inflammation, ureteral stone, or trauma) leads to a cystic dilatation of the distal ureter.

Signs and Symptoms

Urinary tract infection, urosepsis, nephrolithiasis, ureterocele stone [fig. 2.29 on p. 190, fig. 4.18 on p. 452], abdominal tumor or failure to thrive in infants, prolapse from the urethra, urinary retention, hematuria, and incontinence.

Diagnosis

The diagnostic workup is only necessary for patients with symptoms, suspected vesicoureteral reflux, or obstruction.

Ultrasound imaging: Ureteroceles present as thin-walled cysts in the bladder [fig. 4.14]. For imaging, a medium-filled bladder is preferable. A ureterocele might cause urinary obstruction of the contralateral kidney.

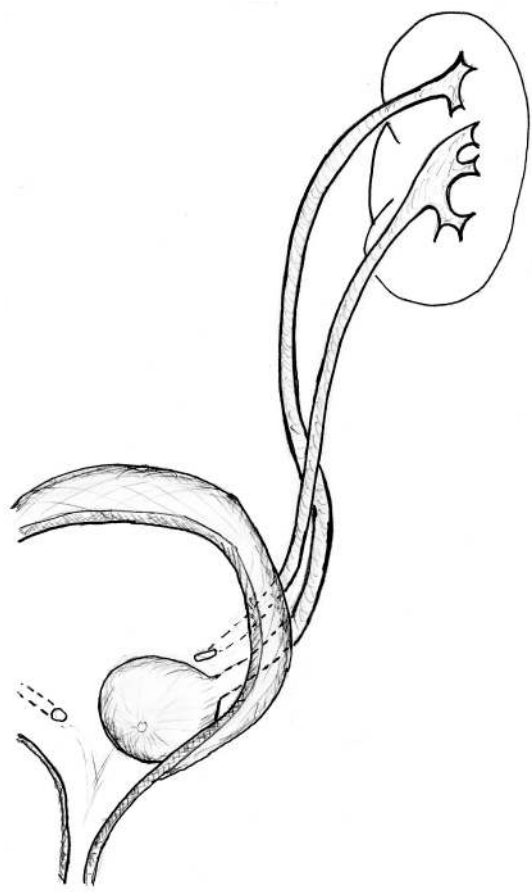


Figure 4.13: Ureteroceles are associated with ureteral duplication (in 80 %); the ureterocele drains the upper pole of the duplex kidney.

Ureteral duplication is often present, and a stenotic ostium of the ureterocele may cause hydronephrosis of the upper pole system.

Intravenous urography: Often missing contrasting of the upper pole of the duplex kidney due to poor function; however, the upper part can contrast in late KUB. Hints for a non-contrasting upper pole are reduced chalices and a greater distance between the kidney system and the spine. The lower pole ureter is serpentine (around the upper pole ureter). Sometimes the ureterocele can be visualized in the bladder [fig. 4.15 and 4.16(b)]. The

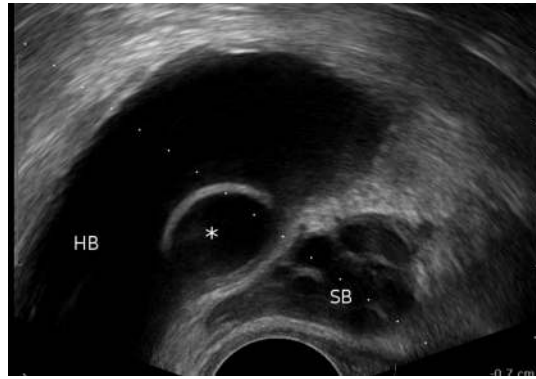


Figure 4.14: Transrectal ultrasound imaging: ureterocele (*) in the filled bladder (HB). In addition, an enlarged seminal vesicle (SB) is seen. With kind permission, Dr. med. H. Kempter, Augsburg.

cystic dilatation of an uncomplicated intravesical ureterocele is called a „cobra head sign “ [fig. 4.16(a)].

Voiding cystourethrography: Filling defect by the ureterocele shows size and location. Imaging the ureterocele is best during early filling; the ureterocele may collapse in a full bladder and present as a bladder diverticulum. Filling the bladder to full capacity is necessary to demonstrate reflux into the lower pole ureter, which is seen in duplex kidneys in up to 50 %.

Cystoscopy: See fig. 4.12.

Retrograde pyelography: See fig. 4.15 and fig. 4.17.

Renal scintigraphy: Evaluates renal function or the significance of hydronephrosis. If a duplex system is present, the renal function of the upper and lower pole must be analyzed separately.

Therapy

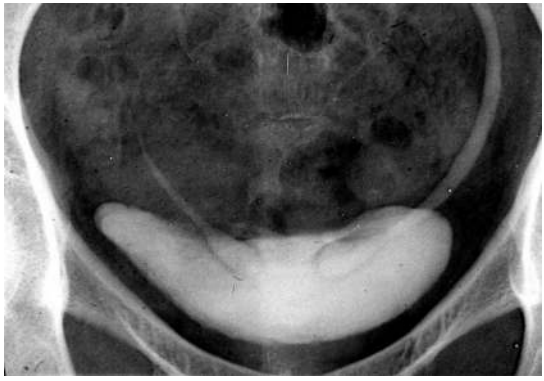
A ureterocele without symptoms does not require therapy. The aim is to treat ureterocele with accompanying problems (prevent vesicoureteral reflux and UTI, relieve obstruction) with a single intervention, if possible.

Transurethral incision: Of the ureterocele is a treatment option with low morbidity [fig. 4.18]. It is often curative for obstructive intravesical

Figure 4.15: Intravenous urography (left): right-sided ureterocele creates a filling defect in the urinary bladder, the associated upper pole chalices are not contrasted. Retrograde pyelography after incision of the ureterocele (right): the upper pole chalices show moderate signs of chronic hydronephrosis. With kind permission, Dr. R. Gumpinger, Kempten.

ureterocele with a single system or with a double system without severe reflux in the lower pole ureter. Extravesical ureteroceles are less likely to be healed by incision, never the less ureterocele incision is often performed as the first treatment attempt. If obstruction or severe postoperative reflux is a problem, ureteropyelostomy or ureteral reimplantation is necessary.

Ureteropyelostomy: treatment option for an obstructive ureterocele (relapse after transurethral incision) of a duplex kidney without relevant reflux in the lower pole ureter. The upper pole ureter is anastomosed to the lower pole ureter (end-to-side). Surgery is possible with a flank incision or laparoscopically. The ureterocele should decompress spontaneously and thus



(a) Bilateral cobra head sign: cystic dilatation of the distal ureters.

(b) Contrasted intravesical ureterocele.

