Hydronephrosis and Hydroureteronephrosis

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1. Introduction

Hydronephrosis is a common reason for a patient to be referred to a pediatric urologist. A prenatal diagnosis of hydronephrosis is made in 1-5% of all pregnancies, so many referrals occur prenatally or during early infancy. [1,2,3,4] Hydronephrosis can be associated with ureteral dilation; if ureteral dilation is present, it is called hydroureteronephrosis. This chapter will review the differential diagnoses, evaluation, and management of hydronephrosis and hydroureteronephrosis. Hydronephrosis can also first be identified in older children, and we will discuss this clinical situation including surgical repair of ureteropelvic junction (UPJ) obstruction with pyeloplasty. Some specific topics related to hydronephrosis including vesicoureteral reflux (VUR), posterior urethral valves (PUV), and neurogenic bladder are discussed in more depth in other sections.

Definitions

Hydronephrosis is dilation of the renal pelvis and/or calyces. Some amount of dilation of the renal pelvis is considered normal, while any dilation of the calyces is considered abnormal (**Table 1**).[5] Hydronephrosis is measured or graded several ways by pediatric urologists and pediatric radiologists, and practices vary.[6,7] The anterior-posterior renal pelvis diameter (APRPD) is measured on a transverse ultrasound image that shows the maximal diameter of the intra-renal pelvis (**Figure 1**). Note that the APRPD measurement does not consider the degree of calyceal dilation.

Table 1: Normal findings on prenatal or postnatal ultrasound

	Time at presentation		
Findings	16-27 weeks	≥28 weeks	Postnatal (>48 hours)
APRPD	<4 mm	<7 mm	<10 mm
Calyceal dilation	None	None	None
Parenchymal thickness	Normal	Normal	Normal
Parenchymal appearance	Normal	Normal	Normal
Ureter	Normal	Normal	Normal
Bladder	Normal	Normal	Normal
Unexplained oligohydramnios	No	No	n/a
Adapted from Nguyen et al 2014.[5]			

The Society for Fetal Urology (SFU) grading system is a common system used by pediatric urologists and is based on subjective assessment of location of hydronephrosis (pelvis vs calyces), how severe or uniform the calyceal dilation is, and whether the renal parenchyma appears normal or not (**Table 2**, **Figure 2**).[8] Based on amount of renal pelvis dilation (**Table 1**), some SFU grade 1 hydronephrosis can be considered normal or physiologic.

Table 2: Society for Fetal Urology hydronephrosis grading system

SFU Grade 0	No urine seen in renal pelvis or calyces
SFU Grade 1	Urine causes slight separation of renal pelvis
SFU Grade 2	Renal pelvis is further dilated and a single or a few calyces may be visualized
SFU Grade 3	Renal pelvis and all calyces are dilated. Renal parenchyma is normal thickness.
SFU Grade 4	Renal pelvis and all calyces are dilated. Renal parenchyma is thinned.

Adapted from Ferbach et al 1993.[8]

The urinary tract dilation (UTD) classification system is a newer classification system that considers the degree of hydronephrosis as well as bladder and ureteral abnormalities (**Table 3**, **Figure 2**).[5] The UTD classification system categorizes prenatal (antenatal) hydronephrosis as UTD A1 (low risk) or UTD A2-3 (increased risk). The “A” in A1-A3 indicates antenatal. Postnatal findings are categorized into UTD P1 (low risk), UTD P2 (intermediate risk), and UTD P3 (high risk). The “P” in P1-P3 indicates postnatal. The SFU and UTD classification systems are similar if there is only hydronephrosis and no hydroureter or abnormal bladder, with SFU 1-2 corresponding to UTD P1, SFU 3 corresponding to UTD P2, and SFU 4 corresponding to UTD P3 (**Figure 2**). Note that the APRPD may affect the UTD grade.

Table 3: UTD classification system

Antenatal presentation	*Postnatal presentation (initial ultrasound > 48 hours)*			
UTD A1 (low risk)	UTD A2-3 (increased risk)	UTD P1 (low risk)	UTD P2 (intermediate risk)	UTD P3 (high risk)
APRPD 4 to <7mm for 16-27 weeks	APRPD ≥7mm for 16-27 weeks	APRPD 10 to <15 mm	APRPD ≥15 mm	APRPD ≥15 mm
APRPD 7 to <10mm for ≥ 28 weeks	APRPD ≥10mm for ≥28 weeks	Central calyceal dilation	Peripheral calyceal dilation	Peripheral calyceal dilation
Central or no calyceal dilation	Peripheral calyceal dilation		Ureters abnormal	Parenchymal thickness abnormal
	Parenchymal thickness abnormal			Parenchymal appearance abnormal
	Parenchymal appearance abnormal			Ureters abnormal

	Ureters abnormal			Bladder abnormal
	Bladder abnormal			
	Unexplained oligohydramnios			

Adapted from Nguyen et al 2014.[5]

The definition of hydroureter is not well established in the literature however, the ureteral diameter in children typically does not exceed 5 mm. Work by Cussen examining the ureteral dimensions in fetuses at 30 weeks gestation showed the upper limit of the range of the diameter was 0.5-0.65.[4] This study was later supported by work performed by Hellstrom, studying excretory urograms in children.[3] Thus, one definition of ureteral dilation which is supported by the British Association of Paedriatric Urologists (BAPU) consensus statement on the management of the primary obstructive megaureter is to consider ureteral dilation of **7 mm** or more in the retrovesical area as abnormal.[2]

The term “megaureter” is not a diagnosis but simply describes ureteral dilation without specifying the cause. The terms **primary obstructive megaureter** or **primary megaureter** is often used to specifically refer to a dilated ureter with an adynamic distal segment and ureterovesical junction (UVJ) obstruction. Therefore when discussing ureteral dilation, hydroureteronephrosis is a more general and appropriate term to use until the underlying pathology is known. Several pathologies can result in hydroureteronephrosis. Ureteral dilation can be primary or secondary and can be grouped into categories as showed in **Figure 1** and as defined by King.[1] The most common causes of megaureter are vesicoureteral reflux (VUR) and primary obstructed megaureter from an adynamic ureteral segment at the UVJ. Rarely, a megaureter can be simultaneously obstructed and refluxing. This typically occurs when there is an ectopic ureter to the bladder neck or sphincter area. With voiding, the ureter will open allowing VUR, but once voiding ends and the bladder neck and/or sphincter contracts, the ureter is again obstructed.

