IN THE NAME OF GOD

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PUV . UPJO. VUR

Congenital Hydronephrosis

Fetal hydronephrosis ➔in 0.5 to

1 percent of pregnancies

Blyth B, Snyder HM, Duckett JW. Antenatal diagnosis and subsequent management of hydronephrosis. J Urol 1993; 149:693.

It can be detected as early as the 12th to 14th week of gestation

Robyr R, Benachi A, Daikha-Dahmane F, et al. Correlation between ultrasound and anatomical findings in fetuses with lower urinary tract obstruction in the first half of pregnancy. Ultrasound Obstet Gynecol 2005; 25:478.

Society of Fetal Urology (SFU) criteria

1. degree of pelvic dilation,
2. presence of caliceal dilation,
3. presence and severity of parenchymal thinning or atrophy

Society of Fetal Urology (SFU) criteria

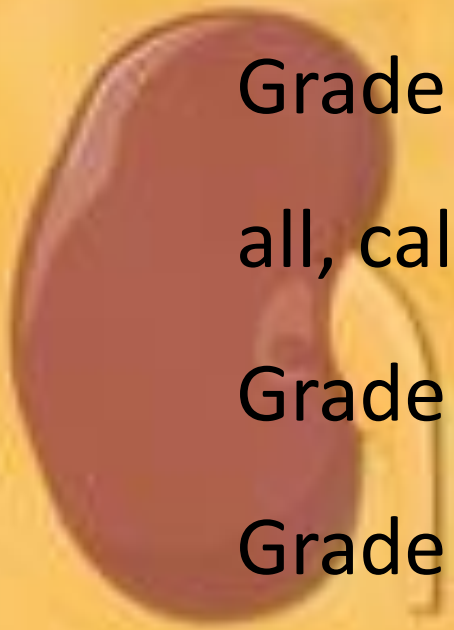
Grade 0: no dilation.

Grade 1: renal pelvis is only visualized.

Grade 2: renal pelvis as well as a few, but not all, calyces are visualized.

Grade 3: virtually all calyces are visualized.

Grade 4: Grade 3 + parenchymal thinning



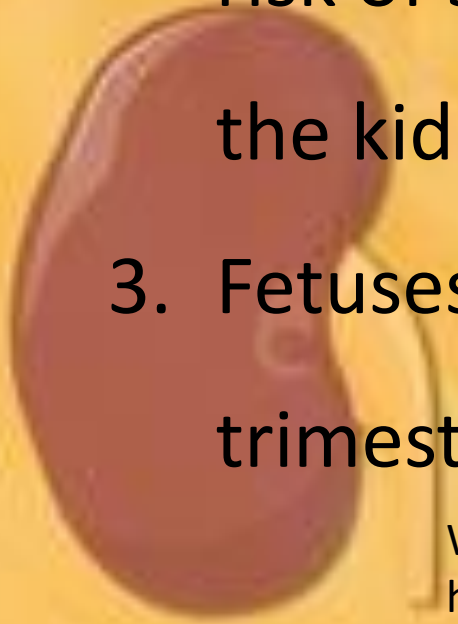
Renal pelvic diameter

the most generally accepted method to define antenatal hydronephrosis

Woodward M, Frank D. Postnatal management of antenatal hydronephrosis. BJU Int 2002; 89:149.

Renal pelvic diameter

1. Mild renal pelvic dilatation: RPD of ≥4 to 10 mm
2. RPD >10 mm is associated with an increased risk of significant congenital anomalies of the kidney and urinary tract (CAKUT)
3. Fetuses with RPD >15 mm during the third trimester are at the greatest risk for CAKUT



Woodward M, Frank D. Postnatal management of antenatal hydronephrosis. BJU Int 2002; 89:149.

A renal pelvic dimension of>5 mm at any gestational age is unusual and dilatation beyond this level should prompt a detailed post-natal urological investigation.

Scott JE, Wright B, Wilson G, et al. Measuring the fetal kidney with ultrasonography. Br J Urol 1995; 76:769.

EPIDEMIOLOGY

- Hydronephrosis occurs approximately **twice as often in males than in females.**
- It is bilateral in 20 to 40 percent of cases

González R, Schimke CM. Ureteropelvic junction obstruction in infants and children. Pediatric Clin North Am 2001; 48:1505.

ETIOLOGY

Based upon postnatal evaluation, most cases of antenatal hydronephrosis are found to be transient (48 percent) or physiologic (15 percent)

Woodward M, Frank D. Postnatal management of antenatal hydronephrosis. BJU Int 2002; 89:149.

ETIOLOGY

Transient — 48 percent

Physiologic — 15 percent

UPJ obstruction — 11 percent

VUR — 9 percent

Megaureter — 4 percent

Multicystic dysplastic kidney — 2 percent

Ureterocele — 2 percent

Posterior urethral valves — 1 percent

Less common causes included ectopic ureter, prune belly, urachal cyst, duplex collecting system , and urethral atresia

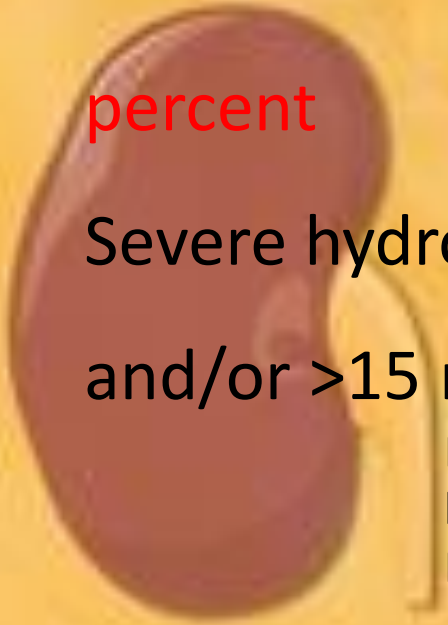
Woodward M, Frank D. Postnatal management of antenatal hydronephrosis. BJU Int 2002; 89:149.

Predictive factors

Mild hydronephrosis (≤7 mm in the second trimester and/or ≤9 mm in the third trimester) — 12 percent

Moderate hydronephrosis (7 to 10 mm in the second trimester and/or 9 to 15 mm in the third trimester) — 45 percent

Severe hydronephrosis (>10 mm in the second trimester and/or >15 mm in the third trimester) — 88 percent

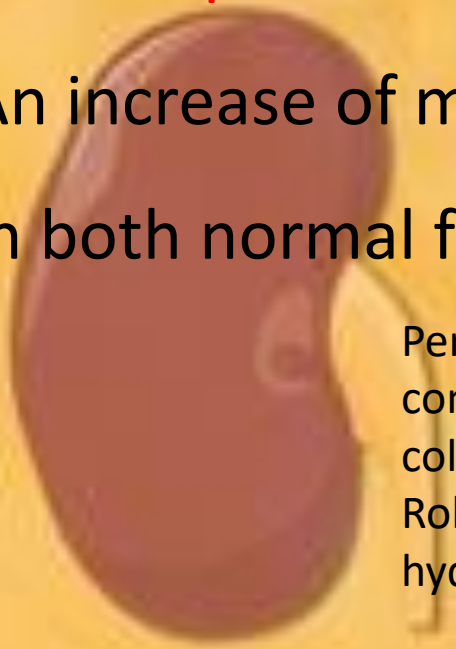


Lee RS, Cendron M, Kinnamon DD, Nguyen HT. Antenatal hydronephrosis as a predictor of postnatal outcome: a meta-analysis. Pediatrics 2006; 118:586.

Ultrasound examination

During the ultrasound examination, the appearance of the fetal renal system can vary in both normal fetuses and those with hydronephrosis. Therefore, this diagnosis should not be based upon a single measurement

An increase of maternal hydration can also increase the RPD in both normal fetuses and those with hydronephrosis



Persutte WH, Hussey M, Chyu J, Hobbins JC. Striking findings concerning the variability in the measurement of the fetal renal collecting system. Ultrasound Obstet Gynecol 2000; 15:186.
Robinson JN, Tice K, Kolm P, Abuhamad AZ. Effect of maternal hydration on fetal renal pyelectasis. Obstet Gynecol 1998; 92:137.

Ultrasound examination

1. Severity of hydronephrosis
2. Unilateral versus bilateral involvement
3. Ureter
4. Renal parenchyma
5. Bladder
6. Amniotic fluid

Routine karyotyping is not recommend for all cases of isolated hydronephrosis.

1-additional fetal anomalies are detected,
2-advanced maternal age,
3-abnormal maternal serum screening tests for Down syndrome.

