Pediatrics- Hydronephrosis, Hydroureteronephrosis, and congenital anomalies of GU tract

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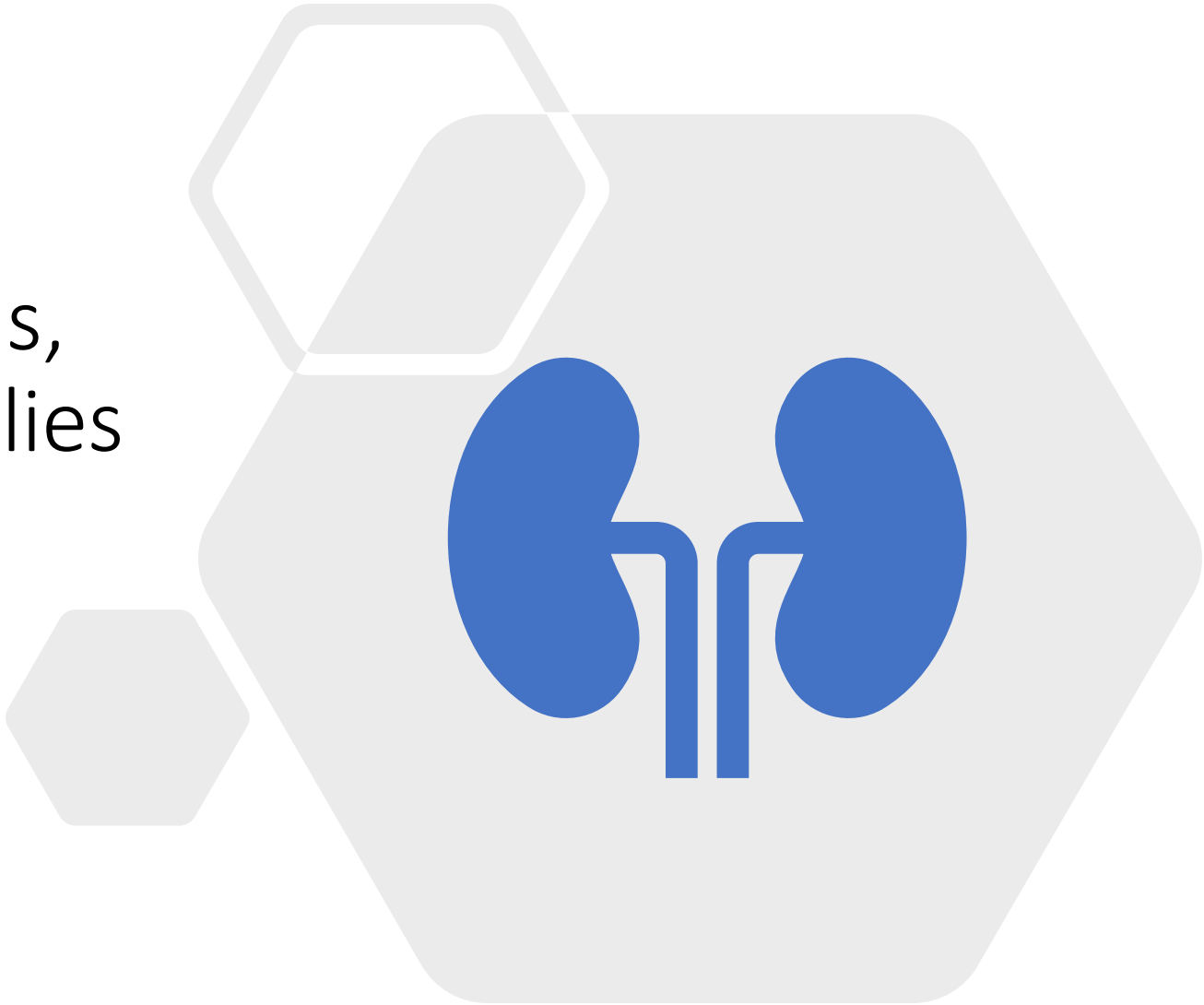
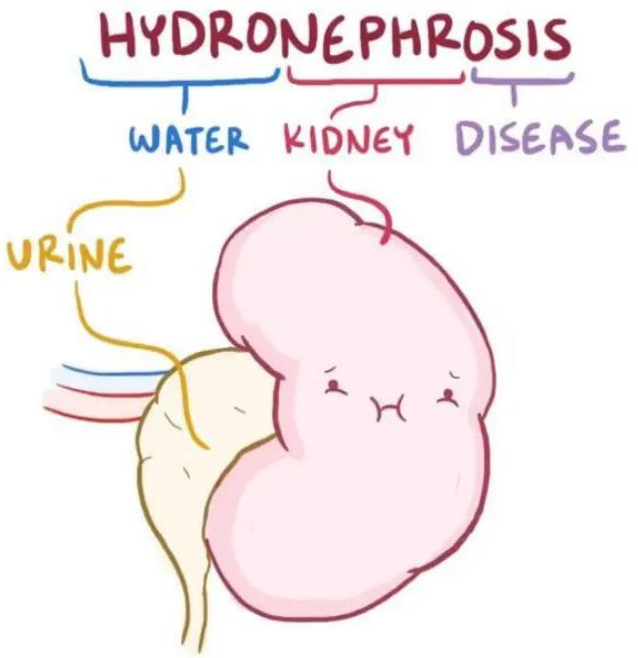


Outline

- **Hydronephrosis**
- **Hydroureteronephrosis/ Megaureter**
- **Ectopic ureter**
- **Ureteroceles**
- **GU anomalies**

Hydronephrosis

- Hydronephrosis is dilation of the renal pelvis +/- calyces.
- Several systems for grading are used in peds



Anterior-posterior renal pelvis diameter

- **Anterior-posterior renal pelvis diameter (APRPD)** is measured on a transverse ultrasound image that shows the maximal diameter of the intra-renal pelvis.
  - does not take into account the degree of calyceal dilation.

Table 1: Normal findings on prenatal or postnatal ultrasound

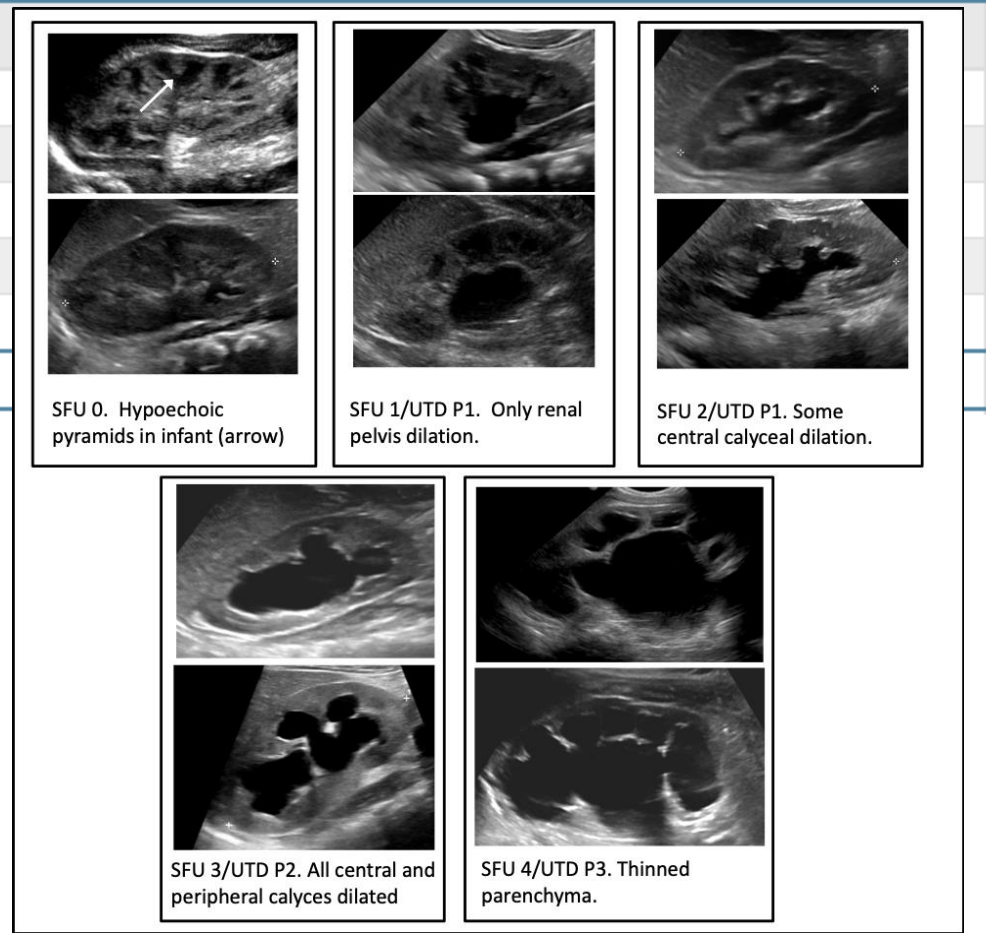
	Time at presentation		
Findings	16-27 weeks	≥28 weeks	Postnatal (>48 hours)
APRPD	<4 mm	<7 mm	<10 mm
Calyceal dilation	None	None	None
Parenchymal thickness	Normal	Normal	Normal
Parenchymal appearance	Normal	Normal	Normal
Ureter	Normal	Normal	Normal
Bladder	Normal	Normal	Normal
Unexplained oligohydramnios	No	No	n/a

Adapted from Nguyen et al 2014.[5]

Society for Fetal Urology grading system

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]



- based on subjective assessment of where the hydronephrosis occurs (pelvis vs calyces), how severe or uniform the calyceal dilation is, and whether the renal parenchyma appears normal or not

Urinary Tract Dilation system

- takes into account degree of hydronephrosis as well as bladder and ureteral abnormalities
- “A” in A1-A3 indicates antenatal.
- “P” in P1-P3 indicates postnatal.

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]

Differential Diagnosis

- Transient hydronephrosis
- UPJ obstruction
- Vesicoureteral reflux*
  - in many cases occurs with hydroureter as well.
- Post urethral valves*
  - Especially consider In males with bilateral hydronephrosis
- Other Bladder outlet obstruction
- Prune bell Syndrome
- Neurogenic bladder

Differential diagnosis

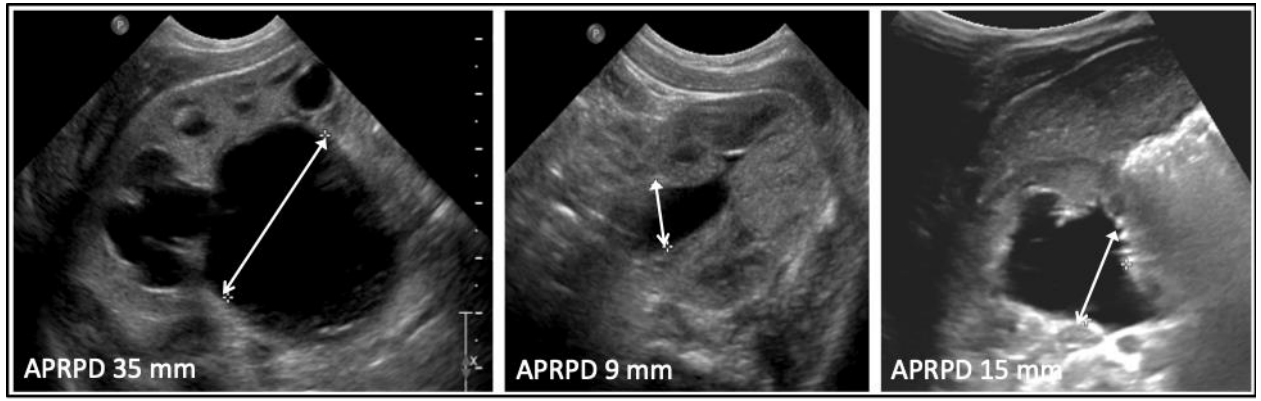
Table 4: Differential diagnosis based on severity of prenatal hydronephrosis and presence of hydroureter or abnormal bladder

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]

Prenatal hydronephrosis

- Prenatal diagnosis of hydronephrosis is made in 1-5% of all pregnancies
- If a fetus at 20 weeks gestation has an APRPD ≥ 4mm or calyceal dilation, they are considered to have prenatal hydronephrosis.
- 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.



