Congenital Ureteropelvic Junction Stenosis

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89.1 Introduction

Congenital ureteropelvic junction (UPJ) stenosis is defined as an impairment of urine flow from the renal pelvis to the ureter resulting in dilation of the pelvis. It has the potential to damage kidney function (Fig. 89.1). UPJ obstruction is the most frequent congenital malformation of the genitourinary system with an incidence of 1 in 1000 live births.

The male-to-female ratio of congenital UPJ stenosis is 2:1. The left side is significantly more frequently affected than the right side. In 30% of cases, both kidneys are affected.

The outcome of a UPJ stenosis is quite good in general, given timely therapy, but the outcome often depends on whether there are concomitant malformations. The most frequent are further anomalies of the urinary tract—for example, duplex kidneys, horseshoe kidneys, mega-ureters, urethral valves or contralateral multicystic renal dysplasia.

The condition has been shown to be associated with congenital cardiac malformations, hydrocephalus, skeletal dysplasias or trisomy 21. In the international literature, nonrenal concomitant malformations have been described in up to 12% of cases (Table 89.1).

89.2 Aetiology

Causes of UPJ obstruction can be divided into congenital and acquired, intrinsic and extrinsic. The commonest cause in the pediatric age group is congenital intrinsic obstruction caused by an aperistaltic section of the ureter at the UPJ usually lacking in interstitial cells of Cajal or pacemaker cells. This may result in a functional obstruction. One of the commonest causes in the adult population is extrinsic UPJ obstruction caused, by accessory or aberrant lower polar vessels crossing the ureter junction.

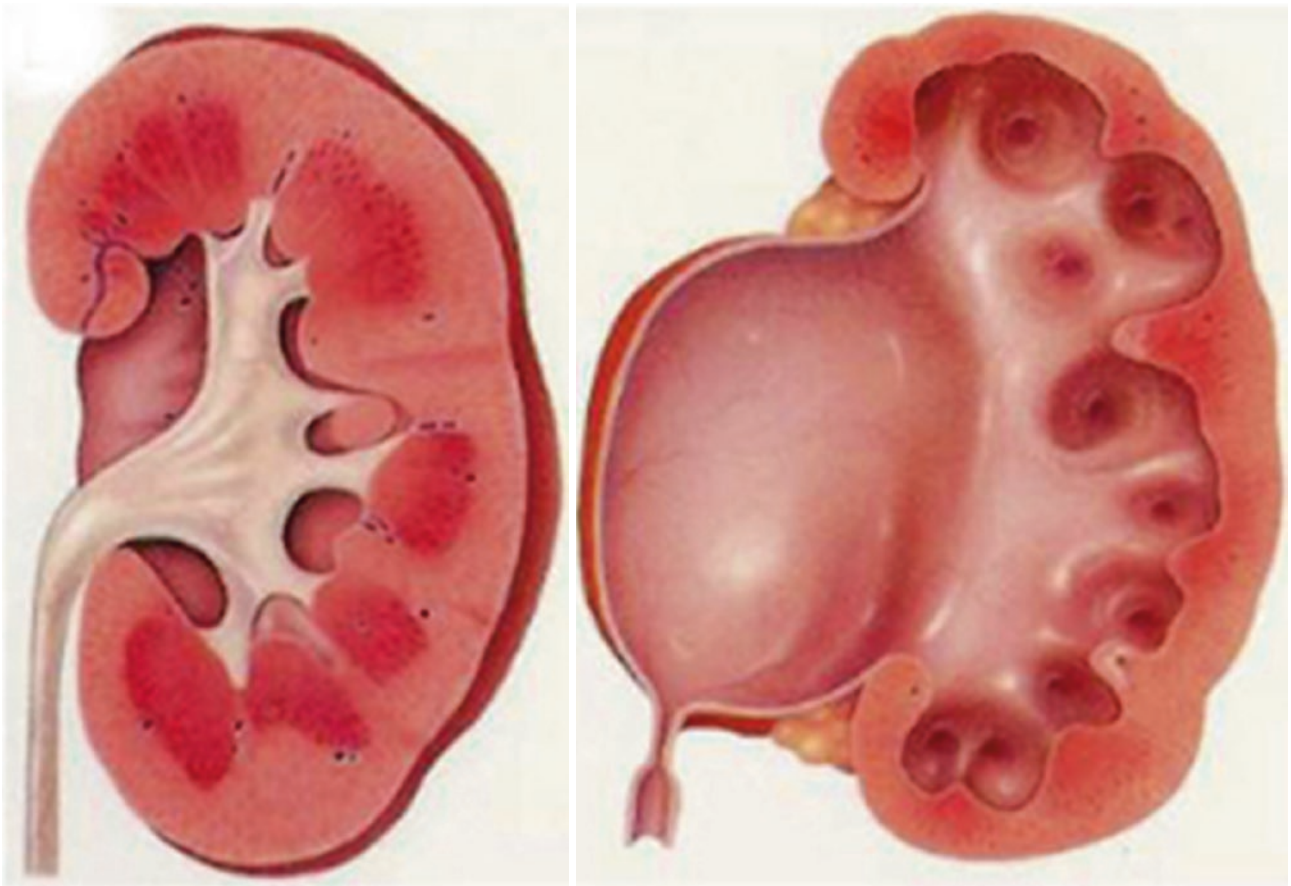
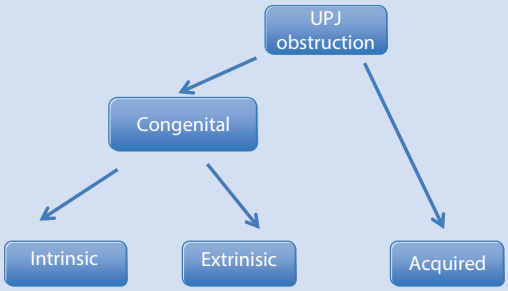