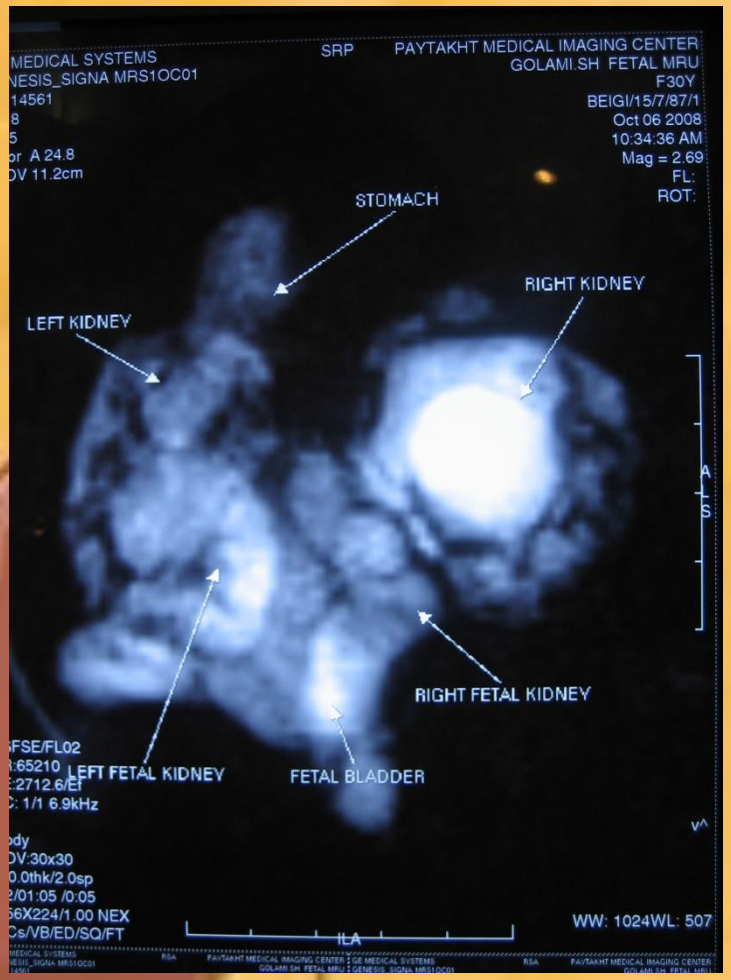
The risk of renal and urinary tract abnormality increases with the

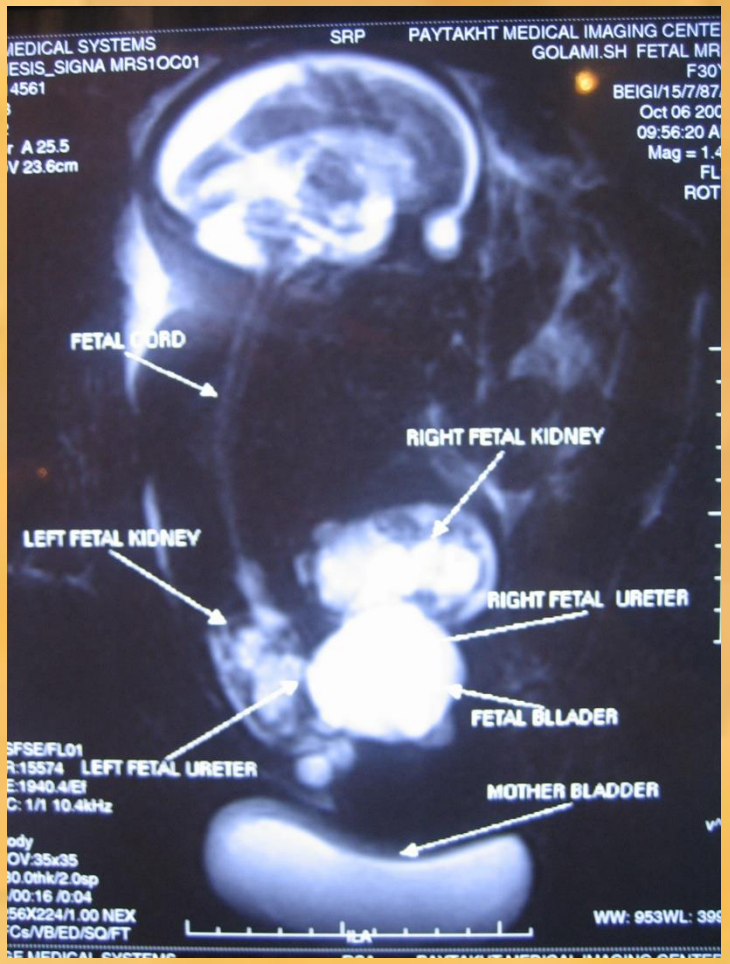
1-**severity** of hydronephrosis,
2-**persistence** of hydronephrosis into the third trimester,
3-**bilateral** involvement, and
4-presence of **oligohydramnios**

MRI

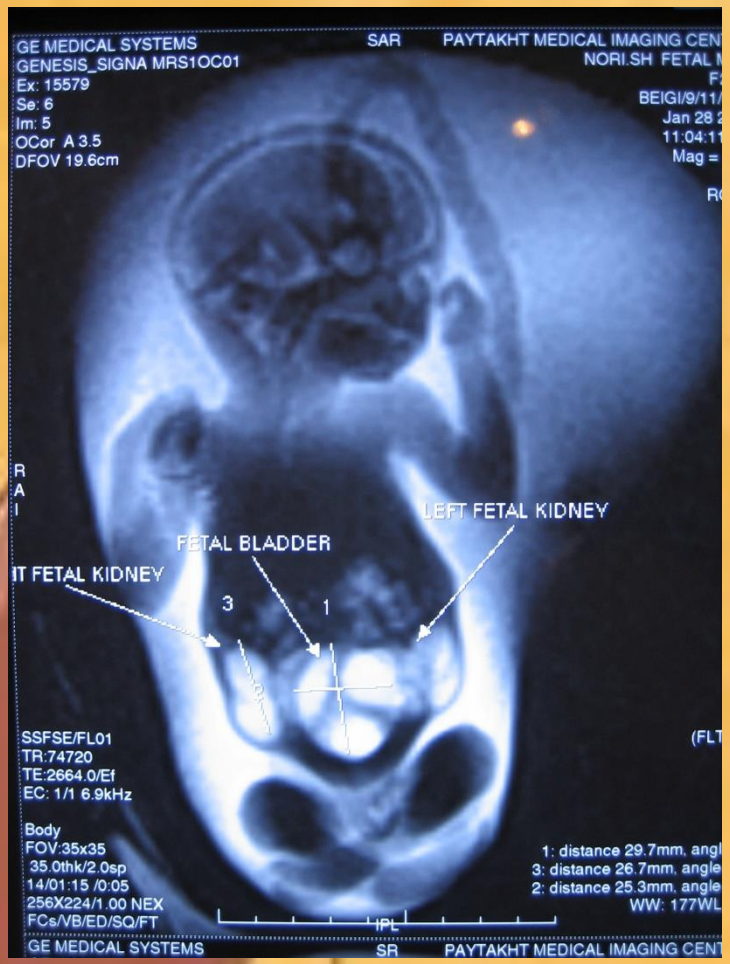
when there is gross oligohydramnios and poor resolution on the ultrasonography examination.

