CHAPTER 89
Congenital Ureteropelvic Junction Stenosis

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Introduction

Congenital ureteropelvic junction (UPJ) stenosis is a disturbance in the flow of urine at the crossover point from the kidney pelvis to the ureter, with a dilatated and obstructed renal-pelvic calyceal system (Figure 89.1). UPJ obstruction is the most frequent congenital malformation of the genitourinary system. Its incidence is around 1 out of 1,000 newborns.

The first sonographically demonstrable sign of URJ stenosis in early gestation is a dilatation of the renal-pelvic calyceal system. In part, this can be observed sonographically, in varying degrees, in the 18th to 20th week of gestation.

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 of the genitourinary system or other organ systems. 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.

More frequent are further anomalies in the area of the urinary tract, for example, kidneys with two urinary track systems, horseshoe kidneys, mega-ureters, urethral valves, or contralateral multicystic renal dysplasia.

Demographics

The male-to-female ratio of congenital UPJ stenosis is 2:1. The left side is significantly more frequently affected in isolated cases than the right side. In 30% of the cases, both kidneys are affected. The literature does not indicate a racial predilection in UPJ stenosis.

Aetiology

At present, the cause of congenital UPJ stenosis has not been clearly determined. Extrinsic and intrinsic stenoses are distinguished. Extrinsic UPJ stenoses are caused, for example, by accessory polar vessels crossing the pelviureteric junction or by pressure exerted by an external tumour. The intrinsic form is characterised by an aetiologically unclear congenital disturbance in the texture of the ureter wall, which forms a stenotic fibrosis at the pelviureteric junction. This is regularly shown at histological investigation. A further cause of an intrinsic obstruction is a rare mucous membrane polyp at the pelviureteric junction.

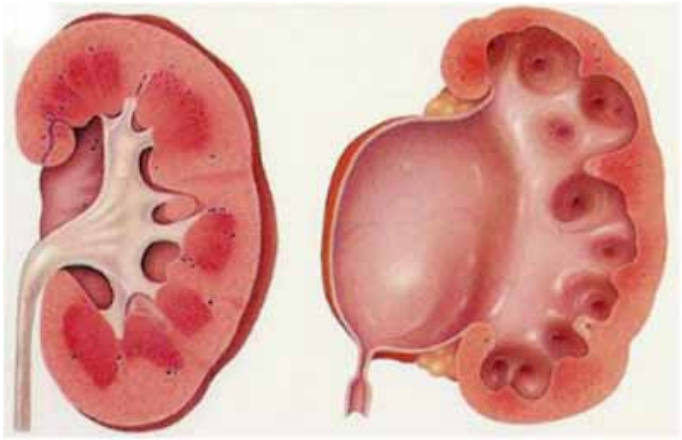
Pathophysiology

The obstruction of the ureter results in a backflow of urine into the proximal urinary tract. When there is a functionally effective stenosis, it leads at first to a progressive dilatation of the renal pelvis and its calyxes. If this dilatation exerts a specific pressure on the renal parenchyma, the wall of the renal pelvis and the blood vessels for a certain amount of time, the result is, at first, a reversible but in time irreversible loss of kidney function 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.

Prenatal Diagnosis

Prenatal ultrasound (US) can show reliably a dilatation of the renal pelvis at the 16th to 20th week of gestation (Figure 89.2), and can be used to follow the development of the dilatation during the course of pregnancy. The most frequent prenatal foetal abnormality is an isolated renal pelvis dilatation. Not every prenatal dilatation of the kidney results in a pathological condition, however. Many low-level prenatal dilatations of the renal pelvis can no longer be detected postnatally.

On the basis of many investigations, a prognostic limit for the prenatal renal pelvis width has been determined. The limit is around 10–11 mm for the posterior-anterior diameter of the renal pelvis. In general, prenatal dilatations below this value do not result in pathological conditions requiring postnatal treatment. Renal pelvis



Normal renal pelvis **UPJ stenosis**

Figure 89.1: Normal and stenosed ureteropelvic junction.