Figure 4.16: Ureterocele in intravenous urography (detailed images of the bladder). With kind permission, Prof. Dr. R. Harzmann, Augsburg.

reduce the reflux in the lower pole ureter. If postoperative vesicoureteral reflux is problematic, ureterocele resection and ureterocystoneostomy of the lower pole ureter are necessary.

Heminephrectomy: Indicated for ureterocele and double system with poor upper pole function and without relevant reflux in the lower pole ureter. Surgery is possible with a flank incision or laparoscopically. The ureterocele should decompress spontaneously and thus reduce the reflux

(a) Cystoscopy: distended edematous ureteral orifice.

(b) Retrograde pyelography: cobra head sign with a filling defect.

Figure 4.17: Small ureterocele with stone.

in the lower part. If postoperative vesicoureteral reflux is problematic, ureterocele resection and ureterocystoneostomy of the lower pole ureter are necessary.

Simultaneous upper and lower urinary tract reconstruction: May be necessary for ectopic ureterocele with severe reflux or obstruction of the contralateral kidney. Heminephrectomy or ureteropyelostomy is done via a flank incision, excision of the ureterocele, and ureteral reimplantation is performed via a separate incision for surgical access to the urinary bladder.

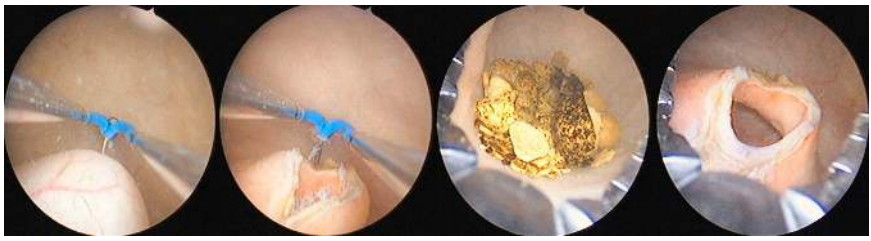


Figure 4.18: Transurethral incision of an intravesical ureterocele to enable treatment of multiple stones in the ureterocele.

4.1.5 Vesicoureteral Reflux

Definition

Vesicoureteral reflux (VUR) is a common congenital or acquired disorder of the vesicoureteral junction with reflux of urine into the upper urinary tract, which can lead to recurrent urinary tract infections, pyelonephritis with scarring, arterial hypertension, and chronic renal insufficiency. Synonyms: reflux uropathy, vesicorenal reflux (Dewan 1999) (Körner et al. 2010). EAU guidelines: (Radmayr et al. 2022).

Epidemiology

The prevalence of VUR in non-symptomatic children is around 1 %. In children with recurrent UTI or lower urinary tract dysfunction, the prevalence of VUR is much higher (up to 70 % in children age < 1, declining to 15 % with age 12). Male to female ratio = 1:4. VUR can be an inherited condition: siblings and children of affected patients have an increased risk (up to 30 % in twins).

Etiology

Please see section **??** on p. **??** for the anatomy of the ureterovesical junction.

Primary reflux: A multifactorial congenital defect leads to premature ureter budding and fusion of the Wolffian duct with the urogenital sinus, overrotation of the ureter bud with lateralized ostia, and impaired trigonal musculature development.

Ureteral duplication: Predisposes the ureter of the lower renal pole to vesicoureteral reflux since the intravesical section is disturbed by an often-existing ureterocele of the upper pole ureter.

Secondary reflux: *lower urinary tract dysfunction* with high micturition pressures (dysfunctional micturition, urethral valves, neurogenic bladder disorders such as spina bifida) are a strong risk factor for high-grade vesicoureteral reflux with renal scarring.

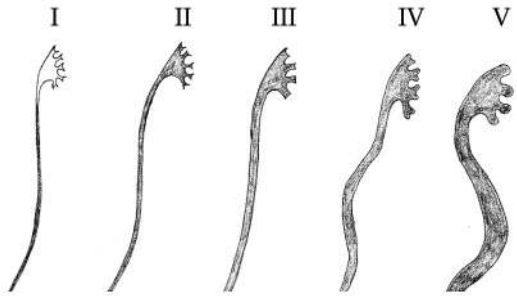


Figure 4.19: Classification of vesicoureteral reflux using the VCUG imaging results (Wingen et al. 1999):
Grade I: Reflux into a non-dilated ureter.
Grade II: Reflux into the renal pelvis without dilatation.
Grade III: Reflux into the renal pelvis, moderate dilatation of the upper tract, and minimal blunting of the calyces.
Grade IV: Reflux with significant dilatation of the upper tract, moderate ureteral kinking, and blunting of the calyces with preserved papillary impressions.
Grade V: Reflux with gross dilatation of the upper tract, ureteral tortuosity, and loss of papillary impressions.

Decompensation of the ureterovesical junction: Borderline functioning ureterovesical transitions can decompensate and cause reflux with ascending infection: e. g., UTI with bladder wall edema or during pregnancy.

Prune-Belly syndrome: Leads to disturbed development of the abdominal wall muscles and the smooth muscles of the ureters and bladder with severe vesicoureteral reflux [p. **??**].

Iatrogenic: Incisions of the bladder trigonum may cause vesicoureteral reflux: prostatectomy, trigonal TURB, or resection of a ureterocele.

Classification

Earlier classifications differentiated between low-pressure reflux (during bladder filling) and high-pressure reflux (caused by micturition). The current classification was proposed by the Int. Reflux Study Group and depicts the possible results of the VCUG into five grades, see fig. 4.19.

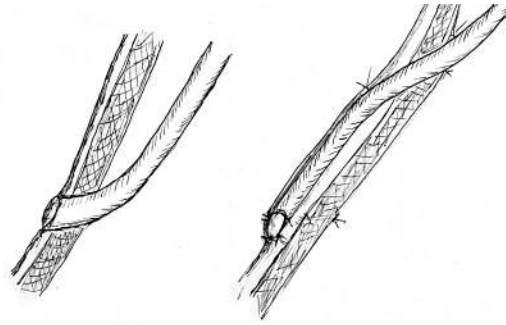


Figure 4.20: Schematic drawing of an antirefluxive ureteral reimplantation: One cause of reflux is insufficient length of the intramural ureter (right). After antirefluxive reimplantation, the ratio between ureter diameter and tunnel length should be at least 1:5 (left).

Pathophysiology

Insufficient vesicoureteral junction: VUR is caused by the reduced length of the intramural ureter (reduced passive reflux protection, see fig. 4.20) and a weak bladder wall at the trigonum (missing active reflux protection by muscle contraction).

Spontaneous healing: Due to the growth of the ureter and the increase in bladder capacity, vesicoureteral reflux improves during childhood (over the years) without therapy. Spontaneous healing is common in mild reflux and normal positioned ureteric orifices.

