Congenital Anomalies of the Kidney and Urinary Tract (CAKUT)

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CAKUT - Categories

- Renal parenchymal anomalies
  - Renal dysplasia, MCDK, ADPKD, ARPKD, renal agenesis
- Collecting system anomalies
  - Hydronephrosis, UPJ obstruction, megaureter, megacalycosis, ectopic ureter, ureterocele, duplicated kidney, VUR
- Renal ascent and fusion anomalies
  - Ectopic kidney, horseshoe kidney, crossed renal ectopia (fused/unfused)
- Lower urinary tract anomalies
  - Posterior urethral valves, anterior urethral valves, urethral atresia, megalourethra, prune belly syndrome, urachal remnants

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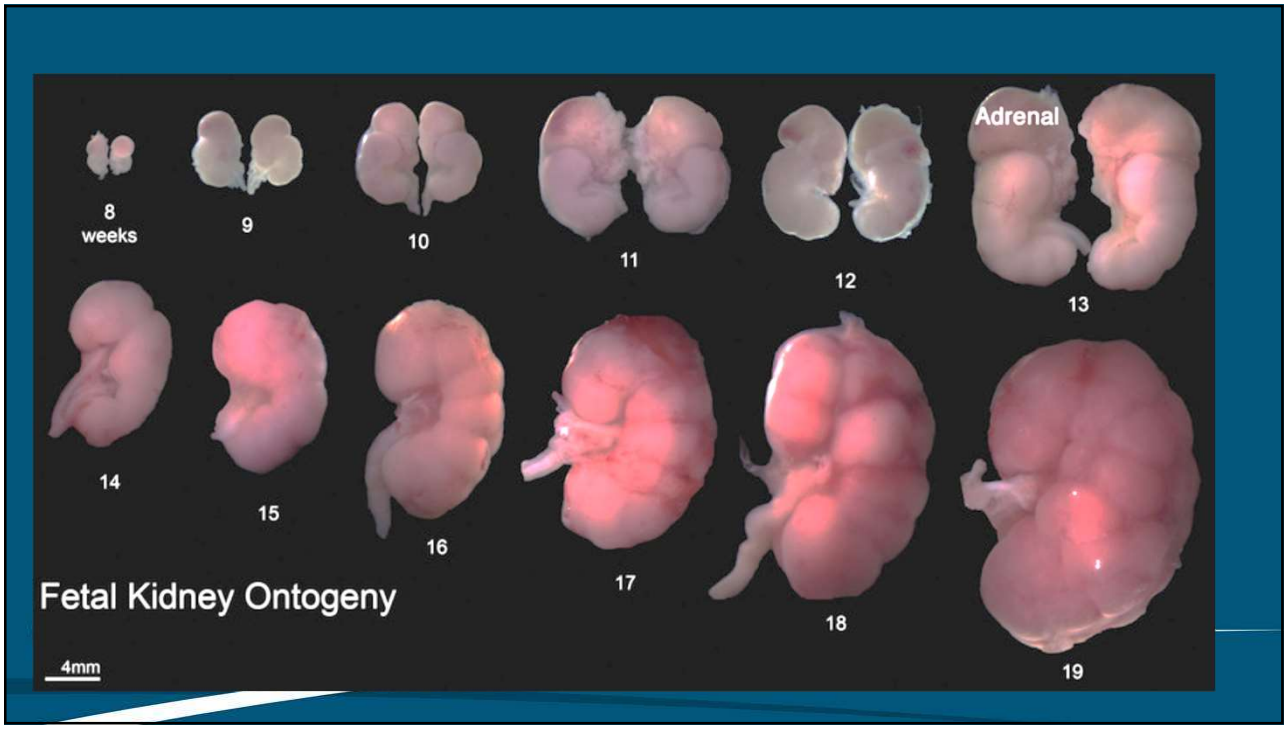

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Embryology Key Points

- 3 kidneys develop from intermediate mesoderm
  - Pronephros -> regresses by week 4, forget about it…
  - Mesonephros (Wolfian duct)-> *caudal end ureteric bud, vas/epididymis/seminal vesical, trigone*
  - Metanephros (starts week 5) = *final kidney!*
    - Intermediate mesoderm in the sacral area
    - Interacts with the ureteric bud to become renal parenchyma
    - While ascending, blood supply changes along great vessels