Fig. 89.1 Normal and stenosed ureteropelvic junction

Table 89.1 Associations with UPJ obstruction

Non-urological (12%)	Urological (50%)
Congenital cardiac defects	Contralateral UPJ obstruction (10–40%)
Hydrocephalus	Ectopic kidney (35%)
Skeletal dysplasia	Horseshoe kidney (15%)
Trisomy 21	Duplex kidney (15%)
VACTERL syndrome	VUR (0.5–5%)
	Unilateral renal agenesis
	Contralateral renal dysplasia or MCDK (multicystic dysplastic kidney)

Table 89.2 Causes of UPJ obstruction



Intrinsic	Extrinisic	Acquired
Adynamic segment (common)	Crossing lower pole vessel (common)	Stricture (stone, trauma, tumour, instrumentation, TB, schistosomiasis)
Intrinsic stenosis	High ureteral insertion into the pelvis	Severe vesicoureteric reflux with tortuosity
Valvular mucosal folds	Horseshoe, ectopic or mal-rotated kidney	
Mucosal polyp	Extrinsic compression from a tumour	

It has been postulated that neuronal innervation defects play a role in the pathogenesis of UPJ; however, a well conducted study showed a heterogeneity of neuronal markers histologically with no significant difference to controls [1] (Table 89.2).

89.3 Pathophysiology

Pelvic dilatation is the hallmark of UPJ. Obstruction of the urinary tract is defined as decreased flow of urine that if untreated will lead to a decrease in renal function. However, only about 1/3 of patients with unilateral UPJ obstruction will develop loss of renal function. Thus, the challenge is often not diagnosing the pelvic dilation but knowing which dilation to treat. True obstruction of the UPJ causes increased pressure on the renal parenchyma, the wall of the renal pelvis and the blood vessels. Initially this decrease in renal function is reversible, but in time irreversible loss of kidney function can occur due to renal atrophy. This process has been shown not to be pure pressure atrophy; the main cause of the hydronephrotic atrophy is circulatory disruption of the blood and lymph vessels due to increased tension in the renal pelvic wall. Many investigations have shown that the loss of kidney function after long-standing obstruction is irreversible, even after relief.

89.4 Prenatal Diagnosis

Prenatal ultrasound (US) can show reliably a dilatation of the renal pelvis at the 16th to 20th week of gestation and can be used to follow the development of the dilatation during the further course of pregnancy. The most frequent prenatal fetal abnormality is an isolated renal pelvis dilatation. In all cases, a postnatal ultrasound is recommended [2]. The timing of the postnatal ultrasound can be done immediately in severe cases; however, it is more accurate to repeat the ultrasound after day 3 as there is a physiological and transient dehydration of the neonate within the first 48 hours of life. However, not every prenatal dilatation of the kidney results in a pathological condition. Many low-level prenatal dilatations of the renal pelvis can no longer be detected postnatally [3–4].

The upper urinary tract dilation classification system proposes using the term dilatation rather than hydronephrosis to emphasize this fact that not all dilatation is obstructive. This classification system aims to use uniform language and criteria for antenatal diagnosis to improve communication and research [5] (Table 89.3).