Fetal MRU PUV

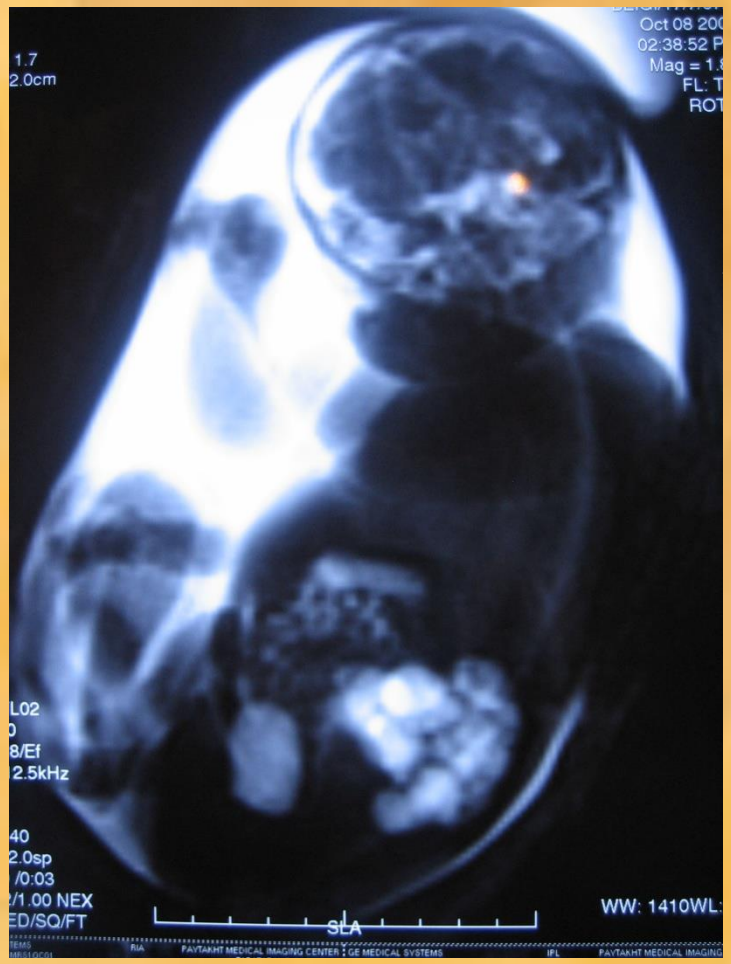




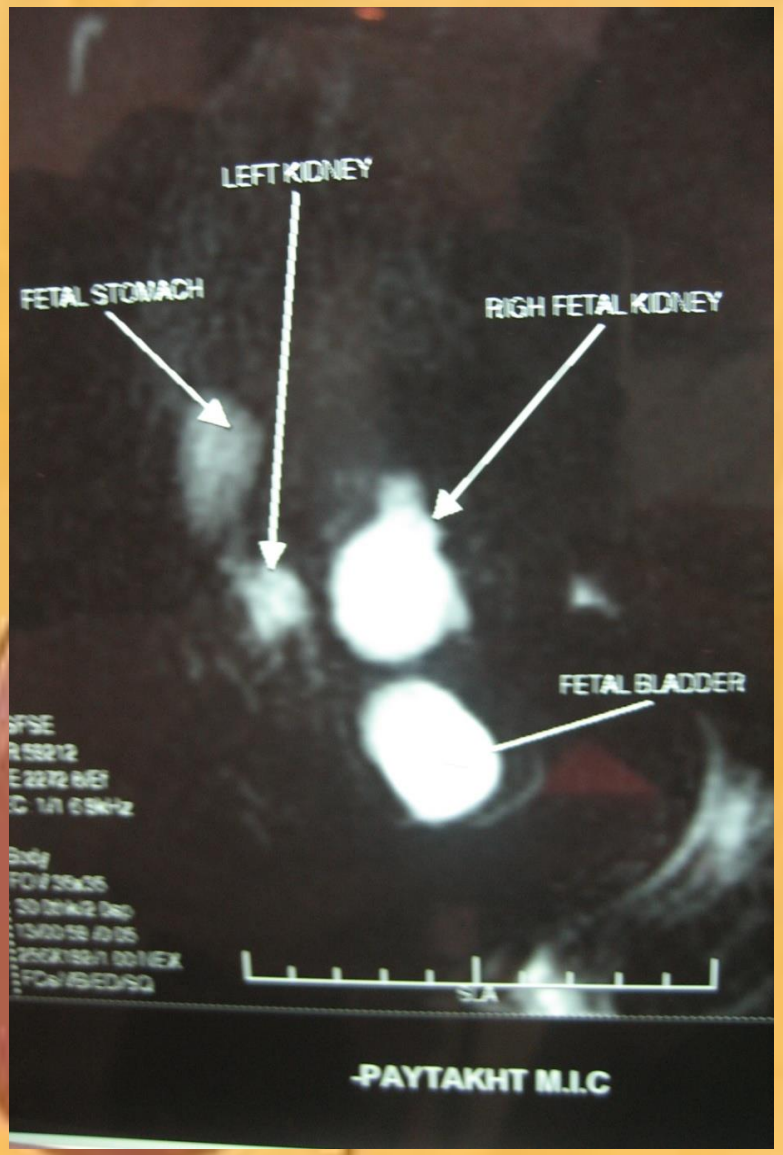
Fetal MRU Prune Belly Synd

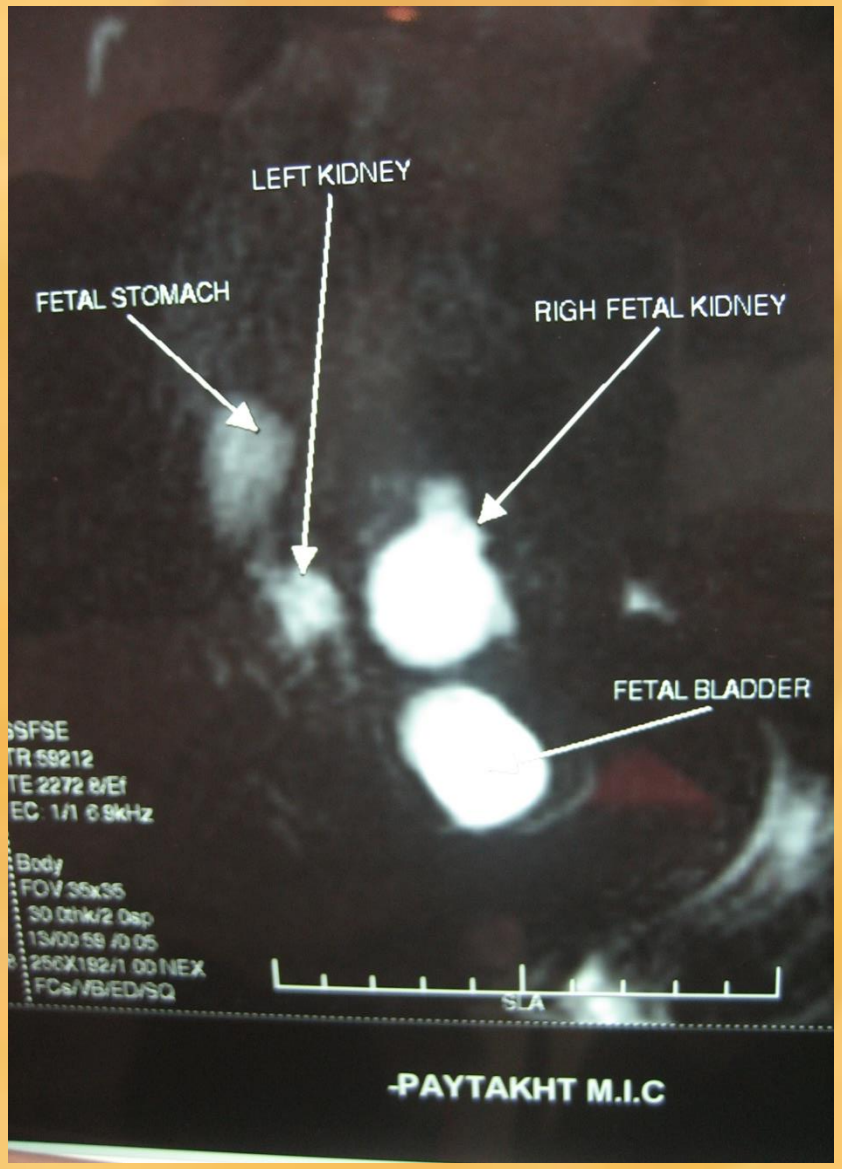


Fetal MRU MCDK



Fetal MRU UPJO





Postnatal Management

Evaluation includes physical examination and the use of radiologic studies to detect renal and urinary tract abnormalities including obstructive uropathy or vesicoureteral reflux (VUR)

POSTNATAL RADIOLOGIC STUDIES

Ultrasonography

- Should be **avoided** in the first two days after birth
- infants with bilateral hydronephrosis and those with a severe hydronephrotic solitary kidney require urgent evaluation on the first postnatal day
- For unilateral hydronephrosis without antenatal bladder pathology :one to four weeks after birth

Voiding cystourethrogram

- to detect VUR and
- in boys, to evaluate the posterior urethra
- Circumcision
- Full-dose coverage during VCUG

Diuretic renography

- in infants with persistent hydronephrosis, & RPD >15 mm
- usually ordered after a VCUG has demonstrated no vesicoureteral reflux
- diuretic renography can be performed after six weeks of life because surgical intervention is rarely required

Chung S, Majd M, Rushton HG, Belman AB. Diuretic renography in the evaluation of neonatal hydronephrosis: is it reliable? J Urol 1993; 150:765.

Diuretic renography

1-Drainage Curve

2- Split Renal Function

3-Postvoid scan

4-Delayed (3 hr) scan

5-T 1/2

If the VCUG is negative (no reflux) and the infant has persistent postnatal hydronephrosis that is not severe (renal pelvis diameter < 15 mm), a follow-up ultrasound is performed at three months of age

Magnetic resonance urography

- ✓MRU can more clearly define the anatomy
- ✓The disadvantage → often requires a general anesthesia or heavy conscious sedation in children.
- ✓Contrast agent gadolinium can only be used if the renal function is normal because of reports of irreversible renal fibrosis in patients with renal insufficiency.

Antibiotic prophylaxis

- ***Infants with antenatal hydronephrosis are nearly 12 times more likely to have pyelonephritis-related hospitalizations in the first year of life.***
- This association is stronger in girls.

Walsh TJ, Hsieh S, Grady R, Mueller BA. Antenatal hydronephrosis and the risk of pyelonephritis hospitalization during the first year of life. Urology 2007; 69:970.