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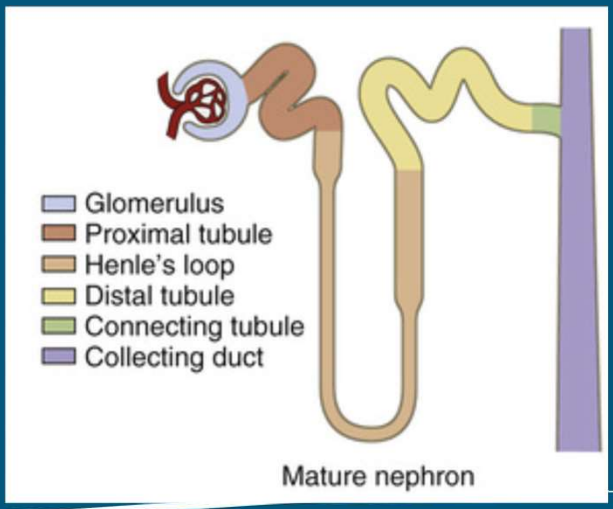
Metanephros and ureteric bud

- Ureteric bud comes off mesonephric duct, AKA Wolfian duct
- Ureteric bud branches -> *collecting ducts and everything distal*
- Metanephros becomes everything else
- Urine at 10-11 wks, fully developed by week 32



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Renal parenchymal anomalies

- Abnormal development of metanephros -> abnormal renal parenchyma = *renal dysplasia*
- Ureteric bud and metanephros interact -> *dysplasia often associated collecting system anomalies*
  - VUR, ectopic ureter, ureterocele
- Multicystic Dysplastic Kidney (MCDK)
- Autosomal dominant polycystic kidney disease (ADPKD)
- ARPKD (Autosomal recessive polycystic kidney disease (ARPKD)
- Renal agenesis

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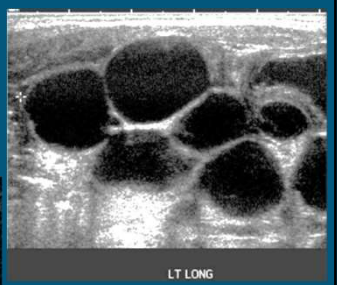
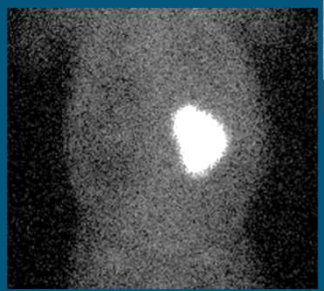
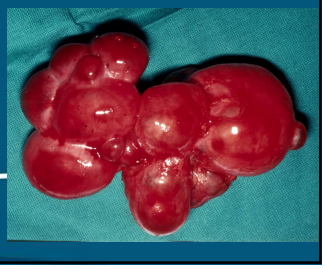
Multicystic dysplastic kidney

- ~1/1000 births, often prenatal detection
- Severe renal dysplasia, often absent or atretic ureter
- Non-reniform shape, *multiple non-communicating cysts*
- No function on renal scans
- Natural history is involution
  - Observe serial ultrasounds
- *Surgery is rarely indicated*
  - *HTN, pulmonary compromise*
- *Risk of malignancy is negligible*







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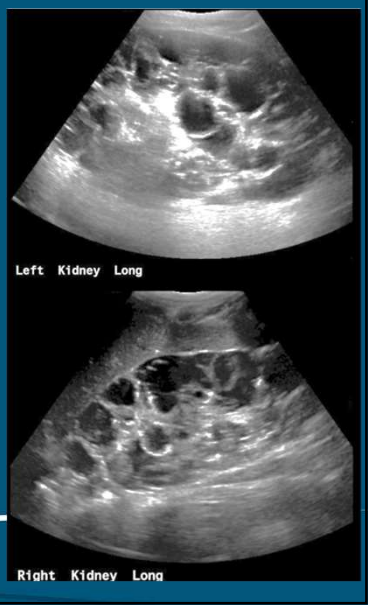
Autosomal Dominant Polycystic Kidney Disease (ADPKD)

- ~1/500. Mutations in PKD1 (85%) or PKD2 gene
- PKD1 mutations
  - More severe, >90% have cysts by age 20, ESRD in 50s typical
- Can present with simple cysts in childhood
  - Multiple cysts, especially bilateral -> testing for ADPKD
- *Large cysts are typical*
- Progressive cysts, HTN, hematuria, ESRD, stones
- Extra-renal manifestations
  - Liver/spleen/pancreatic cysts, intracranial (berry) aneurisms, mitral valve prolapse