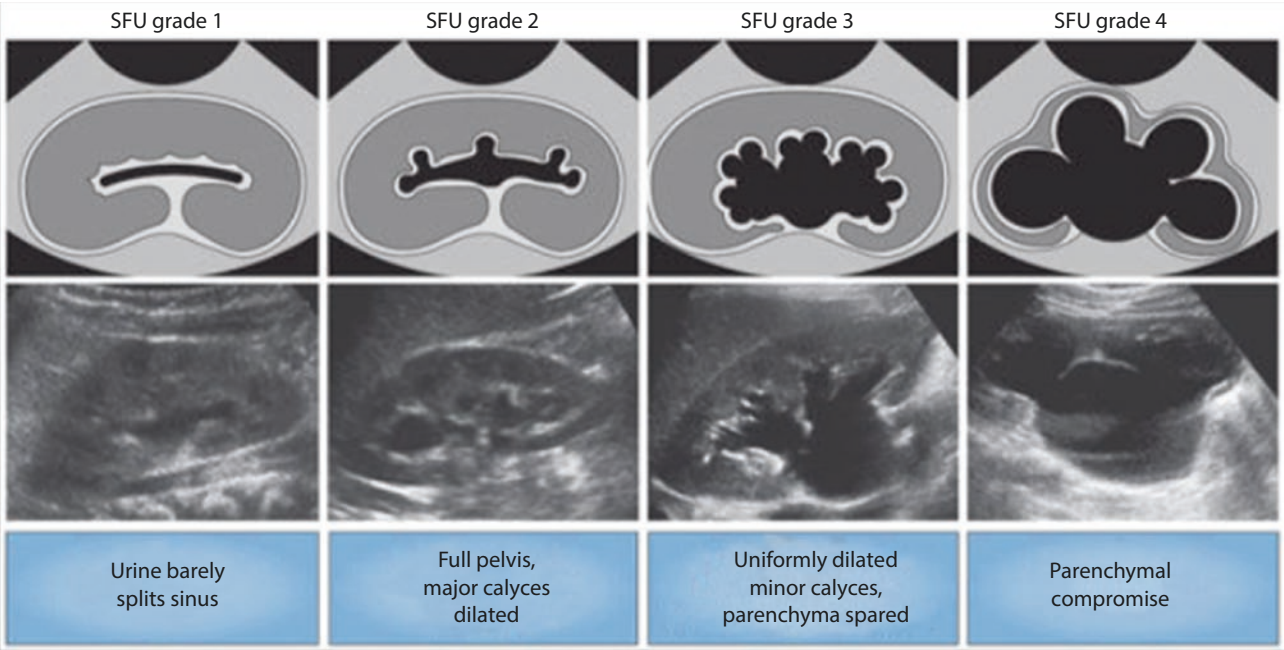
Although the urinary tract dilatation grading system was designed to address potential shortcomings of the Society for Fetal Urology classification, a recent comparison showed that both had good predictive value for resolution of hydronephrosis. Cumulative resolution rates at 3 years were 98% for SFU grade 1, 87% for grade 2, 76% for grade 3 and 57% for grade 4, respectively ($p < 0{,}001$). The resolution rates for the UTD classification at 3 years were 90% for grade 1, 81% for grade 2 and 71% for grade 3, respectively [6].

Table 89.3 Prenatal urinary tract dilation (UTD) classification[a] for UTD A1 and UTD A2-3

		UTD A1	UTD A2–3
APRPD 16–27 weeks	<4 mm	4–7 mm	≥7 mm
APRPD ≥28 weeks	<7 mm	7–10 mm	≥10 mm
Calyceal dilation	None	Central or none	Peripheral[b]
Parenchymal thickness	Normal	Normal	Abnormal
Parenchymal appearance	Normal	Normal	Abnormal
Ureters	Normal	Normal	Abnormal
Bladder	Normal	Normal	Abnormal
Oligohydramnios	None	None	Unexplained[c]

[a]Classification is based on the presence of the most concerning feature. For example, a fetus with an anterior posterior renal pelvic diameter (APRPD) within the UTD A1 range but with ureteral dilation would be classified as UTD A2–3
[b]Central versus peripheral calyceal dilation can be difficult to assess early in gestation
[c]Oligohydramnios thought to be the result of a genitourinary cause

89.4.1 Society for Fetal Urology Classification System [7]



89.5 Clinical Symptomatology

In well-resourced settings, the majority of stenoses of the UPJ are diagnosed prenatally by means of US and are often asymptomatic.

A UPJ obstruction can present with UTI in infancy, which often has non-specific features, such as poor growth, refusal to drink, recurring vomiting and agitation. This should prompt an US examination to evaluate a cause, which may prove to be UPJ obstruction. A

UPJ can present with a flank mass in the neonatal period.