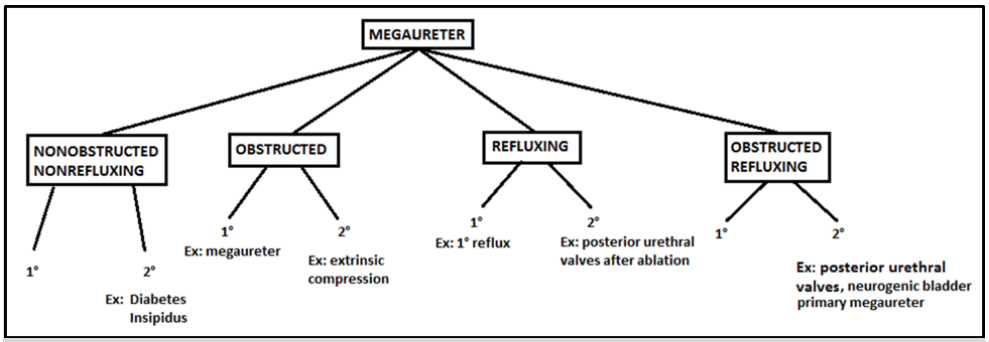

Figure 1: Clinical classification of megaureter

In addition to an adynamic segment of ureter causing UVJ obstruction, ureteral dilation from obstruction can also be caused by an ectopic ureter or a ureterocele. An ectopic ureter is any ureter, single or duplex, that does not enter the trigonal area of the bladder. In a duplex system, this is usually the upper pole ureter, presumably because of its budding from the mesonephric duct later than the lower pole with later incorporation into the developing urogenital sinus. In females, the ectopic ureter may enter anywhere from the bladder neck to the perineum and into the vagina, uterus, and even rectum. In males, the ectopic ureter always enters the urogenital system above the external sphincter or pelvic floor. An ectopic ureter in males can enter the Wolffian structures, including vas deferens, seminal vesicles, or ejaculatory duct. **Therefore, ectopic ureters can present with urinary incontinence in females but not in males.**

Ureteroceles represent a version of the ectopic ureter with a cystic dilation of the distal aspect of the

ureter that is located either within the bladder or spanning the bladder neck and urethra. As with the ectopic ureter, ureteroceles may be associated with a single or duplex system, and in duplex systems are associated with the upper pole. When ureteroceles are associated with a single system, this can result in hydroureteronephrosis of the entire system. When ureteroceles are associated with the upper pole of a duplicated system, this can result in hydroureteronephrosis of the upper pole. However, if ureteroceles are very large, they can obstruct the bladder neck and make bladder emptying difficult and present with lower pole hydronephrosis in a duplicated system or even with contralateral hydronephrosis. Ureteroceles that affect all renal units should prompt some urgency in management, initially typically with a puncture, as they can pose significant risk of urinary tract infection and even renal insufficiency.

Ureteroceles can be classified as intravesical versus extravesical. The intravesical ureterocele is entirely within the bladder and above the bladder neck. Ectopic, extravesical, ureteroceles include those in which some portion of the ureterocele is situated permanently at the bladder neck or in the urethra. An important distinction should be made between the extravesical ureterocele and the intravesical ureterocele that prolapses into the urethra with voiding, as these are two quite different entities with possible implications for treatment. [5] Another notable classification was described by Stephens et al and separates ureteroceles into cecoureterocele, stenotic, sphincteric, sphincterostenotic, blind, and nonobstructed. [6]

3. Differential Diagnoses

When seeing an infant with hydronephrosis or a pregnant mother whose fetus has hydronephrosis, one of the questions you may be asked by the family is what is causing the hydronephrosis. The majority (50-75%) of prenatal hydronephrosis will be mild (SFU grade 1-2, UTD A1) and have a ~90% chance of being transient hydronephrosis that will resolve spontaneously.[4,9,10,11,12] The more moderate and severe cases of prenatal hydronephrosis (SFU grade 3-4, UTD A2-3) have higher chance of being a UPJ obstruction or another diagnosis and going on to have surgery. [9,10,11,12]

In males with bilateral hydronephrosis, a diagnosis of posterior urethral valves (PUV) needs to be considered especially for cases with severe bilateral hydronephrosis, hydroureter, and dilated bladder. PUV is covered in more detail in another chapter. In patients with hydroureter diagnoses such as ureterocele, ectopic ureter and megaureter need to be considered. Vesicoureteral reflux (VUR) can often be found in children with hydronephrosis, although in many cases VUR occurs with hydroureter as well. **Table 4** lists the differential diagnosis in relative order of likelihood for hydronephrosis based on mild (SFU 1-2) and moderate/severe (SFU 3-4) as well as associated hydroureter or bladder abnormalities.