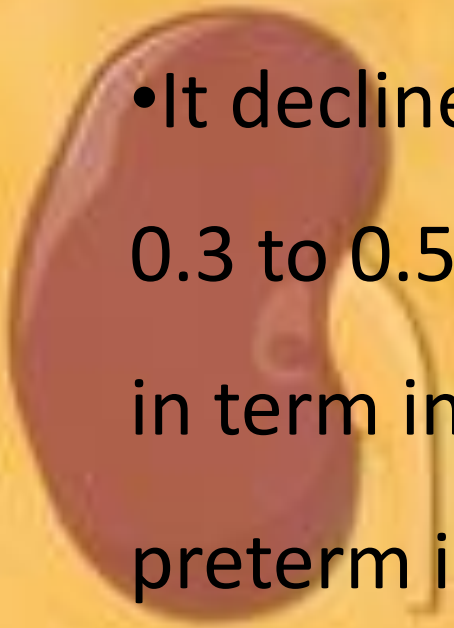
Antibiotic prophylaxis

The risk of infection rises if there is an underlying urologic abnormality, such as VUR or obstructive uropathy, and is greater in girls compared to boys

Coelho GM, Bouzada MC, Lemos GS, et al. Risk factors for urinary tract infection in children with prenatal renal pelvic dilatation. J Urol 2008; 179:284.

Serum creatinine concentration

- At birth is similar to that in the mother (usually ≤1.0 mg/dL).
- It declines to normal values (serum creatinine 0.3 to 0.5 mg/dL) in approximately **one week** in term infants and **two to three weeks** in preterm infants



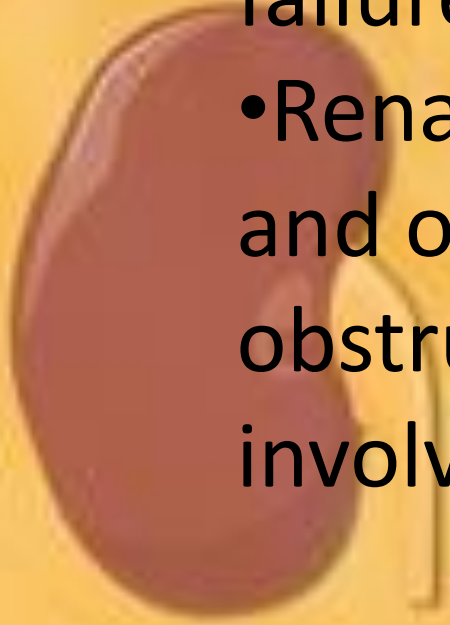
UPJO

- 1 in 500 live births screened by routine antenatal ultrasound
- Lesions are found more frequently on the left than on the right side.
- The reported rate of bilateral involvement ranges from **10 to 40 percent**

Koff, SA, Mutabagani, KH. Anomalies of the kidney. In: Adult and Pediatric Urology, 4th ed, Gillenwater, JY, Grayhack, JT, Howards, SS, Mitchell, ME (Eds), Lippincott Williams and Wilkins, Philadelphia, 2002. p.2129.

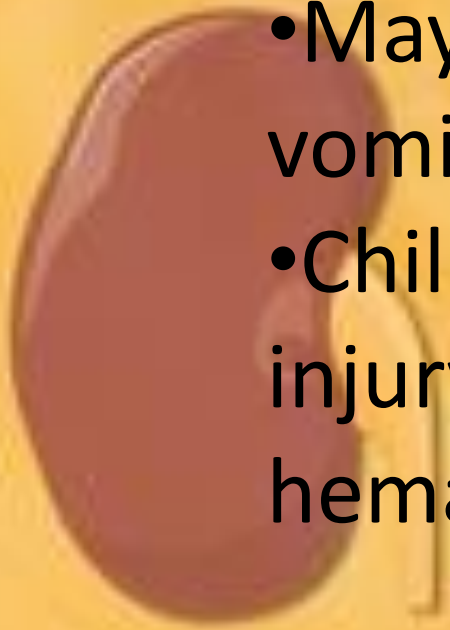
CLINICAL PRESENTATION

- Fetal and neonatal :maternal ultrasound screening
- Palpable abdominal mass
- Urinary tract infection, hematuria, or failure to thrive.
- Renal failure is an unusual presentation, and occurs in infants with a single obstructed kidney or with bilateral involvement.



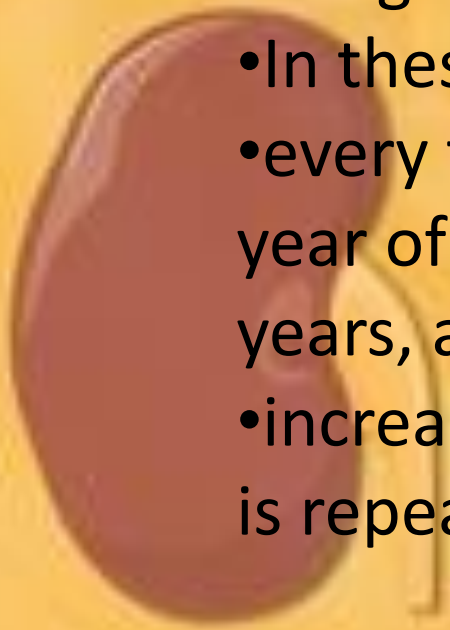
CLINICAL PRESENTATION

- **Older children** :intermittent flank pain or abdominal pain.
- The pain may worsen during brisk diuresis
- May be accompanied by nausea and vomiting
- Children may also present with renal injury after experiencing minor trauma, hematuria, renal calculi, or hypertension



Management - Observation

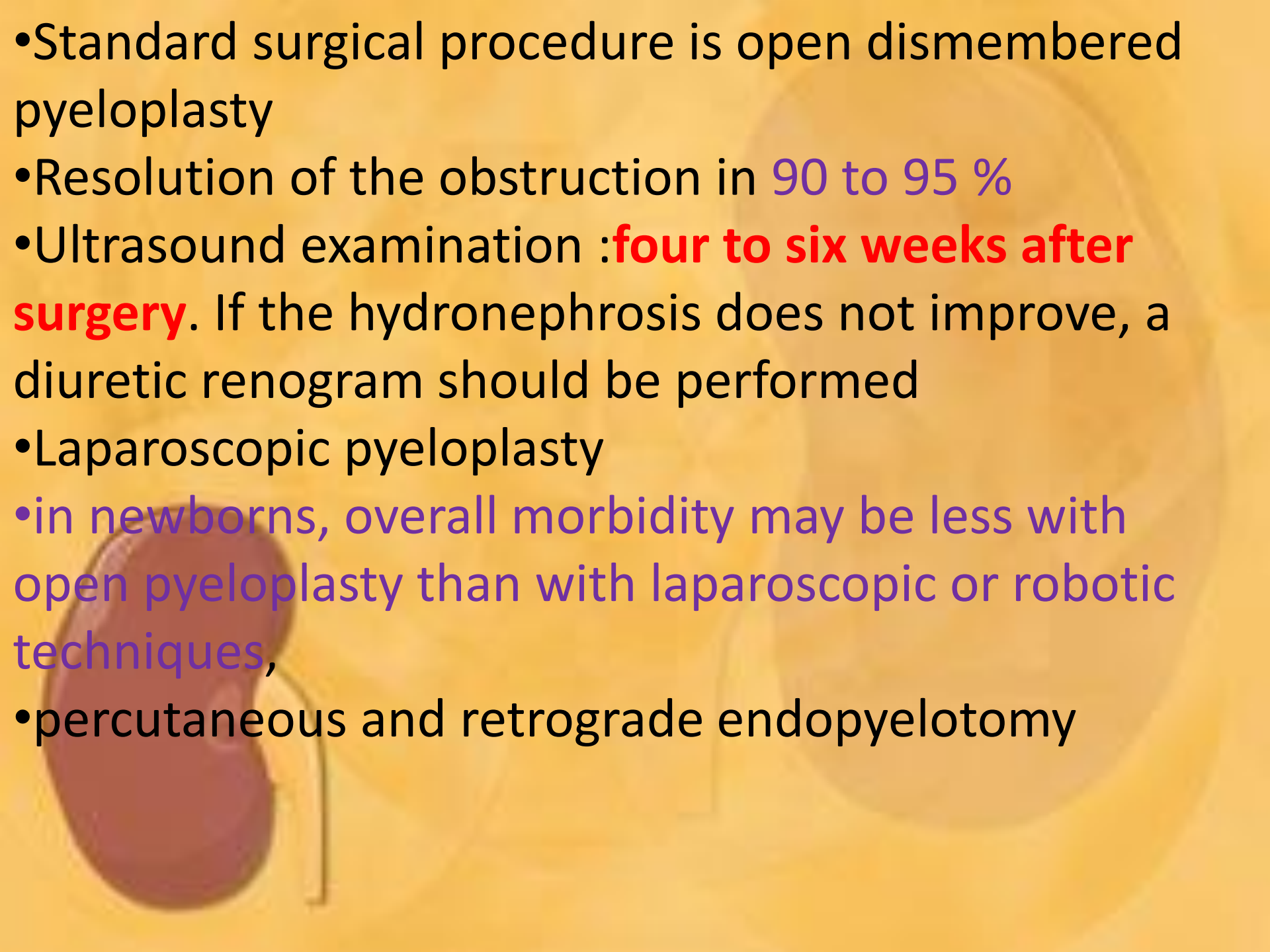
- the main criterion for observation in cases with significant hydronephrosis on sonogram is demonstration of greater than 40 percent of split renal function of the affected kidney by diuretic renography, **even if washout is delayed**
- In these patients → serial renal ultrasounds,
- every four months until the child reaches one year of age, every six months for the next two years, and then annually
- increasing hydronephrosis → diuretic renography is repeated to confirm that renal function is stable.