When UPJ stenosis is not diagnosed in the perinatal period, it can present later with symptoms such as flank pain, haematuria (in 30% of late diagnoses), a palpable intra-abdominal masses and urinary tract infections (UTIs). In severe cases, the patient can have classic symptoms of urosepsis.

Untreated unilateral UPJ obstructions result in a nonfunctional, hydronephrotic, sacculated kidney; as the loss of kidney function progresses, children with untreated bilateral UPJ obstructions begin to show symptoms of renal insufficiency [8, 9]. Delayed presentation in a resource-limited setting was associated with nephrectomy in 37% of cases. This can be mitigated by improved primary health care and wider availability of ultrasound.

89.6 Diagnostics

89.6.1 Clinical Assessment

As with all illnesses, prior to imaging diagnostics, a thorough medical history and physical examination of the child have to be undertaken. A family history is important as there is familial clustering in UPJ obstruction; however, there is no known hereditary chromosomal influence described. Complete blood and urine tests have to be carried out to determine or exclude any accompanying nephrological or urological conditions or an infection.

89.6.2 Imaging

An ultrasound scan is a useful initial investigation, as it has no radiation, it is minimally invasive, widely available and relatively cheap. However, it is operator dependent and there is a significant intraobserver variability. The SFU and UTD are standardized systems, which aim to decrease intraobserver variability and predict the need for intervention (Table 89.4).

Although it has its limitations, AP pelvis diameter does predict the need for surgery. The need for surgery is 80% if the AP pelvis is more than 40 mm, 55% if it is more than 30 mm, 20% if more than 20 mm and 1–3% if less than 20mm [10].

In cases where there is bilateral hydronephrosis, a thickened bladder wall, hydroureteronephrosis, duplex kidney, ureterocele or diagnostic doubt, a voiding cystourethrogram should be performed. It is useful in these cases to exclude vesicoureteric reflux, posterior urethral valves and other pathology; however, it should be used judiciously due to the radiation risk and need for catheterization. There are, however, cases with combined malformations showing a UPJ obstruction and a primary vesicoureteral reflux on the ipsilateral side. It is challenging to decide which problem to address first, and each case should be evaluated on an individual basis.

Table 89.4 Classification of Urinary Tract Dilatation (UTD)

	Normal	UTD P1	UTD P2	UTD P3
APRPD	<10 mm	≥10–15 mm	≥15 mm	>=10 mm
Calyceal dilation	None	Central only	Peripheral	_[b]
Parenchymal thickness	Normal	Normal	Normal	Abnormal
Parenchymal appearance	Normal	Normal	Normal	Abnormal
Ureters	Normal	Normal	Abnormal	_[b]
Bladder	Normal	Normal	Normal	Abnormal

[a]Classification is based on the most concerning ultrasound finding. For example, if the anterior posterior renal pelvic diameter (APRPD) is in the UTD P1 range but there is peripheral calyceal dilation, the classification is UTD P2. The presence of parenchymal abnormalities denotes UTD P3 classification as long as there is urinary tract dilation

[b]Calyceal dilation and ureteral dilation, although frequently present in patients with UTD P3, are not necessarily needed to qualify for UTD P3 if there is urinary tract dilation with either abnormal parenchymal thickness, abnormal parenchymal appearance or abnormal bladder

The decisive investigation is technetium-99 m mercaptoacetyltriglycine (Tc-99 m MAG3) renal function scintigraphy, which not only shows the renal function for both sides but also is especially important for determining the flow relationships in the urinary tract collection system. In the case of an obstruction, Fig. 89.2 shows a mounting curve on the affected side. It is important to carry out the Tc-99 m MAG3 scintigraphy investigation together with a furosemide investigation [11–13].

Many parameters have been proposed to determine the need for surgery based on a renogram. T ½ is the time it takes for 50% of the tracer to be removed from the kidney. A T1/2 of less than 10 mins is considered normal, 10–20 mins is indeterminate and more than 20 mins is suggestive of obstruction. However, although prolonged T ½ is suggestive of obstruction, it is not a reliable predictor and should be taken in context of the images, curves and other quantitative data. Some