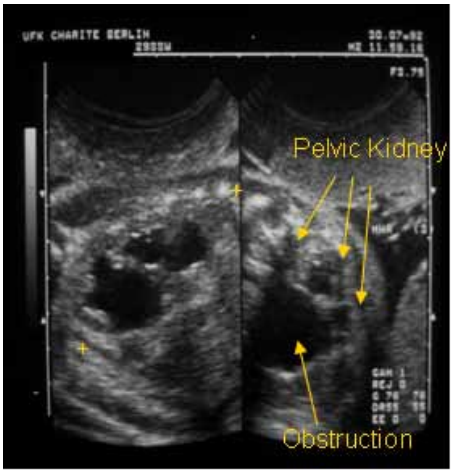


Figure 89.2: Prenatal ultrasound image of UPJ stenosis at the 16th (left) and 20th (right) week; renal pelvis diameter >10 mm.

dilatations during pregnancy that clearly exceed this limit and remain beyond it urgently require postnatal observation and treatment. In these cases, diagnosis frequently shows the presence of an UPJ stenosis requiring surgery.[1–4]

Clinical Symptomatology

The clinical symptoms of a congenital UPJ stenosis present a broad spectrum, depending upon the point in time of diagnosis. The majority of stenoses of the UPJ diagnosed prenatally by means of US show no symptoms either clinically or paraclinically. The newborns show no clinical signs of hydronephrosis. The diagnostic indication here is a conspicuous postnatal ultrasound image of the kidney associated with a prenatally determined dilatation of the renal pelvis.

In the age of prenatal diagnostics, general symptoms, such as poor growth, refusal to feed, recurring vomiting, and agitation, are usually not observed in connection with the stenosis because treatment begins early enough; these symptoms are often observed only in connection with an infection of the urinary tract. If US examination is not possible, infections of the urinary tract are usually indicative and should lead to further diagnostic procedures. Due to early diagnosis and the resulting possibility of timely treatment with antibiotics, the incidence of urinary tract infections has declined significantly.

In cases where a UPJ stenosis is not diagnosed prenatally or neonatally, symptoms other than the general one mentioned above arise during the course of the disease: characteristic pains in the area of the kidneys, haematuria (in 30% of late diagnoses), palpable intraabdominal masses, and infections of the urinary tract. 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.[5,6]

Diagnostics

Clinical Diagnostics

As with all illnesses, prior to imaging diagnostics, a thorough medical history and physical examination of the child have to be undertaken. Particular attention should be paid here to the urination history and the family history. There is no known hereditary chromosomal influence in UPJ obstruction, although there is familial clustering. Furthermore, there are no typical physical malformations that accompany UPJ obstruction. Complete blood and urine tests have to be carried out to determine or exclude any accompanying nephrological or urological conditions or an infection.

Imaging Diagnostics

The goal of imaging diagnosis in children is to obtain reliable results while causing the least possible stress to the child. The following imaging procedures are available for routine diagnosis of a disturbance in the transport system for urine:

- ultrasound;
- voiding cystourethrography (VCUG);
- renal function scintigraphy; and
- uro-magnetic resonance imagery (uro-MRI).

In recent years, excretion urography has been used only for special examinations due to its high level of x-ray exposure.

Diagnostic Procedure

Generally, the following investigations provide a reliable basis for deciding how to proceed with treatment:

- *clinical and paraclinical examination:* medical history, current status, blood and urine tests;
- *ultrasound investigation:* morphology of the urinary tract as initial examination and as a basis for future observation;
- *VCUG:* to exclude a vesicoureteral reflux; and
- *renal function scintigraphy:* technetium-99m mercaptuacetlytriglysine (T-99m MAG3) scintigraphy to evaluate functional processes such as renal perfusion and the renal function of both sides separately, as well as to evaluate an existing obstruction and use of a furosemide load; and dimercaptosuccinic acid (DMSA) scintigraphy to evaluate perfusion, especially after recurring kidney infections.[7–9]

A UPJ obstruction requiring surgery is present when US regularly shows a renal pelvis dilatation of more than 10–15 mm, with progressive dilatation. In general, infections of the urinary tract in cases of UPJ obstruction arise late, so they do not represent an isolated indication for surgery.

The decisive investigation is renal function scintigraphy, which not only shows the renal function for both sides, but is especially important for determining the flow relationships in the urinary tract collection system. In the case of an obstruction, Figure 89.3 shows a mounting curve on the affected side. It is important to carry out the T-99m MAG3 scintigraphy investigation together with a furosemide investigation.

