



Ureteropelvic Junction Obstruction

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Abstract

Ureteropelvic junction obstruction (UPJO) is the most common cause of postnatal hydronephrosis. However, hydronephrosis now is usually diagnosed antenatally and affects approximately 1 in 1500 live births. Ultrasonographic scanning is the imaging modality of choice to diagnose hydronephrosis but is unable to differentiate between obstructed and nonobstructed kidneys. Mercaptoacetyl triglycine (MAG3) scanning is essential to distinguish an obstructed renal pelvis from a renal pelvis that is dilated but otherwise normal. Occasionally, vesicoureteric reflux may give a similar picture. Reflux can usually be ruled out with a well-performed MAG3 scan, but if not, micturating cystourethrography should be performed.

There is no consensus on how to manage patients with UPJO and when to treat them conservatively or surgically. Some indications for surgery include <40% differential function of the hydronephrotic kidney on MAG3 scanning, a >20-mm anterior-posterior diameter of the renal pelvis on ultrasonographic scan, pain, and infection.

Pyeloplasty is the gold-standard treatment if surgery is indicated. This procedure can be open, laparoscopic, or robot-assisted. Endopyelotomy and ureterocalicostomy have also been performed in children with some success.

There is also no consensus on how to follow up patients who have had surgery. Some guidelines recommend 2–3-yr follow-up with ultrasonographic and MAG3 scanning, and if stable, the patient then should be discharged. Follow-up of patients who have conservative treatment must be more rigorous to avoid deterioration of the kidneys.

There is increasing interest in using different diagnostic modalities, including urinary markers and magnetic resonance urography, in the diagnosis of UPJO and in correlating the findings with the best treatment option.

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1. Definition

Ureteropelvic junction obstruction (UPJO), or *pelviureteric junction obstruction*, is defined as a blockage or obstruction of urine flow from the kidney into the proximal upper ureter. This obstruction can lead to an increase in back-pressure on the kidney, hydronephrosis, and progressive

damage to the kidney function. It is therefore important to understand how to diagnose and treat this condition.

2. Epidemiology

The most common causes of antenatal hydronephrosis are either transient or physiologic. UPJO is the most common

pathologic cause of antenatal hydronephrosis and occurs sporadically in 1 in 750–1500 live births, but familial inheritance has been reported [1]. Other causes of antenatal hydronephrosis are listed in Table 1 [2].

UPJO has a ratio of 2:1 in boys compared with girls, and the left side is affected in approximately two-thirds of cases. The condition occurs bilaterally in 10–46% of cases [3].

3. Etiology

There are several theories about the development of UPJO, which may be congenital or acquired. The causes can be divided into intrinsic and extrinsic.

Intrinsic causes of UPJO include (1) scarring of ureteric valves, which results in stenosis and a decrease in nerve terminals [4], and (2) ureteric hypoplasia resulting in smooth muscle discontinuity and replacement with collagen. This process disrupts ureteric peristalsis through the abnormal segment [5]. These causes are thought to be a result of inadequate recanalization in utero at 10–12 wk of gestation. From a molecular point of view, improper innervation with decreased synaptic vesicles and abnormal smooth musculature may be the cause. Several growth factors have been implicated, including protein gene product 9.5 (a general neuronal marker), synaptophysin (a neuromuscular junction marker), and nerve growth factor receptor resulting in decreased nerve growth factor messenger RNA (mRNA) expression [6]. Increased transforming growth factor (TGF)- β 1 mRNA expression and decreased epidermal growth factor (EGF) mRNA expression in the stenotic tissue have also been found [7].

Extrinsic causes of UPJO include the following:

- A crossing lower pole renal vessel (aberrant, accessory, or early branching) causing an impingement on the ureter and obstructing flow—this characteristic can occur in $\leq 40\%$ of cases, although it may be in addition to an intrinsic UPJO rather than the only cause; anterior crossing vessels are more common than posterior ones [8]
- Congenital abnormalities of the kidney, such as horseshoe kidneys or duplex kidneys [9]
- Scar formation secondary to ureteric manipulation by surgery
- Fibroepithelial polyps (a rare cause of UPJO) [10].