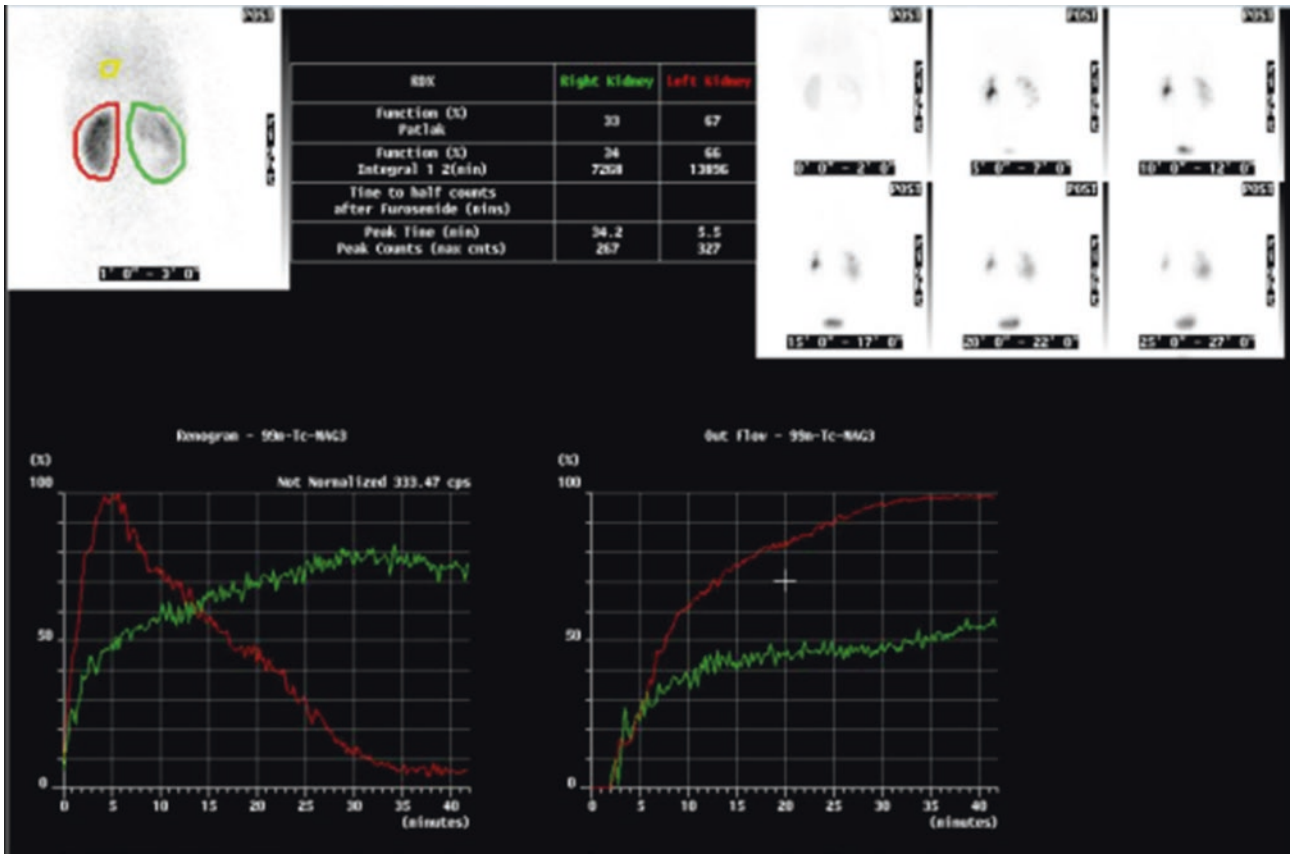


Fig. 89.2 Renal function scintigraphy with a mounting curve for the left kidney, showing obstruction for any uncertain issues that still exist—for example, the presence of different simultaneous renal malformations or of a tumour—imaging procedures such as CT or MRI urography can be employed. It is recommended that an on-table retrograde pyelogram be performed prior to pyeloplasty to confirm the diagnosis of PUJ obstruction and rule out the presence of a vesicoureteric obstruction

authors have suggested that it should be no longer be used as a criteria to predict surgery as it is prone to technical, physiological and anatomical error [14].

The NORA (normalized residual activity) is a ratio of the renogram value at a certain time to the value at 1–2 mins. A ratio of less than 1.0 is considered normal if no diuretics are given, but parameters are dependent on hydration status and the administration of diuretics. Like NORA, renal output efficiency (ROE) was introduced to normalize output to renal function. Normal Mag 3 renal output efficiency is >81.6% at 20 minutes and 89% at 40 minutes [15].

However, in the setting of pelvic dilatation, there are inherent problems with measurements of outflow efficiency given that a large dilated pelvis may appear obstructed when it is simply filling with tracer. Recent data suggests that gravity-assisted drainage post void is a greater predictor of renal obstruction than measurements of outflow efficiency [16].

Renal tissue tracer transit is likewise a useful predictor of obstruction given that there is a drop in GFR in the obstructed kidney. In a recent retrospective analysis, delayed tissue tracer transit was the only independent predictor of improved renal function after pyeloplasty [17].