The main reason for VCUG is to exclude a vesicoureteral reflux, which is associated with a significantly higher incidence of urinary tract infection (UTI). The US image can show a dilatation of the renal pelvis with a mega-ureter. The scintigram in Figure 89.3 shows no obstructive influence on the urinary flow.

There are, however, cases with combined malformations showing a UPJ obstruction and a vesicoureteral reflux on the ipsilateral side.

A UPJ obstruction requiring surgery shows a clear dilatation of the renal pelvis and an obstructive mounting curve with functional scintigraphy on an accompanying furosemide investigation. With timely diagnosis and treatment, the normal division of renal function can be maintained. When there is a clear obstruction, corrective surgery should be carried out; without waiting until there are possible signs of a loss of renal function in the affected kidney shown in a follow-up scintigram. Multiple studies have demonstrated that renal function does not improve, even after normal urinary flow conditions have been restored.

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 uro-MRI can be employed.

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 in Figure 89.4.

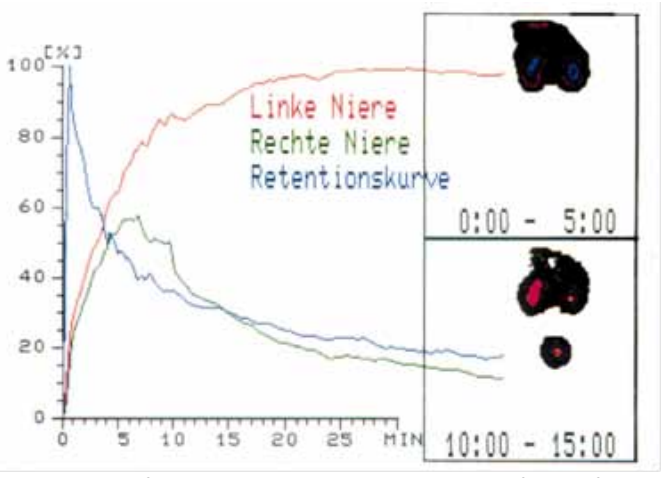


Figure 89.3: Renal function scintigraphy with a mounting curve for the left kidney, showing one obstructed curve (T-99m MAG3, furosemide investigation).