Prenatal management

- If a fetus at 20 weeks has only unilateral hydronephrosis with APRPD <7 mm without any peripheral calyceal dilation, this is considered low risk 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
- If significant bladder outlet obstruction from PUV or another cause, consideration to prenatal intervention can be given

Postnatal Management

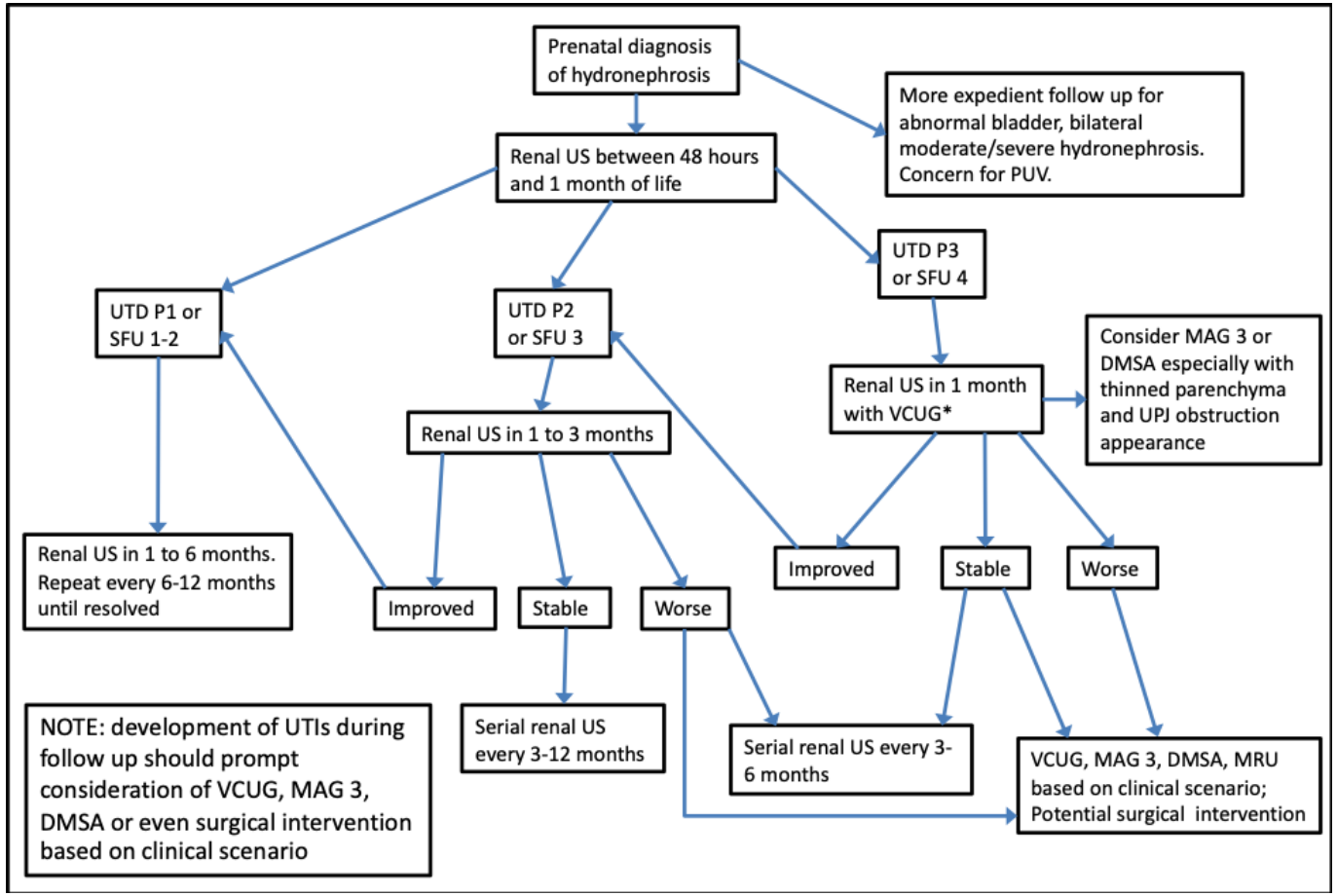
- For majority of cases, an initial postnatal renal ultrasound is recommended sometime between 48 hours and 1 month of life.
- Based on this initial postnatal ultrasound, the hydronephrosis is categorized

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]

Postnatal Follow up

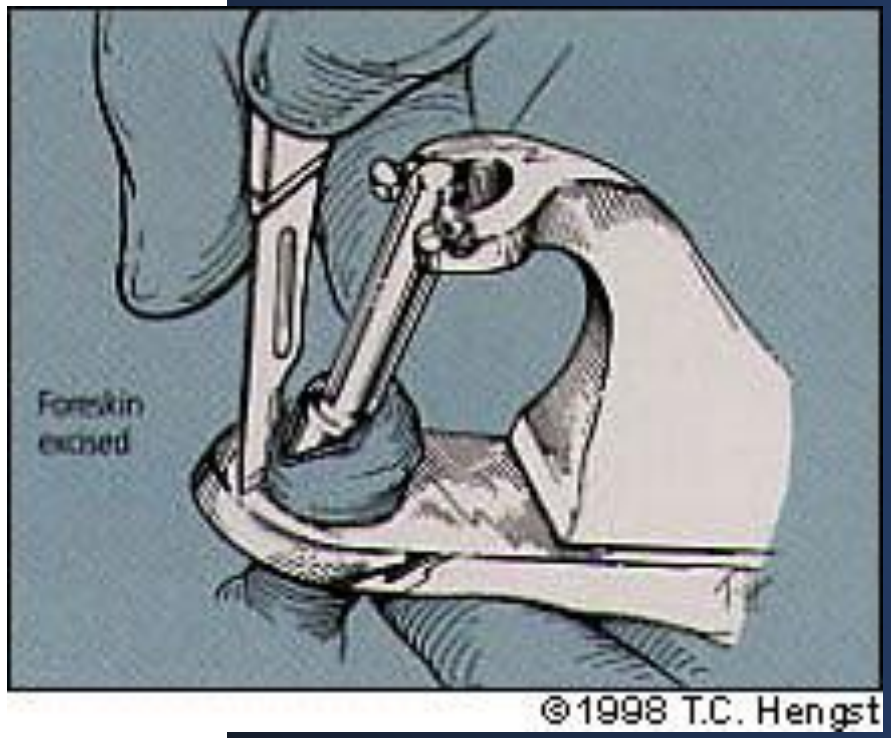


UTI Prevention

- Prophylactic antibiotics should be discussed as an option with the families of children with more severe hydronephrosis or other risk factors
  - Other risk factors for UTI include female gender, uncircumcised status, vesicoureteral reflux, and hydroureteronephrosis.
- 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.
- 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.

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.
- Circumcision is an option to decrease risk of UTI for infants with severe hydronephrosis, especially if significant urinary tract anomalies such as PUV are present.
- OR treat physiologic phimosis with topical steroid cream



Natural course of hydro

- The majority (50-75%) of prenatal hydronephrosis will be mild and have a ~90% chance of being transient hydronephrosis that will resolve spontaneously
- More moderate and severe cases of prenatal hydronephrosis have higher chance of being a UPJ obstruction or another diagnosis and going on to have surgery

Older presentation

- Usually diagnosed incidentally, due to symptoms (flank pain) or UTI
- 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.
- Management depends on etiology

Ureteropelvic Junction Obstruction

- UPJO is the most common pathologic etiology of prenatal hydronephrosis, occurring in 1/500-1000 birth
- It is thought to be caused by either an **intrinsic functional** or anatomical **stenosis of the UPJ** (most often seen in infants) or by **extrinsic compression by a lower pole renal vessel** that crosses the pelvis and obstructs drainage (seen more frequently in older children).
- Diuretic renography is used to confirm the diagnosis of UPJO and can also be predictive of the need for surgical management with pyeloplasty
    - If the differential renal function is near 50%-50% on diuretic renography (indicating lack of renal functional loss due to obstruction), an observational approach may be indicated.

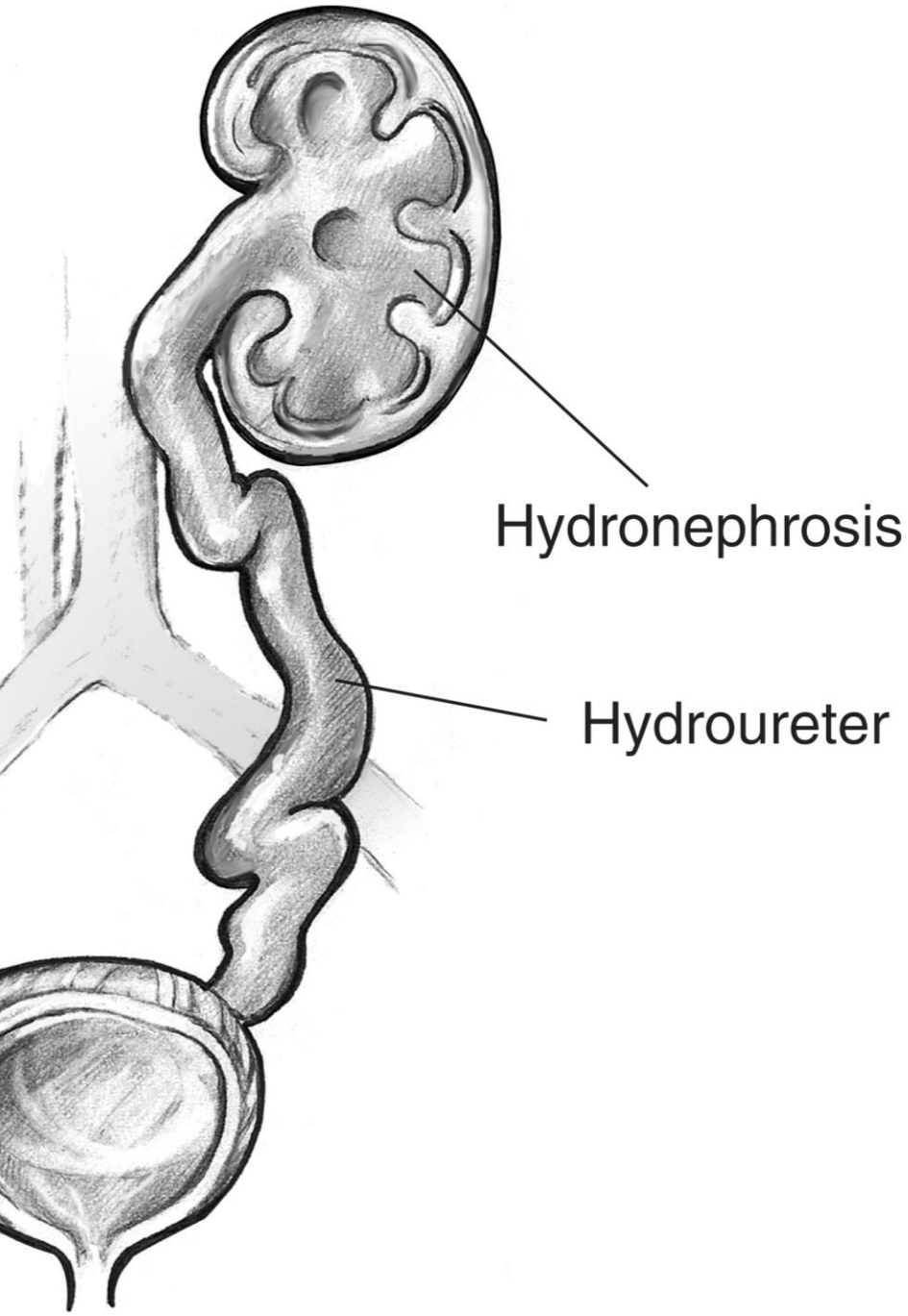
Surgical Treatment

- Indications for **pyeloplasty** include worsening differential renal function, worsening hydronephrosis especially with development of thinning parenchyma, urinary tract infections, pain, or nephrolithiasis.

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]



Hydroureteronephrosis

- Incidence of ureteral dilation prenatally is unknown, likely given the difficulty of identifying the ureter on prenatal imaging
- ureteral diameter in children typically <5 mm
- Megaureter describes ureteral dilation, measuring **> 7 mm in diameter**.
- occurs in **0.4/1000 births.**

Differential Diagnosis

- **Megaureter**
  - UVJ obstruction*
  - Vesicoureteral reflux*
- **Ectopic ureter -** single or duplicated system
- **Ureterocele -** intravesical vs. extravesical

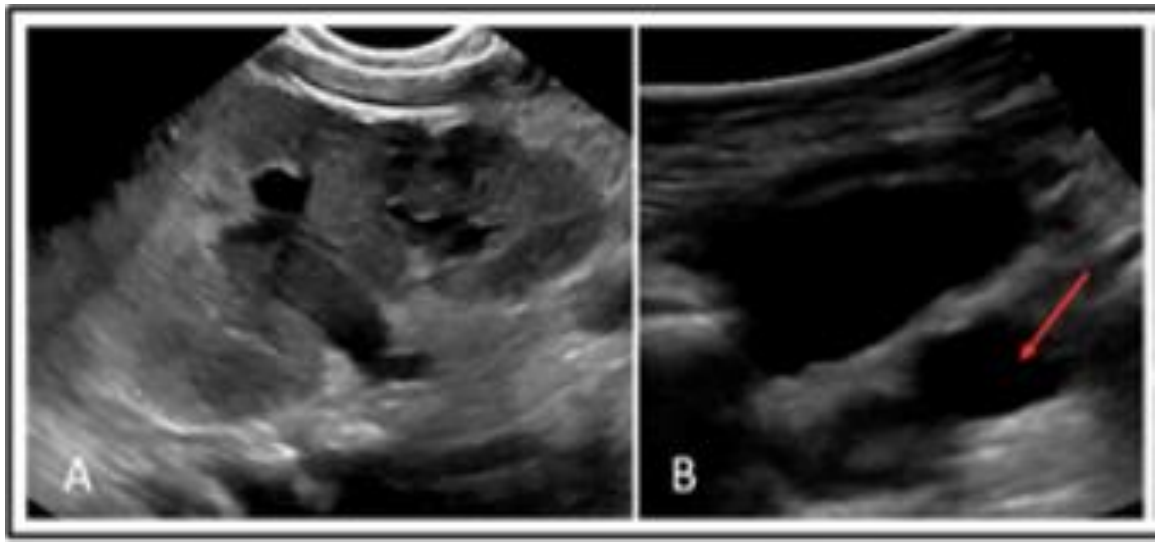
Megaureter

- **Primary obstructive megaureter** or **primary megaureter** refers to a dilated ureter with an adynamic distal segment and ureterovesical junction (UVJ) obstruction.
- **Secondary megaureter** comes from distal obstruction—neurogenic bladder, bladder outlet obstruction

Table 4: Classification of Megaureters	
Refluxing, Nonobstructed Megaureter	**Nonrefluxing, Obstructed Megaureter**
Primary (congenital reflux)	Primary (adynamic segment)
Secondary (urethral valves, neurogenic bladder)	Secondary (urethral obstruction, extrinsic mass, or tumor)
Nonrefluxing, Nonobstructed Megaureter	**Refluxing, Obstructed Megaureter**
Primary (idiopathic, physiologically insignificant adynamic segment)	Occurs with ectopic ureter to urethral sphincter. Reflux during voiding.
Secondary (polyuria, infection, postoperative residual dilation)	

