Pediatric Urinary Disorders

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Objectives:
1-Identify the common congenital anomalies.
2-How to detect this anomaly on radiological investigations.
3-Important steps in management.

Color Index:
-Doctor’s Notes -Surgery Recall -Doctor’s Slides -Important -Extra-Handout notes

Correction File Email: Surgeryteam434@gmail.com
Anatomy of the urinary system is already discussed in **Adult urological disorders**. Please check it out.
Congenital Urinary Disorders

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<th>Anomalies of the Lower Urinary Tract</th>
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<td>Kidney</td>
<td>Urinary Bladder</td>
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Anomalies of the Upper Urinary Tract:

**Kidney:**

1-Renal Agenesis: (absence of the Kidney)

- It has been associated with several genetic syndromes including digeorge, Fraser, Kallmann, trisomy C and D, and cat eye syndromes.

a-Unilateral Renal Agenesis (URA):

- 1 in 1100 births.
- **Male:** Female of 1.8 : 1
- The **left** side is absent more frequently.
- The ipsilateral ureter is completely absent in 50%.
- Anomalies of other organ systems are found frequently in affected individuals.
  - CVS, GIT, MSC

**Complications:**

- 1-absence of the kidney
- 2-not mandatory if the kidney absent that the ipsilateral adrenal gland will be absent because they have different embryology.

**Investigation:**

- 1-do following of blood pressure every year? b/c with hyperfiltration there is risk of hypertension
- 2-dipstick of urine? there is risk of proteinuria

He will continue his life normally (except severe contact sports) because the other kidney will compensate.
continue (URA)...

- Müllerian duct abnormalities occur in 25% to 50% of cases of females with URA compared with wolffian duct anomalies in 10% to 15% of males with URA.

- Approximately one fourth to one third of women with müllerian duct anomalies are found to have URA.

It is very important if you go back to the embryology: urinary system developed at the same time of genitalia system. So, if there is abnormality in one side of one of these system the other system will be affected (if male & have renal agenesis most likely his will be absent.

Diagnosis:
An important differential of URA is renal ectopia, should be ruled out first

How to confirm the absence of the kidney?

1- CT Abdomen. Usually they can not see the kidney absent (not visualize), so we must use DMSA or MRI.

2- DMSA. The diagnostic study, and have two types:

<table>
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<th>Static</th>
<th>Dynamic (the comments one)</th>
</tr>
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<tbody>
<tr>
<td>Renal tissue anatomy, function and shape</td>
<td>Obstruction</td>
</tr>
</tbody>
</table>

One kidney = the diagnosis will be unilateral renal agenesis

a- Bilateral Renal Agenesis (BRA):

Mode of inheritance most likely autosomal dominant

Important environment for the fetus is the amniotic fluid and which is formed by placenta (from 0-16 week), but after that the amniotic fluid formed by urine. So, if there is no kidney = no urine = no amniotic fluid = uterus contraction. Which will cause compression on the whole infant “flat face, nose, and ears” so the lung of the patient will not developed
40% are stillborn.

Do not survive beyond 48 hours due to respiratory distress associated with pulmonary hypoplasia.

The characteristic

*Potter’s syndrome.* they will die in the uterus not from the absence of the kidney but from pulmonary insufficiency / or even if they delivered they will die in the first 24h. nowadays with new supportive of ICU they can live & they need dialysis & transplantation.

Potter Syndrome (PS*) is a term used to describe a typical physical appearance, which is the result of a dramatically decreased amniotic fluid volume (oligohydramnios) secondary to renal diseases such as bilateral renal agenesis (BRA).

Reduced amniotic fluid volume causes increased pressure on the developing fetus, resulting in a sloped forehead, “parrot beak” nose, shortened fingers, and hypoplasia of internal organs, particularly the gut and lungs. Collectively, this sequence of anomalies is known as the Potter sequence.

*Oligohydramnios.*

- Ureters are almost always absent.
- Bladder is either absent or hypoplastic.
- Adrenal glands are usually positioned normally.
- Müllerian duct anomalies are commonly observed.