Recurrent urinary tract infections: VUR leads to pendulum urine, which becomes infected. Pyelorenal reflux causes pyelonephritis, particularly in young children (first years of life) pyelonephritis causes renal scarring.

Flat or concave papillae have an opening of the collecting tubes at a right angle, which is a risk factor for intrarenal reflux. Convex papillae have an oblique slit-like opening of the collecting tubes with less intrarenal reflux. The papillary anatomy predisposes to pyelorenal reflux at the poles.

Reflux nephropathy: Renal scarring causes renal hypertension, proteinuria, focal segmental glomerular sclerosis, and growth delay and may lead to terminal renal insufficiency (see chronic pyelonephritis on p. 302). Reflux

nephropathy is responsible for 10–20 % of all children with chronic renal failure.

Signs and Symptoms

Acute pyelonephritis: Fever, chills, flank pain, vomiting, dysuria and pollakisuria. Sometimes without symptoms (only pyuria and bacteriuria).

Chronic pyelonephritis: Arterial hypertension, growth delay, symptoms of uremia.

Diagnosis

Ultrasound imaging: Record the width of the renal parenchyma, scars of the parenchyma (echogenic areas), the size of both kidneys, the diameter of the renal pelvis and ureter, thickness of the bladder wall, search for ureterocele or standard variants of the renal anatomy and measure postvoid residual volume. An increased resistance index in Doppler sonography indicates kidney damage with scarring (RI usually < 0.7 [p. 169]). Ultrasound imaging cannot exclude vesicoureteral reflux.

Sonography with contrast medium: After the instillation of air-containing contrast medium into the bladder, reflux can be visualized during or after micturition.

Voiding cystourethrography: VCUG is the most important imaging tool for detecting vesicourethral reflux [fig. 4.21]. VCUG can be combined with urodynamics if lower urinary tract dysfunction is suspected [fig. 4.22]. VCUG should only be done after sufficient treatment of urinary tract infection (at least 7–10 days after pyelonephritis); otherwise, there is a risk of false positive results. Imaging with VCUG classifies the severity of VUR; see the fig. 4.19. Two diagnostic problems of VCUG exist: the detection of reflux, which is insignificant for the patient, and false-negative imaging in patients with renal scarring.

Ureteral diameter ratio (UDR): The diameter of the distal ureter is divided by the distance between the cranial endplate L3 and caudal endplate L1. This ratio compensates for varying patient size and magnification factors

Figure 4.21: Vesicoureteral reflux (Grade V) in VCUG: in addition, a small bladder capacity with multiple pseudodiverticles is depicted as a sign of a neurogenic bladder disorder. With kind permission, Dr. G. Antes, Kempten.

on imaging and correlates better with prognosis (spontaneous healing, recurrent UTI, and renal scarring) than the classic classification. Low-grade reflux corresponds to a UDR less than 0.2; any increase in UDR significantly worsens prognosis, and a UDR greater than 0.35 makes spontaneous healing from reflux unlikely (Arlen et al. 2017).

Renal scintigraphy: In addition to assessing kidney function, static kidney scintigraphy with ^{99m}Tc-DMSA can reliably diagnose renal scarring by urinary tract infections. If additional obstruction is suspected, a functional scintigraphy of the kidneys is performed with ^{99m}Tc-MAG3.

Figure 4.22: Urodynamics and VCUG in a girl with vesicoureteral reflux: third to fourth degree reflux due to detrusor-sphincter dyscoordination, the ballooning of the proximal urethra during micturition is visible. With kind permission, Dr. U. Löffler, Augsburg.

The diagnostic advantage of DMSA scintigraphy is the reliable detection of patients at risk of upper renal tract damage by vesicoureteral reflux. Some authors recommend early use of DMSA scintigraphy and do not recommend VCUG if renal scans are inconspicuous.

Diagnosis of neonatal ectasia of the renal pelvis: Perform VCUG or DMSA scintigraphy if there are signs of severe VUR (renal scarring) with ultrasound imaging or after a febrile Urinary tract infections. Whether a

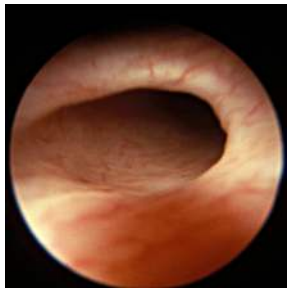


Figure 4.23: Ureteral orifice with reflux (golf hole) in cystoscopy. With kind permission, Prof. Dr. R. Harzmann, Augsburg.

VCUG, DMSA kidney scintigraphy, or both examinations are necessary is controversial.

Diagnostic workup after pyelonephritis: Perform ultrasound imaging of the kidney, ureter and bladder for the first febrile Urinary tract infections. In the case of an unremarkable examination, further workup is unnecessary. Perform VCUG or DMSA scintigraphy after recurrent pyelonephritis or if sonographic abnormalities (e.g., renal scarring) are found (AAP-Guidelines 2011). Whether a VCUG, DMSA kidney scintigraphy, or both examinations are necessary is controversial.

Cystoscopy: The shape and location of the ureteric orifices can indicate vesicoureteral reflux (horseshoe or golf hole shape) and prognosis: the higher the grading, the greater the ureterotrigonal insufficiency and the worse the spontaneous healing.

- Grade 0 (Normal): like a cone or volcano with a small opening
- Grade 1 (Stadium): oval form with a circular wall
- Grade 2 (horseshoe): oval form, no medial wall
- Grade 3 (golf hole): open ostium without elevation [Abb. 4.23].

PIC cystography: uses a cystoscope to instill the contrast medium near the ostium (PIC, positioning the instillation of contrast). PIC cystography is more sensitive than conventional VCUG. The indication for PIC cystography is children with recurrent febrile urinary tract infections and no reflux

in normal VCUG (Rubenstein et al. 2003). Endoscopic treatment can be done in the same session if there is evidence of reflux.

Urodynamics: Indicated for suspected elevated detrusor pressure or lower urinary tract dysfunction [fig. 4.22].

Intravenous urography: Rarely indicated. The following radiological signs are suspicious for vesicoureteral reflux: dilated ureter and pyelocalyceal system, signs of healed pyelonephritis (blunted calyces, thin cortex), ureteral duplication, and ectopic ureter. A normal IVU does not rule out reflux.

Watchful Waiting and Conservative Therapy

The therapeutic goal in treating VUR is to maintain and secure renal function by avoiding pyelonephritis. At least 50 % of the children with primary reflux can be treated by watchful waiting, as trigonal insufficiency is prone to spontaneous healing, and recurrent infections are rare without risk factors. Children with recurrent urinary tract infections can remain infection-free with low-dose long-term antibiotics, reducing the risk of further renal scarring.