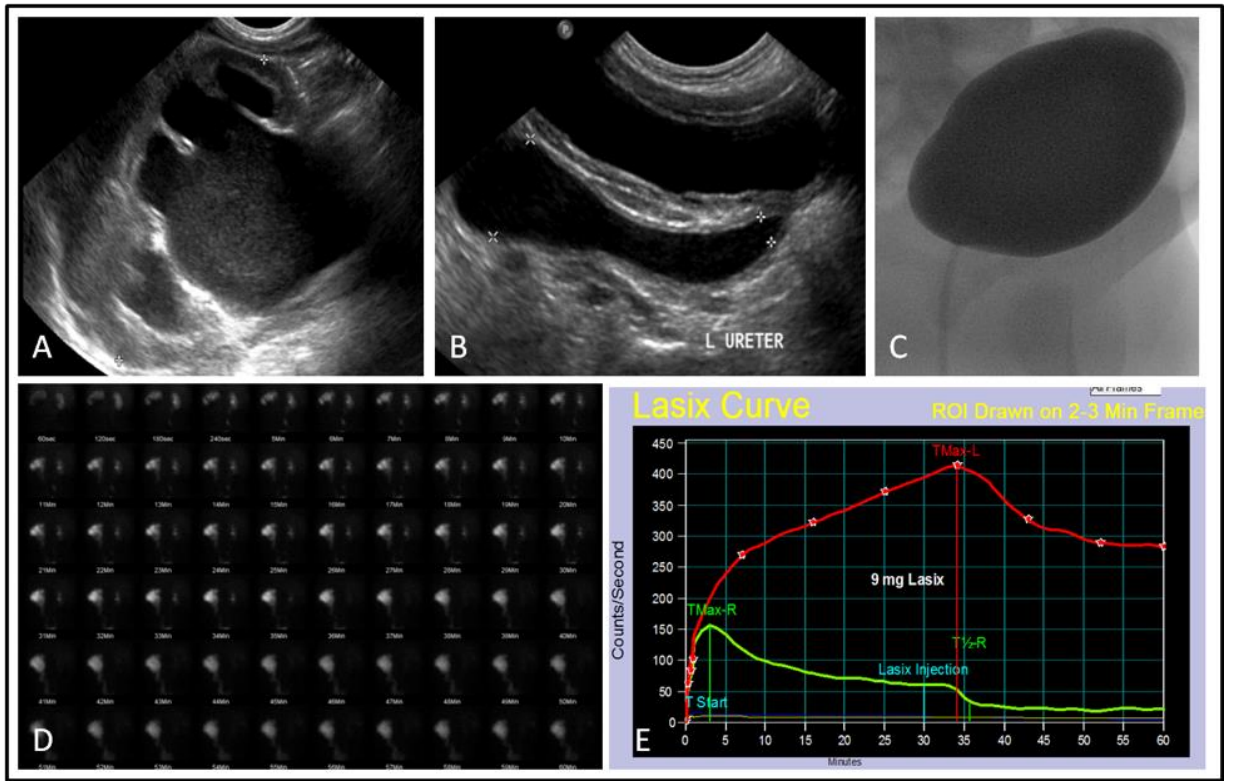
Evaluation

- Prenatal evaluation is same as hydronephrosis
  - Prenatal ultrasound may be able to identify not only hydronephrosis with or without ureteral dilation but also evidence of duplicated collecting system, ectopic ureter, and ureterocele.
- Postnatal renal/bladder ultrasound



Other Imaging

- **Voiding cystourethrogram (VCUG)**- evaluate for VUR and BOO
- **Renal function study (MAG3)**- assess differential renal function and drainage
- **MRI urography**- can provide detailed images for complex anatomy, may require sedation

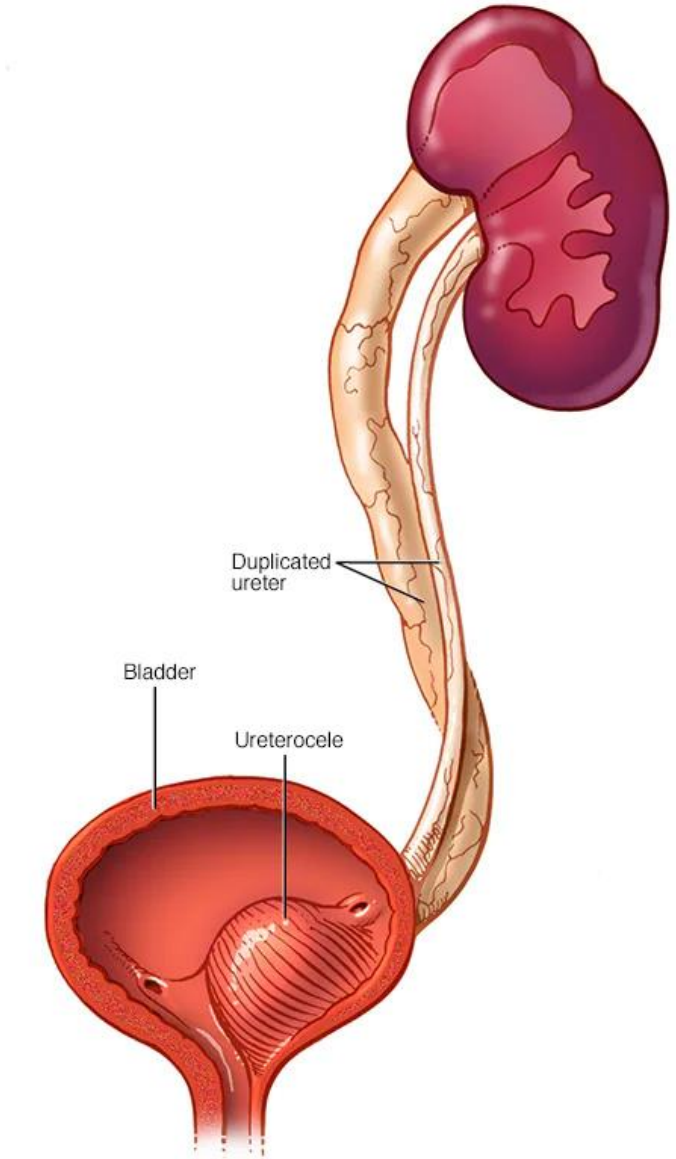


Primary obstructed megaureter

- Management has become more observational and less interventional
- Ureteral reimplantation.
  - Ureteral tapering may be indicated if the ureteral dilation is >1 cm.
  - typically performed after 9-12 months of age
- Other options : percutaneous nephrostomy tube, cutaneous ureterostomy, or ureteral stent placement
  - Considered when ureteral reimplantation is thought to be difficult due to young age/small bladder size or thought to be urgent due to significant infection

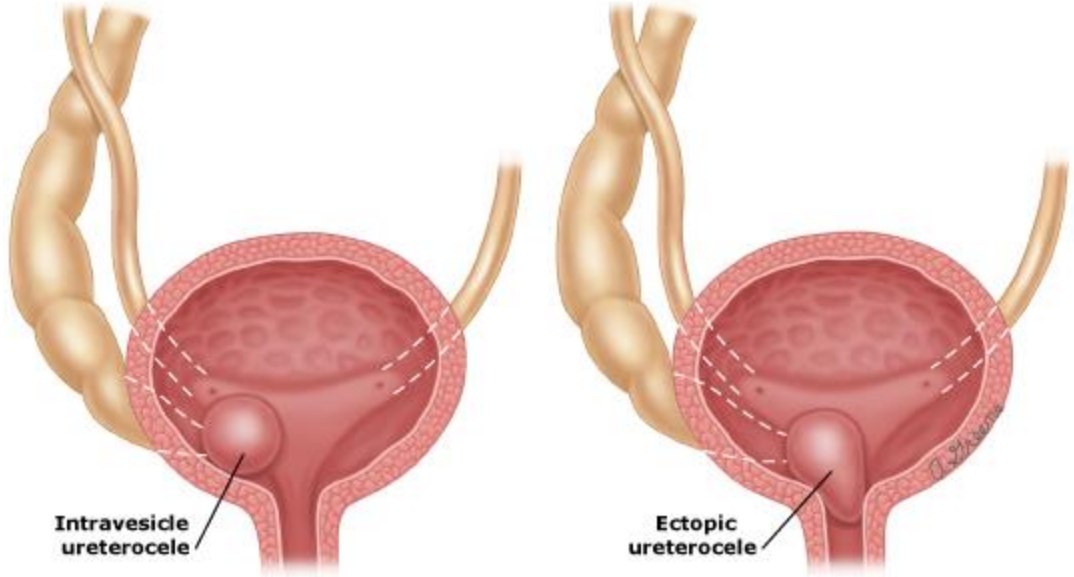
Ureteroceles

- Describes a cystic dilation of the distal aspect of the ureter that is located either within the bladder or spanning the bladder neck and urethra.
- May be associated with a single or duplex system; in duplex systems they are associated with the upper pole.



Intravesical vs Extravesical

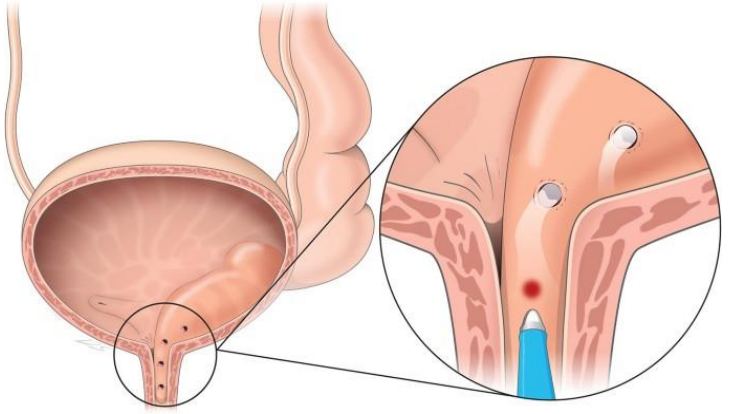
- **Intravesical ureterocele** is entirely within the bladder and above the bladder neck.
- **Ectopic or Extravesical ureteroceles** include those in which some portion of the ureterocele is situated permanently at the bladder neck or in the urethra.



Treatment

- Small ureteroceles in orthotopic systems without significant obstruction can be monitored and may never need any intervention.
- Children with a urinary tract infection can generally be acutely managed with antibiotics and careful monitoring
- If the child does not quickly improve or deteriorates, urgent drainage of the system affected by the ureterocele is indicated, most frequently with a ureterocele puncture

Treatment



- Ureterocele puncture/incision
  - If the renal function in the obstructed kidney is poor or deteriorating or if the ureterocele is large and there is concern for risk for infection and interest in protecting renal parenchyma
  - Provides adequate drainage while minimizing the risk of new VUR in the renal unit associated with the ureterocele.
  - low transverse incision in the ureterocele or a “watering can technique” of puncture using a laser.
- Ureterocele excision with ureteral reimplantation, ureteroureterostomy/pyelostomy, or upper pole heminephrectomy

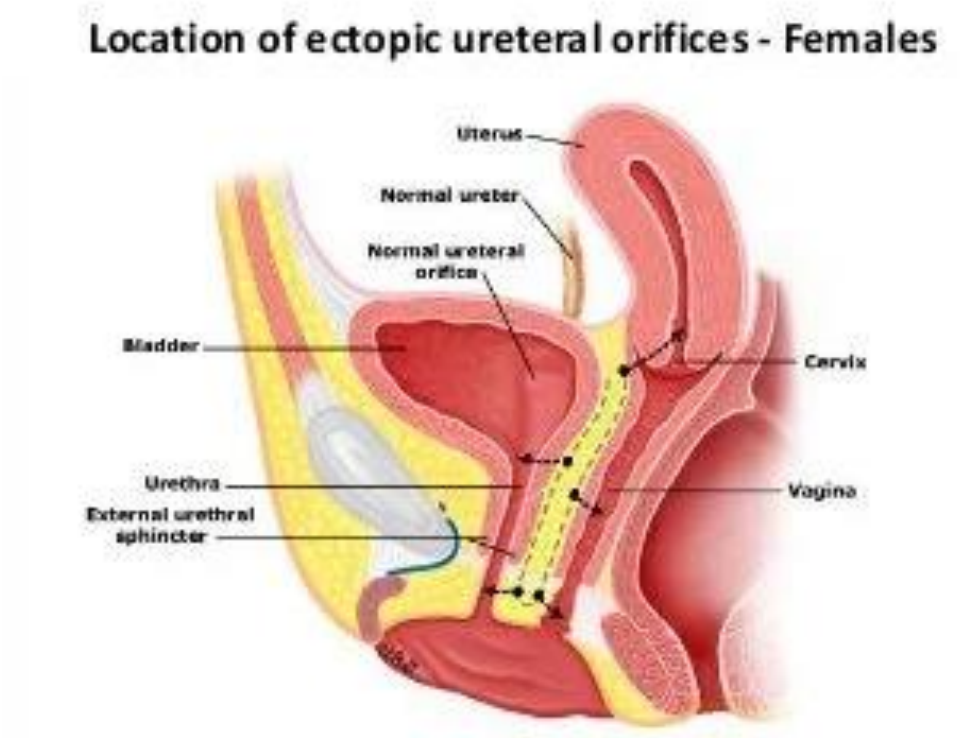
Table 1: Options for ureterocele management