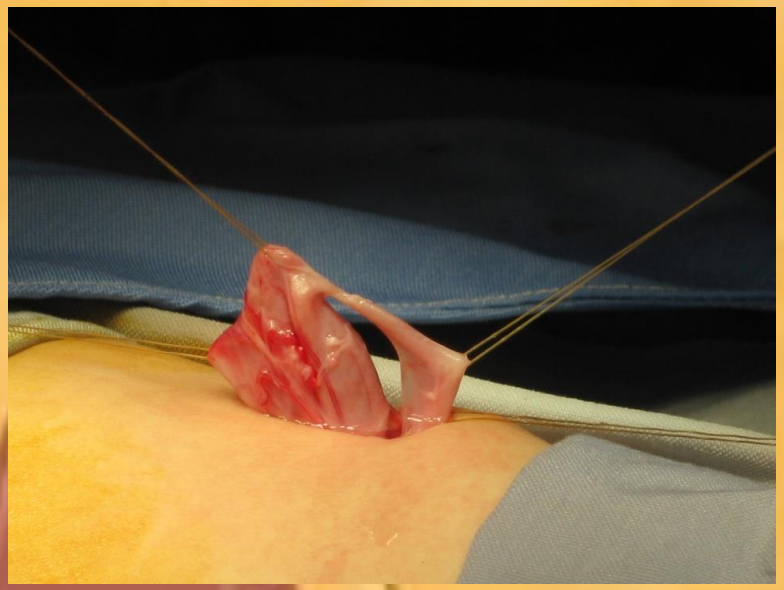
Management - Surgical

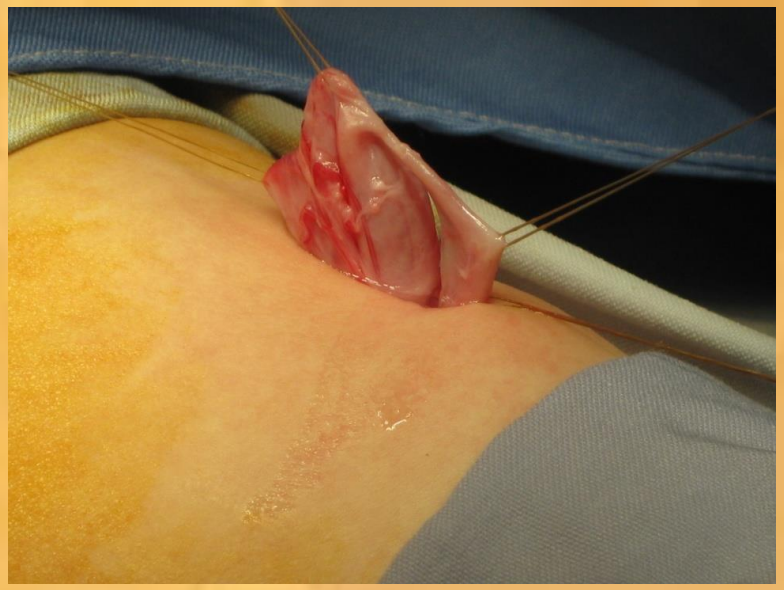
- Symptoms including pain, infection, and renal stones
- Increasing grade of hydronephrosis, and decrease in function of the affected kidney so that it is less than the threshold of 40 percent of split renal function, or there is a serial loss greater than 10 percent
- Parental and/or patient preference to avoid continued invasive testing, and allow definitive correction of the problem.
- Massive hydronephrosis with a renal pelvic diameter >50 mm

- Standard surgical procedure is open dismembered pyeloplasty
- Resolution of the obstruction in 90 to 95 %
- Ultrasound examination :**four to six weeks after surgery**. If the hydronephrosis does not improve, a diuretic renogram should be performed
- Laparoscopic pyeloplasty
- in newborns, overall morbidity may be less with open pyeloplasty than with laparoscopic or robotic techniques,
- percutaneous and retrograde endopyelotomy