The third robust indicator for pyeloplasty is a drop in differential renal function. An ipsilateral split function of below 40% is recommended as an indication for pyeloplasty although some would consider a lower threshold of 30%. However, the proviso is that the contralateral kidney is functioning normally. If not, the split function can appear normal despite a dramatic drop in global renal function. A nuclear medicine-based GFR will identify this. A decrease of more than 10% in split function on the subsequent renogram is an indication for intervention.

Should the investigations show no flow obstruction of the urinary tract, despite sonographically detected dilatation, the child should undergo periodic examinations, and, if the dilatation of the renal pelvis progresses or UTIs develop, the diagnostic procedure should be repeated, as shown in the algorithm.

89.7 Treatment of a Congenital Ureteropelvic Junction Obstruction

The spectrum of treatment for a congenital UPJ obstruction reaches from conservative observation to the application of a temporary, percutaneous nephrostomy in acute emergency situations; to plastic reconstructive organ-preserving surgery methods; and to the rare necessary excision of a nonfunctional organ.

All moderate renal pelvis dilatations without indications for surgery should be followed up with regular US, urine and blood tests and renograms. Should UTIs occur, or should the US show an increase in the renal-pelvis calyceal system dilatation, a new renogram is warranted.

The goal of a temporary, percutaneous nephrostomy is to decompress an infected hydronephrotic kidney. Normally, this is performed percutaneously under general anaesthesia, with US monitoring. This temporary drainage of urine results in a reduction in pain, allows an efficient antibiotic treatment of the infection and enables an adequate diagnosis. The required surgical procedure can then be carried out on an organ that is not infected and with the patient in the best possible general condition.

An indication for nephrectomy is given only when the renal parenchyma is so damaged by obstructive and/or infectious processes that the organ has become functionally worthless. The decision to perform a nephrectomy can be made only after a thorough diagnostic evaluation and the reliable demonstration that the kidney is no longer functional. With acute hydronephrosis, especially in early infancy, it is advisable, after primary percutaneous relief of the kidney, to perform a renal function scintigraphy examination once again. In the literature, renal function below 10% is frequently given as an indication for nephrectomy.

Surgical techniques can be divided into those that involve resection and those that do not. Due to the relatively high rate of recurrence, the nonresectional flap techniques have not found acceptance internationally. In Fig. 89.3, the stenotic region at the ureter junction is left in place and is extended by means of a folded-in ureteral flap.

In recent decades, resectional surgical techniques—in particular, that of Anderson-Hynes [18, 19]—have gained acceptance. Common to all resectional methods is the removal of the dysplastic portion of the ureter followed by a microsurgical anastomosis to connect the renal pelvis and the ureter. The Anderson-Hynes technique is seen today as the standard. It can be carried out both in open surgery and—with older children, from around age 24 months—laparoscopically [28, 19]. Once there is a surgical indication, the operation should be carried out regardless of the age of the child. Figures 89.4, 89.5, and 89.6 show the essential steps in the Anderson-Hynes procedure.

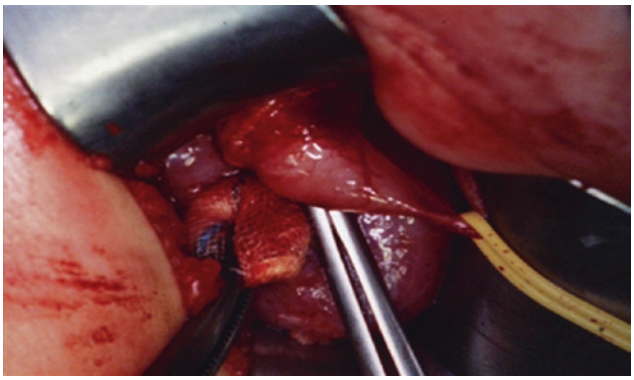


Fig. 89.3 Surgical site uretero. The goal of all plastic-reconstructive surgical methods is the removal of the hindrance to the urinary flow with retention of the organ