Table 4: Differential diagnosis based on severity of prenatal hydronephrosis and presence of hydroureter or abnormal bladder

Mild: SFU 1-2 or UTD A1	*Mod/severe: SFU grade 3-4 or UTD A2-3*	*Hydroureter*	*Abnormal bladder*
Transient hydronephrosis (~90%)	UPJ obstruction (~40%)	Transient hydronephrosis (non-obstructive megaureter)	Posterior urethral valves
Vesicoureteral reflux (~5%)	Transient hydronephrosis (~30%)	Vesicoureteral reflux	High grade VUR (can lead to large bladder)
UPJ obstruction (~5%)	Vesicoureteral Reflux (~10%)	UVJ obstruction or other form of ureteral obstruction (e.g. ureterocele or ectopic ureter)	Prune belly syndrome
UVJ obstruction (~1%)	PUV (~5%)	PUV (especially if bilateral hydroureter)	Neurogenic bladder
PUV (<0.5%)	UVJ obstruction (~10%)	Other cause such as prune belly syndrome	Other form of bladder outlet obstruction (e.g. urethral atresia)

Adapted from Lee et al 2006.[3]

4. Prenatal Evaluation

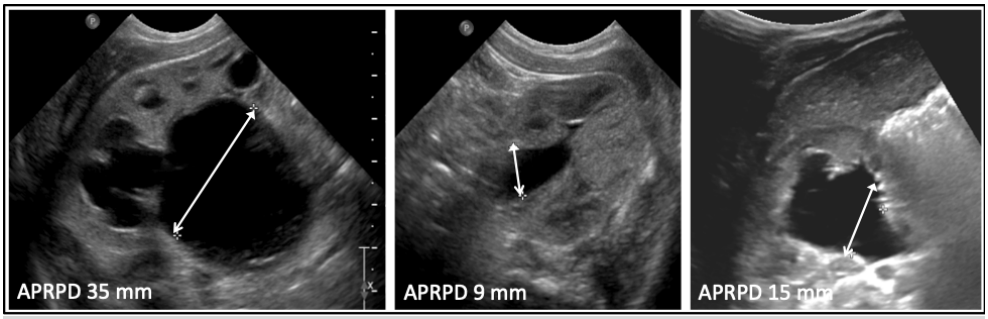

Figure 2

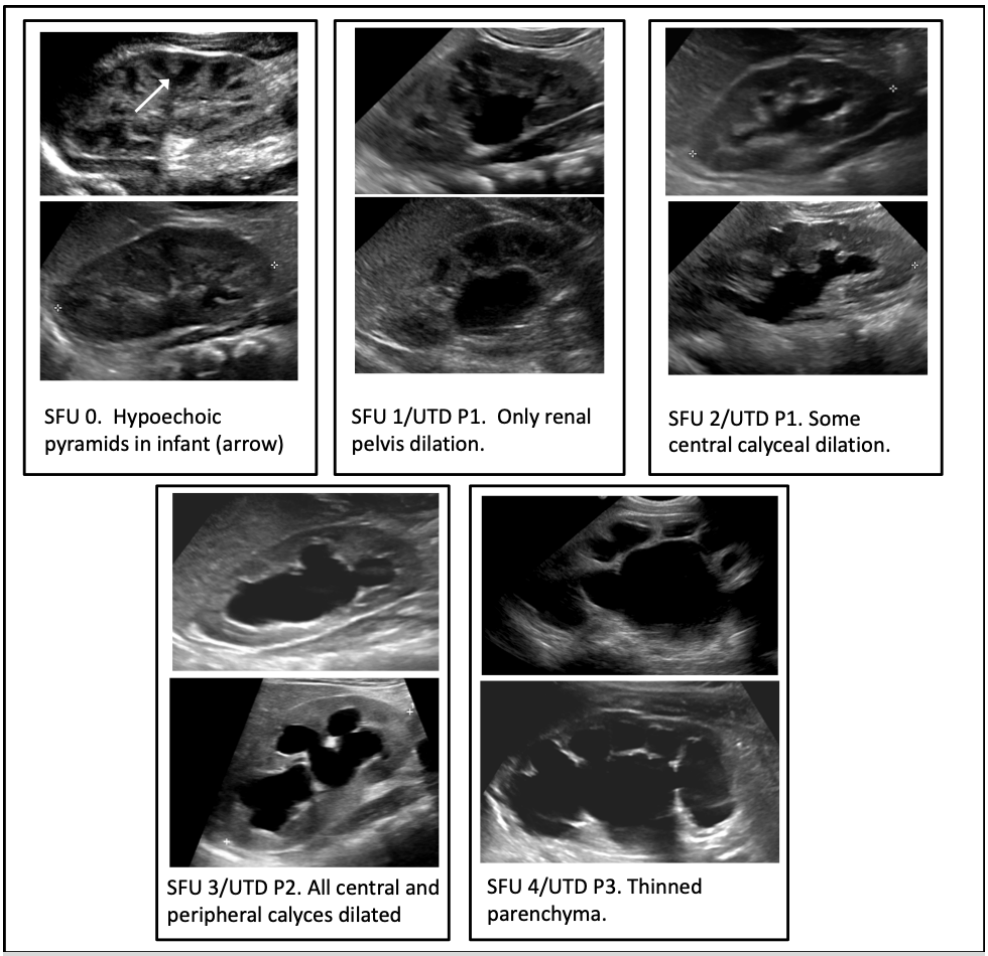

Figure 3

For most pregnancies, a routine anatomical ultrasound is obtained around 20 weeks of gestation. National practice guidelines in the United States include evaluation of kidneys and bladder as a required component. [13] If a fetus at 20 weeks gestation has an APRPD ≥ 4mm or calyceal dilation, they are considered to have prenatal hydronephrosis.[5] Prenatal hydronephrosis may resolve before birth, worsen, or remain stable. Milder forms of prenatal hydronephrosis (4-8 mm APRPD) have been reported to resolve before birth 80% of the time. [10] It is also important to note on the prenatal ultrasound if amniotic fluids levels are normal, low (oligohydramnios), or absent (anhydramnios). Most of the amniotic fluid is from fetal urine production, and a normal amount of amniotic fluid is essential

for fetal lung development. Low levels of amniotic fluid would suggest an obstruction along the fetal urinary tract. In addition, ureteral dilation, bladder abnormalities, or any other congenital anomalies are important to note on the prenatal ultrasound.