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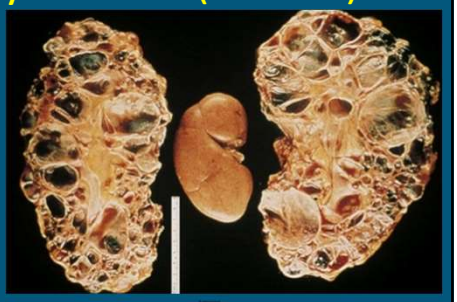
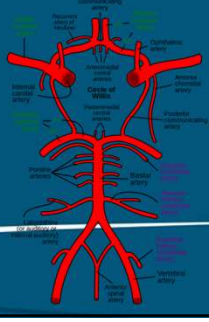
Autosomal Dominant Polycystic Kidney Disease (ADPKD)

- Simple cysts in children followed a couple years
  - If more cysts develop -> consider ADPKD
- Rarely surgery needed in childhood for ADPKD
- Treatment goal is to delay ESRD
  - HTN control; ACE inhibitors
- Adulthood
  - Renal transplants, *large kidneys -> native nephrectomy*
- ~20% have aneurisms and ~10% fatal
  - Berry aneursyms of Circle of Willis -> subarachnoid hemorrhage





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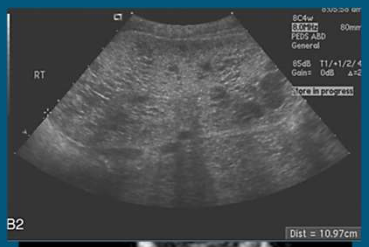
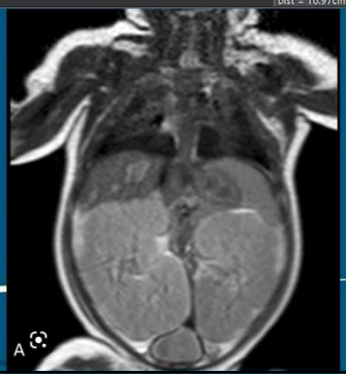
Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- ~1/20000, prenatal detection. Mutation in PKHD1.
- *Very large, homogeneous, echogenic kidneys*
- *Small cysts (mm) compared to large cysts in ADPKD*
  - Cysts involve collecting tubules
- More lethal than ADPKD:
  - 50% of newborns do not survive
- Less severe cases exist with delayed diagnosis and better survival





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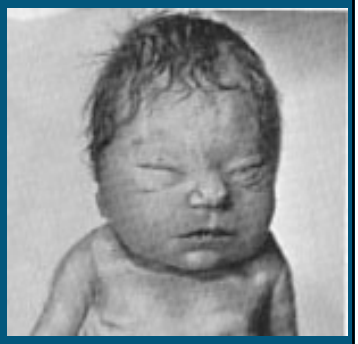
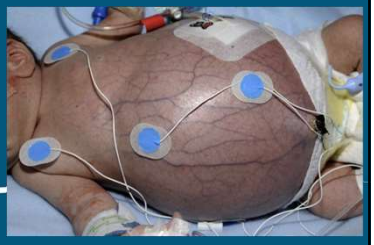
Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- One cause of Potter syndrome
  - Due to lack of amniotic fluid
  - Low set ears, wide set eyes, recessed chin, broad nose, short limbs, *pulmonary compromise*
- Multiple comorbidities and medical issues
  - Hepatic fibrosis/failure, *pulmonary issues*, HTN, resulting CHF, esophageal varices, portal HTN, developmental delays
- Newborns (if survive) often require unilateral/bilateral nephrectomy due to large size of kidneys
  - Pediatric urology consultation





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Renal agenesis

- ~1/4000 births. Theory: failure of ureteral bud formation
- Compensatory hypertrophy of contralateral kidney
- Bilateral renal agenesis usually fatal in-utero or after birth
  - Another cause of potter syndrome
- ***Can have ipsilateral absence of vas and epididymis***
  - ***Why? Wolfian duct gives rise to ureteral bud and vas/epididymis***
  - ***Random fact: bilateral absence of vas seen with cystic fibrosis and CFTR gene mutations (often heterozygous)***