4. Pathophysiology of upper tract obstruction

Obstruction of the upper tracts in the acute phase leads to an increase in ureteric and renal pelvic pressures and renal blood flow. As ureteric pressure continues to rise, the renal pelvis dilates and renal blood flow decreases as a result of efferent arteriole vasoconstriction. In the long run, the ureteric pressure falls and renal blood flow decreases because of afferent arteriole vasoconstriction, leading to a decrease in the overall glomerular filtration rate. The dilation of the renal pelvis dampens the effect of the increase in pressure and results in tubular dilation, glomerulosclerosis, inflammation, and fibrosis of the kidney secondary to UPJO. There is an increase in fibronectin, type 4

Table 1 – Causes of antenatal hydronephrosis [2]

Frequency, %	Abnormality
48	Transient
15	Physiologic
11	Pelviureteric junction obstruction
4	Vesicoureteric reflux
4	Megaureter, obstructed or nonobstructed
2	Multicystic kidneys
2	Ureterocele
1	Posterior urethral valves

collagen, laminin, and expression of the B-cell CLL/lymphoma 2 gene (*BCL2*) at the intrafascicular space of smooth muscle and the matrix of stroma [11].

5. Presentation

Most hydronephroses are diagnosed antenatally using ultrasonographic scans at 18–20 wk. Prior to the advent of ultrasonographic scanning, the most common presentation of UPJO was pain, especially with excessive drinking. Urinary tract infections that may have progressed to pyonephrosis were sometimes seen with an end-stage kidney, especially in the elderly. In children, infection in UPJO is rare unless there is coincident reflux. Some children may present with an abdominal mass or hematuria following a minor trauma. Finally, some hydronephroses only come to light as an incidental finding when investigating for a cause of abdominal pain [3].

A special diagnostic dilemma has arisen in the last 20 yr with the finding of unilateral or bilateral hydronephrosis in the fetus in an otherwise normal pregnancy, which is now the most common presentation. Providing there is no evidence of oligohydramnios, the pregnancy is allowed to continue to term, and the baby's condition is investigated further after delivery.

6. Investigations

The main aim of an investigation is to diagnose obstruction to aid in planning treatment of the kidneys, which are likely to deteriorate in terms of function. The currently available methods to diagnose obstruction are not sensitive or specific enough.

6.1. Ultrasonography

During the antenatal period, at 16–20 wk, ultrasonography is performed to assess the amniotic fluid volume to rule out oligohydramnios or any associated abnormalities and to measure bladder volume, kidney size, and the anteroposterior diameter (APD) of the renal pelvis. The most sensitive time for urinary tract evaluation is 28 wk. Ultrasonographic scanning cannot assess the degree of obstruction but only the presence or absence of hydronephrosis. Attempts have been made to use ultrasonography to predict obstruction and the need for surgery. The APD is one parameter used and should not be >6 mm. The most sensitive, but least

Table 2 – Society of Fetal Ultrasound hydronephrosis grading system

Grade	Pattern of renal sinus splitting
Grade 0	No splitting
Grade 1	Urine in pelvis barely splits sinus
Grade 2	Urine fills <i>extrarenal</i> pelvis and <i>major</i> calyces dilated
Grade 3	SFU grade 2 and <i>minor</i> calyces uniformly dilated and <i>parenchyma</i> preserved
Grade 4	SFU grade 3 and <i>thin</i> parenchyma

SFU = Society of Fetal Ultrasound.

specific, criteria for the diagnosis of fetal hydronephrosis are the following [12]: (1) calyceal dilation of grade 2 or greater using the Society of Fetal Ultrasound (SFU) scoring system (Table 2), (2) a renal pelvis diameter >4 mm at <33 wk of gestation and >7 mm at >33 wk of gestation, and (3) a renal pelvis-to-kidney ratio of >0.28 . Clinically significant obstruction is more likely if grade 3 or 4 hydronephrosis is present, the renal pelvis diameter is >10 mm, or the renal pelvis-to-kidney ratio is >0.5 [13].

The renal parenchyma–pelviccalyceal area has also been used. If the ratio is <1.6 , there is good correlation with an obstruction and the need for pyeloplasty, whereas patients with a ratio >1.6 can be observed [14].

The resistive index (RI) can also be used with duplex Doppler ultrasonography. RI is defined as the peak systolic velocity minus the lowest diastolic velocity divided by the peak systolic velocity. Patients with an $RI \geq 0.75$ have an obstructive pattern on diuretic renography [15].