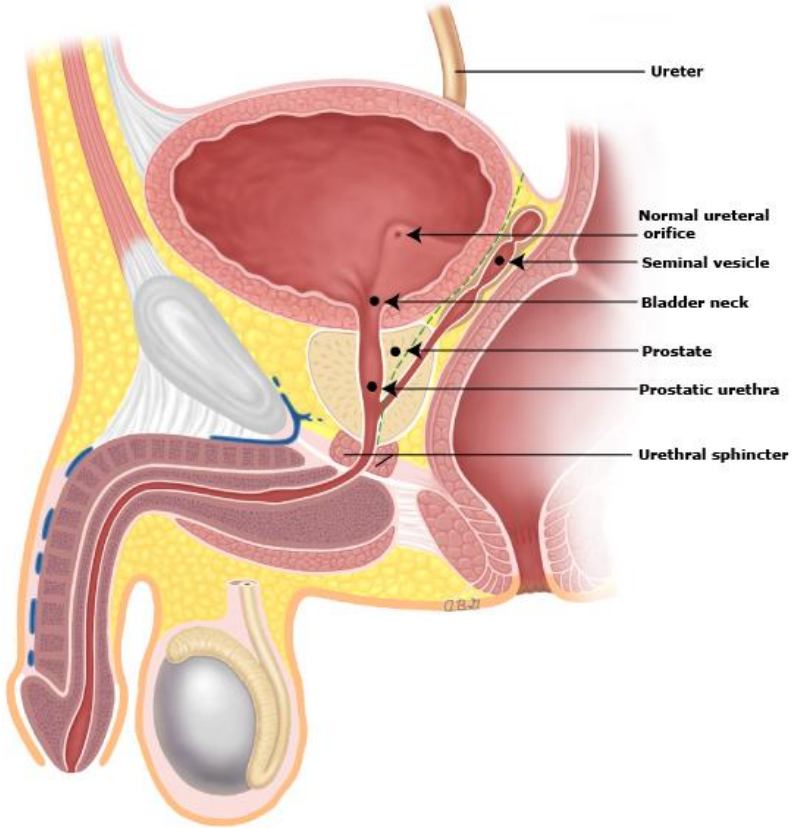
PROCEDURE	IDEAL INDICATIONS	ADVANTAGES	RISKS
Observation	Small ureterocele with preserved renal function and no UTIs Minimal hydronephrosis No or minimal obstruction demonstrated on MAG3	No anesthetic No risk of inducing VUR	May develop UTI and present with sepsis May develop long term renal deterioration or stones in ureterocele Need for follow up
Transurethral incision/puncture	Acute infection Infant with indication for drainage Older child with small ureterocele in single system with indication for intervention (eg UTI)	Minimally invasive, often outpatient procedure Effective decompression Occasionally definitive	May produce VUR into ureterocele segment or ipsilateral upper pole, necessitating further bladder surgery to remove ureterocele and reimplant ureter May not produce definite drainage
Upper pole heminephrectomy (without ureterocele excision)	Older patient with largely dilated, non-functioning upper pole No VUR into upper pole ureter	May be definitive Removes pathology Avoids bladder level surgery	May not be definitive as child may also necessitate bladder surgery in the future Small risk of injury to lower pole
Ureterocele excision and common sheath reimplantation	Refluxing lower pole Functioning upper pole without significant dilation	Eliminates obstruction and reflux, removes ureterocele No significant operative risk to the kidney	Complex surgery Small risk to vagina and bladder neck May require ureteral tapering May develop VUR Small risk of ureteral obstruction.
Ureteroureterostomy or ureteropyelostomy	Functioning upper pole No VUR into lower pole ureter	Drains obstructed segment with little risk of obstruction or UTI No bladder level or renal surgery.	Leaves ureterocele in bladder Potential injury to the healthy, non-involved lower pole ureter May develop VUR Small risk of ureteral obstruction.

Ectopic ureters

- Describes any ureter, single or duplex, that does not enter the trigonal area of the bladder. Ectopic ureters drain to an abnormally caudal location
- The estimated incidence is 1/1000 births with a female predominance.
- In a duplex system, ectopic ureter 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.**





Treatment

- Ectopic ureters can be managed with prophylactic antibiotics
- Ectopic ureters presenting with infection may need to be acutely drained with percutaneous drainage or cutaneous ureterostomy if clinical status does not improve as expected with antibiotics alone.
- If there is no indication for emergent intervention for ectopic ureter, treatment can usually be delayed until the child can undergo definitive surgical management.
- **Definitive Surgery**:
  - Ureteral reimplantation in the case of the single system ectopic ureter, with or without ureteral tapering.
  - In the case of a duplicated collecting system with an ectopic upper pole, an upper pole to lower pole ureteroureterostomy is an option assuming there is no VUR into the lower pole ureter.
  - If the associated renal unit (entire kidney or upper pole moiety) is non-functional, a nephrectomy or upper pole heminephrectomy can be considered.

GU anomalies

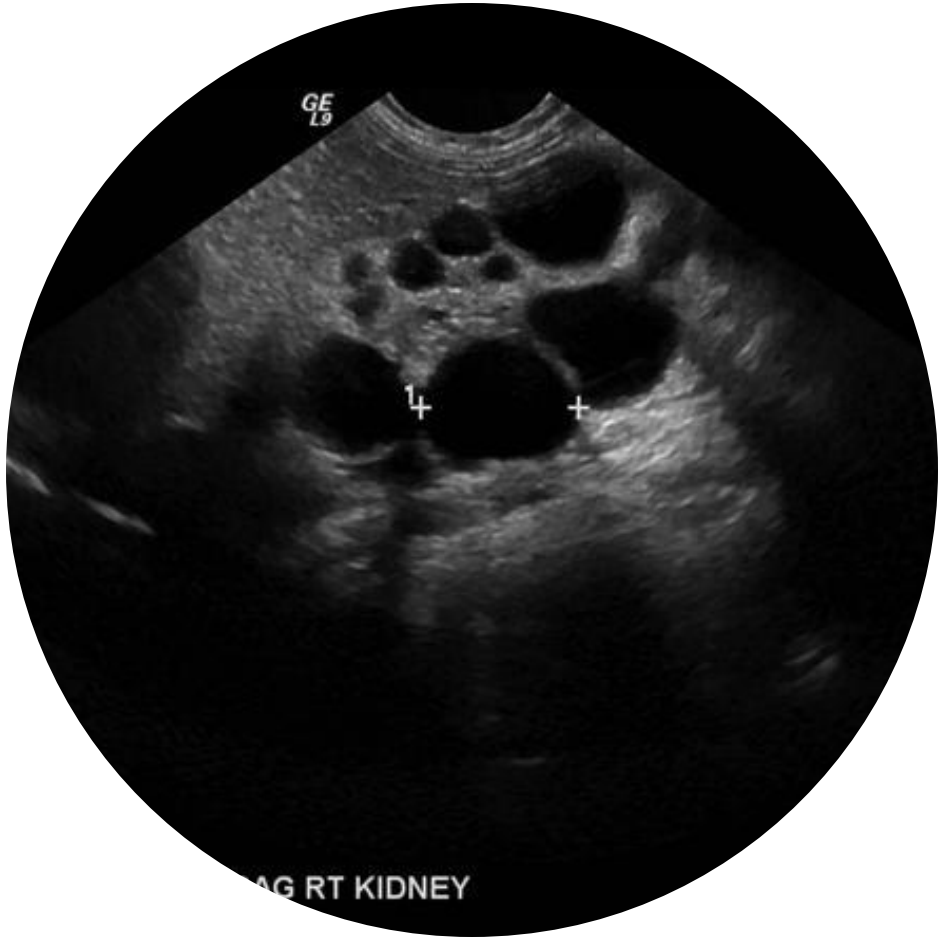
Table 3: Summary of congenital anomalies of the kidney and urinary tract

	Epidemiology	Characteristics	"Pearls" to remember
Renal parenchyma anomalies			
Renal dysplasia	3/1000 births	Many different appearances	Often associated with collecting system anomalies.
MCDK	~1/1000 births	Multiple non-communicating cystic structures. Most involute.	Confirm diagnosis with serial ultrasound or DMSA scan postnatal. Not associated with increased Wilm's risk.
ADPKD	1 in 400 to 1000 people	Large cysts. Progressive. Autosomal dominant. Renal failure in adulthood (50s – 60s) common.	In a child with 1-2 simple cysts, ADPKD is possible diagnosis.
ARPKD	1 in 10000 to 40000 births	Small cysts. Autosomal recessive. Often fatal.	Cysts arise from collecting ducts.
Renal agenesis	1 in 3000 to 5000 births	Lack of renal parenchyma. Failure of ureteral bud formation.	Often with ipsilateteral Wollfian (males) or Mullerian (females) abnormalities.
Collecting system anomalies			
Hydronephrosis	1 to 5% of all pregnancies	Dilation of renal collecting system. Can be associated with thinning of parenchyma.	Transient or physiologic hydronephrosis most common diagnosis. Severity correlates with outcomes.
UPJO	1 in 500 to 1000 births	Hydronephrosis. Delayed drainage on diuretic renography often with decreased differential function.	Indications for treatment include worsening split renal function, UTIs, pain, nephrolithiasis. Crossing vessels are more common in older children than in infants
Megaureter	0.4 in 1000 births	Large ureter > 7 mm. Classified according to presence or absence of reflux and/or obstruction.	Majority of primary megaureters resolve with time and diffential function of 50% predicts resolution. Similar indications for surgery as UPJO.
Megacalycosis	Rare	Increased number of calyces that are dilated, medullary pyramid hypoplasia, and drainage on diuretic renography.	Consider diagnosis over UPJO when calyces are increased and dilated, there is drainage but it is delayed, and the renal pelvis is small in proportion to calyceal dilation.
Ecoptic ureter	Less than 1 in 1000 births	Dilated ureter with varying degree of renal dysplasia in associated renal moiety. Often associated with upper pole of duplex kidney.	Management is quite variable but surgery is generally needed (**Table 4**)
Ureterocele	1 in 1000 births	Cystic dilation of distal ureter in bladder. Often associated with upper pole of duplex kidney.	Consider function of renal segment and ipsilateral lower pole reflux in management decisions.
Duplex kidney	1-5% of population	Separate ureter draining upper and lower renal poles. Can be partial or complete.	The way the mesonephric duct and ureteral buds are incorporated into the bladder explains the Meyer-Weigert rule.
VUR	30% of children with febrile UTI	Retrograde flow of urine from bladder into upper tracts due to failure of adequate mucosal coaptation and closure of UO during voiding.	Voiding dysfunction should be treated prior to surgical intervention. Most low grade VUR resolves.
Renal migration or fusion anomalies			
Ectopic kidneys	1 in 500 to 1200 births	Kidney in ectopic location along route of ascent. Pelvic kidney is most common.	Increased risk of collecting system anomalies such as VUR and UPJO. Anomalous blood supply.
Horseshoe kidney	1 in 600 births	Inferior poles of kidneys fused prior to renal asenct and ascent limited by inferior mesenteric artery.	Increased risk of collecting system anomalies such as VUR and UPJO. Anteriorly oriented pelvis with often medial facing calyces. Anomalous blood supply.
Crossed fused ectopia	1 in 1000 to 2000 births	Both kidneys on same side and fuxed.	Ureter from ectopic kidney crosses back to other side. Anomalous blood supply.
Crossed unfused ectopia	Rare	Only 5% of crossed renal ectopia.	Ureter from ectopic kidney crosses back to other side. Anomalous blood supply.
Bilateral crossed renal ectopia	Rare	Both kidneys are on wrong side.	Ureters from each kidney cross to other side.
Lower urinary tract obstruction anomalies			
Posterior urethral valves	1 in 5000 to 8000 births	Obstruction at posterior urethra by membranous folds. Upper tract damage and bladder dysfunction.	These patients need long term monitoring of bladder and renal function. High incidence of renal failure.
Anterior urethral valves	Less than 300 reported cases	Characteristic web of tissue from ventral anterior urethra. Often associated with urethral diverticulum.	Endoscopic ablation is usually initial treatment. Occasionally urethroplasty needed if diverticulum. Has similar long term concerns as PUV.
Urethral atresia	Very rare	Failure of urethral cannulization.	Fetal survival associated with patent urachus or other method of urine drainage.
Prune belly syndrome	3-4 in 100,000 births	Abdominal wall muscle deficiency with "prune" appearance, bilateral undescended testis, GU tract dilation.	GU tract dilation generally worse distally. Need for CIC because of urinary retention with UTIs is not uncommon. Abdominoplasty controversial.

View Image

Renal dysplasia

- A **multicystic dysplastic kidney (MCDK)** is the consequence of severe renal dysplasia and is characterized by a non-reniform shaped kidney composed of non-communicating cysts, no functional renal tissue, and an atretic or absent ureter
  - **MCDK occurs with a frequency of 0.3-1/1000 births.**
  - Unilateral renal dysplasia can result in compensatory hypertrophy of the contralateral kidney.
  - **Most MCDK kidneys involute with time and do not require surgical intervention**.



Polycystic Kidney Disease

Autosomal Dominant

- **incidence of 1/400-1000 infants**.
- The **PKD1 or PKD2 genes** encode the polycytin-1 and polycytin-2 proteins, respectively. A mutation in PKD1 (~85% of cases) or PKD2 leads to ADPKD.
- cysts in autosomal dominant disease tend to be larger (centimeters), involve the entire nephron, and appear later in life.
- Clinical features in children with ADPKD ranges from normal appearing kidneys to numerous bilateral renal cysts, and potentially, hypertension, hematuria, and infection.