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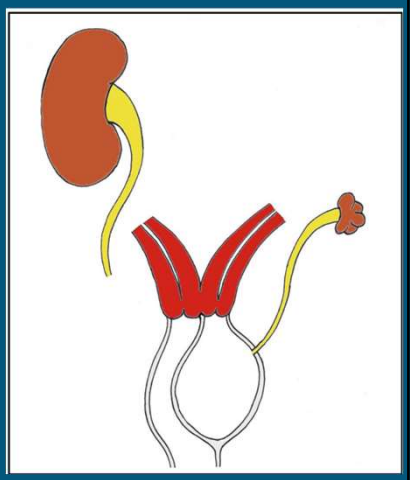
Obstructed Hemivagina and Ipsilateral Renal Anomalies (OHVIRA)

- Duplicated uterus/vagina, ipsilateral obstructed
- Mullerian and Wolfian ducts close to each other
  - Something goes wrong -> OHVIRA can occur
- Presentations
  - *Infant: dilated hemivagina/solitary kidney on imaging*
  - *Puberty: cyclic pain with possible vaginal bulge*
- Treatment: incision/excision of vaginal septum
- Nephrectomy if vaginal leakage after incision



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Antenatal Hydronephrosis

- Most common fetal abnormality
  - 1-3% of fetuses
- ***Majority will be mild and transient***
- Unilateral mod/severe hydronephrosis should be observed
  - Surgery was done after birth early on with prenatal US experience -> overtreatment of UPJ obstructions
- Some diagnoses important to not miss!
  - posterior urethral valves, ureterocele, ectopic ureter

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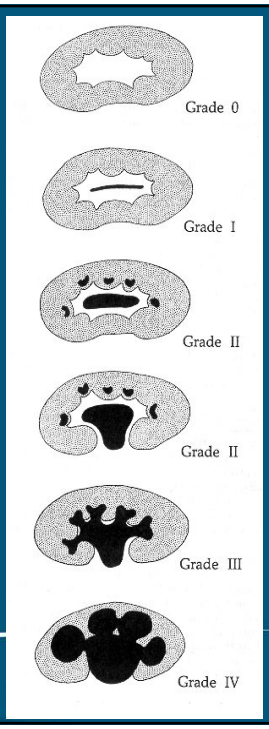
Hydronephrosis Grading - SFU

- SFU Grade 1- only renal pelvis/ ***no caliectasis***
  - "Mild"
- SFU Grade 2- dilated pelvis and ***single or few calices***
  - "Mild"
- SFU Grade 3- dilated pelvis and ***uniformly dilated*** calyces
  - "Moderate"
- SFU Grade 4- grade 3 dilation with ***thin parenchyma***
  - "Severe"



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Pediatr Radiol. 1993; 23: 478-480

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Hydronephrosis Grading – UTD Classification

- Introduced in 2014; UTD = urinary tract dilation
- 6 elements
  - Anterior-posterior renal pelvis diameter (APRPD), degree of dilation of calyces, parenchyma thickness, parenchyma appearance, hydroureter, bladder
- Findings = risk score
- Antenatal and postnatal classifications
  - Antenatal: UTD A1 = low risk, UTD A2-3 = increased risk
  - ***Postnatal: UTD P1 = low risk, UTD P2 = intermediate, UTD P3 = high***
- Management based on UTD risk score

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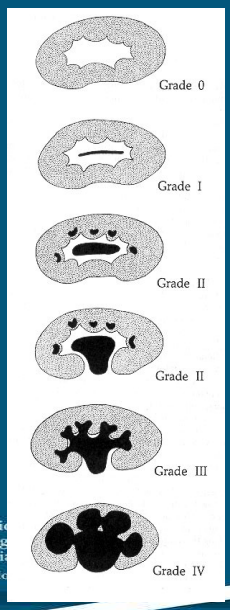
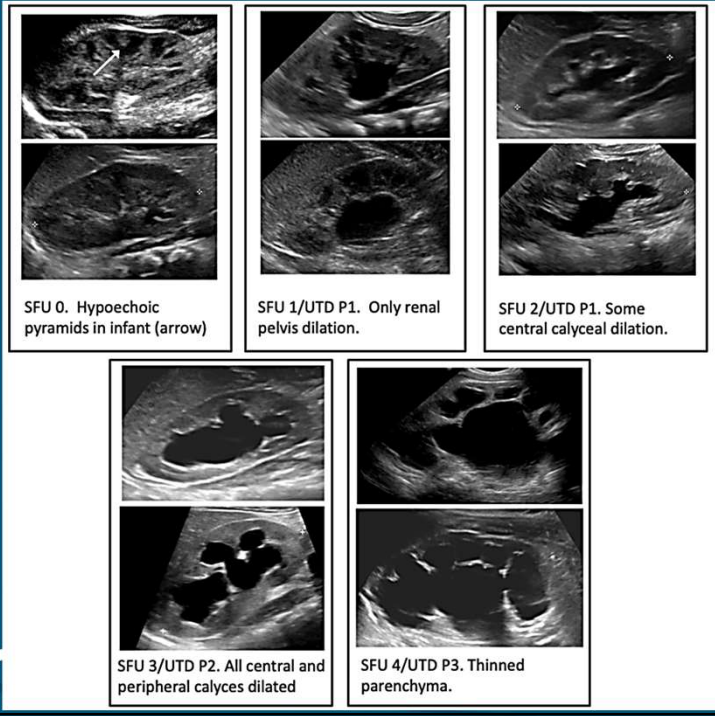
Hydro Examples