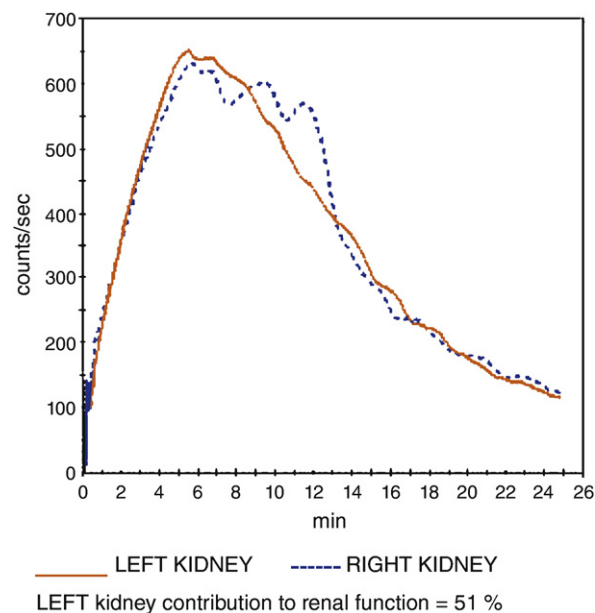
Following delivery, ultrasonographic scanning is performed within 48–72 h. If performed earlier, false-negative values are obtained because of neonatal dehydration and physiologic oliguria. If there is bilateral hydronephrosis, a solitary kidney, or oligohydramnios antenatally, immediate ultrasonography is performed. The ultrasonographic scan is used to assess the APD, pelviccalyceal dilation, renal cortical thinning, ureteric dilation, kidney size, cortical echogenicity, bladder wall, and residual urine.

If postnatal ultrasonography does not show any hydronephrosis, the test should be repeated after 4–6 wk.

6.2. Diuretic renography

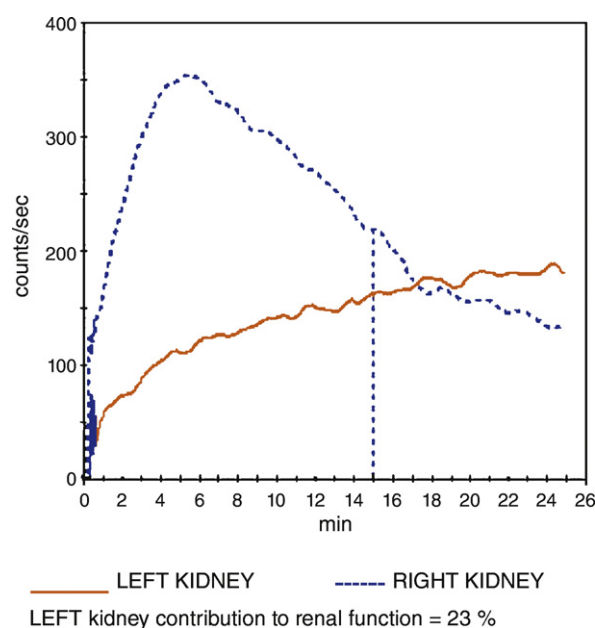
Diuretic renography is the most commonly used diagnostic tool to assess for obstruction. Technetium Tc 99m MAG3 is the radiopharmaceutical agent of choice for this purpose and has largely replaced Tc 99m diethylenetriamine penta-acetic acid (DTPA). MAG3 has a better gamma image than DTPA, as well as a faster clearance rate and lower background activity. The advantage of DTPA is that it can be used to measure the glomerular filtration rate.

MAG3 scanning will provide differential renal function by comparing isotope uptake in the two kidneys, which in turn is a reflection of renal blood flow. Renogram curves looking at uptake and drainage of MAG3 have been defined by O'Reilly et al [16]. Normally, the time required for clearance of 50% of the accumulated radionuclide ($t_{1/2}$) is <10 min, while a $t_{1/2}$ of >20 min is suggestive, but not

**Fig. 1 – Type 1 renogram.**

diagnostic, of obstruction. Four types of renogram curves are defined:

- Type 1: normal uptake with prompt washout (Fig. 1)
- Type 2: a rising uptake curve with no response to diuretics, which suggests obstruction (Fig. 2)
- Type 3a: an initially rising curve that falls rapidly in response to diuretics, which suggests nonobstructive dilatation (Fig. 3)
- Type 3b: an initially rising curve that neither falls promptly nor continues to rise (equivocal).