Autosomal Recessive

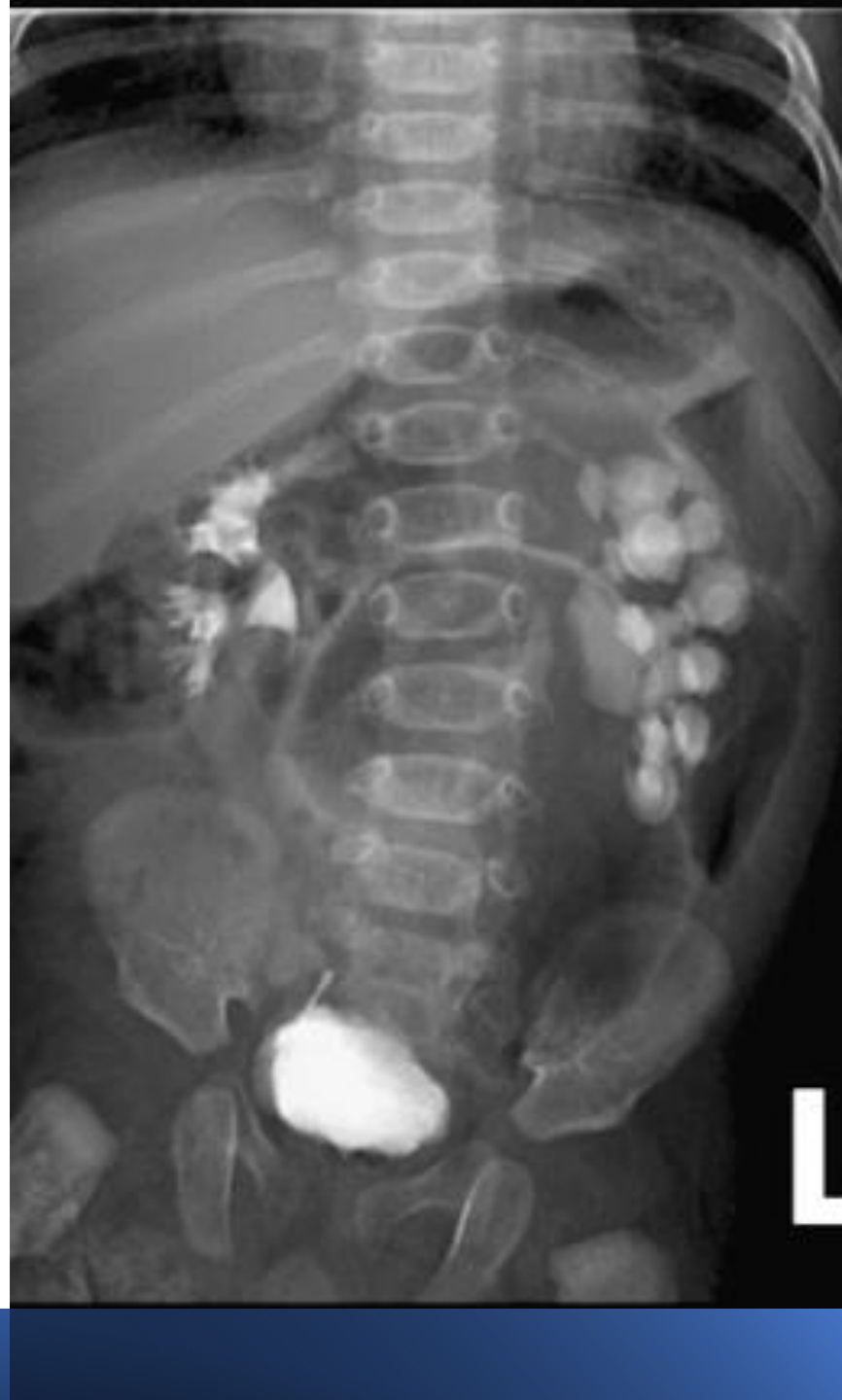
- **incidence of 1/10,000-40,000** infants
- Mutations in the **PKHD1 gene** that encode the protein fibrocystin thought to be causative.
- Cysts tend to be smaller (millimeters), arise from the collecting duct, and appear early in life.
- ARPKD can cause in utero or perinatal demise due to oligohydramnios and pulmonary hypoplasia, end stage renal disease in early life requiring dialysis, or, relatively normal renal function with delayed diagnosis in adulthood.

Renal Agenesis

- Renal agenesis describes the absence of renal parenchymal tissue unilaterally
- occurs at **1/3000-5000 births.**
- It is thought to result from a **failure of ureteral bud formation**.
- Males often have associated abnormalities of the vas deferens, seminal vesicles, and epididymis (but a normal ipsilateral testis).
- Females have an increased risk of ipsilateral Mullerian abnormalities such as uterine didelphys, duplicated vagina, and potentially obstructed ipsilateral hemivagina.[11]
- **In both males and females, the presence of genital duct abnormalities should prompt renal ultrasound to look for ipsilateral renal agenesis**.

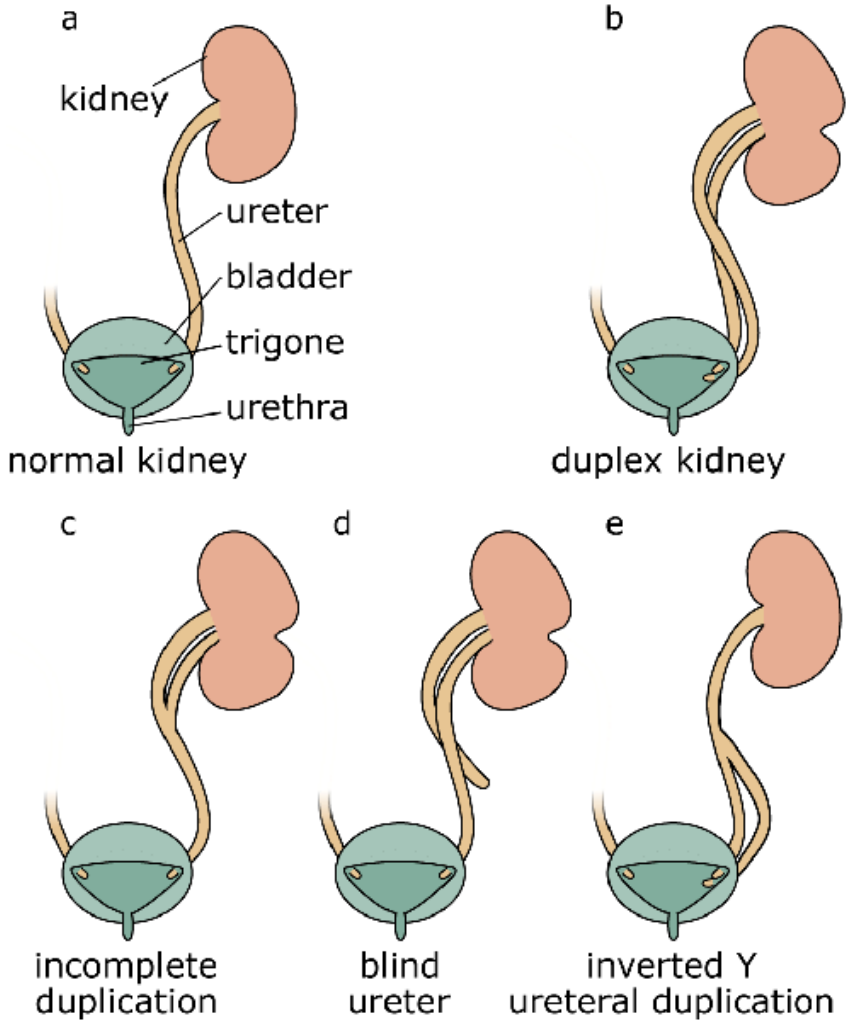
Collecting system anomalies

- Megacalycosis
  - diagnosis of unknown etiology
  - characterized by increased number of calyces, dilation of calyces without obstruction, and medullary pyramid hypoplasia
  - It is usually detected on prenatal ultrasound with postnatal evaluation revealing increased number of dilated renal calyces on ultrasound and lack of obstruction on diuretic renography.
- Other examples: Hydronephrosis, Megaureter, UPJO, ectopic ureter, ureterocele



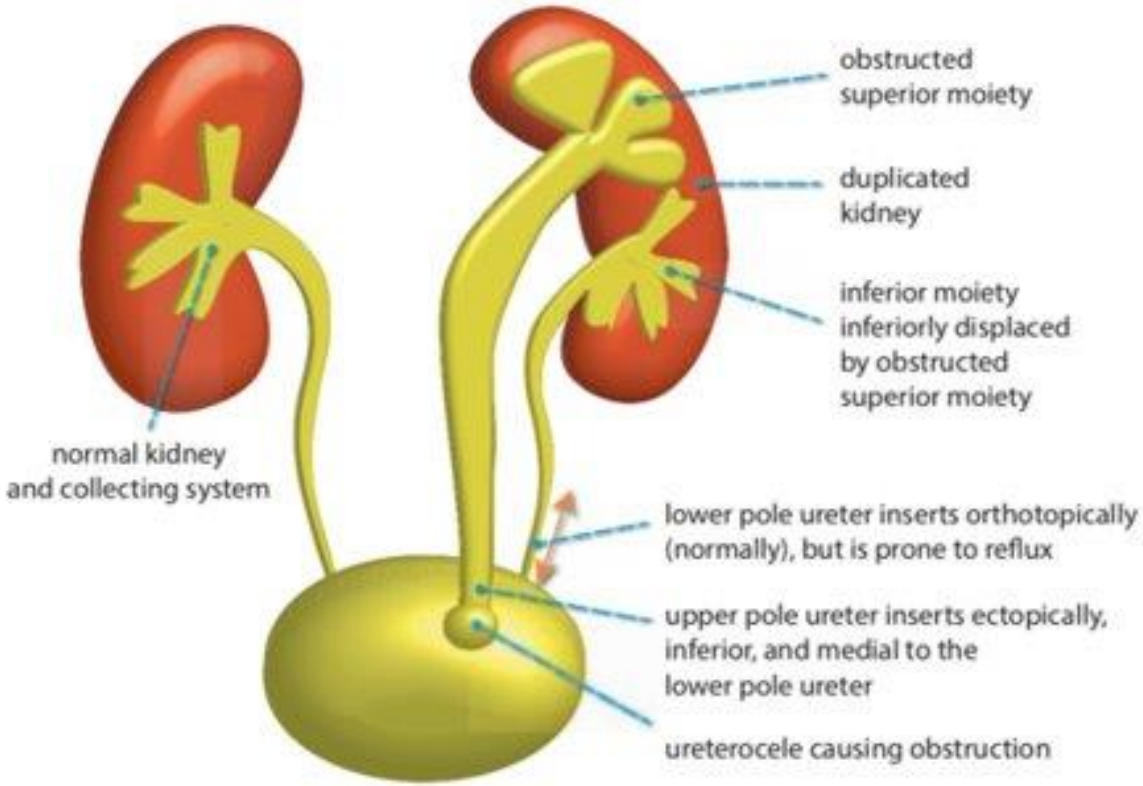
Pyeloureteral Duplication

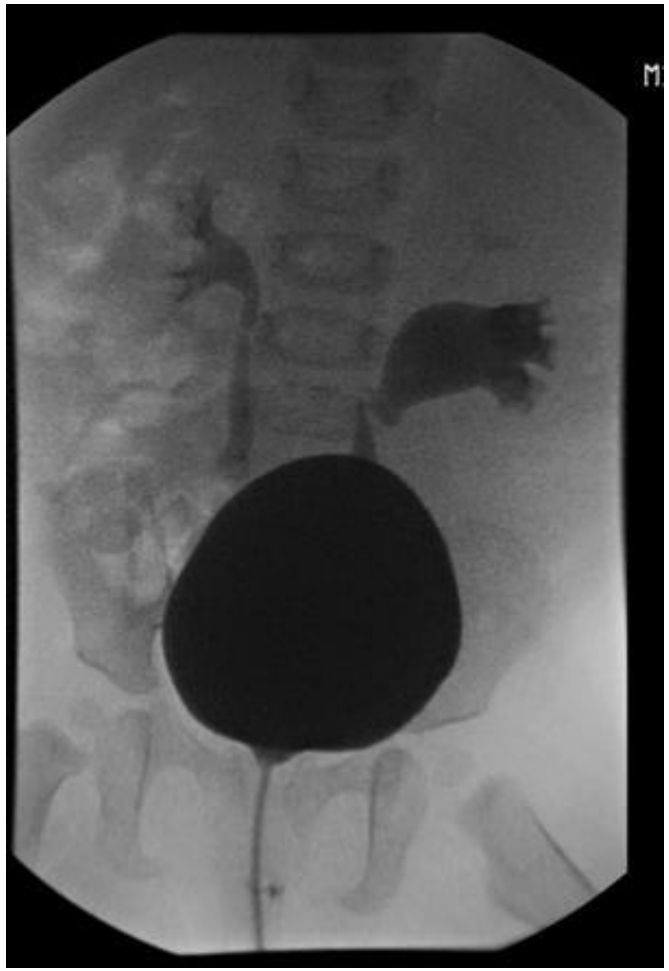
- Complete or partial pyeloureteral duplication is the most common collecting system anomaly with an incidence of **1-5%**.
- Complete duplication results in two distinct ipsilateral ureteral orifices, two ureters and two collecting systems.
- Incomplete duplication results in a Y-shaped ureter or bifid renal pelvis.