Prenatal hydronephrosis has been associated with some genetic syndromes such as trisomy 21, especially in fetuses with other associated anomalies.[14,15]

The likelihood ratio of isolated prenatal hydronephrosis for increasing posttest probability of trisomy 21 has been estimated to be around 1.5-2.5.[14,15]

Therefore, the vast majority of fetuses (>99%) with isolated prenatal hydronephrosis will not have trisomy 21.[15] Some studies suggest that in low risk pregnancies, isolated hydronephrosis should not be an indication for prenatal karyotyping.[4]

In 2014, eight societies (including 2 urology societies) with interest in the management of fetuses and infants with hydronephrosis created guidelines regarding the classification of hydronephrosis as well as the prenatal and postnatal management.[5] Prenatal hydronephrosis was categorized into low risk (UTD A1) and increased risk (UTD A2-3) (**Table 3** and **Figure 2**). If a fetus at 20 weeks has only unilateral hydronephrosis with APRPD <7 mm without any peripheral calyceal dilation, this is considered low risk (UTD A1) and an ultrasound in the 3rd trimester is recommended. If the hydronephrosis has resolved during the 3rd trimester, no addition prenatal or postnatal imaging is recommended. If a fetus has an APRPD ≥ 7 mm, peripheral calyceal dilation, abnormal renal parenchyma, hydroureter, abnormal bladder, or oligohydramnios (UTD A2-3) this was considered increased risk and a follow up ultrasound in 4-6 weeks during gestation was recommended (**Table 3**).

5. Prenatal Intervention

If there is a concern for significant bladder outlet obstruction from PUV or another cause, consideration to prenatal intervention can be given. This is most appropriately done in the setting of a multidisciplinary team in a fetal center consisting of a maternal fetal medicine specialist, pediatric intensivist, pediatric urologist, and pediatric nephrologist ideally in the setting of a prospective study[16] The most common and most studied prenatal intervention is a vesicoamniotic shunt.[17,18] Essentially, this is a tube similar to a double J stent that is inserted through the abdominal wall of the fetus into the bladder. This tube then drains the fetus' bladder into the amniotic cavity. This may improve lung development in fetuses with significant oligohydramnios or anhydramnios potentially leading to improved survival but it is unclear if it improves long term kidney function.[17,18] Problems with vesicoamniotic shunts such as malposition or becoming dislodged are common, and many fetuses will undergo more than one intervention.[17] In addition, there is a risk of preterm labor or intra-uterine death due to procedure.[17] Thus, patients (both mother and fetus) must be carefully selected, as not all cases are candidates for prenatal intervention. Other prenatal interventions such as fetal cystoscopy with valve ablation are experimental and should be undertaken in the setting of a multidisciplinary fetal center participating in a prospective study.[19] PUV including prenatal intervention is covered in more detail in another chapter.

6. Postnatal Evaluation and Management

As mentioned in a previous section, the UTD grading system was developed in 2014 to serve as a guideline for prenatal and postnatal evaluation and management of hydronephrosis.[5] For the vast majority of cases of prenatal hydronephrosis, an initial postnatal renal ultrasound is recommended sometime between 48 hours and 1 month of life. Based on this initial postnatal ultrasound, the hydronephrosis is categorized as low risk (UTD P1), intermediate risk (UTD P2), or high risk (UTD P3) (**Table 3**). For low risk (UTD P1) cases, a follow up ultrasound is recommended in 1-6 months. For intermediate risk (UTD P2) cases a follow up ultrasound is recommended in 1-3 months. In high-risk cases (UTD P3), a voiding cystourethrogram (VCUG) is recommended as well as a renal ultrasound in 1 month. A functional scan such as a DMSA or MAG 3 scan can be considered for intermediate risk or high-risk cases. Often a MAG 3 scan is obtained if the hydronephrosis involves all of the calyces and the parenchyma is abnormally thin (SFU grade 4), and a MAG 3 scan is often considered in cases with normal parenchyma (SFU grade 3) if the hydronephrosis persists or worsens. See **Table 5** for summary of the recommended postnatal evaluation by the multidisciplinary consensus.[5] Note that many decisions are at the discretion of the clinician, and there are not recommendations for long-term follow up.

Table 5: Recommended management after initial postnatal ultrasound according to multidisciplinary consensus

	UTD P1 Low risk	UTD P2 Intermediate risk	UTD P3 High risk
Follow up ultrasound	1 to 6 month	1 to 3 months	1 month
VCUG	Discretion of clinician	Discretion of clinician	Recommended
Prophylactic antibiotics	Discretion of clinician	Discretion of clinician	Recommended
Functional scan such as MAG 3 or DMSA	Not recommended	Discretion of clinician	Discretion of clinician

Adapted from Nguyen et al 2014.[5]

The postnatal evaluation of a solitary kidney with severe hydronephrosis or bilateral severe hydronephrosis with abnormal bladder is different, as the implications of these findings are more serious. More expedient follow-up with early ultrasound, VCUG, and potentially intervention may be needed depending on the clinical scenario. It is very important not to miss cases of PUV, ureteroceles, ectopic ureters, or neurogenic bladder which may need more expedient intervention. These topics are covered in more detail in other sections such as posterior urethral valves (PUV), hydroureteronephrosis, and neurogenic bladder.

7. IMAGING MODALITIES

7.1 Renal ultrasound

The first postnatal renal/bladder ultrasound often provides the anatomic diagnosis and allows visual assessment of renal parenchyma in infants with hydroureteronephrosis. There are no characteristics that permit differentiation between a dilated ectopic ureter and ureterocele above the level of the bladder. The bladder images therefore are critically important because management of ectopic ureter and ureterocele can be very different. Bladder views are typically diagnostic in differentiating ureterocele from ectopic ureter by revealing a thin-walled, cystic dilation within the bladder and not extending beyond its walls. The laterality of the ureterocele is usually apparent but may appear midline if large. Differentiating primary obstructive megaureter from an ectopic ureter by ultrasound can be difficult. In general, the renal parenchyma of a primary obstructive megaureter is more normal appearing while the renal parenchyma for an ectopic ureter is more dysplastic appearing. **Figure 4** shows an example of a duplicated collecting system with an ectopic upper pole ureteral insertion. Note should be made of appearance of ureter on the imaging showing the bladder, which shows the ureter posterior to the bladder contrasting the appearance of a ureterocele which appears as a thin walled intravesical structure.