**Fig. 2 – Type 2 renogram of left kidney (40 mg frusemide administered intravenously at 15 min).**

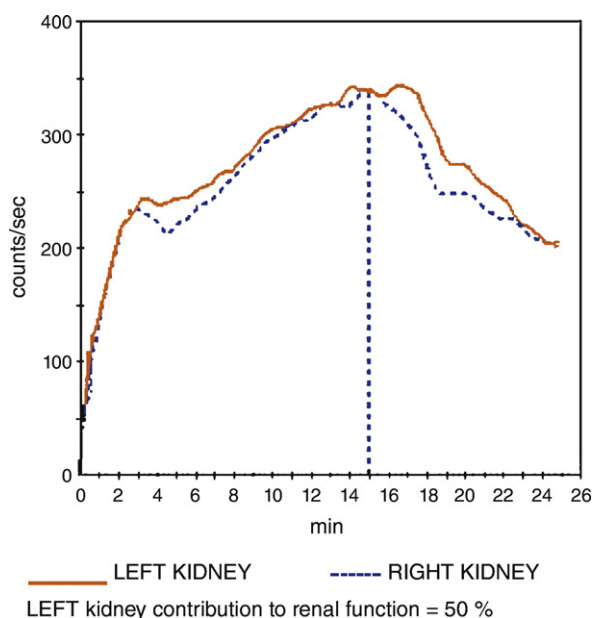


Fig. 3 – Type 3a renogram bilaterally (5 mg furosemide administered intravenously at 15 min).

In practice, the curves can be difficult to interpret, and the test may need to be repeated with a diuretic given 15 min prior to the start of an assessment for possible obstruction.

It is important that a MAG3 scan be performed under standard conditions in the fourth to sixth weeks of life. These conditions include adequate oral hydration prior to the test and infusion of normal saline intravenously at a rate

of 15 ml/kg over 30 min and then at a maintenance rate of 4 ml/kg per hour throughout the whole investigation. The recommended dose of furosemide is 1 mg/kg for infants aged <1 yr and 0.5 mg/kg for individuals aged 1–16 yr, with a maximum dose of 40 mg. A urethral catheter may also need to be inserted in some cases. MAG3 scanning can also be combined with indirect radionuclide cystography to detect reflux in any child, but ideally in children who are potty trained [17].

6.3. Voiding/micturating cystourethrogram

The voiding/micturating cystourethrogram helps exclude other causes of upper tract dilation, including vesicoureteric reflux (VUR), urethral valves, and ureteroceles. VUR coexists with UPJO in 8–14% of cases [2]. VUR, however, may not be clinically significant, so this test is not always recommended, and parents should be counseled about it [18]. The indications for a voiding cystourethrogram include bilateral hydronephrosis (or solitary kidney), duplex kidney, small kidney, abnormal echogenicity, dilated ureter, ureterocele, suspected infravesical obstruction, and abnormal bladder [19].

6.4. Other imaging modalities

Computed tomography (CT) and magnetic resonance imaging (MRI) are the newer modalities and have been used in some centers [20]. CT is used in the trauma setting but is limited by radiation exposure and is not required for the “routine” cases. MRI can evaluate renal blood flow, anatomy, and urinary excretion but is limited by cost, the