2-Supernumerary Kidney: (one more extra kidney)(Have its own collecting system)

- Definitive accessory organ with its own collecting system, blood supply, and distinct encapsulated parenchyma.
- Either completely separate or loosely attached to the kidney on the ipsilateral side.
- The ureteral inter-relationships on the side of the supernumerary kidney can be variable.
**Anomalies of Ascent and Form and Fusion**

### Types of Ectopia

<table>
<thead>
<tr>
<th>Simple Renal Ectopia (Pelvic)</th>
<th>Cephalad Renal Ectopia</th>
<th>Thoracic Kidney</th>
<th>Crossed Renal Ectopia</th>
<th>Horseshoe Kidney</th>
</tr>
</thead>
<tbody>
<tr>
<td>(commonest) lower lumbar/Sacral</td>
<td></td>
<td></td>
<td></td>
<td>consider within types of ectopia (kidneys mate together either in the lower pole 90%, or in the upper pole 10%)</td>
</tr>
</tbody>
</table>

Instead of being in its usual position it’s in the pelvis or somewhere else.

Ectopia can be either simple or CROSS, in cross ectopia the ureters cross over to the other side.

- The left is more than the right.
- Pelvic ectopia has been estimated to occur in 1 of 2100 to 3000 autopsies.
- 50% have a **hydronephrosis**:  
  - ✓ Obstruction: UPJO and UVJO  
  - ✓ Reflux: grade III or greater  
  - ✓ Malrotation
- VUR is found in 30%
- The incidence of genital anomalies in the patient with ectopia is about 15%.
- Most ectopic kidneys are clinically **asymptomatic** found with rotin US or the patient present with any other reasons like vague abdominal pain.

### *with Fusion*  
### *without Fusion*

- **Crossed ectopic**: kidney is located on the side opposite from that in which its ureter inserts into the bladder.  
  - 90% are fused with their mate  
  - the superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney.  
  - The ureter from each kidney is usually orthotopic.

Who to differentiate between Simple and Crossed Ectopia?

In crossed ectopic, ureter will **cross** the other side while in the simple the kidney will not be ascend to normal position.

**CT**: ureter **crossing** the other side

- Occurs 1 in 400 persons.  
- The **isthmus** is bulky and consists of parenchymatous tissue.
- The calyces: normal in number atypical in orientation.  
  - pelvis remains in the vertical or obliquely lateral plane
- The blood supply can be quite variable.
- **Horseshoe kidney** is frequently found in association with other congenital anomalies.
- **UPJ obstruction** in one third.
- 60% asymptomatic.
Continue Anomalies of Ascent and Form and Fusion:

In Simple Ectopia When the kidneys ascend from the pelvis to their permanent location in the upper lumbar region, they come into apposition with the adrenal glands, which develop in situ. During ascent, the kidneys rotate medially so that the hilum, which initially faced anteriorly, (the pelvis is posteriorly and the calyces are anterior), now faces medially, (the pelvis is medial and the calyces are lateral). The segmental vessels supplying the kidney are added cranially and lost caudally during ascent.

Most of horseshoe ectopic and not go to flank area b/c once they fuse together the blood supply from inferior mesenteric artery will not allow the kidney to go up in the normal position.

The blood supply can be quite variable.
In the normal kidney there is one renal artery or sometimes 2 to supply each kidney, but in horse kidney there are several arterial supply b/c it take U shape & every area of the aorta giving blood supply when you want to do nephrectomy put on mind you will not ligate 1 artery (several)

Multicystic Dysplastic Kidney(MCDK)

Doctor did not explain anything here.
The kidney and renal pelvis normally rotate 90 degrees ventro medially during ascent:

- the calyces point laterally.
- the pelvis faces medially.

Fused from the poles usually ectopic and doesn't reach the flank area.

When this alignment is not exact, the condition is known as malrotation.


During ascent, the kidneys rotate medially so that the hilum, which initially faced anteriorly, (the pelvis is posteriorly and the calyces are anterior), now faces medially, (the pelvis is medial and the calyces are lateral). The segmental vessels supplying the kidney are added cranially and lost caudally during ascent.