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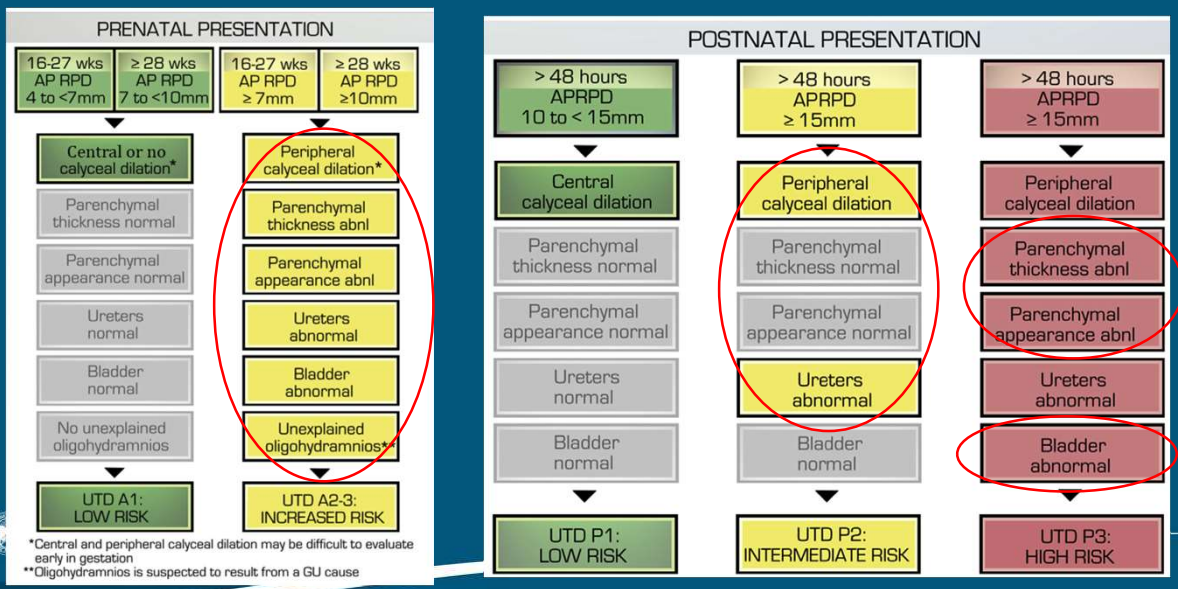

Journal of Pediatric Urology (2014) 10, 982e999

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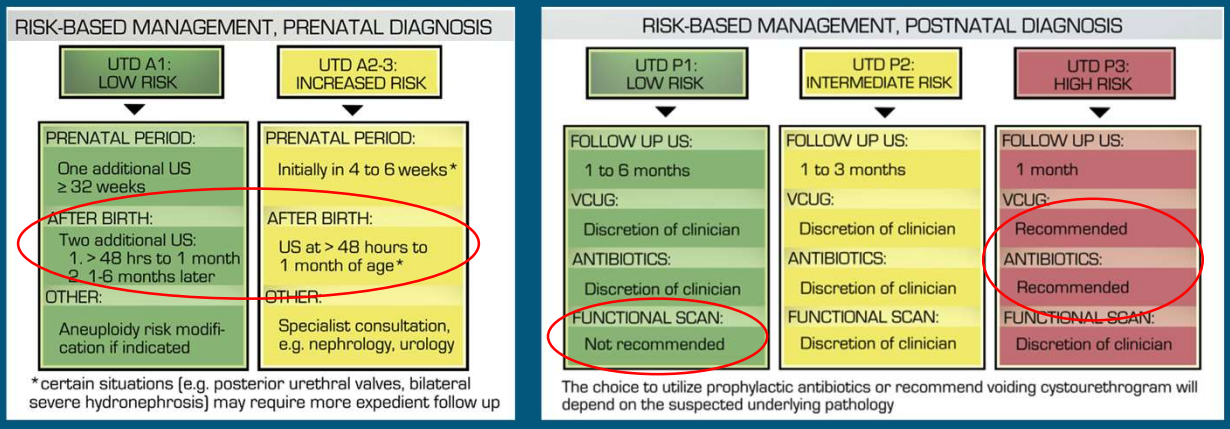
Hydronephrosis Grading – UTD Classification