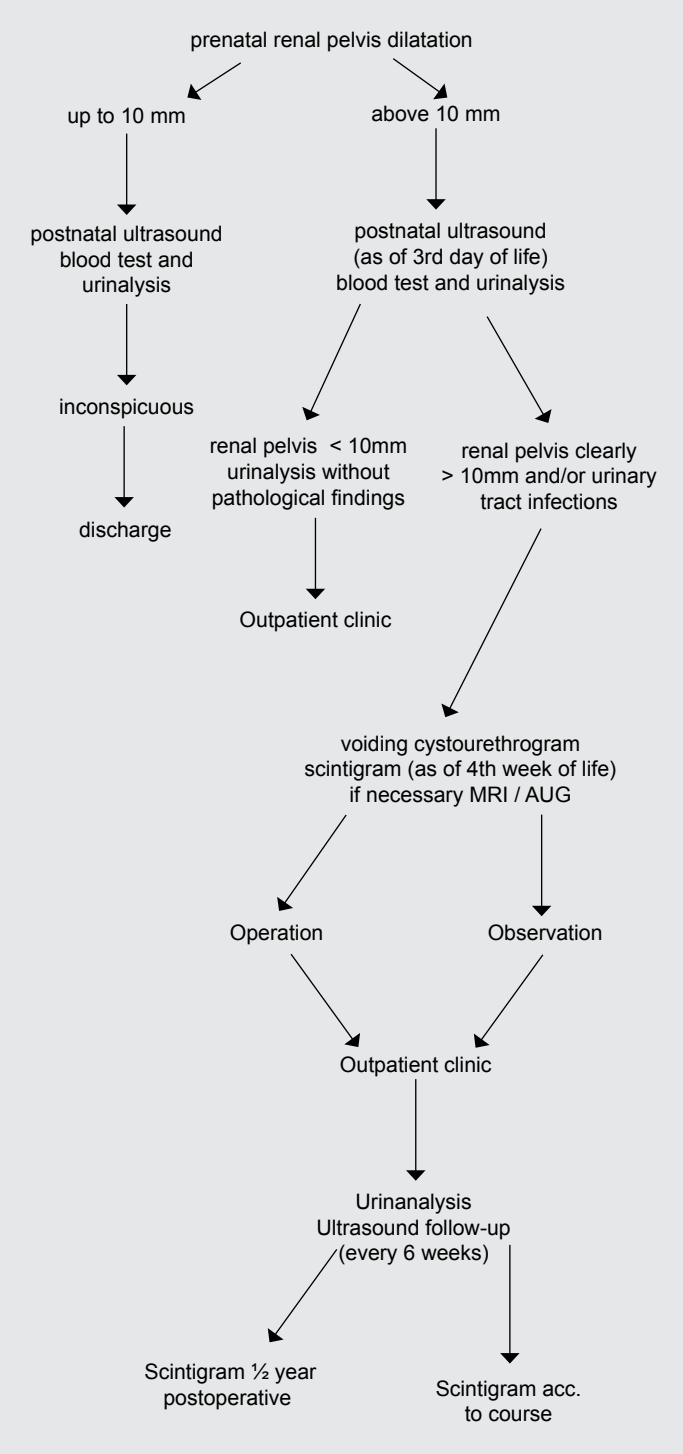


Figure 89.4. Algorithm for the diagnosis of UPJ stenosis.

Treatment of a Congenital Ureteropelvic Junction Obstruction

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

All moderate renal pelvis dilatations for which the clinical, paraclinical, and imaging diagnoses have shown no further pathological condition beyond the dilatation—in particular, no obstruction with or without a loss of renal function—should be kept under regular US observation in an outpatient clinic. In addition, regular urine and blood tests are necessary. Should UTIs occur, or should the US show an increase in the renal-pelvis calyceal system dilatation, a new series of urological imaging investigations should be carried out.

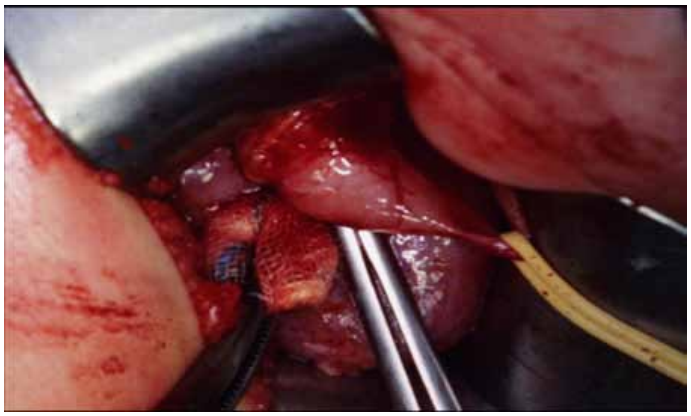


Figure 89.5: Surgical site intraoperative. The goal of all plastic-reconstructive surgical methods is the removal of the hindrance to the urinary flow with retention of the organ.

The goal of a temporary, percutaneous nephrostomy is immediate relief of the kidney, which is usually infected and painful due to the hydronephrosis. 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 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 Figure 89.5, 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[10,11]—have gained acceptance. Common to all resectional methods is 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 standard. It can be carried out both in open surgery and—with older children, from around age 24 months—laparoscopically.[10,11] Once there is a surgical indication, the operation should be carried out regardless of the age of the child. Figures 89.6 to 89.8 show the essential steps in the Anderson-Hynes procedure.

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 anatomosis is sutured continuously microsurgically by using thin, absorbable suture material corresponding to the age of the child (e.g., vicryl 6-0 or 7-0).

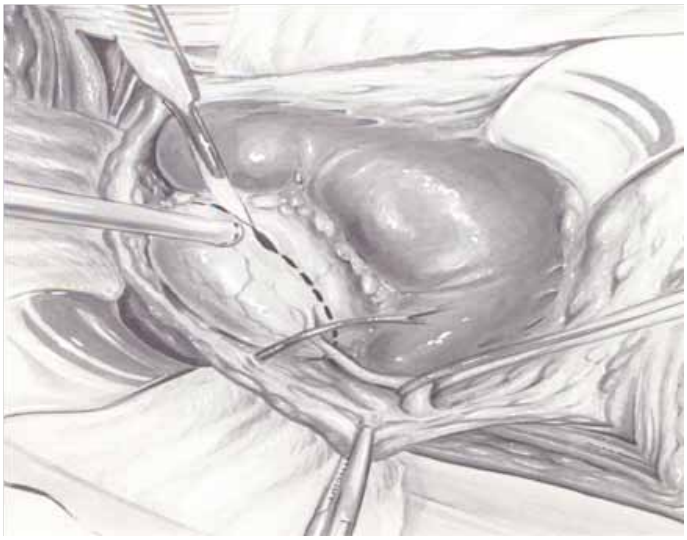


Figure 89.6: Resection line renal pelvis, ureter identified by ligature pelvic junction obstruction.