Usually it is asymptomatic. If there are symptoms, we treat accordingly, but we don’t treat the abnormal kidney itself. If there is hydronephrosis, UTI or reflex, treat them not the malrotated kidney itself.

Ureter:

Presentation:

- Incidental in Neonates
- Incidental in Children
- Symptomatic:
  - UTI, Pain, Mass, Hematuria and Stone
  - usually it is
  - intrinsic: segment is not formed
  - outside: apparent vessels

Routine for any antenatal pregnancy doing US (22,32 weeks of pregnancy)

- Most of the anomalies are detected during pregnancy.
- If missed diagnosed pt. present with abdominal pain, hematuria, UTI, stone formation.
**Dx.** Investigation start with US.
Renal pelvis dilated & communication with calyces to diagnose (mickey mouse sign) pt. as ureteropelvic obstruction junction & ureter usually not dilated. Anything distal to obstruction is normal.

**Multicystic dysplastic kidney:** completely replaced by cyst & there is no renal pelvis & no communication with calyces. Hydronephrosis is significant. 80% of hydronephrosis caused during pregnancy spontaneously go by itself.

**How to confirm? Renogram**

- **Static:** assess anatomy, function e.g. DMSA to see if the kidney is absent or ectopic.
- **Dynamic:** to see if there is obstruction.

Which is significant before taking pt. to surgery? Renogram

- **Green:** curve coming down ⇒ no obstruction
- **Yellow:** contrast (isotope) staying in the kidney ⇒ the kidney is obstructed

**PUJO...**

**Dismembered Pyeloplasty**

[Diagrams of surgical procedures]

1 did not explain
**Ureter:**

**Different between UJP & UVJ in US:** Ureter dilation in UVJ

- Ureteropelvic obstruction: only renal pelvic dilated
- Ureterovesical obstruction: renal pelvic dilated & ureter dilated

**Rx.** Is it significant? Is it affecting kidney function? Is the pt. asymptomatic in the form of loin pain or UTI or US shows increase in hydronephrosis? Function affected or system obstructed or urological anomalies!

**Surgery:** urethral reimplantation (disconnect the ureter & put it again in the bladder)

**An ectopic ureter is any ureter, single or duplex, that doesn't enter the trigonal area of the bladder.**

- Each ureter going to the kidney at the edge of trigone & it opens in the bladder & should be at the triangle of trigone in right & left side. If it’s anywhere except this place ectopic
  - In the bladder = orthotopic
  - Outside the bladder = ectopic
- Male: ectopic site proximal to the sphincter
- DDx. Of recurrent epididymitis or …. ectopic ureter (presented with infection ex.epididymitis) distal to the sphincter...
- Female: it will open in area there is no sphincter (vagina) (distal to sphincter)
- DDx. Of continuous urinary incontinence ectopic ureter (most of the pt. with infection (not a rule))
Ectopic Ureter

In a *duplex* system the ectopic ureter is inevitably the **upper pole ureter** due to its budding from the mesonephric duct later (more cephalad) than the lower pole ureteral bud.

<table>
<thead>
<tr>
<th>Ectopic Ureter</th>
<th>femal</th>
<th>male</th>
</tr>
</thead>
<tbody>
<tr>
<td>ectopic ureter may enter anywhere from the bladder neck to the perineum and into the vagina, uterus, and even rectum.</td>
<td><img src="image1" alt="Image of female ureter" /></td>
<td><img src="image2" alt="Image of male ureter" /></td>
</tr>
<tr>
<td>One of the classic symptoms is <strong>continuous wetting</strong>.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>the ectopic ureter always enters the urogenital system above the external sphincter or pelvic floor, and usually into the wolffian structures including vas deferens, seminal vesicles, or ejaculatory duct.</td>
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</tbody>
</table>

Ureter:

- A cystic dilation of the distal aspect of the ureter
- Located either within the bladder or spanning the bladder neck and urethra.
- Types:
  - Intravesical: Orthotopic, simple, adult type.
  - Extravesical: Ectopic, duplex system, infant type.