Meyer-Weigert rule

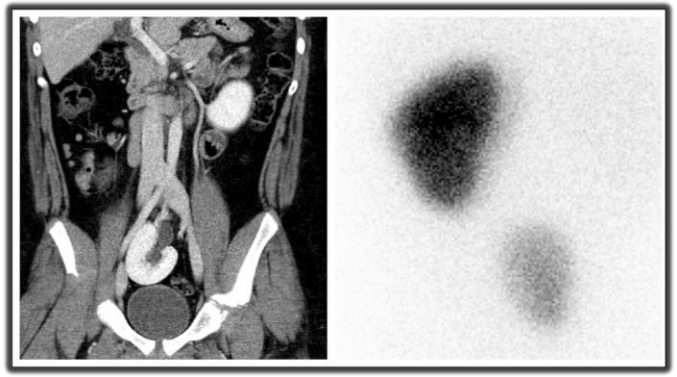
- Upper pole ureter inserts ectopicall into the bladder and prone to obstruction
- Lower pole ureter inserts orthotopically and prone to reflux
- SOLR



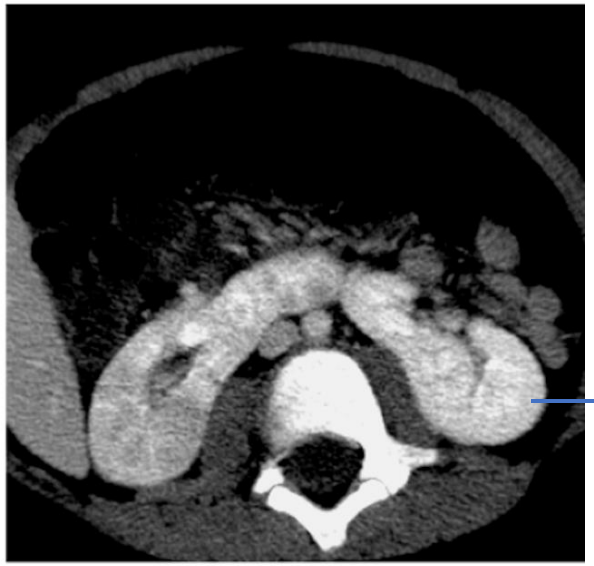


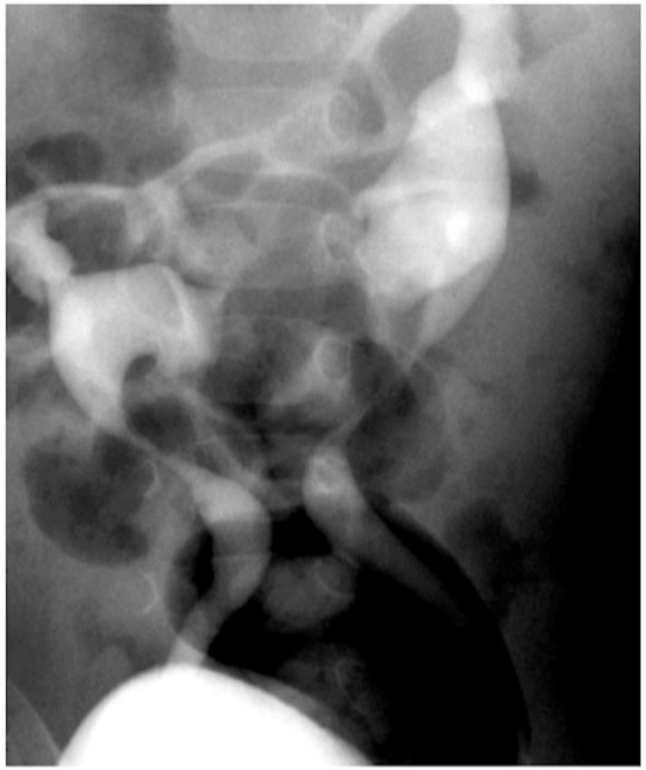
- **When VUR is present in the lower moiety ureter, there is a characteristic “drooping lily” appearance on the VCUG**

Ectopic Kidney



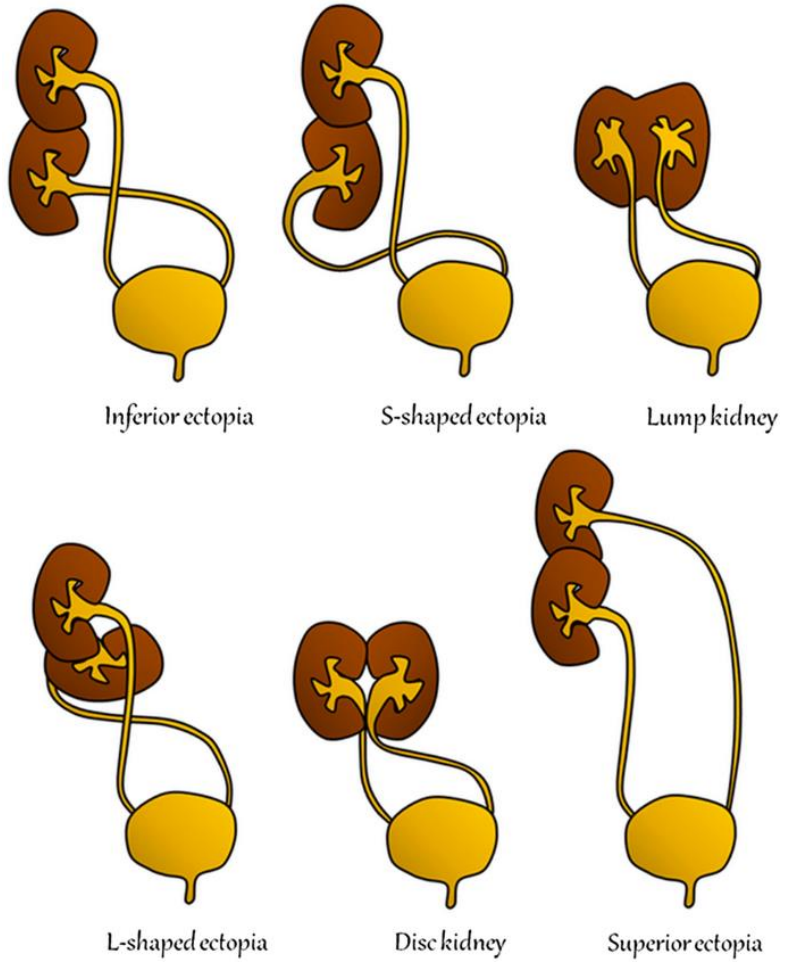
- Disruptions in renal migration lead to ectopic kidneys
- The kidneys normally ascend from a pelvic to lumbar location by 8 weeks of gestation. Failure of ascent leads to an ectopic kidney, with the bony pelvis representing the most common location (in 1/500-1200 births)
- Ectopic kidneys have an anomalous blood supply and tend to have anteriorly oriented renal pelvis.
- Have increased risk of ureteropelvic junction obstruction (UPJO), renal dysplasia, vesicoureteral reflux (VUR), nephrolithiasis, and urinary tract infection (UTI).





Renal Fusion

- right and left renal units being abnormally paired together within the retroperitoneum.
- The most common fusion anomaly is a **horseshoe kidney**, occurring in 1/600 births
  - The inferior poles of the kidneys fuse prior to ascent and the inferior mesenteric artery limits further ascent of the isthmus.
  - do not rotate normally or ascend completely, so they have an anteriorly oriented pelvis as well as anomalous blood supply.
  - increased risk of VUR, UPJO, nephrolithiasis, and UTI.

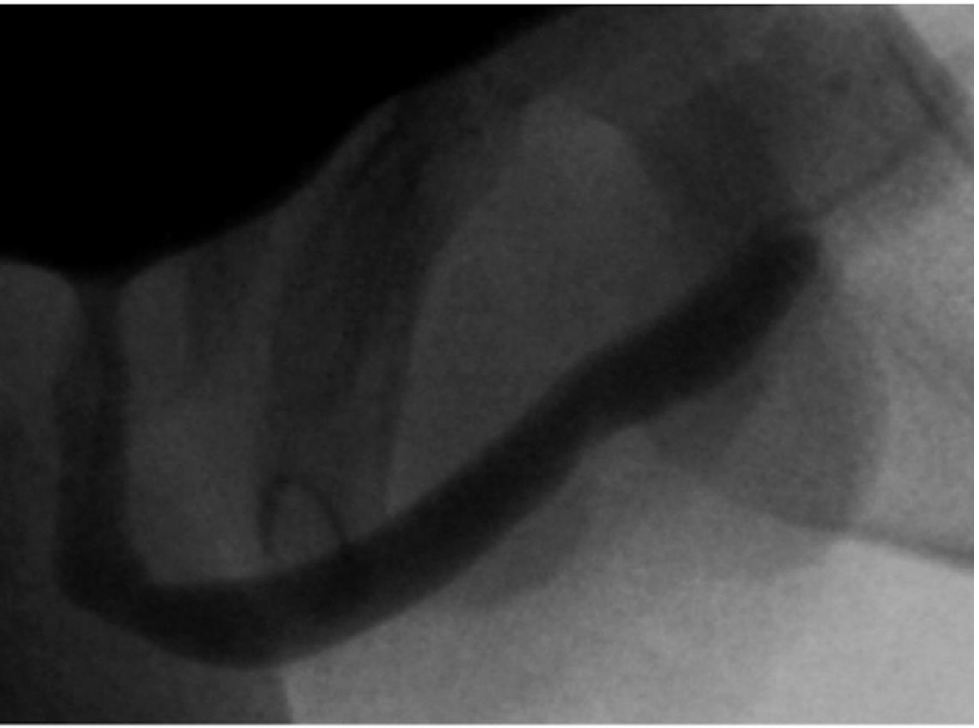


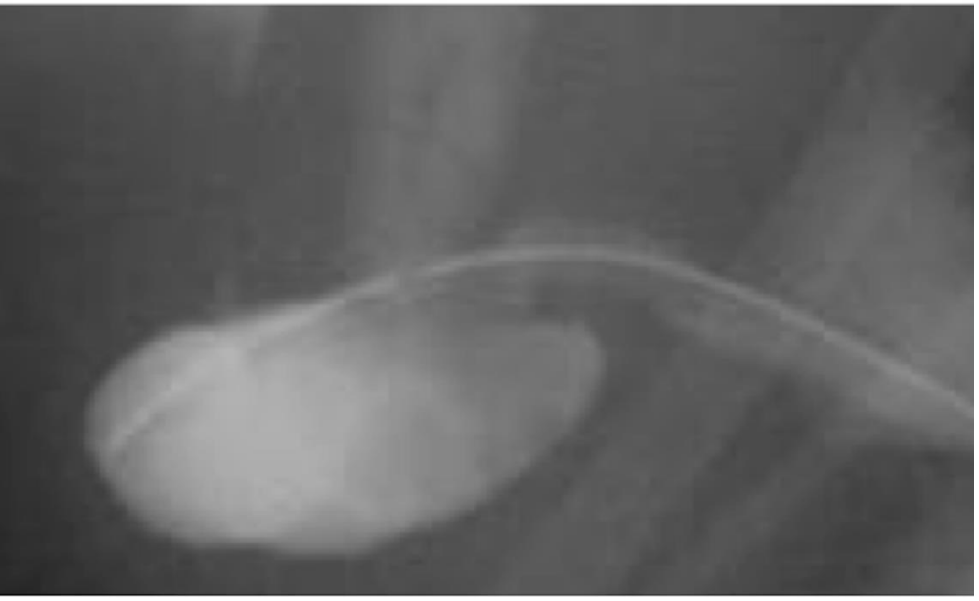
Renal Fusion

- **Crossed fused renal ectopia** occurs when both renal units are fused on the same side of the body, occurring in 1/1000 to 2000 births.

Lower urinary tract anomalies

- Congenital lower urinary tract obstruction can lead to poor clinical outcomes including severe bilateral hydroureteronephrosis with oligohydramnios or anhydramnios with fetal demise.
- The most common cause of congenital lower urinary tract obstruction is **posterior urethral valves (PUV)**.
- Less common causes are **Anterior urethral valves** and **urethral atresia**.
- **Prune Belly or Eagle Barrett syndrome (PBS)** is an additional congenital anomaly that can be confused with PUV, especially on antenatal ultrasound.



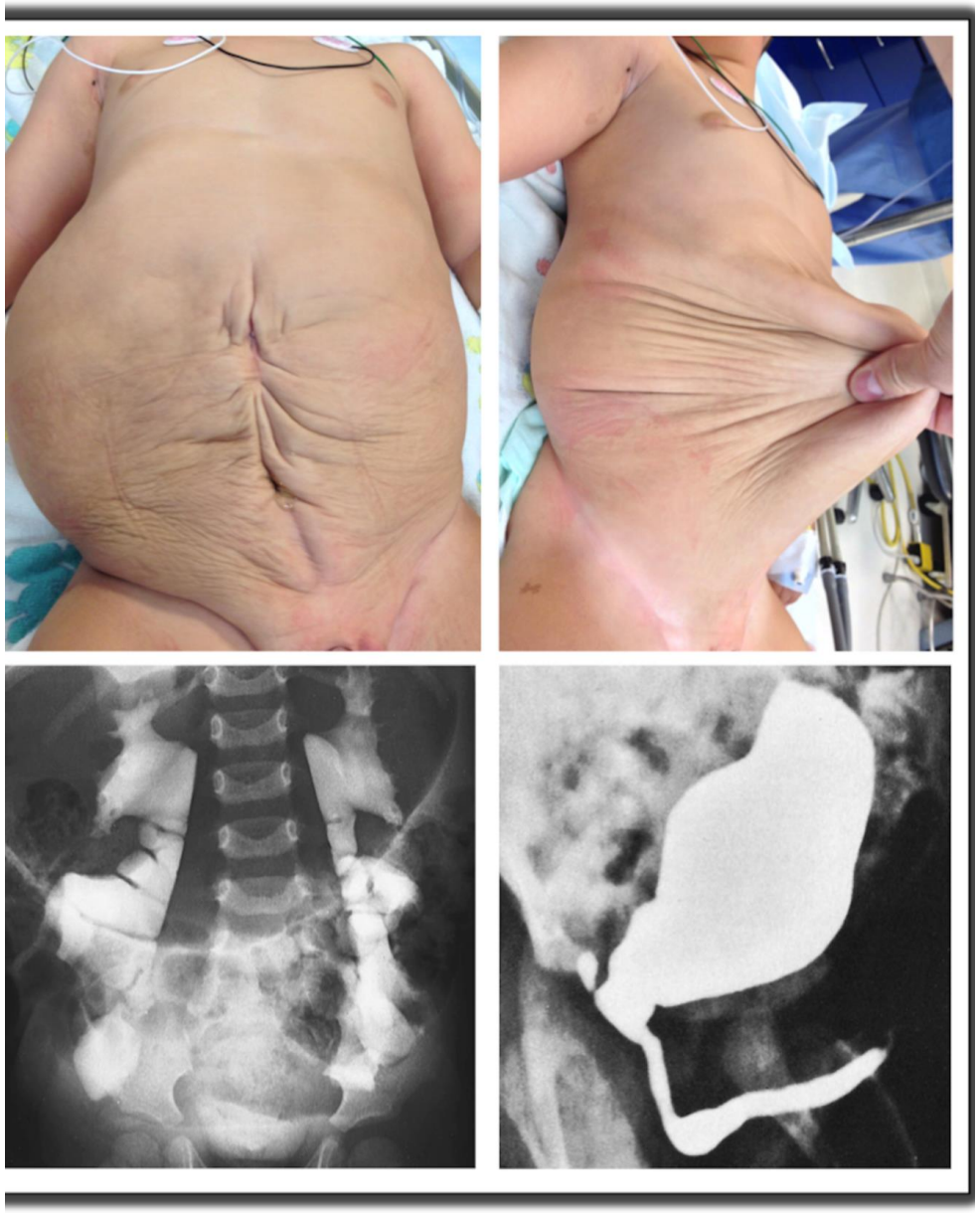


Anterior urethral valve

- very rare entity (less than 300 reported cases) caused by an <u>obstructing web of ventrally-arising tissue in the anterior urethra.</u>
- often associated with a **ventral urethral diverticulum**.
- associated with similar upper tract and bladder consequences as PUV and require close follow up.
- Treatment: **endoscopic ablation**. If there is a large associated diverticulum, excision of that diverticulum with urethroplasty and/or ureteral reimplantation may be needed

Urethral atresia

- Very rare anomaly which may be associated with oligohydramnios, pulmonary hypoplasia, and fetal demise.
- A tragic outcome may be averted if the urine has an alternate route for drainage via a patent urachus or rectourethral fistula.
- In surviving patients, urethral atresia is typically managed with cutaneous vesicostomy until formal urethral reconstruction or urinary diversion can be undertaken
- Urethral atresia is most commonly identified in patients with prune belly syndrome, though rare even in this group.