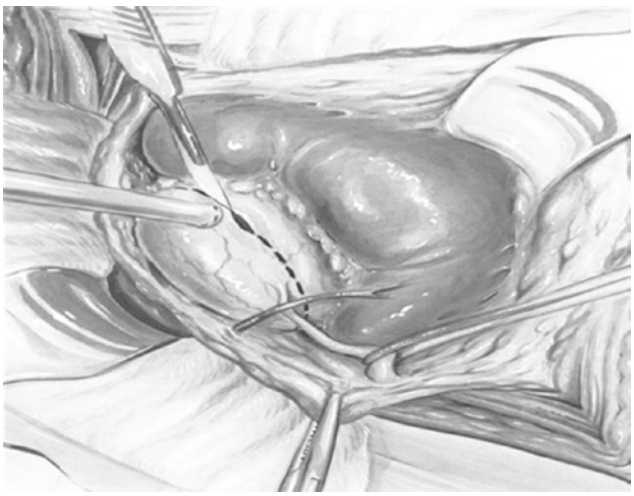


Fig. 89.4 Resection line renal pelvis, ureter identified by ligature pelvic junction obstruction

Especially with small children, conventional open surgical retroperitoneal-dorsal access is usually preferred, for example, access according to the Bergmann-Israel procedure. After freeing the kidney, the renal pelvis and the ureter, the dysplastic constriction at the ureter junction with the prestenotic dilated renal pelvis can be clearly seen. After placement of retention stitches, the resection of the ureter restriction is undertaken, together with a small part of the dilated renal pelvis, in order to avoid a Windkessel function postoperatively. Then the renal pelvis-ureter spatulated anastomosis is sutured continuously microsurgically by using a thin, absorbable suture material corresponding to the age of the child (e.g. Vicryl 6–0 or 7–0). The need for a reduction pyeloplasty in the era of laparascopic pyeloplasty is

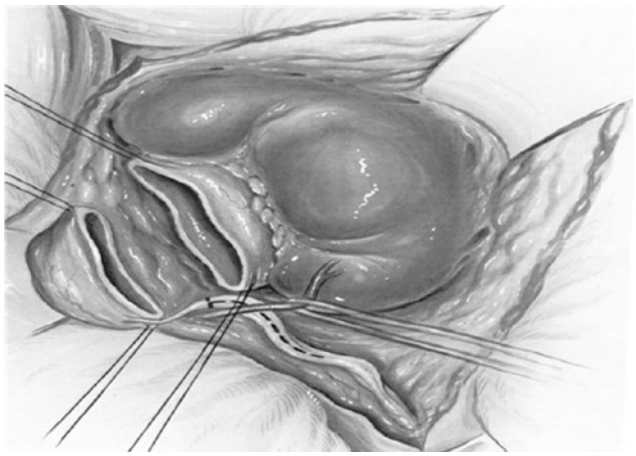


Fig. 89.5 Resection line: ureter-renal pelvis cut through

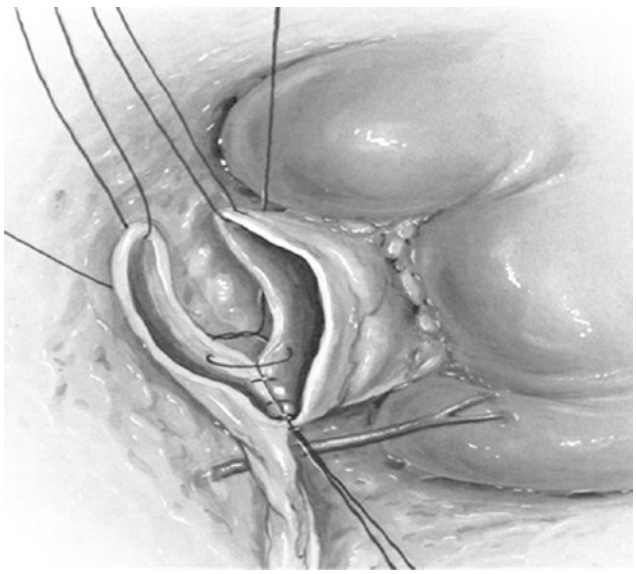


Fig. 89.6 Continuous ureter-renal pelvis anastomosis with inserted pyelostomy

debatable with outcomes being equivalent weather the pelvis is tapered or not.

Intraoperative insertion of a pyelostomy or a ureteral stent for postoperative securing and splinting of the anastomosis can be carried out in different ways. Both methods and whether they should be used at all are subjects of debate. If the surgeon decides to install a pyelostomy, it should be removed between the seventh and tenth postoperative day. Before its removal, the pyelostomy enables one to monitor the flow control of the anastomosis by means of x-ray contrast imaging or to influence the flow by a temporary "training" of the anastomosis by means of pinching it off for a time and then releasing it. For the removal of an intraoperatively inserted double-J ureteral stent about 3 months postoperative, a cystoscopy under general anaesthesia is required.

According to the international literature, a completely drainage-free surgery shows no inferior results—in particular, no higher rate of anastomosis leakage [20, 21].