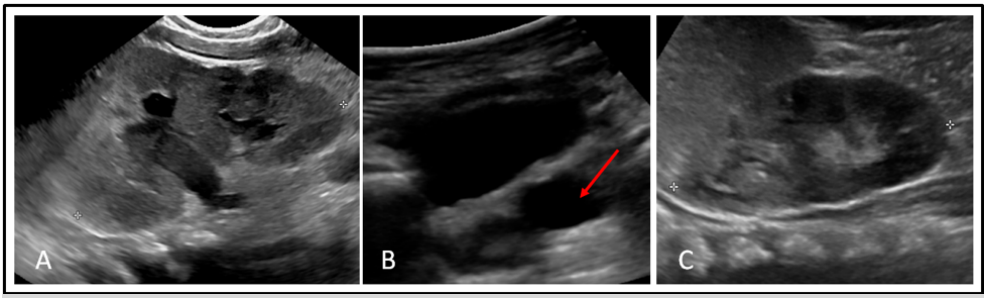

Figure 4: Infant presented with febrile UTI. Renal ultrasound showed duplicated collecting system with upper pole hydroureteronephrosis and mild lower pole hydronephrosis. (Panel A) At the level of the bladder, a dilated ureter (red arrow) is seen posterolateral to the bladder (Panel B) VCUG was normal. The child underwent ureterostomy of upper pole ureter with plans for an upper pole to lower pole ureteroureterostomy at a later date. Post operation renal ultrasound revealed resolution of upper pole

hydronephrosis. (Panel C)

7.2 VCUG

The voiding cystourethrogram (VCUG) provides the most definitive evaluation of the bladder and urethra and is recommended for infants with history of prenatal hydroureter by the AUA guideline statement on VUR.[{{7466}] Society for Fetal Urology consensus statement states that VCUG for hydroureter is at discretion of clinician as long as hydroureteronephrosis is unilateral and there are no bladder or renal parenchymal anomalies on renal/bladder ultrasound.[11] In particular, if a male infant is noted to have bilateral or even unilateral hydroureteronephrosis, an early VCUG (prior to discharge from the hospital) should be strongly considered in order to rule out bladder outlet obstruction from posterior urethral valves. Presence of only unilateral dilation does not rule out bladder outlet obstruction as it has been noted that posterior urethral valves may be associated with only unilateral dilation in up to 14% of patients. [12] A VCUG will also assess for presence of vesicoureteral reflux as the cause of ureteral dilation.

A VCUG should be considered before any intervention such as ureterocele incision to define the baseline anatomy and presence of lower pole or contra lateral VUR. The unusual situation in which a VCUG may not be obtained would be if decompression of a ureterocele that is causing bladder outlet obstruction in an infant is urgently indicated. It is unlikely that the findings on VCUG would alter treatment, which would nearly always be transurethral puncture of the ureterocele. The presence of reflux may determine initial treatment for some practitioners and is an important parameter in clinical management after initial decompression of the ureterocele. In the setting of an ectopic ureter, ipsilateral lower pole reflux is unlikely to resolve spontaneously and will influence definitive treatment options.

7.3 Renal function study (MAG-3)

In the presence of hydroureteronephrosis and absence of bladder outlet obstruction or vesicoureteral reflux, a MAG-3 renal function study is often obtained to assess differential renal function and drainage. The BAPU consensus statement recommends MAG-3 renal function study if ureteral dilation is >10 mm. Mild ureteral dilation can be monitored with renal ultrasound assuming there is no renal parenchymal thinning. When a MAG-3 study is performed, a Foley catheter is generally advisable in order to keep the bladder drained during the procedure and improve the accuracy of the exam. An initial differential function of <40% or a drop of >5% in function between studies is considered abnormal and significant. Care must be taken when interpreting this study and it is useful to draw drainage curve around entire ureter rather than just kidney in order to assess for obstruction at the UVJ. [3] **Figure 5** shows an example of a child with hydroureteronephrosis suggestive of obstructed megaureter in which MAG3 demonstrated obstruction in the left kidney at the level of the ureterovesical junction (red curve in panel E.)

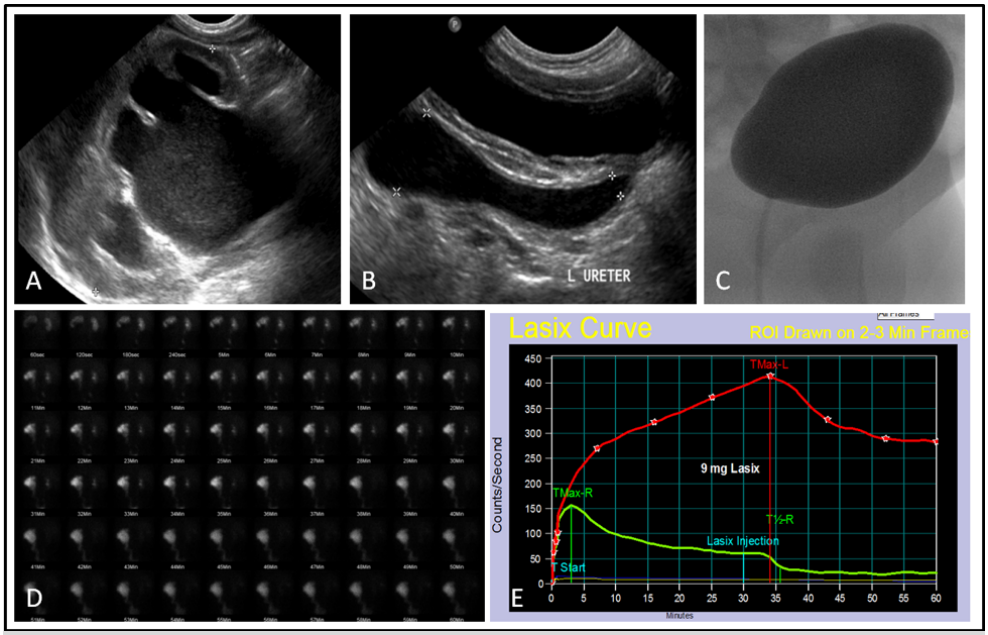

Figure 5: 4 year old child diagnosed with hydroureteronephrosis presented with worsening dilation and febrile UTI (Panel A and B). VCUG was normal (Panel C). Renal function study revealed evidence of obstruction at the Ureterovesical junction (Panel D and E).