**Presentation:**
- Most present with dilatation detected is US & 80% of ureterocele coming with duplicity (1 kidney with 2 ureters ⇒ chick the kidneys)
- Antenatal (U/S)
- Urine retention
- Infection
- Calculus formation

If you see a cystic mass in the bladder (ureterocele) you have to look to the upper tract is it single or duplicity

**Diagnose:** usually during pregnancy we can do implantation because the bladder capacity of a newborn is 50 cc, later on we follow & treat accordingly.

**Treatment:** Puncture (cystoscopy) like a balloon
**Ureter:**

<table>
<thead>
<tr>
<th>Normal anti-reflux mechanism “flap valve”</th>
<th>Presentation</th>
<th>Diagnostic modality</th>
<th>Management</th>
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<tr>
<td></td>
<td>Asymptomatic</td>
<td><strong>MCUG:</strong> to rule out VUR</td>
<td>● Prophylactic antibiotic (to prevent infection, give once/day, dose: 1/3 of the therapeutic dose)</td>
</tr>
<tr>
<td>1. Oblique course as it enters the bladder.</td>
<td>Prenatal</td>
<td>X-ray with contrast in urinary tract: usually the contrast stay in the bladder, if you see any contrast going to the upper tract → reflux</td>
<td>Select abx which is concentrated in the urine.</td>
</tr>
<tr>
<td>2. Proper muscular attachments to provide fixation.</td>
<td>Fluctuated dilatation</td>
<td>● Endoscopic treatment: 1st step before intervention because it’s a morbidity.</td>
<td></td>
</tr>
<tr>
<td>3. Posterior support to enable its occlusion.</td>
<td>Febrile UTIs</td>
<td>● Ureteral reimplantation: (indication to do it: if the patient got breakthrough infection while he is taking antibiotics, if the high grade reflected by 4 &amp; 5 or the upper tract is affected e.g. DMSA show the function of the kidney come &lt; 40)</td>
<td></td>
</tr>
<tr>
<td>4. Adequate submucosal length</td>
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</tbody>
</table>

**Why is it important to know VUR?**

Recurrent UTI lead to → pyelonephritis→ destruct renal function Reflux with infection is the most dangerous one.

Urine usually come from kidney → ureter → bladder → urethra But if it’s going back we call it VUR
The study to rule out reflux is MCUG and it is also used for grading:
Normal: contrast in bladder
Grade I: confined to ureter, contrast is in the distal part of the ureter
Grade II: contrast reaches the kidney but there is no dilation
Grade III: Mild dilation of the renal pelvis and ureter without loss of calyces
Grade IV: moderate dilation but there is loss of calyces
Grade V: severe dilatation and tortuous dilated ureter

**Anomalies of the Lower Urinary Tract:**

**Bladder:**

- Urachal: is the connection between the **umbilicus** and the **bladder**.
- Normally it is closed in male and female.
- If it is not closed --> **patent urachal**.
- Urachal anomalies are usually detected postnatally due to umbilical drainage.
- **Imaging possibilities:**
  - ultrasound, CT and VCUG (voiding cystourethrogram). to confirm urachal abnormalities.
Treatment:
- Asymptomatic cases: conservative treatment with observation due to possible spontaneous resolution.
- Infected urachal remnants:
  - initially treated with drainage & antibiotics
  - followed by surgical excision.
- Nonresolved urachal remnants: excised due to the increased risk of later adenocarcinoma formation.
Bladder:

- Bladder Diverticulum: pouch in the bladder.
- Bladder diverticula can be detected on prenatal ultrasound.
- The gold standard → VCUG, which will reveal possible accompanying VUR.
- Primary diverticula → Most likely caused by a congenitally deficient bladder wall.
  - Arise as a localized herniation of bladder mucosa at the ureteral hiatus.
  - Secondary para-ureteral diverticula → acquired
  - Develop due to existing infra-vesical obstruction.
  - Symptomatic diverticula
    - especially in conjunction with VUR
    - should be treated surgically.
  - Management: if there is indication (if becomes very large) → excise it.

VCUG is the gold standard

Bladder:

- Often associated with duplication anomalies of the external genitalia and lower gastrointestinal tract
  (2 bladders, each ureter open in 1 bladder, 1 or 2 urethra).
- Initial treatment is directed toward
  - renal preservation.
  - prevention of infections.