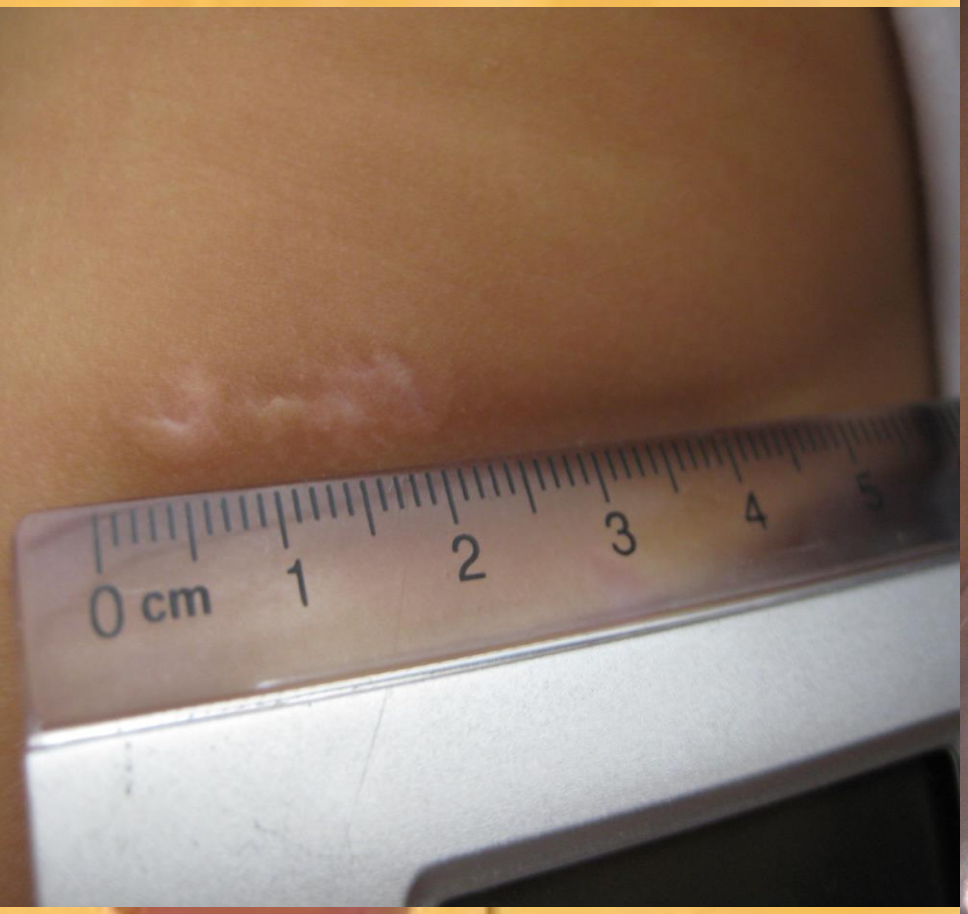
Miniature Pyeloplasty

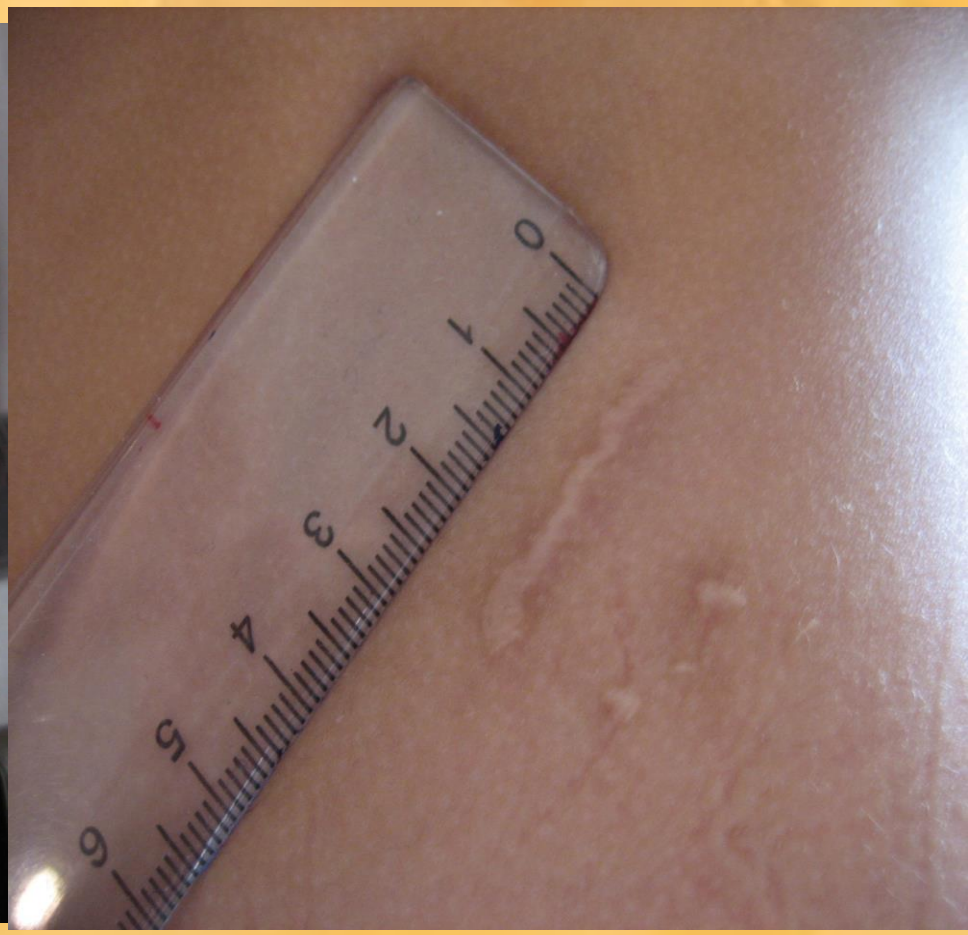




Miniature Pyeloplasty

Postoperative images





vesicoureteral reflux

- occurring in approximately 1 percent of newborns, and as high as 30 to 45 percent of young children with a urinary tract infection
- Primary vs Secondary
- High pressure vs. Low pressure

Indications for imaging

- Girls younger than **three years of** age with a first UTI
- Boys of **any age** with a first UTI
- Children of any age with a **febrile UTI**
- Children with **recurrent UTI**
- First UTI in a child of any age with a family history of renal disease, abnormal voiding pattern, poor growth, hypertension ,or abnormalities of the urinary tract

DIAGNOSIS

• 2010 revised guidelines from the AUA state that an initial evaluation of the child's renal status, growth parameters, and blood pressure should be performed in any child with VUR

- VCUG
- RNC
- DMSA renal scan
- Serum creatinine
- Urinalysis to detect proteinuria

Bladder and bowel (voiding) dysfunction is associated with VUR and recurrent UTI.

Elder JS, Peters CA, Arant BS Jr, et al. Pediatric Vesicoureteral Reflux Guidelines Panel summary report on the management of primary vesicoureteral reflux in children. J Urol 1997; 157:1846.

Spontaneous Resolution

Grades I and II : 80 percent of patients; Not dependent upon age at presentation or whether the reflux was unilateral or bilateral (laterality of reflux).

Grade III : The oldest group of children (5 to 10 years of age) with bilateral reflux had less than 20 percent resolution over five years ;70 percent resolution rate in the youngest group of children (one to two years) with unilateral disease.

Grade IV : 60 percent unilateral disease -- less than 10 percent resolution rate for bilateral

Grade V — Spontaneous resolution rarely

Medical therapy

- daily prophylactic administration of antibiotics
- Dosage 1/4 - 1/2 of full dose
- Nitrofurantoin, trimethoprim, bactrim
- Nitrofurantoin is contraindicated in infants < 2 months old.
- Breakthrough Febrile UTI =>Surgery
- Breakthrough no febrile UTI => change medication or double Px
- Voiding schedule every 2-3 hr
- Complete bladder emptying during micturition
- Elimination of constipation
- Genital & perineal hygiene

Indications for surgery

- breakthrough febrile UTI,
- renal scarring detected by renal radionuclide scan,
- nonresolution, and
- parent preference.

Open reimplantation

- Open surgical :95.9% success rate for correcting VUR in children.
- However, for grade V reflux the success rate is estimated to be 80.7% and
- surgery in infants is also more technically challenging.
- Open surgery is not without its risks with an estimated 2% risk of obstruction and a 2% reoperation rate.

Elder JS, Peters CA, Arant BS Jr, et al. Pediatric Vesicoureteral Reflux Guidelines Panel summary report on the management of primary vesicoureteral reflux in children. J Urol 1997; 157:1846.

- Ureteral reimplantation is a major surgical procedure necessitating a few days of hospitalization.
- Surgery is not free of complications and it has a failure rate of about 20% for grade V VUR.
- A 0.3% to 9.1% rate of obstruction after reimplantation requiring reoperation has been reported

Endoscopic correction

Routh JC, Inman BA, Reinberg Y. Dextranomer/hyaluronic acid for pediatric vesicoureteral reflux: systematic review. Pediatrics 2010; 125:1010.

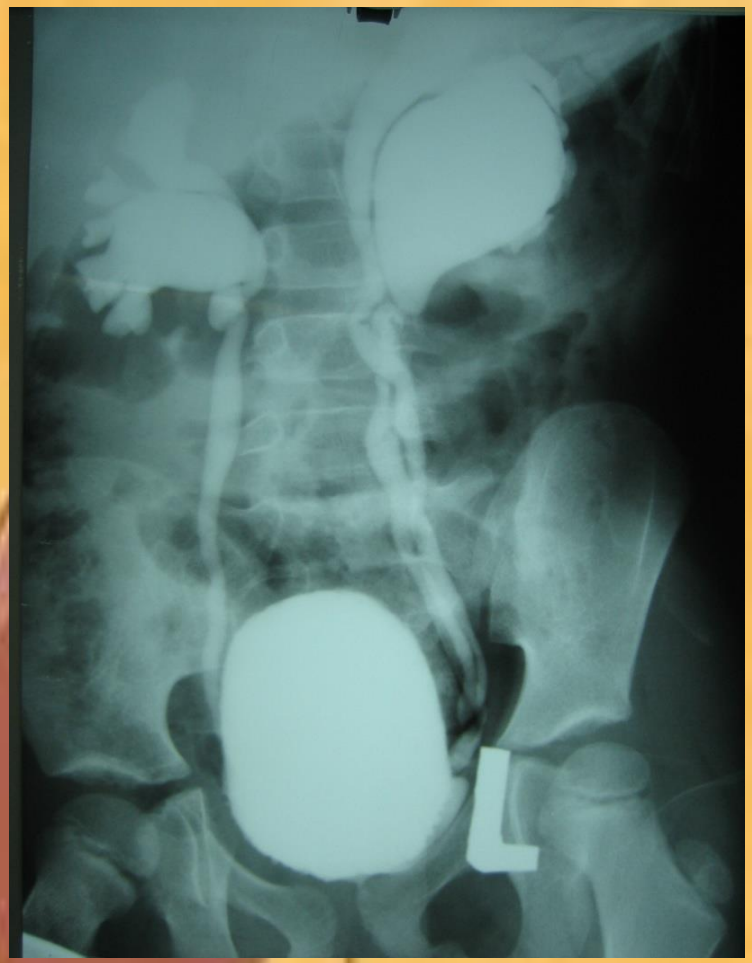
The overall per-ureter Dx/HA success rate was 77% after 3 months

Prem Puri J Urol 2007

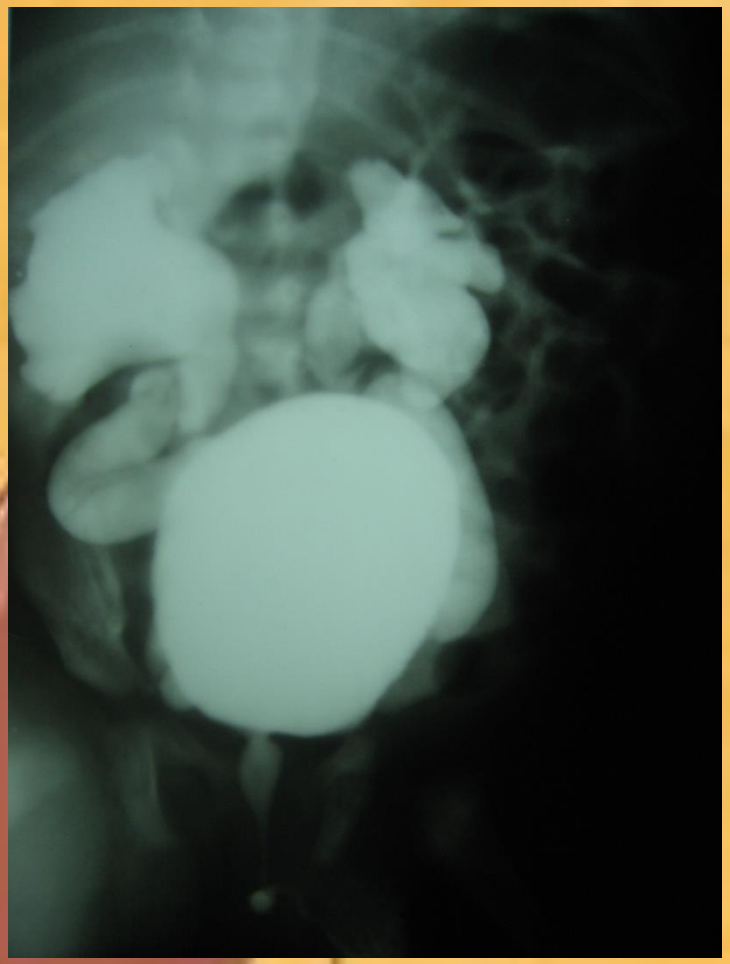
Endoscopic Treatment of Moderate and High Grade Vesicoureteral Reflux in Infants Using Dextranomer/Hyaluronic Acid

- Reflux resolved in 373 ureters (79.9%) after a single endoscopic injection and it was downgraded to grade I in 21 (4.4%). Of the ureters 65 (14%) required a second injection and 8 (1.7%) required a third injection to resolve VUR.