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Infants with hydronephrosis key points

- ***Initial renal US should be done > 48 hours after birth***
  - Newborns can be dehydrated -> underestimation of hydronephrosis
- Most prenatal hydronephrosis will be unilateral and transient
- ***Don't miss PUV, ectopic ureter, ureterocele -> expedite evaluation***
- No randomized controlled trials for antibiotic prophylaxis
  - Observational studies suggest benefit with hydroureter
  - Consider if VCUG planned
- If no calyceal dilation and APRPD < 10 mm = normal
- Even moderate to severe hydronephrosis can improve/resolve

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Antenatal Hydronephrosis - etiology

Table 5 The etiology of ANH.

Etiology	Incidence
Transient hydronephrosis	41–88%
UPJ obstruction	10–30%
VUR	10–20%
UVJ obstruction/megaureters	5–10%
Multicystic dysplastic kidney	4–6%
PUV/urethral atresia	1–2%
Ureterocele/ectopic ureter/duplex system	5–7%
Others: prune belly syndrome, cystic kidney disease, congenital ureteric strictures and megalourethra	Uncommon

Table 4 Risk of specific postnatal pathologic conditions by the degree of ANH.

	% ANH [95% CI]		
	Mild	Moderate	Severe
UPJ	4.9 [2.0–11.9]	17.0 [7.6–33.9]	54.3 [21.7–83.6]
VUR	4.4 [1.5–12.1]	14.0 [7.1–25.9]	8.5 [4.7–15.0]
PUV	0.2 [0.0–1.4]	0.9 [0.2–2.9]	5.3 [1.2–21.0]
Ureteral obstruction	1.2 [0.2–8.0]	9.8 [6.3–14.9]	5.3 [1.4–18.2]
Other	1.2 [0.3–4.0]	3.4 [0.5–19.4]	14.9 [3.6–44.9]

Journal of Pediatric Urology (2010) 6, 212e231

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Antenatal Hydronephrosis – Etiology Key Points

- Transient hydronephrosis is most common by far
- Vesicoureteral reflux is a possibility for any severity
- UPJ obstruction is most common obstruction
  - more likely from mild to severe
- PUV, ectopic ureter, ureterocele are rare, but do not miss

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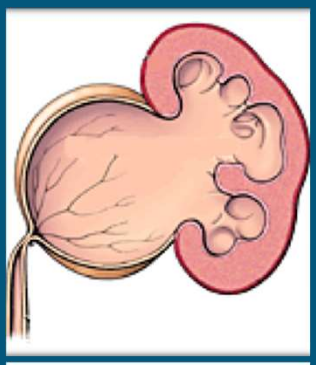
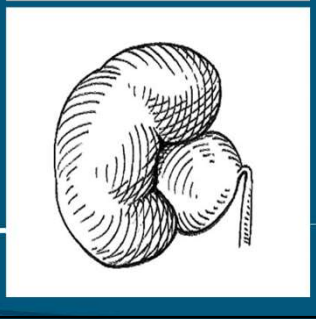
UPJ Obstruction - Infants

- Commonly intrinsic narrowing and/or high insertion
- SFU grade 4 -> ~25-50% will get pyeloplasty
- SFU grade 3 -> ~15% or less
- SFU grade 1-2 -> rare
- UTIs, stones, or symptoms are rare