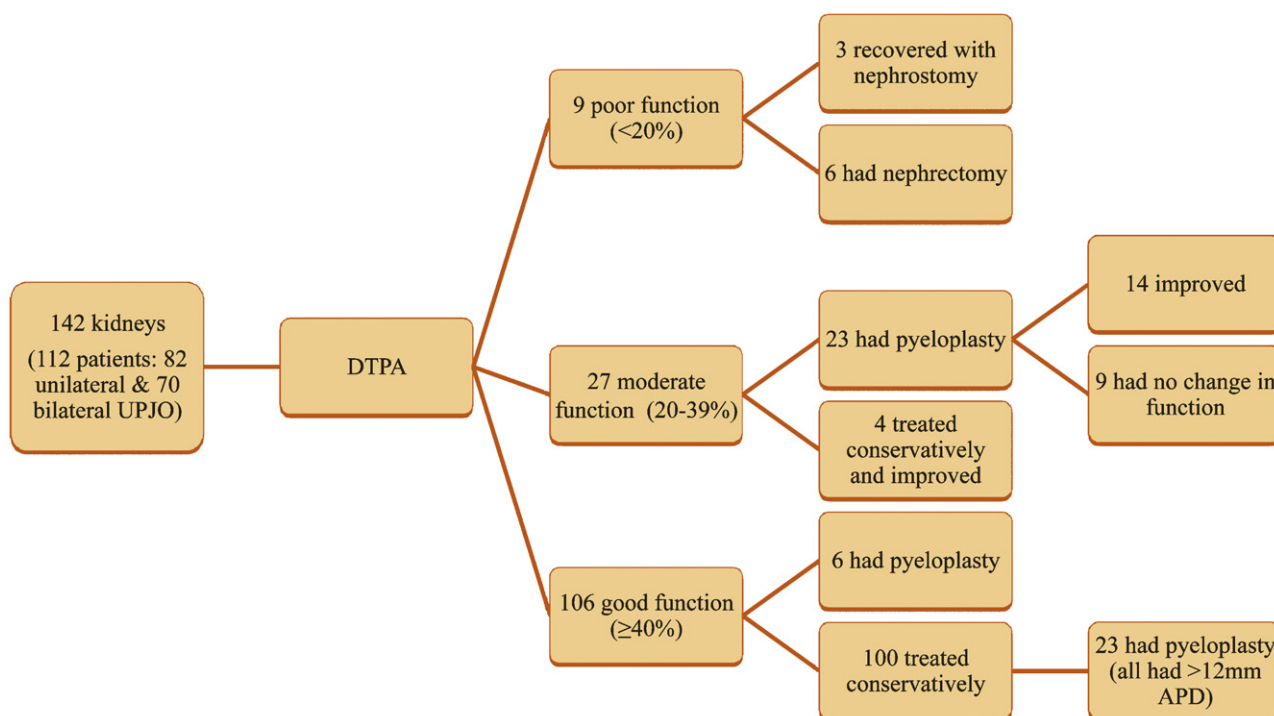


Fig. 4 – Outcome of treated patients with ureteropelvic junction obstruction (UPJO) based on diethylenetriamine penta-acetic acid (DTPA) results. APD = anteroposterior diameter.

need for sedation in infants, and the higher frequency of adverse reactions to the contrast agent. However, magnetic resonance urography may become the standard test in the future.

The Whitaker test, an antegrade pressure/flow study, has been used in equivocal cases of obstruction. The test is not recommended for children because it is invasive and requires anesthesia to insert the pressure lines [21].

6.5. Biochemical parameters

Some studies are looking at urinary biomarkers to define obstruction in hydronephrotic kidneys [22]. These biomarkers include urinary EGF produced by tubular cells, monocyte chemotactic protein-1, TGF-beta 1, β 2-microglobulin, carbohydrate antigen 19-9, urinary kidney injury molecule-1, and neutrophil gelatinase-associated lipocalin [23,24]. These biomarkers are not widely available, and larger studies are required to assess their use.

7. Treatment

The aims of treatment are to prevent deterioration of renal function and relieve pain (if present). The difficulty lies in determining which kidneys need surgical treatment. The natural history of UPJO is not clearly defined, and using the investigative modalities previously described, it is not possible to fully agree on a treatment algorithm. Ransley et al showed that 23% of kidneys with >40% differential function on the hydronephrotic side needed pyeloplasty, with the rest being treated conservatively (Fig. 4) [25]. Conservative treatment measures, therefore, can be used if the differential renal function of the obstructed kidney is >40% and the APD is <12 mm [26].

Antibiotic prophylaxis—such as trimethoprim, 1–2 mg/kg at night, or cephalexin, 5 mg/kg at night—is started in infants with antenatal hydronephrosis until VUR has been excluded. The indications for surgical intervention include (1) pain and infection, (2) asymptomatic obstruction with a differential function <35–40% and an APD >19 mm [27], (3) failure of conservative management resulting in >10% deterioration of renal function, and (4) grade 3 or 4 dilation as defined by the SFU. Poor surgical prognostic factors include (1) renal function <30% [28], (2) APD >50 mm and dilated calyces [29], and (3) progressive hydronephrosis on two consecutive ultrasonographic scans [30].

7.1. Surgical options

Since the first description of the dismembered pyeloplasty by Anderson and Hynes in 1949 for the management of retrocaval ureter [31], open reconstructive surgery has been considered to be the gold standard for the treatment of UPJO [32]. Other procedures have been aimed at reducing the size of the scar (laparoscopic procedures performing the same reconstruction) or avoiding a scar altogether (endoscopic procedures). The question, then, is how much deterioration in outcomes is acceptable to achieve these goals. It is important to remember that with all reconstructions, the

first operation is the easiest, and subsequent operations will be hampered by the effects of a failed first one.