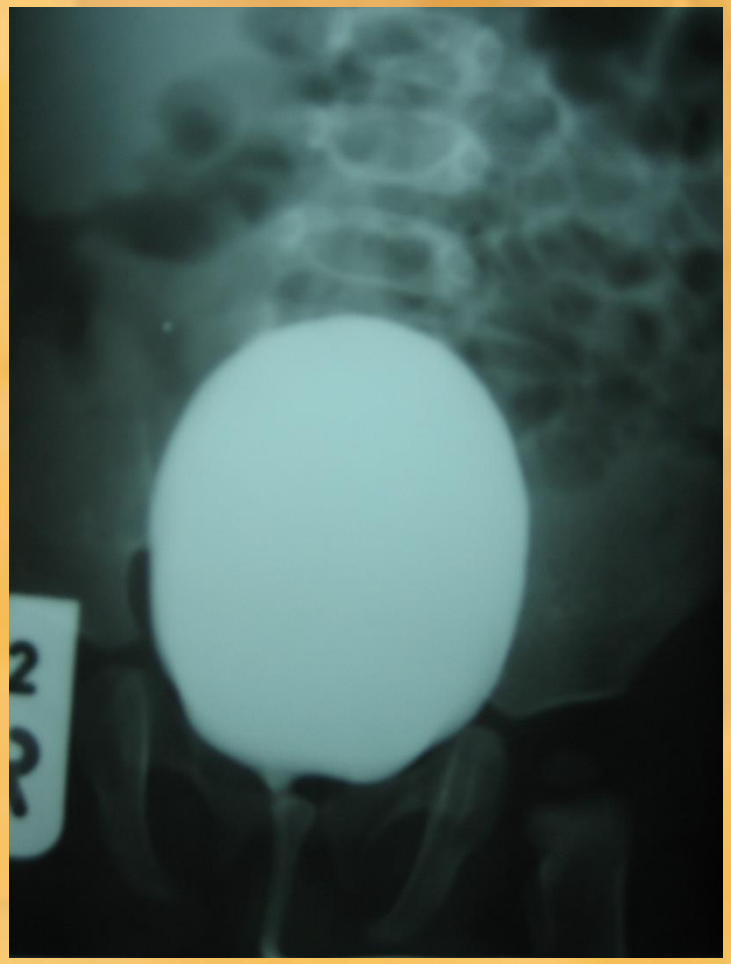
Before & after STING



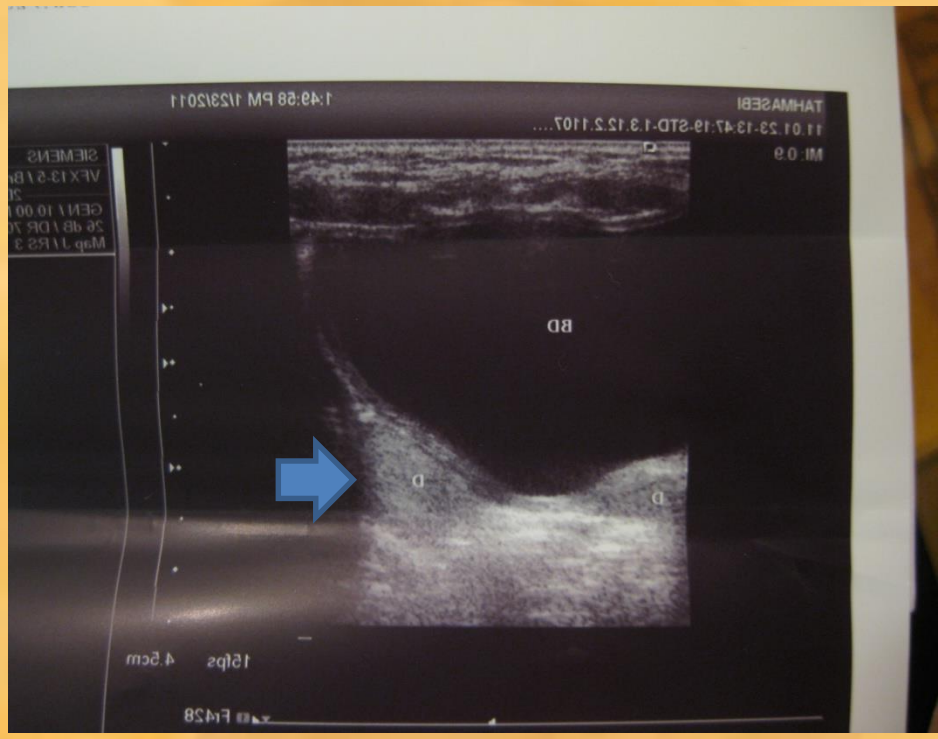
Before STING

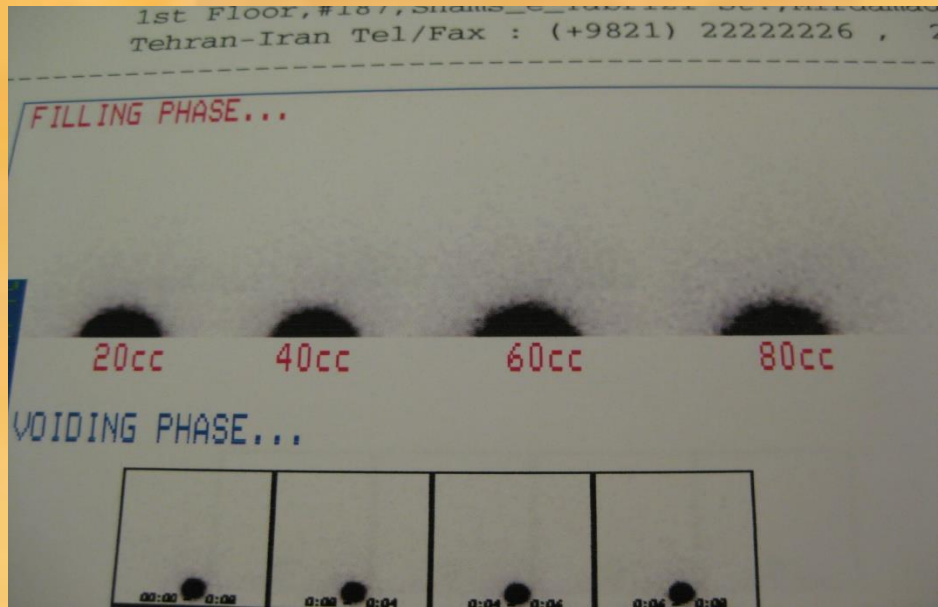


After STING



Before & after STING





Endoscopic Therapy for **Vesicoureteral Reflux:** A Meta-Analysis. I. **Reflux** Resolution and Urinary Tract Infection
J UROL feb 2006

5,527 patients and 8,101 renal units.
Following 1 **treatment the reflux** resolution rate (by ureter) for grades I and II **reflux** was 78.5%, grade III 72%, grade IV 63% and grade V 51%.
Second **treatment** had a success rate of 68%, and the third **treatment** 34%.
The aggregate success rate with 1 or more injections was 85%.
The success rate was significantly lower for duplicated (50%) vs single systems (73%), and neuropathic (62%) vs. normal bladders (74%).