Prune Belly Syndrome

- Thought to potentially be a defect in the development of the lateral plate mesoderm affecting the abdominal wall.
- Most (95%) cases occur in males and occurs in 3-4/100,000 births.
- Constellation of findings including abdominal wall muscle deficiency, genitourinary tract malformation characterized by dilation, and bilateral undescended testis
- Collecting system anomalies seen with PBS include large bladders that are not obstructed, dilated ureters, prostatic hypoplasia, and variable amounts of renal dysplasia.
- Management generally includes bilateral orchiopexies, and possible use of clean intermittent catheterization depending on presence of UTIs and bladder function, and abdominoplasty in select cases. There is varying degree of renal dysplasia in patients with PBS and chronic kidney disease is common.

2022

69. The most likely etiology for hydronephrosis in an ectopic kidney is:

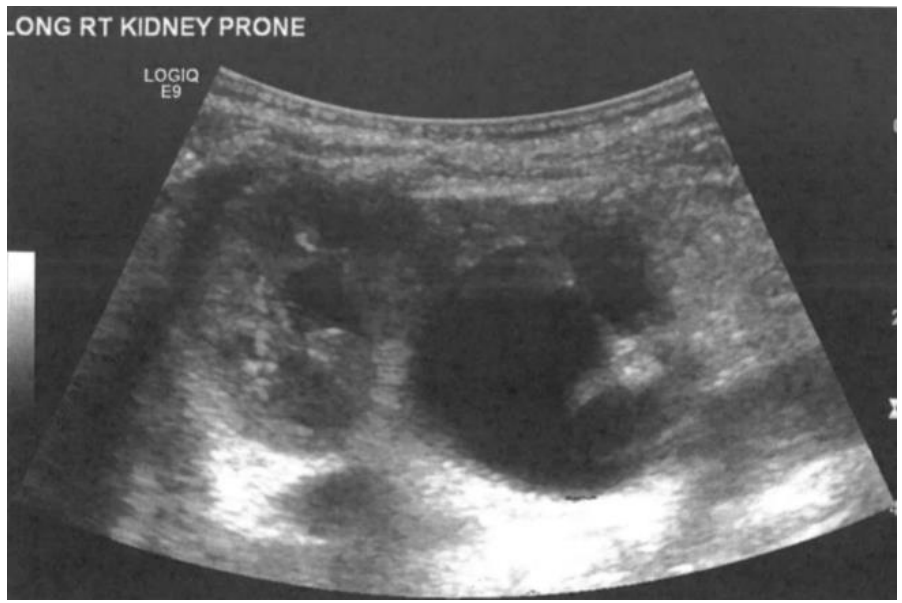
A. VUR.
B. aberrant crossing vessels.
C. UPJ obstruction.
D. ureterovesical junction obstruction.
E. redundant ureter.

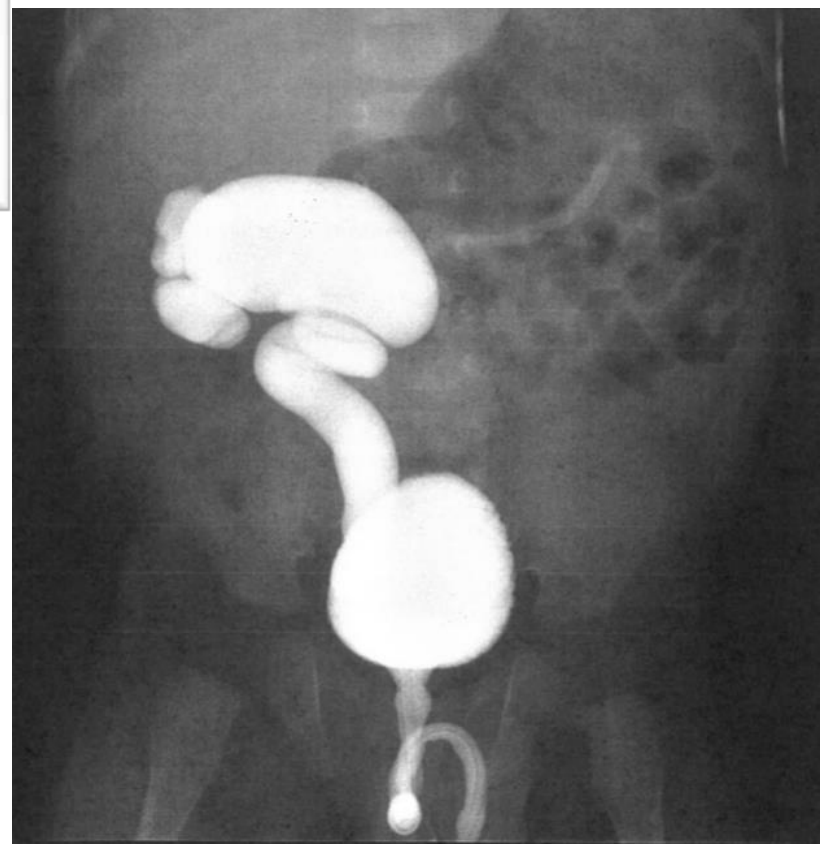
Question #69 ANSWER=C

The axis of the ectopic kidney is slightly medial or vertical, but it may be tilted as much as 90 degrees laterally so that it lies in a true horizontal plane. The renal pelvis is usually anterior (instead of medial) to the parenchyma because the kidney has incompletely rotated. As a result, 56% of ectopic kidneys have a hydronephrotic collecting system. Half of these cases are a result of obstruction of the ureteropelvic or ureterovesical junction (70% and 30%, respectively), 25% from VUR grade 3 or greater, and 25% from the malrotation alone. VUR has been found in 30% of children with ectopic kidneys. The length of the ureter usually conforms to the position of the kidney; the ureter is occasionally slightly tortuous, but it is rarely redundant. The ureter usually enters the bladder on the ipsilateral side with its orifice positioned normally, except for those unusual cases with ectopic ureters. The arterial and venous network is anomalous, and its vascular pattern depends on the ultimate position of the kidney. There may be one or two main renal arteries arising from the distal aorta or from the aortic bifurcation, with one or more aberrant arteries emanating from the common or external iliac or even the inferior mesenteric artery. The kidney may be supplied entirely by multiple anomalous branches, none of which arise from the aorta. In no instance has the main renal artery arisen from the level of the aorta that would be its proper origin if the kidney were positioned normally.

73. A five-month-old boy is treated for his first febrile UTI. Ultrasound of the right kidney and VCUG are shown. The left kidney is normal. Diuretic renal scan with an indwelling catheter shows diminished function in the right lower pole with a T1/2 time of 20 minutes. The next step is antibiotic prophylaxis and:

A. observation.
B. lower pole pyeloplasty.
C. lower pole to upper pole pyeloureterostomy.
D. common sheath ureteral reimplant.
E. lower pole heminephrectomy.





Question #73

ANSWER=A

This boy has high-grade VUR into the lower pole of a duplicated system. The reduced renal function that is seen on the renal scan is likely secondary to renal dysplasia. Renal dysplasia is not unique to primary isolated VUR but may also occur in a variety of urologic settings. Duplex renal moieties, prune-belly syndrome, and PUVs may all exhibit reflux-associated renal dysmorphism, particularly when the grade of VUR has been high. The incidence of a concomitant UPJ obstruction in patients with VUR ranges from 1-5%, with the incidence being on the higher end in children with high-grade VUR. When significant hydronephrosis is seen in patients with VUR, one may suspect a UPJ obstruction. However, the hydronephrosis is usually secondary to the VUR as opposed to an obstructive defect. Three radiologic signs on a VCUG that might suggest the existence of UPJ obstruction in the setting of VUR are: 1) if the pelvis shows little or no filling of contrast whereas the ureter is filled with contrast; 2) contrast that does enter into the pelvis is poorly visualized because of dilution of the contrast in a large amount of retained urine; 3) poor drainage of retained contrast in the renal pelvis is seen on a drainage film. In this patient, none of these signs are present; thus, UPJ obstruction is very unlikely. In this boy, there is no dilution of the contrast in the pelvis on the VCUG. The delay that is noted on the drainage portion of the renal scan is more likely due to reduced renal function, as opposed to obstruction. In addition, drainage times on diuresis renography have not been found to directly correlate with obstruction in children. With no clear evidence of obstruction, surgery to promote drainage by either pyeloplasty or ureteropyelostomy would not be appropriate at this time. Lower pole nephrectomy is also not indicated at this age after one UTI. The next best course of action in this boy is to observe him on prophylactic antibiotics. Ureteral reimplantation would be too premature in this setting, especially in light of the patient's young age and the size of his bladder.

Khoury AE, Wehbi E: Management strategies for vesicoureteral reflux, in Partin AW, Peters CA, Kavoussi LR, Dmochowski RR, Wein AJ (eds): CAMPBELL WALSH WEIN UROLOGY, ed 12. Philadelphia, Elsevier, 2020, vol 1, chap 29, p 499.

134. A three-month-old girl with antenatal hydronephrosis has persistent severe right pelvicaliectasis and no ureteral dilation on postnatal ultrasound. The left kidney is normal. A diuretic renal scan shows 48% differential function and T1/2 of 27 minutes on the right. The next step is:

A. MR urography.
B. repeat renal ultrasound in three months.
C. retrograde pyelography.
D. ureteral stent placement.
E. pyeloplasty.

Question #134

ANSWER=B

This patient has delayed drainage based on mildly elevated T1/2 (greater than 20 minutes is considered abnormal) with retained renal function. The delayed drainage may be from reservoir effect alone, or there could be some dysfunction or obstruction of the UPJ. MR urography would offer both anatomical and functional information that might help confirm the presence of UPJ obstruction, but the retained renal function on the renal scan is not consistent with a severely obstructed kidney. Infants also require anesthesia to achieve adequate MR urography results. Similarly, a retrograde pyelogram would outline the anatomical findings but expose the patient to general anesthesia. Managing this with ureteral stent placement at this time is not indicated given the excellent differential relative renal function but may be of use in equivocal cases when they are followed by a renal scan to assess a change in renal function. Indications for pyeloplasty include worsening dilation on repeat ultrasound, low or decreasing renal function, urinary tract infections, urolithiasis, or symptoms such as severe flank pain. Infants may also have feeding difficulties or failure to thrive. The majority of cases resolve over time and will not require surgical intervention. Therefore, patients who do not meet indications for immediate surgery should be observed with serial ultrasound and/or repeat renal scan.

Olsen LH, Rawashdeh YFH: Surgery of the ureter in children: Ureteropelvic junction, megaureter, and vesicoureteral reflux, in Partin AW, Peters CA, Kavoussi LR, Dmochowski RR, Wein AJ (eds): CAMPBELL WALSH WEIN UROLOGY, ed 12. Philadelphia, Elsevier, 2020, vol 1, chap 42, pp 826-827.

Koff SA: Neonatal management of unilateral hydronephrosis. Role for delayed intervention. UROL CLIN NORTH AM 1998;25:181-186.

2021

89. A four-month-old boy has a history of prenatal bilateral hydronephrosis. Postnatal ultrasound reveals severe left hydroureteronephrosis. VCUG is normal. A diuretic renal scan shows normal symmetric uptake bilaterally; T1/2 of the right kidney is 8 min; T1/2 of the left kidney is 28 min. The next steps are prophylactic antibiotics and:

A. renal ultrasound in three months.
B. MRI urogram.
C. cystoscopy and distal ureteral dilation.
D. cutaneous ureterostomy.
E. ureteral reimplantation.

Question #89 **ANSWER=A**

The patient has a primary obstructive megaureter. The majority of non-refluxing megaureters run a benign course and resolve spontaneously within the first few years of life. Surgery is to be considered for this condition when patients are symptomatic or have recurring UTIs, progressive unremitting dilation on ultrasound, differential renal function < 40%, and/or significant decreases in differential renal function of 5% or greater on sequential renal nuclear function studies. Since this patient has equal function on renal scan and is asymptomatic, no surgical intervention is warranted at this time. Observation with a renal ultrasound in three months is reasonable to assess changes in the degree of dilation. If it appears to be getting worse, then a repeat renal scan could be obtained to assess any change in function which would also indicate the need for surgical intervention. An MRI urogram at this age would require anesthesia and is unnecessarily invasive and expensive. Ureteral dilation, ureterostomy, or reimplantation are treatment options if intervention is indicated for the development of symptoms or loss of renal function.

Olsen LH, Rawashdeh YFH: Surgery of the ureter in children: Ureteropelvic junction, megaureter, and vesicoureteral reflux, in Partin AW, Peters CA, Kavoussi LR, Dmochowski RR, Wein AJ (eds): CAMPBELL WALSH WEIN UROLOGY, ed 12. Philadelphia, Elsevier, 2020, vol 1, chap 42, pp 849-850.