7.4 MRI

Magnetic resonance urography can provide the most detailed images of an affected urinary tract. However, MRI in young children often requires sedation or even anesthesia and may not add information not already obtained with less expensive methods. At present the value of MRI rests with patients in whom other imaging cannot define complex anatomy. This may occur in duplicated collecting systems with a non-dilated upper pole, one with massive dilation in which the existence of duplication may be uncertain, or in which anatomic relationships are significantly distorted. If this type of anatomic delineation is needed, MRI offers the addition of functional information that is an important aspect of evaluation. In detecting the presence of an ectopic ureter in incontinence children suspected to have ectopic ureter as the cause of incontinence, MR urography's sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) to be 88.8-100%, 70-90%, 75-88.8% and 90-100% for the detection of ectopic ureter. High resolution MR offers even more detailed anatomy when compared with standard MR urography. [13,14,15]

8. Long Term Postnatal Follow Up

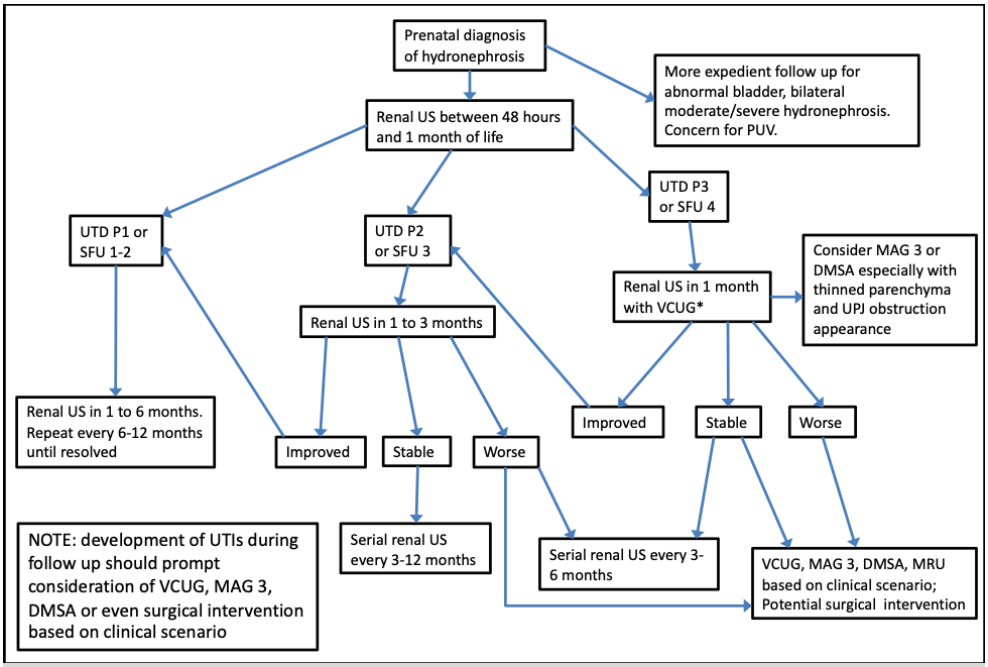

Figure 6: Potential follow up algorithm for prenatal hydronephrosis. Wide variation in practices exist.

After initial postnatal evaluation, patients will continue to follow up in clinic for their hydronephrosis. The general goals of management are to preserve renal function, minimize infection risk, correct obstruction or VUR when appropriate, and achieve urinary continence. While there are no guidelines for long term follow up, some clinical principles can guide follow up protocols. One principle is that mild hydronephrosis in an infant (SFU grade 1-2, UTD P1) has around 90% chance of resolution over the first 2-3 years of life and very low risk of surgical intervention.[{{7466},20] Another principle is that more severe forms of hydronephrosis (SFU grade 3-4, UTD P2-3) are more likely to have issues with UTIs and undergo surgery, especially for UPJ obstruction.[3,21,22] Yet another principle is that it is important to not miss rare diagnoses that can lead to significant morbidity such as PUV, ectopic ureter, neurogenic bladder, and ureterocele.[5] **Figure 6** shows a practical algorithm, but practice varies widely among pediatric urologists.

For milder forms of hydronephrosis (SFU 1-2, UTD P1), typically a follow up ultrasound is recommended 1 to 6 months after the first postnatal ultrasound; if resolved (APRPD <10 mm, no calyceal dilation) this may be the last follow up. If the hydronephrosis is stable (SFU grade 1-2, UTD P1), follow up renal ultrasounds are obtained every 6-12 months until resolution (APRPD <10 mm, no calyceal dilation) or until toilet trained. If mild hydronephrosis (SFU grade 1-2, UTD P1) persists after toilet training, the follow up ultrasounds are typically spaced out or stopped.

For more severe forms of hydronephrosis (SFU grade 3-4, UTD P2-P3), a follow ultrasound is recommended 1-3 months after initial postnatal ultrasound. VCUGs are commonly obtained for SFU grade 4, UTD P3, or significant hydroureter. For severe hydronephrosis that does not resolve or improve, an initial or repeat MAG 3 or DMSA scan is considered. If there is initially <40% differential renal function, a decline in differential renal function, worsening hydronephrosis, or urinary tract

infections a pyeloplasty for UPJ obstruction is considered (see section below on pyeloplasty). For patients with persistent SFU grade 4 hydronephrosis with stable differential function > 40%, many families will elect to proceed with pyeloplasty after no improvement with observation for 2-4 years.