Indications: Patients with vesicoureteral reflux up to grade IV, stable kidney function, and without breakthrough febrile urinary tract infections are suitable for conservative therapy.

Bacteriuria: Must be treated with antibiotics, possibly with low-dose long-term antimicrobial prophylaxis with nitrofurantoin, trimethoprim, or an oral cephalosporin. The goal is to avoid febrile UTIs. Dosage: Trimethoprim 2 mg/kg bw/d, Nitrofurantoin 1–2 mg/kg bw/d, Cefaclor 10 mg/kg bw/d.

Double voiding: Recommend regular micturition every three hours. Another voiding, after a few minutes, empties the refluxed urine.

Circumcision: Lowers the rate of UTI in boys.

Anticholinergics: For overactive bladder to increase the functional bladder capacity.

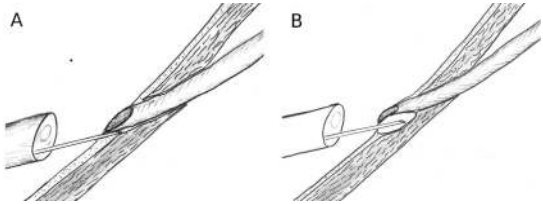


Figure 4.24: Endoscopic treatment of vesicoureteral reflux (STING technique). The implant is submucously injected under the ureteral orifice at the 6 o'clock position until closure (0.5–1.5 ml volume).

Close surveillance: Urine culture is done in cases of suspected UTI and after therapy. Some centers regularly test (every three months) even symptom-free children. VCUG or renal scintigraphy are done depending on the clinical risk every 6–24 months.

Results of controlled trials: Conservative arm of the international reflux study, randomized, n=149, follow-up 10 years of long-term antimicrobial prophylaxis in VUR III–V: 52 % no longer have reflux, 25 % VUR without dilatation, 23 % have VUR with dilatation. Spontaneous healing of reflux is likely for VUR grade $<$ IV, unilateral reflux, children $<$ 5 years (Smellie et al. 2001).

Conservative arm of the Swedish reflux study, randomized, n=203 children between 1–2 years old with dilating reflux grade III–V, follow-up 2 years: 39–47 % spontaneous improvement in VUR. Boys showed a good prognosis even without antibiotic prophylaxis. Girls often had recurrent UTIs, which could be avoided by antibiotic prophylaxis (Brandströmm et al. 2010a) (Holmdahl et al. 2010) (Brandströmm et al. 2010b) (Brandströmm et al. 2010c) (Sillen et al. 2010).

Surgical Therapy

Indications: Several randomized multicenter studies have established successful conservative therapy of VUR (see above). Surgical therapy is only indicated for severe reflux grade V, recurrent pyelonephritis, deterioration of kidney function, or if long-term antibiotic prophylaxis is not accepted.

Endoscopic treatment: Endoscopic injection of bulking agent to close the refluxive orifice is the least invasive surgical treatment option. The implant is injected in the submucosa layer; several injection techniques exist. The STING technique injects the bulking agent at the 6 o'clock position of the orifice until the implant closes the opening [fig. 4.24]. The HIT technique uses two injection positions 12 and 6 o'clock. After the injection, the success is checked by a cystography; further injections are possible if necessary. The HIT technique has a higher success rate controlled by cystography, but no long-term clinical data exists.

Compared to open surgery, retrospective trials of endoscopic treatment report almost comparable results (90 % healing after two injections). However, there are no direct randomized trials. Since endoscopic treatment offers minimally invasive treatment and does not preclude open surgical treatment options in the event of failure, endoscopic therapy is a first-choice option for uncomplicated anatomy.

Ureteral reimplantation: either extravesical (Lich-Gregoir) or transvesical techniques (Leadbetter-Politano or Cohen). The affected kidney should have sufficient renal function (over 10 % split function in renal scintigraphy). Renal function will remain stable or even improve in the long term after reimplantation. For surgical techniques and complications, see p. 1161.

Extravesical Lich-Gregoir technique: extravesical approach to the distal ureter, the incision of the bladder wall creates an extravesical tunnel without opening the mucosa; the ureter is placed in the tunnel, and the bladder wall is closed again. Possible complications are pelvic nerve injury with urinary retention after bilateral surgery.

Transvesical Leadbetter-Politano technique: a transvesical approach to the bladder trigone, a circumferential incision around the orifice, and mobilization of the ureter is done. A submucosal tunnel is created from the old ostium to the new hiatus 3 cm cranially. The ureter is passed through the new hiatus in the bladder and through the new tunnel to the old orifice, where the ureter is implanted with several sutures.

Transvesical Cohen technique: a transvesical approach to the bladder trigone, a circumferential incision around the orifice, and mobilization of the ureter is done. A submucosal tunnel is created in the contralateral direction just above the contralateral orifice. The ureter is pulled into the bladder, passed through the new tunnel, and implanted with several sutures.

Further surgical options: The following procedures are indicated depending on the renal function and associated malformations:

Heminephroureterectomy: For ureteral duplication with a refluxive upper pole ureter and missing renal function of the upper pole system.

Uretero-ureterostomy: For ureteral duplication with a refluxive upper pole ureter and significant renal function of the upper pole system: anastomosis of the caudal end of the refluxing ureter end-to-side to the competent ureter and resection of the refluxive orifice.

Nephroureterectomy: (laparoscopic) nephroureterectomy is an option under 10 % renal split function to avoid recurrent urinary tract infections. Some authors question the need for ureter resection.

Urinary diversion: For severe disorders affecting bladder capacity and function (e.g., spina bifida).

Prognosis

Spontaneous healing: The following factors support spontaneous healing of vesicoureteral reflux or a benign course: low-grade reflux, missing dilatation of the distal ureter, male sex, unilateral reflux, no disorders of urinary bladder or bowel function, no renal scarring, and high urinary bladder volume at the onset of reflux.

4.1.6 Megaureter

Definition

A megaureter is an anomaly with a dilated ureter of more than 7–8 mm (A. A. Shokeir et al. 2000) (Wilcox et al. 1998) (Radmayr et al. 2022); the further classification is made according to the cause:

Figure 4.25: Primary obstructive megaureter: retrograde pyelography. With kind permission, Dr. R. Gumpinger, Kempten.

Primary obstructive megaureter (POM): Congenital ureteral dilatation caused by an aperistaltic ureteral segment at the ureterovesical junction. An embryological stop of the ureter development causes disorientation of the muscle fibers and impaired urine flow.

Figure 4.26: Bilateral primary obstructive megaureter: intravenous urography. With kind permission, Prof. Dr. R. Harzmann, Augsburg.

Secondary obstructive megaureter: Increased intravesical pressure, increased bladder wall tension, and scarring lead to the obstruction of the ureterovesical junction.