Surgical options include the following procedures:

- Pyeloplasty, the gold standard treatment of a UPJO, may be a dismembered Anderson-Hynes [33], Culp, or Foley Y-V pyeloplasty. This treatment can be used in long strictures, in severe hydronephrosis, or in the presence of crossing vessels [34]. Open pyeloplasty can be approached through a lumbotomy incision, an incision above the 12th rib, or an anterior abdominal wall incision [35]. The success rate is $\geq 95\%$, and the procedure has stood the test of time. Laparoscopic pyeloplasty (retroperitoneal or intraperitoneal) is technically challenging in children and has a $\leq 95\%$ success rate in the best hands [36]. Pyeloplasty also can be performed robotically [37].
- Endopyelotomy has an approximately 80% success rate but only in the absence of a crossing vessel. The JJ stent must stay in situ for 6 wk postoperatively. The stricture must be <1.5 cm long. This option may also be used in failed pyeloplasty [38]. Antegrade endopyelotomy requires access to the kidney through a midpole posterior calyx. The stricture is incised using a cold knife, electrocautery, or contact laser fiber [39]. Retrograde endopyelotomy is performed using a 5F ureteric cutting balloon catheter [40]. Ureterocalicostomy involves the ureter being detached from the renal pelvis and anastomosed directly to the most dependent lower pole calyx. This option may be useful in recurrent UPJO [41].
- Simple nephrectomy (open, laparoscopic, robotic) is an option in patients with <10–15% differential function of the hydronephrotic kidney or association with another pathology, such as renal tumors.
- An internal ureteric stent is rarely used, as it does not provide a definitive treatment and is only a temporizing measure.
- Nephrostomy is an option, especially if pyonephrosis is present.
- Endopyeloplasty involves the horizontal suturing of a standard vertical endopyelotomy incision performed through a percutaneous tract via a 26F nephroscope [42]. This option is rarely used.

7.2. Complications of surgery

Complications of pyeloplasty include urinary tract infections, pyelonephritis, urinary extravasation and leakage, recurrent UPJO, and stricture formation. Minor urinary extravasation can be treated conservatively initially for 10–14 d. If this treatment fails or if the extravasation is large, a JJ stent or nephrostomy tube is inserted. Of recurrent UPJO and/or strictures, 2–5% will need to be treated with further surgery, be it redo pyeloplasty, endopyelotomy, or ureterocalicostomy [43].

Complications of endopyelotomy include significant intraoperative bleeding if the endoscopic incision is made inadvertently into a major polar vessel (treated immediately with arteriography and embolization if there is hypotension), postoperative infection, and recurrence of obstruction.

8. Follow-up

Open pyeloplasty is considered a curative procedure. If drainage from the kidney is normal on a renogram at 1 yr, patients may be discharged from further follow-up [44]. There is no obvious reason why the same protocol should not be applied to patients having a pyeloplasty by a minimally invasive route, but data are lacking [36]. A renal ultrasonographic scan is obtained 6 wk after pyeloplasty or

after stent removal to ensure that the hydronephrosis is resolving.

There are fewer data available for the long-term follow-up of patients having endoscopic procedures. Long-term imaging may be performed at 2–3 yr to look for the rare situation of delayed cicatrization and restenosis of the ureteropelvic junction [45].

If a conservative approach is followed for UPJO, careful follow-up is required. This follow-up includes repeat