- Long-term goals: achieving continence and reconstructing the internal and external genitalia.
- Surgeries must be individualized due to:
  - the rarity of the disease.
  - the large variety of presentations.
**Bladder:**

**Classic Bladder Exstrophy:**
- The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.
- Bladder covered by skin, subcutaneous tissue, prelayers of abdominal walls, peritoneum & anterior wall of abdomen.
- In exstrophy all of these are absent. So, if you look to the patient anteriorly you will see the posterior wall of the bladder.
- Bladder exstrophy → you will see the anus

**Cloacal Exstrophy:**
- Urinary tract & GI will open together.
- Cloacal exstrophy → no anus

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Urethral anomalies:

• Emergency, disease of males.
• 1 in 8000 to 25,000 live births.
• Make up 10% of urinary obstructions diagnosed in utero.
• Most common cause of urine retention in male infants.
• 50% have renal impairment.
• The bladder and the kidneys developed under high pressure and resistance.

Between anterior & posterior urethra during embryologically there is canalization from distal to proximal and from caudal to cephalic leading to complete tube without any narrowing, But in Posterior urethral valve incomplete canalization of urethra and leave small membrane (posterior urethral valve) which cause obstruction.
### Presentation
- Antenatal
- Urine retention
- UTI
- Poor urinary stream
- Urinary incontinence
- CRF (ESRD): 40%

### Associated findings
- **Oligohydramnios**
  - Low amount of Amniotic fluid
  - No output of urine or little → Amniotic fluid
  - Low in Ultrasound "because there is no secretion but there is absorption".
  - Obstruction of esophagus → no absorption → Polyhydramnios.
- Bilateral renal dilatation
- VUR: 40%
- Valve bladder
- Renal impairment

### Management:
- **Initial treatment**
- Feeding tube insertion
- Start antibiotic prophylactic
- Ultrasound
- Bilateral hydroureteronephrosis
  - (Dilated posterior urethra & trabeculated bladder (Christmas tree bladder))
  - **MCUG**: filling defect, posterior urethra dilated.
- Treatment: Immediately after birth Endoscopic ablation Or vasectomy if → preterm or low birth weight or there is azotemia or severe infection

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**MCUG**

**Surgical treatment**
- Endoscopic valve ablation
- Cutaneous vesicostomy
## Congenital Genital Disorders:

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<tr>
<th>Hypospadias</th>
<th>Epispadias</th>
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<td>Abnormal position of the EUM (external urethral meatus) on the <strong>ventral surface</strong></td>
<td>Ectopic opening of EUM on the <strong>dorsal surface</strong></td>
</tr>
<tr>
<td>Opening toward scrotum</td>
<td>Opening toward abdomen</td>
</tr>
</tbody>
</table>
| **Types:**  
  Distal hypospadias *(from mid shaft to Glans)*  
  Proximal hypospadias *(from proximal penile “proximal shaft” to the perineal)* | More dangerous commonest type: peno-pubic epispidias it’s opened in bladder neck. Present with urine incontinence. |
| **No circumcision** because the skin will be used in the reconstruction | |
| **When to do repair?**  
  Age 6 to 9 months | |

*Be on the safe side & delay circumcision*
**Prune-Belly Syndrome:**
The incidence: 1 in 29,000 to 1 in 40,000 live births
- Musculoskeletal, urinary & genital tracts are involved.
- The three major findings are
  - deficiency of the abdominal musculature
  - bilateral intra-abdominal testes (undescended)
  - anomalous urinary tract (bilateral ureterohydronephrosis)
- Other names
  - Triad syndrome
  - Eagle-Barrett syndrome
  - abdominal musculature syndrome

**NEUROSPINAL DYSRAPHISMS:**
- The most common cause of neurogenic bladder dysfunction in children is abnormal development of the spinal canal and internecine spinal cord.
- Cutaneous lesions occur in 90% of children with various occult dysraphic states.
- These lesions vary from
  - small lipomeningocele
  - hair patch
  - dermal vascular malformation
  - sacral dimple
  - abnormal gluteal cleft.
- Usually the bladder nerve will be affected.
Antenatal Hydronephrosis (ANH)

Causes:
- Pelviureteric junction obstruction (41%)
- Ureterovesical junction obstruction (23%)
- Vesicoureteric reflux (7%)
- Duplication anomalies (13%)
- Posterior urethral valves (10%)
- MCDK
- Others (6%)

SFU Grading
The doctor didn’t go through these slides

Weigert- Meyer Rule
In complete renal duplication there are two different renal moieties each with its own renal pelvis and ureter and it’s more common in female.