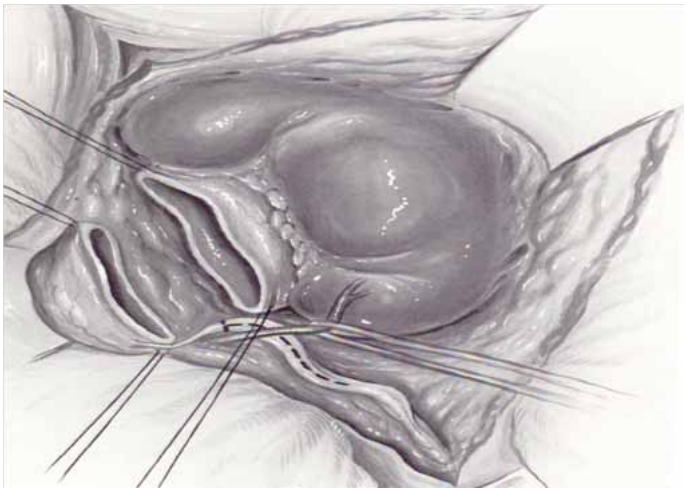


Figure 89.7: Resection line: ureter-renal pelvis cut through.

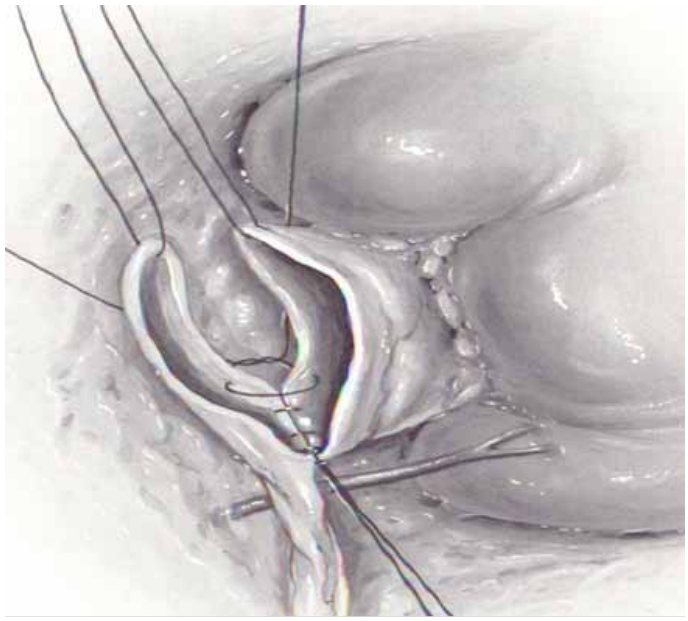


Figure 89.8: Continuous ureter-renal pelvis anastomosis with inserted pyelostomy.

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 7th and 10th 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 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.[12,13]

Postoperative Complications

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

Intraoperative insertion of drain serves to reduce pressure postoperatively in the area of the anastomosis and can prevent a backflow 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 antibiotic sensitivity or by using a wide-spectrum antibiotic such as cefuroxime, adapted for weight.

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 pelviureteric 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 to 6 weeks. If no complications arise, the time between follow-up examinations can be extended.[14–19]

Concluding Comments

Congenital UPJ stenosis is a pathological condition of the genitourinary system that can be detected early on 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.

Evidence-Based Research

Table 89.1 presents a landmark retrospective review of 1,000 children with hydronephrosis. Table 89.2 presents a natural history series of children with severe antenatal hydronephrosis managed nonoperatively.

Table 89.1: Evidence-based research.

Title	Prenatally diagnosed hydronephrosis: the Great Ormond Street experience
Authors	Dhillon HK
Institution	Great Ormond Street Hospital for Sick Children, London, United Kingdom
Reference	Br J Urol 1998; 81(suppl 2):39–44
Problem	Natural history series of 1,000 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 89.2: 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 a ureteropelvic junction obstruction requires 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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