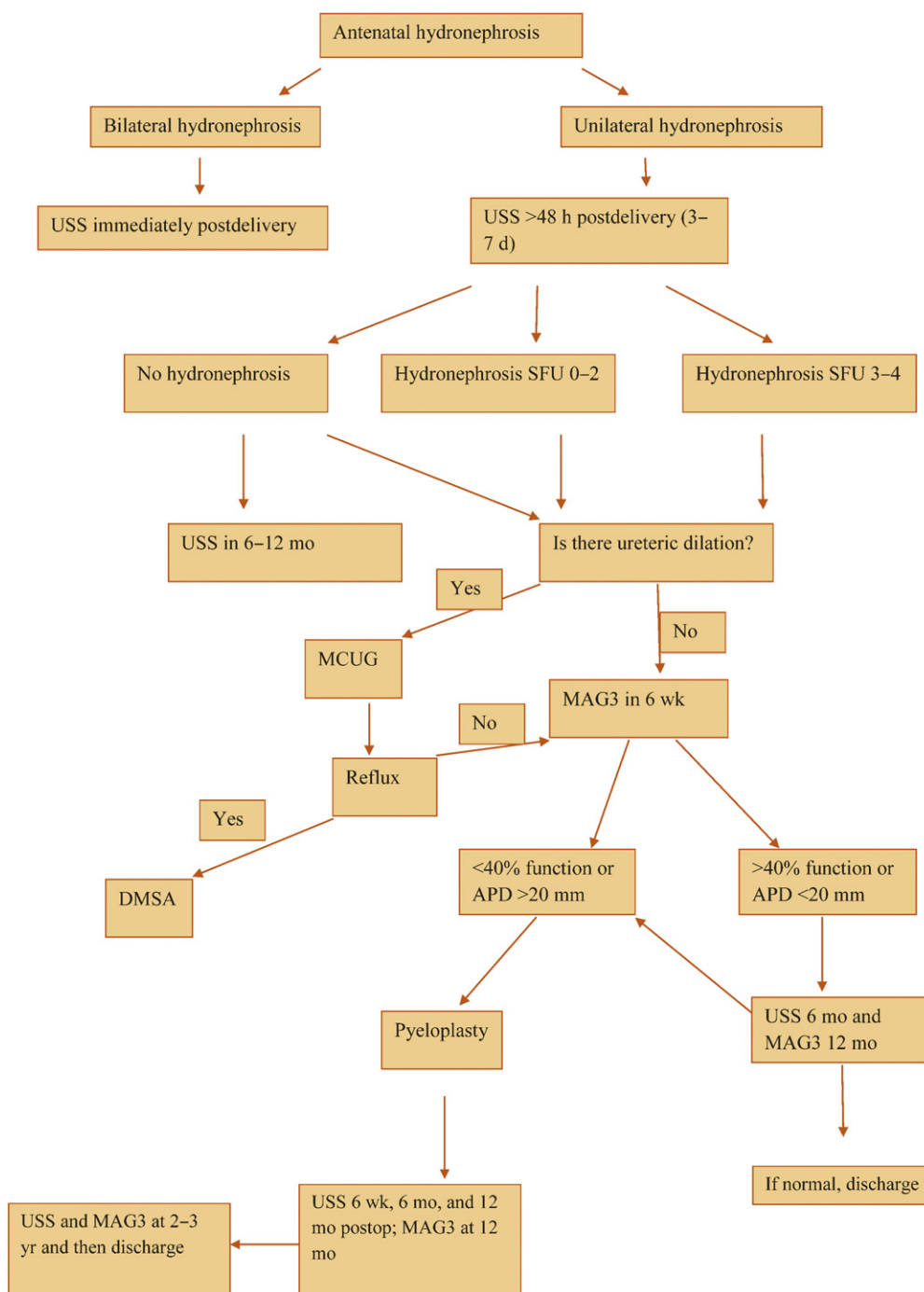


Fig. 5 – Suggested algorithm for the treatment of ureteropelvic junction obstruction.

APD = anteroposterior diameter; DMSA = dimercaptosuccinic acid; MAG3 = mercaptoacetyl triglycine; MCUG = micturating cystourethrogram; SFU = Society of Fetal Ultrasound; USS = ultrasonographic scan.

ultrasonography in 1–3 mo and repeat scintigraphy in 3–12 mo [46].

After 6 mo to 1 yr, MAG3 and renal ultrasonographic scans are obtained to provide a relative assessment of the overall renal function. There is no consensus on the long-term follow-up of infants and children with asymptomatic hydronephrosis and a differential function of >40%. However, there are some data to suggest that there is a 40–45% overall probability that conservative management will be successful, with complete resolution or sustained improvement in the obstruction and a 5% probability of stable but significant persisting dilatation, at the age of 16 yr [47].

9. Conclusions

Further studies are required with better methods of diagnosing obstruction to aid in the management decision concerning who needs conservative treatment and who needs surgery. These methods may include measurements of urinary growth factors or better imaging modalities such as magnetic resonance urography. Long-term experience with newer surgical modalities, such as robot-assisted pyeloplasty, is also required. A universal algorithm for the treatment of UPJO is required. Figure 5 shows an algorithm that we have devised.

Conflicts of interest

The authors have nothing to disclose.

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