Refluxing megaureter: Vesicoureteral reflux causes dilatation of the ureter [fig. 4.21].

Idiopathic (non-obstructive and nonrefluxing) megaureter: Most newborn megaureters are idiopathic; the cause often remains unclear. Increased urine production, a delay of ureteral maturation, or subclinical obstruction may contribute to the development of a megaureter.

Refluxing and obstructive megaureter: Caused by the combination of distal stenosis and vesicoureteral reflux (rare).

Epidemiology

Megaureters account for 20 % of cases with prenatally diagnosed hydronephrosis.

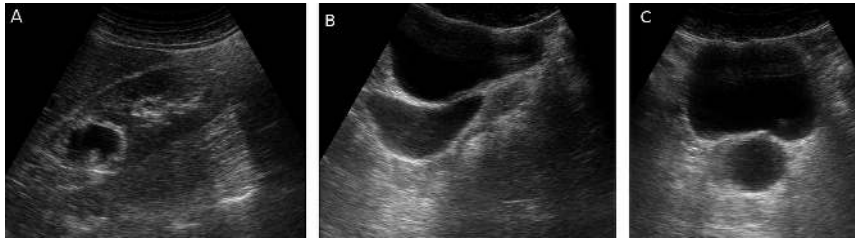


Figure 4.27: Megaureter of the upper pole system in a duplex kidney: ectatic upper pole calyces (A), dilated retrovesical ureter, longitudinal section (B) and horizontal section (C) through the bladder. With kind permission, Dr. M. Fretschner, Augsburg.

Diagnosis

Ultrasound imaging: May differentiate between ureteropelvic junction obstruction and megaureter [fig. 4.27]. The ureter in children is usually less than 5 mm wide.

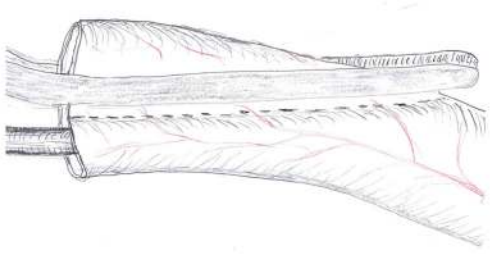
Intravenous urography: For the differential diagnosis between ureteropelvic junction obstruction and megaureter. Furthermore, imaging provides information about split kidney function.

Voiding cystourethrogram: Confirms or excludes vesicoureteral reflux or posterior urethral valves.

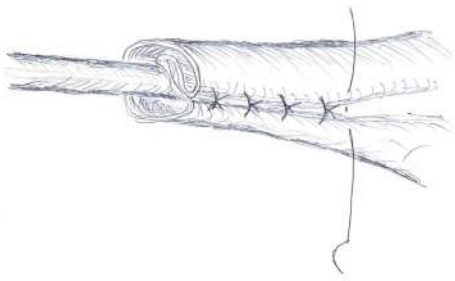
Renal scintigraphy: Determines split renal function and distinguishes between real obstruction and idiopathic (non-obstructive) megaureter. The nuclide wash-out 20 minutes after furosemide injection should be more than 50 % to rule our significant obstruction.

Retrograde pyelography and URS: Are indicated for unclear cases, e.g., to rule out malignancy or ureterolithiasis after diagnosis in adult life [fig. 4.25].

Whitaker perfusion test: with the help of a percutaneous nephrostomy, the renal pelvic pressure is determined during a flow rate of 10 ml/min. A Whitaker test is indicated if renal scintigraphy is unclear, especially in poor kidney function.



(a) Excisional tapering is used to reduce the severely dilated ureter. The lumen is reduced by resection of the excess portion and longitudinal closure of the ureter. Attention must be paid to the ureteral vasculature.



(b) Plication of the ureteral wall (Starr technique): suitable for moderately dilated ureters less than 1.75 cm in diameter without risk for ureteral blood supply or urine leakage.

Figure 4.28: Ureter remodeling before ureterocystoneostomy: the stenotic segment of ureter is resected, the MJ splint is again fixed with thin rapidly absorbable suture. Remodeling of a pronounced dilated ureter is done with plication or resection, use absorbable monofilament sutures 6-0 for suturing.

Therapy

Primary obstructive megaureter: The primary obstructive megaureter in children has a relevant spontaneous healing rate. Indications for an intervention (balloon dilatation or surgery) are a significant obstruction, recurrent febrile UTIs, or progressive loss of kidney function.

Balloon dilatation: And temporary DJ ureteral stent over several months, success rates of 25–90 % are reported (Kassite et al. 2018).

Surgical therapy: Includes excision of the narrow ureteral segment, ureter modellage (folding or vessel-sparing longitudinal resection, see fig. 4.28), and ureteroneocystostomy [p. 1161].

Secondary obstructive megaureter: Treatment of the underlying disease should be sufficient.

Refluxing Megaureter: Please see section vesicoureteral reflux on p. 460; in most cases, medical management is sufficient. If surgery is necessary, perform ureteroneocystostomy with ureter modellage depending on the ureter's diameter [p. 461 and 1161].

Idiopathic megaureter: Observation and conservative treatment (e.g., infections) are sufficient.

4.1.7 Retrocaval Ureter or Preureteral Vena Cava

Definition: A retrocaval ureter, also called a circumcaval ureter, is an abnormal development of the vena cava and ureter. Due to the persistence of the right subcardinal vein in the lumbar area, the vena cava develops ventrally of the right ureter (Zhang et al. 1990).

Epidemiology: 1:1500

Etiology: The ureter lies between the following fetal veins: dorsally, the supracardinal vein, and posterior cardinal vein; ventrally, the subcardinal vein. Usually, the portion caudal of the renal vein of the inferior vena cava develops from the right supracardinal vein and the ureter lies anterior to the inferior vena cava. If the subcardinal vein persists, the inferior vena cava develops ventral of the ureter and may cause hydronephrosis by compression between the vena cava and spine [fig. 4.29].

Signs and symptoms: Right-sided flank pain, nephrolithiasis, and pyelonephritis. Loss of right-sided kidney function is possible.

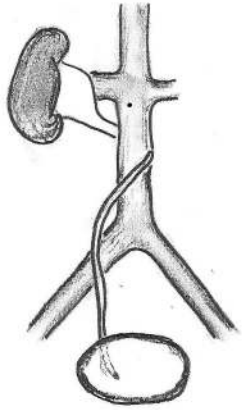


Figure 4.29: Schematic drawing of a retrocaval ureter.

Diagnosis: *intravenous urography:* The right mid-ureter suddenly turns to the medial; there may also be a change in diameter. Caudally from the ureteral kinking, there is often no contrast in the ureter. Thus, the ureter looks like a „J“. Signs of hydronephrosis or nephrolithiasis may be present.

CT or MRI: The abnormal position of the ureter to the inferior vena cava is easily identified.