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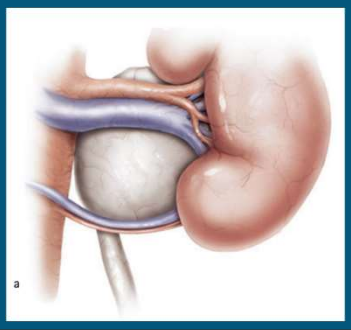
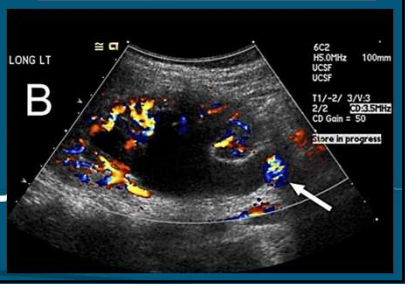
UPJ Obstruction – Older children

- More likely to have symptoms
  - Pain/nausea (Dietl's crisis) with fluid intake
  - Pain poorly localized, esp. younger children
  - *Ultrasound at time of pain can be helpful*
- More commonly lower pole crossing vessel
  - Often unknown until surgery
- Fibroepithelial polyp rare
  - Seen on ultrasound as vascular area
- Intrinsic narrowing or high insertion possible





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UPJ Obstruction – MAG 3 scan

- Bladder catheter if not toilet trained/Sedation for younger children
- Various protocols for Lasix
  - At 30 minutes, at time of peak activity, at 20 minutes
- Normal findings
  - Time to peak activity: 3-5 minutes
  - Drainage time: T1/2 to clear after peak activity = 8-12 minutes
  - Differential update of radiotracer: 45-55%
- Important to look at actual images and the curves
- *Cut-offs of drainage (ex. T ½ > 20 minutes) not used as much in peds*

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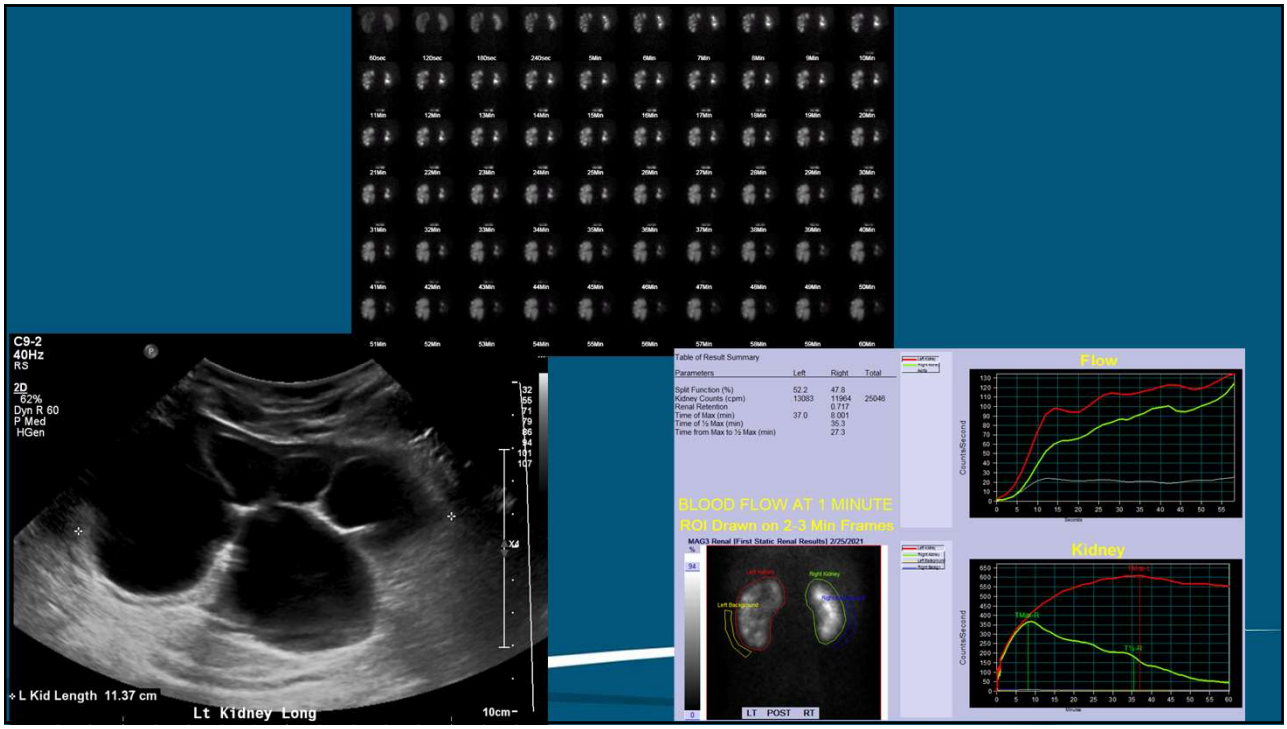

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UPJ obstruction – indications for surgery

- Practices and family preferences vary widely
- In order of strength of indication
  - Symptomatic (pain, UTIs, stones), UTIs rare
  - Declining differential function over time
  - Initial low differential function (<40%) and severe hydronephrosis
  - Worsening hydronephrosis over time with stable function
  - Stable severe hydronephrosis and function with no improvement with observation (2-4 years?)