Most of duplex systems are not complicated and do not need consulting BUT if it’s associated with UPM or LPM --> consult

Weigert-Meyer law: UPM --> more distal and medial (longer) --> ectopic ureter and ureterocele  LPM --> more proximal and lateral (shorter) --> VUR, UPJO.

both UPM and LPM commonly present with recurrent UTI’s, hydronephrosis, and flank pain.

1) Upper Pole Moiety:
- Ectopic ureter: ureter that inserts in the bladder neck or urethra (in males in vas deferens/semenal vesicles, in females in vagina)
Clinical presentation: mentioned above +/- urinary incontinence in girls and acute epididymitis in boys.

Investigation: RENAL ULTRASOUND. UPM is usually hydronephrotic and associated with a tortuous hydroureter. VCUG can be used in cases of recurrent UTI.
Management: UMP heminephrectomy is performed to remove poorly functioning UPM and as much of its ectopic ureter as possible.

- Ureterocele: is a cystic dilatation in the distal ureter which can be intravesical OR ectopic. Duplex systems are associated with ECTOPIC ureteroceles while single systems have intravesical. More common in girls and left sided.
Investigation: Renal ultrasound and VCUG, hydronephrosis and tortuous hydroureter are also found here.

2) Lower Pole Moiety:
- VUR: is the most common urinary tract abnormality associated with duplex systems. Diagnosis is by VCUG. --> drooping lily appearance of the collecting system.
Management: prophylactic antibiotics.
Surgical management includes:
reimplantation of the ipsilateral ureters and heminephrectomy of the LPM.

- UPJO: is the most common congenital obstruction in a single system. It’s often associated with crossing of vessels of the renal pedicle. Usually there’s no dilation of the ureter unless there’s high grade VUR

The presence of obstruction can be assessed by using a diuretic renal scan. If there’s significant obstruction with decreased LPM function --> surgical correction (open surgery or laparoscopic pyeloplasty)

Pyeloplasty is the surgical reconstruction or revision of the renal pelvis to drain and decompress the kidney. Most commonly it is performed to treat an uretero-pelvic junction obstruction if residual renal function is adequate.
**SUMMARY:**

<table>
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<tr>
<th><strong>PUJO</strong></th>
<th><strong>US:</strong> isolated hydronephrosis</th>
<th><strong>Pyeloplasty.</strong></th>
<th><strong>Most common cause of antenatal hydronephrosis</strong></th>
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<td><strong>URETEROVESICAL JUNCTION OBSTRUCTION</strong></td>
<td>US: hydroureteronephrosis</td>
<td>ureteral reimplantation</td>
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<td><strong>URETROCELE</strong></td>
<td><strong>MCUG:</strong> filing defect. US: thin-wall cyst in bladder</td>
<td>Endoscopic puncture of ureterocele</td>
<td>Commonest cause of urine retention in female infants</td>
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<td><strong>ECTOPIC URETER</strong></td>
<td>IVP, VCUG, cystoscopy</td>
<td>-</td>
<td>In a female → continuous wetting, but a male will not present with that</td>
</tr>
<tr>
<td><strong>VESICOURETERIC REFLUX</strong></td>
<td><strong>MCUG</strong></td>
<td>Medical management: UTI → prophylaxis Surgical management: Ureteral reimplantation</td>
<td></td>
</tr>
<tr>
<td><strong>POSTERIOR URETHRAL VALVE</strong></td>
<td><strong>MCUG:</strong> Posterior urethra dilated, Christmas tree bladder US: Bilateral renal dilatation, Oligohydramnios</td>
<td>Endoscopic primary valve ablation</td>
<td></td>
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</tbody>
</table>

Thank you