Retrograde pyelography: Spiral (S-shaped) curve of the ureter around the vena cava [fig. 4.30].

Therapy: Surgical treatment is necessary for complications, such as intermittent flank pain, nephrolithiasis, recurrent infections, or significant hydronephrosis with loss of kidney function. Surgery includes the division of the ureter after complete mobilization, spatulation of the ureteral ends, and reanastomosis ventral of the vena cava (ureteroureterostomy). The technical difficulty of the operation is the necessity of extensive dissection of adhesions between ureter and vena cava with the corresponding risk of injury and bleeding. If the fibrotic adhesions between the ureter and vena cava are severe, the atretic ureter segment can be left behind the vena cava. Open surgery requires a significant retroperitoneal or transperitoneal approach to the vena cava due to the central position within the abdominal

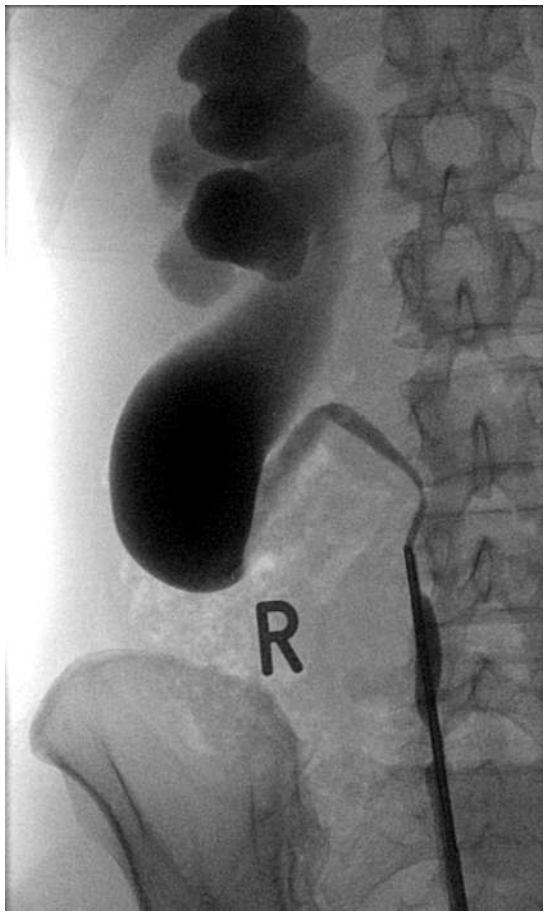
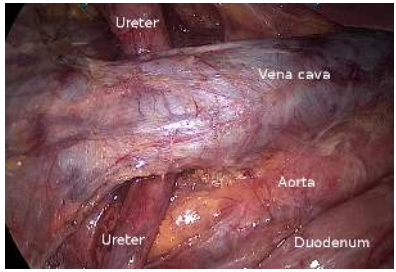


Figure 4.30: Retrocaval ureter: retrograde pyelography shows a spiral course of the right ureter around the vena cava with hydronephrosis. Please see fig. 4.31 for intraoperative findings of the same patient.

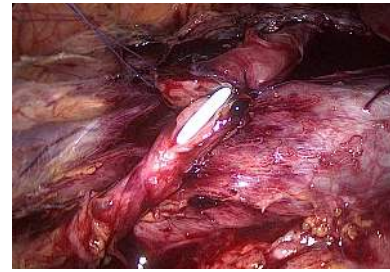
cavity. Laparoscopy can reduce the morbidity of the surgical procedure [fig. 4.31].

4.1.8 Retroiliac Ureter or Preureteral Iliac Artery

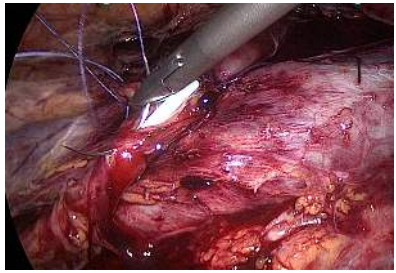
A retroiliac ureter is very rare, and associated malformations are common. The retroiliac ureter is caused by the development of the iliac vessels



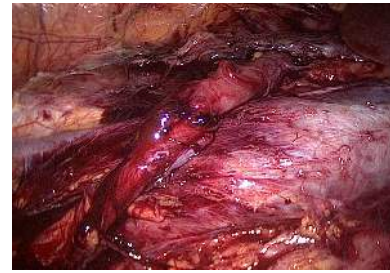
(a) Laparoscopic findings after exposure of the vena cava and ureter.



(b) Spatulation of the ureteral ends and placement of the first corner stitch.



(c) Ureteroureterostomy with an interrupted suture.



(d) Situs after ureteroureterostomy ventral of the vena cava.

Figure 4.31: Laparoscopic ureteroureterostomy of a retrocaval ureter, see fig. 4.30 for retrograde pyelography of the same patient.

from the anterior branch of the umbilical artery (instead of the ordinarily dorsal branch). The ureter can be compressed by iliac vessels, causing hydronephrosis [Abb. 4.32].

4.1.9 Ureteral Diverticula

Definition: Congenital ureteral diverticula can be divided into three groups: blind-ended ureter fissus, true ureteral diverticulum (all wall layers) and saccular outpouching of the mucosa (false diverticula).

Signs and symptoms: Urinary tract infection, hydronephrosis, flank pain, abdominal tumor.

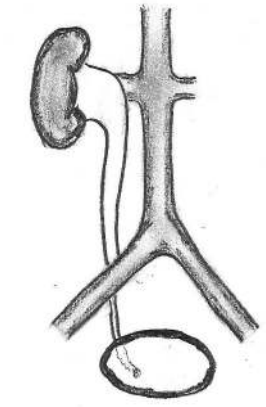


Figure 4.32: Schematic drawing of a retroiliac ureter.

Diagnosis: Ultrasound imaging, intravenous urography, CT or MRI, retrograde pyelography.
Therapy: If necessary resection (diverticulectomy), ureteroureterostomy or ureteral reimplantation.

4.1.10 Congenital Ureteral Stenosis and Ureteral Valve

Ureteral valve: Rare obstructive transverse fold of transitional epithelium containing smooth muscle bundles with prominent signs of hydronephrosis proximal of the valve and a normal ureter distally.
Ureteral stenosis: Congenital ureteral stenosis without transverse fold, with prominent signs of hydronephrosis and a normal ureter distally.
Therapy: Resection and ureteroureterostomy or ureteral reimplantation depending on the location of the obstruction.

4.2 Benign Diseases of the Ureter

4.2.1 Ureteral Trauma

Definition: Ureteral injury is rare and usually caused by penetrating trauma (gunshot or stab wounds). Another mechanism of injury is ureteral

tearing due to deceleration trauma (Elliott et al. 2003) (Elliott et al. 2006). EAU guidelines: (Kitrey et al. 2022).