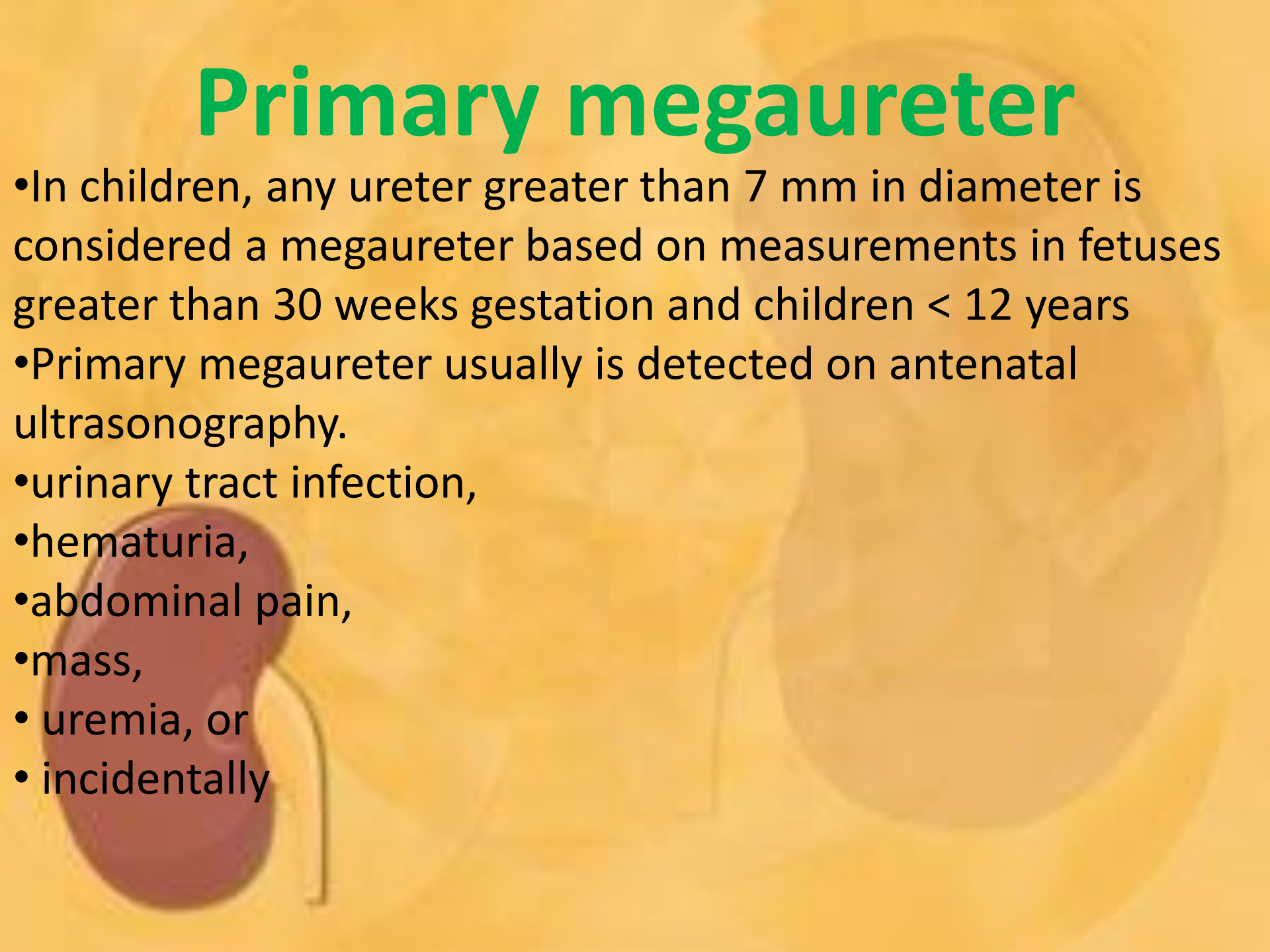
Table 1 Comparison of the main treatment options for VUR

Treatment option	Advantages	Disadvantages	Reflux resolution rate	UTI occurrence post treatment (5-year frequency)
Antibiotic prophylaxis	• No surgical procedure • Low risk of serious adverse events	• Continued need for VCUGs • UTIs continue to occur • Risk of poor tolerability • Risk of poor compliance • Requirement for daily medication • Resolution may not occur for many years, if at all • Risks of antibiotic resistance	16—49% over 4—5 years, depending on reflux grade	29—42%
Ureteral reimplantation	• Highly successful • Little need for subsequent VCUG testing	• Major procedure requiring hospitalization • Postoperative pain is likely • Adverse events such as obstruction are possible	80—99%, depending on reflux grade and surgeon's experience	25—40%
Endoscopic injection (NASHA/Dx gel)	• Minimally invasive procedure • Low risk of adverse effects • Outpatient procedure • Little chance of recurrence after initial cure • Little need for subsequent VCUG testing	• Cure is generally less certain than with open surgery • More than one procedure may be required	70—90%, depending on the injection technique, number of procedures performed, and surgeon's experience	8%

Journal of Pediatric Urology (2006) 2, 539-544 Kirsch

Primary megaureter

- In children, any ureter greater than 7 mm in diameter is considered a megaureter based on measurements in fetuses greater than 30 weeks gestation and children < 12 years
- Primary megaureter usually is detected on antenatal ultrasonography.
- urinary tract infection,
- hematuria,
- abdominal pain,
- mass,
- uremia, or
- incidentally

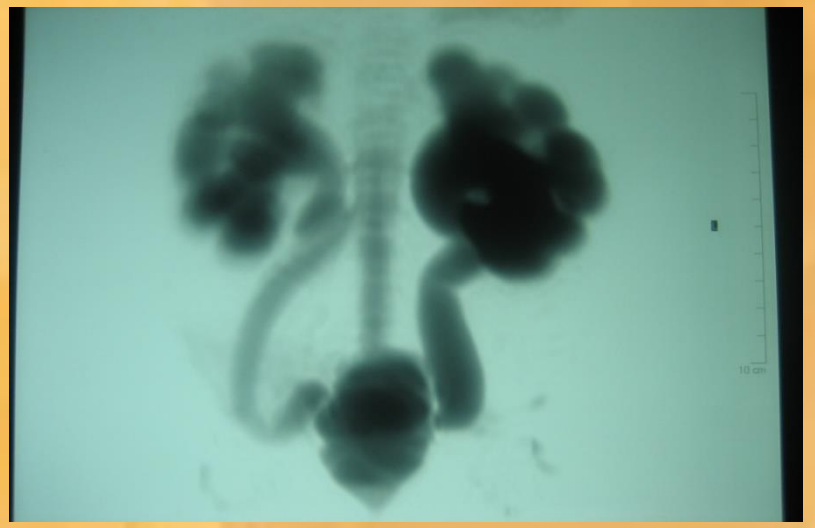


Management

- In symptomatic patients → surgical intervention
- In asymptomatic patients with nonrefluxing, non-obstructed megaureters → nonoperative management ; ***annual ultrasounds and prophylactic antibiotics until the patient is toilet trained. Surgery is indicated if there is subsequent development of symptoms or evidence of obstruction.***
- In asymptomatic patients with refluxing, non-obstructed megaureters → medical treatment with prophylactic antibiotics and ongoing monitoring, or surgical correction
- In asymptomatic patients with obstructed megaureter → surgical correction

Early reimplantation/ before

After



Posterior urethral valves

•The most common cause of lower tract obstruction in the newborn male, occurring in 1 in 5000 to 8000 pregnancies

• USG: persistently distended bladder, and a dilated posterior urethra with a typical keyhole appearance

•The urinary bladder may distend and fill the entire abdomen and may show wall hypertrophy (>2 mm).

•Sometimes the urinary bladder ruptures leading to urinary ascites, urinothorax, and peritoneal calcifications

CLINICAL MANIFESTATIONS

- Prenatal :bilateral hydronephrosis, dilated bladder, and a dilated posterior urethra (keyhole sign), bladder wall thickness In severe cases of obstruction→oligohydramnios ,urinary ascites or a perinephric urinoma.
- Other prenatal renal ultrasound findings may include increased renal echogenicity and cortical cysts suggestive of renal dysplasia,

CLINICAL MANIFESTATIONS

- Neonates :respiratory distress due to lung hypoplasia,abdominal distension due to enlarged overdistended bladder or urinary ascites, difficulty with voiding, or a poor urinary stream
- Infants :failure to thrive, urosepsis, poor urinary stream, and straining or grunting while voiding
- Older boys: urinary tract infections, day time and nocturnal incontinence (enuresis), and other symptoms of voiding dysfunction including frequency, straining to void, a poor urinary stream, and a large urinary volume at each void

About 15 to 20 percent of patients progress to end-stage renal disease (ESRD)

DeFoor W, Clark C, Jackson E, et al. Risk factors for end stage renal disease in children with posterior urethral valves. J Urol 2008; 180:1705.

- early valve ablation can be considered as the primary treatment in the majority of patients, without the need for preoperative drainage or diversion

Sarhan O, Zaccaria I, Macher MA, et al. Long-term outcome of prenatally detected posterior urethral valves: single center study of 65 cases managed by primary valve ablation. J Urol 2008; 179:307.

- VUR is present in about one-third to one-half of patients with PUV
- VUR will resolve in at least one-third of patients with relief of the obstruction.

DeFoor W, Clark C, Jackson E, et al. Risk factors for end stage renal disease in children with posterior urethral valves. J Urol 2008; 180:1705.

- The diagnosis is made by VCUG
- USG : degree of hydroureteronephrosis, BWT, renal echo.

POSTNATAL MANAGEMENT

- Medical management
- Temporary drainage of the urinary tract → soft feeding tube
- **Virgin urethra** Suprapubic Drainage in our ward
- careful monitoring of serum electrolyte including serum bicarbonate, and fluid status

POSTNATAL MANAGEMENT

- ***Primary ablation during cystoscopy is the preferred initial surgical treatment***
- The timing of this procedure is dependent on the overall health status of the neonate and issues with general anesthesia
- Vesicostomy
- Higher diversions

Posterior urethral valves before & after ablation