Endopyelotomy is an alternative procedure; however, it only has a success rate of 71%, which is significantly inferior when compared to pyeloplasty. It has also been associated with a higher complication rate [22]. It is contraindicated when UPJ obstruction is due to a crossing vessel. In cases where a crossing vessel is present, a laparascopic vascular hitch can be contemplated. In small series, it has shown to be appropriate if there is a type 3 anatomic abnormality (when the UPJ lies below the crossing vessel causing the ureter to curl back or swan neck over the vessel) and when the pelvis is seen to decompress when the vessel is retracted away from the PUJ (the urine charge test) with the assistance of fluid and diuretics [23].

89.8 Postoperative Complications

During the immediate postoperative phase, the focus with regard to complications is on anastomosis anastamotic leakages, swelling of the anastomosis with resultant backing up of urine and UTIs.

Intraoperative insertion of drainage serves to reduce pressure postoperatively in the area of the anastomosis and can prevent a backing up or leakage of urine, which aids in recovery.

The patient should be protected against possible postoperative infections by perioperative and postoperative intravenous administration of antibiotics. This is carried out either on the basis of a resistogram or by using a wide-spectrum antibiotic, such as cefuroxime, adapted for weight.

89.9 Outcome

The long-term results of the Anderson-Hynes procedure are very good, with the rate of recurrence of a stenosis being 3–5% in the international literature. The most frequent reason for a new operation is the development of a stenosis in the area of the anastomosis. In rare cases, suture granulomas or connective tissue accretions in the area of the surgery, which narrow or deform the ureter junction, can be found.

Decisive for the success of the operation is not the complete reduction of the renal pelvis dilatation but the free, unobstructed flow of urine, as shown by scintigraphy. This follow-up investigation should be carried out

6 months to 1 year after surgery. All children operated on for a UPJ obstruction must remain in outpatient care. At the outset, regular urinalysis and US examinations should be carried out every 4–6 weeks. If no complications arise, the time between follow-up examinations can be extended [24–29].

89.10 Concluding Comments

Congenital UPJ stenosis is a pathological condition of the genitourinary system that can be detected early during pregnancy by US examination. These early prenatal signs make possible a timely diagnosis and treatment postnatally. Only in very severe cases does the condition result in a complete loss of the kidney and serious complications, despite timely treatment. As a rule, given timely diagnosis and the indicated treatment, it is possible to maintain kidney function at the level of the point in time of surgery without further complications.

89.11 Evidence-Based Research

Table 89.5 presents a landmark retrospective review of 1000 children with hydronephrosis. Table 89.6 presents a natural history series of children with severe antenatal hydronephrosis managed nonoperatively.

Table 89.5 Evidence-based research

Title	Prenatally diagnosed hydronephrosis: the Great Ormond Street experience
Authors	Dhillon HK
Institution	Great Ormond Street Hospital for Sick Children, London, UK
Reference	Br J Urol 1998; 81(suppl 2):39–44
Problem	Natural history series of 1000 children with antenatal hydronephrosis
Intervention	Anderson-Hynes pyeloplasty
Comparison/control (quality of evidence)	Retrospective review
Outcome/effect	Demonstrated the essentially benign nature of antenatally picked hydronephrosis. Only 5% of children with an AP measurement of <20 mm required pyeloplasty
Historical significance/comments	Landmark paper

Table 89.6 Evidence-based research

Title	The long-term follow-up of newborns with severe unilateral hydronephrosis initially treated nonoperatively
Authors	Ulman I, Jayanthi VR, Koff SA
Institution	The Ohio State University, Columbus, Ohio, USA
Reference	J Urol 2000; 164:1101–1105
Problem	Natural history series of children with severe antenatal hydronephrosis managed nonoperatively
Intervention	Nonoperative
Comparison/control (quality of evidence)	Retrospective review
Outcome/effect	Highlights the benign nature of most cases of antenatal hydronephrosis, but close follow-up is required to pick up children needing pyeloplasty
Historical significance/comments	Excellent natural history series

Key Summary Points

1. Congenital hydronephrosis is the most common cause of a palpable neonatal abdominal mass, of which ureteropelvic junction obstruction is the most likely cause.
2. Not all dilatation seen on ultrasound and thought to be due to ureteropelvic junction obstruction requires surgery. Split function below 40%, a decrease in split function by 10%, poor gravity-assisted drainage and delayed tissue transit time on renogram as well as symptoms are indications for surgery.
3. True ureteropelvic junction obstruction must be attended by worsening dilatation on sonar and declining renal scan function.
4. An open dismembered pyeloplasty (Anderson-Hynes technique) offers excellent surgical outcomes.

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