Classification: Of ureteral injuries according to AAST, advance one grade for bilateral injuries:

- Grade I: hematoma without devascularization
- Grade II: laceration with under 50 % transection
- Grade III: laceration with over 50 % transection
- Grade IV: laceration with complete transection and less than 2 cm devascularization
- Grade V: laceration or avulsion with more than 2 cm devascularization

Signs and symptoms: The mechanism of injury usually dominates the symptoms of ureteral injury. Additional signs of ureter trauma are urinoma, abdominal pain, fever, urinary obstruction, hematuria, peritonism or urinary secretion via the wound.

Diagnosis: Ultrasound imaging and abdominal CT (with imaging of the excretion phase). Intravenous urography has largely been replaced by CT [fig. 4.33]. Signs of ureteral injury are urinary obstruction or leakage of urine (urinoma). Cystoscopy, retrograde pyelography, and ureteroscopy are only necessary, if endoscopic treatment is deemed possible.

Therapy: Minor ureteral injuries can be managed with endoscopic treatment (insertion of a ureteral stent or percutaneous nephrostomy). Surgical treatment is necessary for significant ureteral injury (suture repair, ureteroureterostomy, transureteroureterostomy, or ureteral reimplantation).

4.2.2 Iatrogenic Injury of the Ureter

Causes: Urological operations may jeopardize the ureter: ureteroscopy, retropubic prostatectomy, simple prostatectomy, transurethral resection of the prostate or bladder, pelvic or retroperitoneal lymphadenectomy. Frequent operations of other departments with a risk for ureteral injury

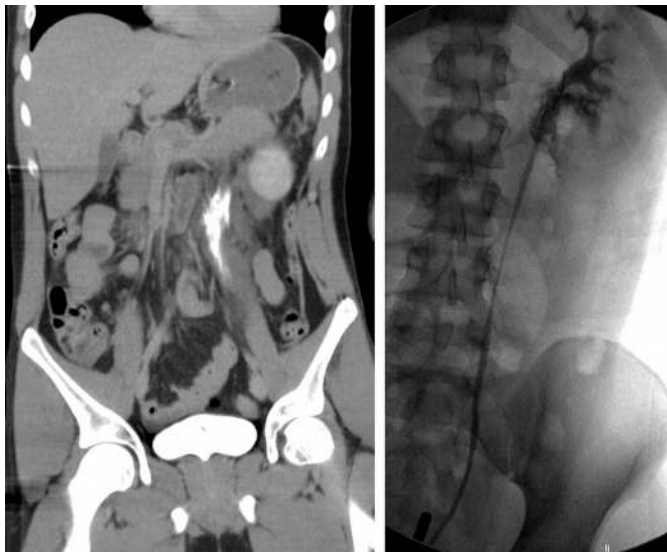


Figure 4.33: Left-sided ureteral injury: abdominal CT (excretion phase) and retrograde pyelography after fall from great height. Additional injury were a ruptured spleen and grade IV kidney injury. With kind permission (left figure), Prof. Dr. K. Bohndorf, Augsburg.

are hysterectomy, colon surgery and vascular surgery (Elliott et al. 2006) (Preston 2000).

Mechanisms of injury: Dissection, suture, ligature, contusion, devascularization, infection, or compression by hematoma or lymphocele.

Signs and symptoms: Flank pain, abdominal pain, fever, upper urinary tract obstruction, hematuria, urinoma, ascites, peritonitis, or urine secretion via the wound or vagina, elevated creatinine.

Diagnosis: Laboratory tests (creatinine, CRP), imaging with ultrasound, intravenous urography or better CT, cystography, retrograde or antegrade pyelography.

Therapy

Endoscopic treatment: An option if antegrade or retrograde placement of a ureteral stent is possible. Depending on the healing of the ureter, the ureteral stent can be withdrawn. Surgical treatment is necessary for

persisting ureteral stricture, see next section on p. 476. Percutaneous nephrostomy is an option when ureteral splinting is not possible.

Surgical Treatment: Indications for surgical treatment are frustrate endoscopic treatment, urinous ascites, ureteral-vaginal fistula, and persistent ureteral stricture after endoscopic treatment. Surgical reconstruction depends on the location and extent of the ureteral injury, see p. 476. After succesful endoscopic treatment, final reconstruction must be postponed until the patient is in good condition, e.g., after peritonitis, sepsis or shock. Depending on the circumstances, a two-stage procedure after the insertion of a percutaneous nephrostomy may be wise.

4.2.3 Ureteral Stricture

Definition

Ureteral stricture is the narrowing of the ureter, caused by various diseases and leads to upper urinary tract obstruction (Hafez et al. 2003).

Etiology

Diseases of the ureter (intrinsic causes): Ureteral stones, infections (e.g., urogenital tuberculosis and schistosomiasis), upper urinary tract carcinoma, idiopathic or congenital diseases.

Iatrogenic causes: Irradiation or intra-operative injury (e.g., ureteroscopy, rectal surgery, hysterectomy, or vascular surgery).

Extraureteral diseases (extrinsic causes): Various diseases may lead to obstruction of the ureter by compression or infiltration, please see the differential diagnosis of hydronephrosis on p. 484.

Signs and Symptoms

Ureteral stricture may develop unnoticed, a slowly progressive ureteral stricture is often asymptomatic. Flank pain, fever, and hematuria occur depending on the underlying disease or infectious complications.

Diagnosis

Imaging: Renal ultrasound: grade of hydronephrosis? Intravenous urography or better abdominal CT: etiology of ureteral stricture? Renal scintigraphy: renal function? Significant obstruction?

Endoscopy: Retrograde pyelography and ureterorenoscopy to confirm the diagnosis and assess the stricture length. Biopsy of the stricture is necessary in case of unclear etiology.

Therapy

Ureteral stenting: Initial treatment of symptomatic hydronephrosis after endoscopic diagnosis (see above). In conditions with possible spontaneous healing and resolution of the ureteral stricture, the ureteral stent can be removed after a few weeks (e.g., after infections).

Endoscopic Balloon Dilatation: A balloon dilatator is introduced after retrograde (or antegrade) pyelography and the insertion of a guide wire. Dilatation is done to a diameter of 4–6 mm, and a ureteral stent (DJ) is placed for 2–4 weeks. The success rate is 50–75 %. Balloon dilatation is especially suitable for short segment strictures of less than 2 cm and for strictures after lithotripsy of ureteral stones.