9. Other Management Issues

9.1 Prophylactic antibiotics

The use of prophylactic antibiotics is controversial but is an option in infants with more severe forms of hydronephrosis. The risk of urinary tract infection has been shown to be higher in children with hydroureteronephrosis suggestive of ureterovesical junction obstruction than with children with ureteropelvic junction obstruction and no ureteral dilation. UTIs occur most frequently in the first 6 months of life. Thus, antibiotic prophylaxis may be advisable for the first 6-12 months of life in this patient population.[2,9] The prophylactic antibiotic of choice is amoxicillin and the typical dose is 10-15 mg/kg once daily for the first two months of life. Nitrofurantoin and trimethoprim/sulfamethoxazole are frequently used prophylactic antibiotics, however neither can be used in the immediate postnatal period. Nitrofurantoin cannot be used in the first month of life and trimethoprim/sulfamethoxazole cannot be used until the infant is >2 months of age.There are no randomized, controlled trials that show a benefit to the use of prophylactic antibiotics for infants with hydronephrosis not attributable to VUR. However, there is an ongoing randomized trial on this topic (clinical trials registry number: NC50114516) where infants with SFU grade 3-4 hydronephrosis were randomized to placebo and trimethoprim and results may be available in the near future. There are some observational studies that suggest prophylactic antibiotics are associated with a decreased risk of UTI in patients with severe forms of hydronephrosis (SFU grade 3-4, UTD P3). [23,24] Other potential factors associated with increased risk of UTI include female gender, uncircumcised status, vesicoureteral reflux, and hydroureteronephrosis. [22,23,24] Prophylactic antibiotics should be discussed as an option with the families of children with more severe hydronephrosis or other risk factors such as hydroureteronephrosis. This should also include a discussion about the potential risk of bacterial resistance if a UTI does occur.[25] Prophylactic antibiotics were recommended by the UTD group for UTD P3 or high risk patients.[5] If a VCUG is planned, one option is to use prophylactic antibiotics around the time of VCUG or until a VCUG is obtained as VCUGs do carry a small risk of causing UTI. [26]

9.2 Circumcision

Males who are uncircumcised have approximately a 1% risk of developing a UTI during the 1st year of life, compared to 0.1% for circumcised males.[27,28] Circumcision certainly is an option to consider to decrease risk of UTI for infants with severe hydronephrosis, especially if significant urinary tract anomalies such as PUV are present.[29] However, it is not a requirement, and parental preference should be taken into account.

9.3 Treatment of physiologic phimosis

For children with severe hydronephrosis, significant anomalies such as PUV, or who have had a UTI and whose family does not want circumcision, the use of a steroid cream to release normal physiologic phimosis is an option. Topical steroid management of phimosis has been shown to decrease risk of recurrent UTI in a small randomized study and was associated with decreased risk of recurrent UTI in a retrospective cohort study.[30,31] A common option is betamethasone valerate 0.1% cream BID until the physiologic phimosis has resolved, or up to 2 months.

9.4 Urinary tract infections during follow up

When a child with any degree of hydronephrosis develops a UTI, a VCUG should be considered and discussed with the family. If vesicoureteral reflux is present, a daily prophylactic antibiotic is typically the option discussed with families as first line management. Antireflux procedures such as ureteral reimplant or subureteric injection of bulking agent can be considered if UTIs recur while on prophylactic antibiotics, renal scarring develops, high grade VUR does not resolve, or if family preference is to proceed with antireflux surgery. Please see section on vesicoureteral reflux for more details.

If there is concern for possible UPJ obstruction or UVJ obstruction based on degree of hydronephrosis and a patient develops a UTI, a functional study such as MAG 3 scan or DMSA scan should be considered.

9.5 Complex anatomy

In patients with complex anatomy or unclear diagnosis, an MR urogram (MRU) is being used more commonly as an option to define anatomy.[32] MR urogram can help distinguish ectopic ureter from UVJ obstruction and is being used to assess function and drainage characteristics in UPJ obstruction.[32,33] Cystoscopy can also be helpful in distinguishing UVJ obstruction from ectopic ureter. These topics are covered in more detail in the section on hydroureter.

10. Older children with hydronephrosis

Occasionally, older children will be diagnosed with hydronephrosis. This can be detected incidentally or because of symptoms of flank pain or UTI. The evaluation and management depends on the presentation and ultimate diagnosis. If a patient presents with flank pain, mild hydronephrosis, and hematuria on urine analysis, then a ureteral stone should be considered. If a patient presents with intermittent flank or abdominal pain, severe hydronephrosis, and normal urine analysis, consideration should be given to the possibility of symptomatic UPJ obstruction. Younger children with symptomatic UPJ obstructions often cannot localize the pain and may complain of generalized abdominal pain with nausea, and it is not uncommon to see a younger child with UPJ obstruction to present with a reported history of recurrent gastrointestinal illness. In older children and adolescents, the pain can often be localized to the flank. Some adolescents will present with intermittent flank pain that occurs with intake of large amounts of fluids (such as hydration while playing sports) or with intake of caffeinated beverages, as urine production is increased during those times causing a bolus