2020

46. A four-year-old girl voids normally but is continuously wet. A renal ultrasound shows normal-appearing kidneys bilaterally. The next step is:

A. MAG-3 renal scan.
B. VCUG.
C. MRI scan.
D. cystoscopy and vaginoscopy.
E. retrograde pyelogram.

Question #46 ANSWER=C

The clinical scenario of dribbling despite normal voiding creates suspicion of an ectopic ureter. Often the ectopic upper pole moiety of the duplex kidney is very small and not easily identified on ultrasound. In these cases, an MRI scan or MR urogram are the best imaging tests to localize the difficult to identify small, dysplastic upper poles and their ureters. MR urogram is not always required since the T2-weighted images of a standard MRI are particularly suited for finding and defining fluid-filled structures like an ectopic ureter. Sagittal imaging may demonstrate the exact termination of the ectopic ureter. DMSA scan is most useful in the identification of small ectopic kidneys but is unlikely to be useful when the renal ultrasound is normal. If the moiety is small, a MAG-3 renal scan will appear normal because the upper pole often has no function and the lower pole will not deviate. VCUG will sometimes show VUR into an ectopic ureter depending on the location of the orifice. Cystoscopy and vaginoscopy can identify the ectopic orifice, but the orifice is often difficult to identify endoscopically and is not as sensitive as an MRI scan. Retrograde pyelogram is also limited due to difficulty in identifying the ectopic ureteral orifice.

Peters CA, Mendelsohn C: Ectopic ureter, ureterocele, and ureteral anomalies, in Wein AJ, Kavoussi LR, Partin AW, Peters CA (eds): CAMPBELL-WALSH UROLOGY, ed 11. Philadelphia, Elsevier, 2015, vol 4, chap 134, pp 3080-3082.

89. A 34-week gestation male fetus has worsening bilateral hydronephrosis and new bilateral ureteral dilation with normal amniotic fluid. The bladder is full with a wall thickness of 5 mm. The next step is:

A. renal ultrasound at one month after delivery.
B. renal ultrasound and VCUG after delivery.
C. renal scan after delivery.
D. early delivery.
E. fetal MRI scan.

Question #89 ANSWER=B

There is no role for early delivery in the setting of normal amniotic fluid volume. In terms of evaluation, this patient is at risk of UTI because of high-grade kidney dilation, ureteral dilation, and increased bladder wall thickness. Because of male gender, the diagnosis of bladder outlet obstruction, most commonly posterior urethral valves, should be excluded prior to discharge with renal ultrasound and VCUG. Fetal MRI scan would not impact decision making during pregnancy at this juncture for counseling regarding intervention or termination of pregnancy. Finally, a renal scan would not be in order at this stage, but may have a role in follow-up at some point to determine renal function and/or ureteral drainage.

Nguyen HT, Benson CB, Bromley B, et al: Multidisciplinary consensus on the classification of prenatal and postnatal urinary tract dilation (UTD classification system). J PED UROL 2014;10:982-998.

Nguyen HT, Herndon CD, Cooper C, et al: The Society for Fetal Urology consensus statement on the evaluation and management of antenatal hydronephrosis. J PED UROL 2010;6:212-231.

123. A 13-year-old girl has a 2.5 cm stone in the left mid-pole of a horseshoe kidney. Anatomical consideration(s) for surgical treatment is/are:

A. ureters usually insert in a caudal, ectopic location in the bladder.
B. renal pelvises are positioned posteriorly.
C. calyces are fewer than normal.
D. calyces point posteriorly.
E. isthmus is located adjacent to S2-S4.

Question #123 ANSWER=D

In a horseshoe kidney, calyces are normal in number and point posteriorly as the kidney fails to rotate. This consideration is relevant as this anatomical feature guides the technique of percutaneous access. The ureter usually inserts in a normal position on the bladder. Renal pelvises are also anteriorly placed, not posterior due to failure of rotation. The isthmus is inferiorly placed, just below the inferior mesenteric artery, adjacent to L3 or L4.

Shapiro E, Telegrafi S: Anomalies of the upper urinary tract, in Wein AJ, Kavoussi LR, Partin AW, Peters CA (eds): CAMPBELL-WALSH UROLOGY, ed 11. Philadelphia, Elsevier, 2015, vol 4, chap 130, p 2975.

2019

38. A two-month-old girl with prenatal hydronephrosis is on antibiotic prophylaxis and has a serum creatinine of 0.3 mg/dL. Bilateral hydroureteronephrosis is identified on ultrasound and VCUG shows no VUR. MAG-3 diuretic renal scan reveals equal function with a T 1/2 of 21 minutes on the right and 87 minutes on the left. The next step is:

A. left cutaneous ureterostomy.
B. left ureteral reimplantation.
C. bilateral ureterostomy.
D. repeat ultrasound in one month.
E. MR urogram.

Question #38 **ANSWER=D**

Over 90% of antenatally detected megaureters will improve with conservative management. The T 1/2, or Lasix washout time, especially in neonatal megaureters is not a reliable indicator of obstruction, so a normal creatinine and symmetric renal function support initial observation in this child. An end cutaneous ureterostomy is indicated in the neonate with a megaureter and sepsis, ipsilateral reduced function (less than 35% in a neonate), or in cases of marked or increasing hydroureteronephrosis. A tapered reimplant is almost never indicated in a neonate with a megaureter. MR urography gives improved anatomic detail, but the site of narrowing is fairly constant in megaureters and exact delineation is not required to determine the best management. Conservative management with follow-up ultrasonography is the best next step since this will likely spontaneously improve.

Olsen LH, Rawashdeh YFH: Surgery of the ureter in children, in Wein AJ, Kavoussi LR, Partin AW, Peters CA (eds): CAMPBELL-WALSH UROLOGY, ed 11. Philadelphia, Elsevier, 2015, vol 4, chap 133, pp 3066-3067.

79. A ten-year-old girl has a 1.8 cm renal pelvic stone in a horseshoe kidney with moderate hydronephrosis. The next step is:

A. SWL.
B. ureteroscopy and laser lithotripsy.
C. PCNL.
D. laparoscopic pyelolithotomy.
E. open pyelolithotomy.

Question #79 **ANSWER=C**

Patients with horseshoe kidneys and stone burden > 1.5 cm are best managed with PCNL. If stone burden is < 1.5 cm, both SWL and ureteroscopy have been successful, but the single procedure stone clearance rates are lower than those reported with PCNL. The approach for PCNL is usually through a superior, posterior calyx in a horseshoe kidney. Laparoscopic or open pyelolithotomy would be not be indicated and may be technically difficult due to aberrant vasculature.

Matlaga BR, Krambeck AE, Lingeman JE: Surgical management of upper urinary tract calculi, in Wein AJ, Kavoussi LR, Partin AW, Peters CA (eds): CAMPBELL-WALSH UROLOGY, ed 11. Philadelphia, Elsevier, 2015, vol 2, chap 54, p 1260.

2018

21. A six-month-old boy has moderate hydronephrosis in a solitary kidney diagnosed antenatally. A VCUG is normal. MAG-3 diuretic renal scan shows a washout time of 25 minutes. His urinalysis is normal and his serum creatinine is 0.7 mg/dL. The next step is:

A. MAG-3 diuretic renal scan in three months.
B. ultrasound in three months.
C. antegrade pressure perfusion study (Whitaker test).
D. percutaneous endopyelotomy.
E. pyeloplasty.

Question #21

ANSWER=E

The patient has a solitary kidney with a prolonged washout and elevated serum creatinine. Pyeloplasty is the correct response. All the other conservative options would prolong his obstruction and a Whitaker test has potential complications, especially in a solitary kidney. Endopyelotomy has not proven equivalent in terms of success rates nor as safe as pyeloplasty in this age group.

Peters CA: Congenital urinary obstruction: Pathophysiology, in Wein AJ, Kavoussi LR, Partin AW, Peters CA (eds): CAMPBELL-WALSH UROLOGY, ed 11. Philadelphia, Elsevier, 2015, vol 4, chap 132, p 3055.

49. An eight-year-old girl has urinary urgency, urge incontinence, and constant leakage of urine. She is started on timed voiding and has improvement with the urge incontinence but still has constant urinary leakage. Renal and bladder ultrasound are normal. The next step is:

A. MRI urogram.
B. MRI scan of the spine.
C. VCUG.
D. videourodynamics.
E. MAG-3 renal scan.

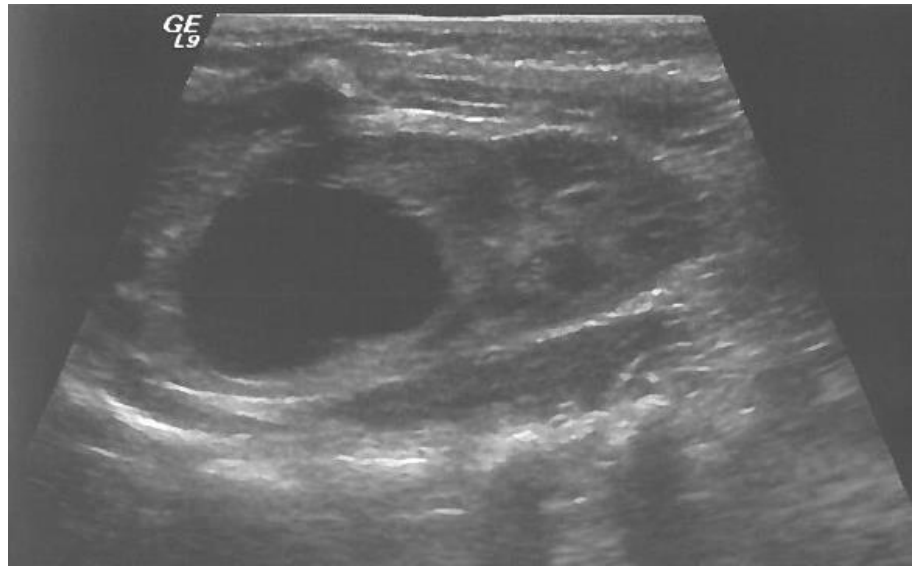
Question #49

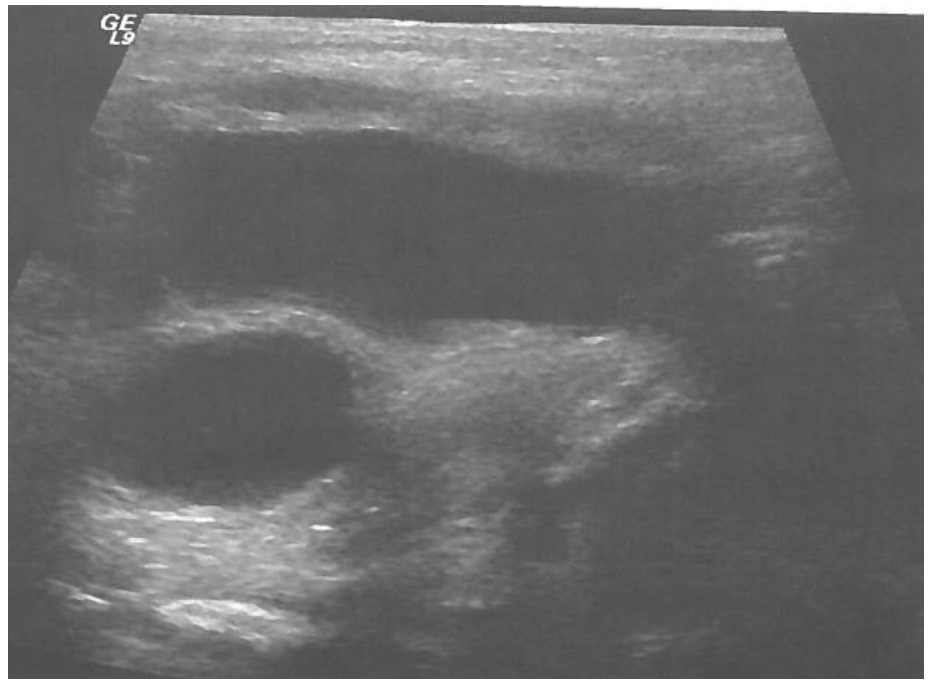
ANSWER=A

The clinical history strongly suggests that this girl has an ectopic ureter even though the ultrasound does not show evidence of this. The absence of an abnormality on ultrasound does not rule-out an ectopic ureter. Occasionally, the renal parenchyma from the upper pole of the kidney that is associated with the ectopic ureter is difficult to visualize on ultrasound and may be identified only by alternative imaging studies. In cases in which an ectopic ureter is strongly suspected because of incontinence, yet no definitive evidence of the upper pole renal segment is found on ultrasound, MRI scan will likely demonstrate the

51. A two-month-old girl has a prenatal history of right hydronephrosis. Neonatal ultrasound images of the right kidney and bladder are shown. The most likely embryologic event to explain this anomaly is:

A. an adynamic distal ureteral segment.
B. a persistence of Chwalla membrane.
C. a cephalad origin of a ureteral bud off the mesonephric duct.
D. a muscular weakness of the trigone of the bladder.
E. early bifurcation of the ureteral bud.





Question #51

ANSWER=C

The images demonstrate a duplicated collecting system of the right side with significant hydronephrosis of the upper pole. The bladder image demonstrates a large cystic structure lateral and posterior to the bladder indicative of a largely dilated ureter. The presence of distinct bladder wall between the lumen of the ureter and bladder distinguishes this as an ectopic ureter rather than a ureterocele. This scenario is most commonly explained by a complete duplication of the right system with an ectopic upper pole ureter. The embryology that explains the pathology of an ectopic ureter is a cephalad origin of the ureteral bud on the mesonephric duct. With an abnormally long common excretory duct, the ureter never becomes incorporated into the bladder, and, therefore, remains ectopic. In the female, the most common locations of an ectopic ureter are the bladder neck, urethra, or Gartner's duct which lies between the urethra and the anterior vaginal wall. An adynamic distal segment would result in a ureterovesical junction obstruction. Persistence of Chwalla membrane would result in a ureterocele. A muscular weakness of the trigone of the bladder would create a diverticulum. Early bifurcation would create a partially duplicated collecting system.

Sources

- https://university.auanet.org/core/pediatric/hydronephrosis/index.cfm?&ct=4952ae3a2fef2b502a756646a9e6a75149418be3978101762340116ddefbb7e4381b90106a3a98c11d7aea6ea26e9511468a56720a45ce387663e96f2765d894
- https://university.auanet.org/core/core.cfm?sectionid=111