Endoscopic ureterotomy: The stricture is incised under endoscopic vision after retrograde (or antegrade) pyelography and insertion of a guide wire. A full-thickness cut through the ureteral wall is done until the periureteral fat is seen. Visualization is possible with retrograde (URS) or antegrade (nephroscope) techniques. Ureterotomy is possible with a cold knife (without cauterization) and laser fibers (holmium or Neodym:YAG). Care has to be given to vessels near the ureter. Proximal strictures (above the iliac vessels) should be cut latero-dorsally. Strictures near the iliac vessels and below should be cut anterior-medially. After the ureterotomy, a thick ureteral stent is placed for 8–10 weeks. The success rate is slightly higher than balloon dilatation, especially after strictures of ureteral anastomosis. The stricture length is the most important prognostic factor.

Ureteroureterostomy: is a simple operation for treating short segment strictures (< 3 cm) of the proximal and mid ureter. For details see p. 1158.

Ureteral reimplantation: ureteroneocystostomy is suitable for the treatment of distal ureteral strictures up to 4–5 cm in length [p. 1161]. With the help of the psoas hitch technique, 6–10 cm of the ureter can be replaced. With the Boari-flap technique and depending on bladder capacity, 12–15 cm of the ureter can be replaced [p. 1168]. An additional distance of 6 cm can be bridged by mobilization of the kidney and fixation of the lower pole to the psoas muscle.

Transureteroureterostomy: strictures of the mid ureter in combination with a low bladder capacity may be treated with transureteroureterostomy. A short donor ureter (proximal stricture) and a diseased ureter on the opposite side are contraindications. Furthermore, transureteroureterostomy is unsuitable for Ormond disease, after retroperitoneal radiation, upper tract transitional cell carcinoma, and in patients with recurrent nephrolithiasis.

Renal autotransplantation: explantation of the kidney and anastomosis with the iliac vessels is possible if the renal pelvis with a short proximal ureter can be anastomosed with the bladder.

Use of buccal mucosa grafts: A longitudinal incision of the strictured ureter segment and onlay grafting with buccal mucosa is used for multifocal ureteral strictures or after failed pyeloplasty.

Intestinal interposition: Reconstruction of long-segment ureteral strictures is possible with ileum (as a last resort). An appropriate segment of the ileum is delivered to the retroperitoneum and is anastomosed with full-thickness watertight sutures to the renal pelvis and bladder in an isoperistaltic fashion for adequate urine transport. Contraindications for ileal ureteral substitution are significant chronic kidney disease, bladder dysfunction, inflammatory bowel disease, or radiation injury of the bowel. Modified techniques to reduce complications (mucus obstruction, metabolic changes, and stone formation) and the length of used bowel include taper-

ing the bowel graft, using a reconfigured ileal ureter (Yang-Monti), and combining it with a Boari flap (Xiong et al. 2020).

4.2.4 Retroperitoneal Fibrosis (Morbus Ormond)

Definition

Ormond disease (retroperitoneal fibrosis) is a disease with an unknown etiology, leading to increasing fibrosis of the retroperitoneum and compression of ureters and retroperitoneal vessels (Vaglio et al. 2006).

Epidemiology

Male to female ratio = 2:1. Prevalence $^{1-2}/_{100\,000}$. Age of onset 40–60 years.

Etiology

Unknown etiology: In 70 % of patients with retroperitoneal fibrosis, the etiology of Ormond disease remains unclear. One assumed disease mechanism is an autoimmune process with periaortitis as the initial pathophysiological mechanism. Later, the inflammatory-fibrotic mass extends into the retroperitoneum and compresses the ureters. There is also evidence of a local manifestation of a systemic increased willingness to develop autoimmune diseases (see below association with fibrotic diseases).

Known triggers: *Drugs:* Ergot alkaloids, β-blocker, phenacetin, LSD, haloperidol, amphetamine, reserpine.

Infections: Lymphangitis, chronic inflammatory bowel disease, gonorrhea, syphilis, urogenital tuberculosis, chronic urinary tract infection, sarcoidosis.

Association with fibrotic diseases: Sclerosing mediastinitis, sclerosing cholangitis, orbital pseudotumor, Riedel's thyroiditis. In children, association with systemic lupus erythematosus or juvenile rheumatoid arthritis.

Other risk factors: Irradiation, retroperitoneal bleeding, pelvic surgery, Purpura Hennoch.

Pathology

Retroperitoneal fibrosis leads to a substantial mass of connective tissue, which surrounds the large retroperitoneal vessels and may include the ureter. The retroperitoneal mass extends from the renal hilum to the sacral promontory in the craniocaudal direction. At the edges of the active fibrosis are signs of inflammation in the connective tissue (lymphocytes, plasma cells, histiocytes). In the center of the mass, inactive connective tissue is found without signs of inflammation.

Signs and Symptoms

- Flank pain: constant dull pain, starting in the flanks with radiating to the lower abdomen.
- Nonspecific symptoms such as weight loss, malaise, nausea, and vomiting may occur at the beginning of the inflammatory process. Lymphadenopathy is not typical.
- In untreated patients, symptoms of uremia may develop.

Diagnosis

Laboratory tests: Elevated ESR and CRP. Further necessary tests are creatinine, electrolytes, full blood count, autoantibodies (ANA, ANCA), immunoelectrophoresis, and rheumatoid factor. Additional tests in men: PSA, AFP, HCG, LDH.

Ultrasound imaging: Reliably identifies hydronephrosis. Reduced thickness of the kidney parenchyma is an sign of chronic renal failure. The retroperitoneal connective tissue mass around the aorta can be visualized in good conditions.

CT or MRI of the abdomen: Imaging reveals a flat mass surrounding the great vessels and the ureters with bilateral hydronephrosis [fig. 4.34(b)]. The renal function or damage can be estimated with the thickness of the renal parenchyma.

(a) Intravenous urography (late KUB after 90 min): medial displacement of both ureters with hydronephrosis (right>left).

(b) CT (frontal plane): circular mass around the aorta (arrows mark the left-lateral border).

Figure 4.34: Imaging of a patient with retroperitoneal fibrosis. With kind permission, Dr. N. Dreger, Prof. Dr. P. Haage and Prof. Dr. S. Roth, Wuppertal.

Intravenous urography: Reveals bilateral hydronephrosis and displacement of the proximal and mid-ureter to the middle [fig. 4.34(a)]. IVP is only necessary if cross-sectional imaging is not available.

Retrograde pyelography: Indicated in patients with contraindications to contrast medium and before endoscopic therapy. Radiological signs are similar to IVP; intrinsic ureteral obstructions are easily identified.

Renal scintigraphy: To determine spit renal function and to objectify urinary obstruction at the onset and later in the course of the disease.

Percutaneous biopsy: Of the retroperitoneal mass should be done to exclude a malignant etiology, especially with suspected retroperitoneal lymphoma (pathological blood count, lymphadenopathy) or cancer in the history. With typical clinical and radiological criteria of retroperitoneal