of urine passing through a narrowed UPJ segment. The intermittent nature of the pain and hydronephrosis can pose a diagnostic conundrum. Children can have minimal or even no hydronephrosis at the time of pain and MAG3 renal functions studies can be indeterminate for obstruction or even normal. Imaging the kidney in question at the time of pain can aid significantly with diagnosis. If a child presents with acute onset flank pain, nausea and vomiting, especially if brought on by intake of large amount of fluid (all signs of Dietl's crisis) imaging with ultrasound in the acute setting may show worsened hydronephrosis and confirm the diagnosis of UPJ obstruction. More sophisticated imaging such as MRU has also been proposed as being beneficial although often ultrasound is sufficient to make a diagnosis and proceed with therapy/surgery. For symptomatic UPJ obstructions, a pyeloplasty is typically recommended. In older children, an accessory lower pole renal vessel passing anterior to the UPJ ("crossing vessel") is a more common cause of UPJ obstruction than in infants who typically have an intrinsic stenosis of the UPJ. [34] This may explain why many of the older children who present with symptomatic UPJ obstructions do not have a history of prenatal hydronephrosis. Polyps of the ureter, typically identified at the UPJ but can be anywhere along the ureter, may manifest clinically with flank pain or hematuria or by incidental detection of hydronephrosis especially in older children. The most common presentation is one of Dietl's crisis as would be seen in older children with crossing vessels. Polyps are a rare case of UPJ obstruction, accounting for approximately 0.5% of cases. Polyps may be identified on retrograde pyelography or they may not be identified until a pyeloplasty procedure is performed for UPJ obstruction. When polyps are identified at the time of pyeloplasty, [35] one must ensure the ureter is spatulated widely well beyond the site of polyps in order to minimize chance of recurrence. Identification of polyps as the cause of UPJ obstruction or in conjunction with UPJ obstruction does not significantly alter a pyeloplasty procedure. Recurrence of polyps is rare and can often be managed endoscopically.

Late presentation with hydronephrosis and/or UTI can be seen with PUV, VUR, or other anomalies such as ureterocele. In addition, neurogenic bladder from undiagnosed causes such as tethered cord or sacral agenesis can present with hydronephrosis. Severe forms of voiding dysfunction referred to as non-neurogenic neurogenic bladder or Hinman's syndrome also can present with hydronephrosis and other symptoms such as incontinence and UTIs. [36] Rarely ureteral obstruction from malignancies can lead to hydronephrosis.

11. Pyeloplasty for UPJ obstruction

The most common cause of severe prenatal hydronephrosis and severe hydronephrosis in an older child is a UPJ obstruction. Pyeloplasty is the most common treatment for infants and children with UPJ obstruction. While some indications are controversial and practice patterns vary, commonly accepted indications for pyeloplasty commonly considered among pediatric urologists are listed in **Table 6**.[37] Note that specific cut offs for drainage time on MAG 3 scan are not typically used as indications for pyeloplasty by pediatric urologists. Despite the limitations of retrospective series and variable surgical indications; in general, infants with SFU grade 4 (UTD P3) hydronephrosis have around 25-50% chance of undergoing pyeloplasty, those with SFU grade 3 (UTD P2) have a lower

risk (likely 15% or less), and the risk is close to 0% for infants with SFU grade 1-2 (PTD P1). [9,10,21,38,39,40] The most common type of pyeloplasty performed is a dismembered pyeloplasty. This can be performed via several approaches including open, laparoscopic, or laparoscopically with robotic assistance. The robotic assisted laparoscopic pyeloplasty (RALP) is becoming more common over time, including in infants. [41,42] Specific risks of pyeloplasty include urine leak and recurrent obstruction or failure (**Table 6**). Most groups report risk of failure after open pyeloplasty or RALP below 5%. [43]

Table 6: Indications for pyeloplasty and risks of pyeloplasty

Indications for pyeloplasty	*Risks of pyeloplasty*
Recurrent pain crises (typically only seen in older children)	Urine leak
Initial differential renal function < 40%	Recurrent obstruction
Decline in renal function over serial functional scans	Injury to surrounding structures
Urinary tract infections	Major bleeding
Nephrolithiasis	Infection
Worsening hydronephrosis	Injury to ureter
Non-resolving severe hydronephrosis after 2-4 years of observation with stable differential function	

Adapted from Nguyen et al 2014.[5]

The surgical steps of dismembered pyeloplasty include exposure of ureter and renal pelvis with isolation of the UPJ, disconnecting the ureter from the renal pelvis, spatulation of the ureter, and anastomosis of spatulated ureter to dependent portion of renal pelvis. [37] If there is a lower pole crossing vessel, the ureter is brought anterior to the vessel and the anastomosis performed anteriorly to the crossing vessel. If there is a very large redundant renal pelvis, some surgeons will excise the redundant pelvis, especially with open pyeloplasty. The anastomosis should be performed in the most dependent portion of the renal pelvis. Many surgeons will place either an indwelling double J stent into the ureter or a temporary externalized nephroureteral stent. Some surgeons will not place a stent but will leave a drain near the kidney, particularly in open pyeloplasty. The use of double J ureteral stent is more common in RALP than open pyeloplasty but some groups are doing RALP without a ureteral stent or perinephric drain.[44] The use of postoperative antibiotic prophylaxis when stents are used is controversial and practices vary.

An AUA update series on UPJ obstruction and pyeloplasty is available here.

12. Conclusions

The evaluation and management of children with hydronephrosis and hydroureteronephrosis is something that all urologists need to be familiar with, as not all hospitals with newborn nurseries will have pediatric urology coverage. While most cases of prenatal hydronephrosis will be mild and transient, it is important to not miss important diagnoses such as posterior urethral valves and to arrange appropriate follow up evaluation after birth. In the older child with hydronephrosis, appropriate evaluation for UPJ obstruction and other causes is also essential. Since UPJ obstruction is the most common cause of severe hydronephrosis in infants and older children, and because and pyeloplasty is the standard treatment, all urologists should be familiar with this diagnosis and surgery. Infants and children with primary obstructive megaureter, ectopic ureter and ureterocele are often best served by a referral to a pediatric urologist given the complexities and intricacies in diagnosis and management and the need for long term follow up.

Videos

Robotic Pyeloplasty Step By Step: Benjamin R Lee CORE

Robotic ureterocalycostomy: Laura Bukavina et al

Robotic Ureterocalicostomy

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