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UPJ obstruction - treatment

- Dismembered pyeloplasty
  - Open, laparoscopic, robotic assisted
  - Robotic likely easier recovery for older children
- Risks: urine leak, persistent or recurrent obstruction (~5%)
- Ureteral stent and drain usage vary
- ***If lower pole crossing vessel, anastomosis anterior to vessels***

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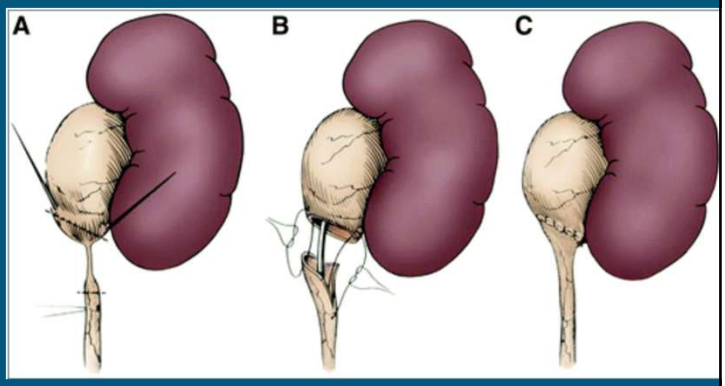
Dismembered Pyeloplasty Key Steps

- Reflect colon or transmesenteric
  - For robotic
- Dissect ureter up to renal pelvis
- Identify any crossing vessels
- Place hitch stitch in renal pelvis
- Open renal pelvis, spatulate
- Anastomosis
- +/- ureteral stent or drain
- Foley catheter in overnight



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Journal of Endourology.May 2018.S-68-S-72.

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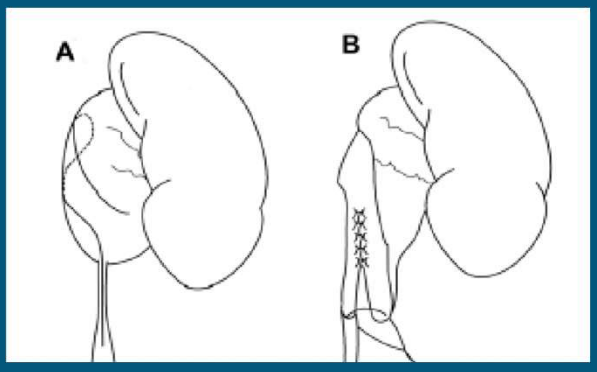
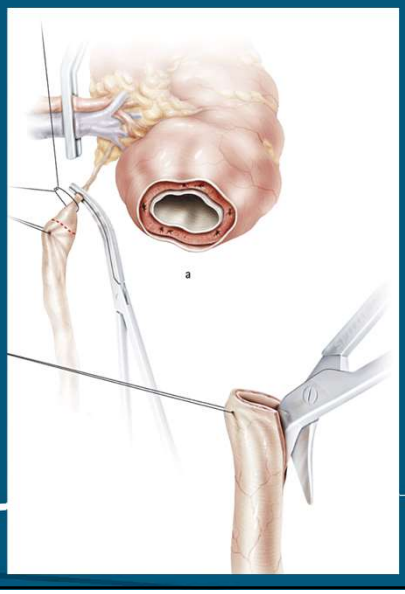
UPJ obstruction – special situations



Spiral flap pyeloplasty
- ***Long stricture***
- Failures



Ureterocalycostomy
- Failures
- ***Thin parenchyma***

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Megaureter

- ***Megaureter is just descriptive term***
  - refluxing or non-refluxing
  - obstructed or non-obstructed
  - Ectopic ureter, ureterocele, VUR, primary megaureter (UVJO)
  - High pressure bladder: PUV, neurogenic bladder
- *What causes a refluxing and obstructed megaureter?*
  - *An ectopic ureter to the bladder neck*

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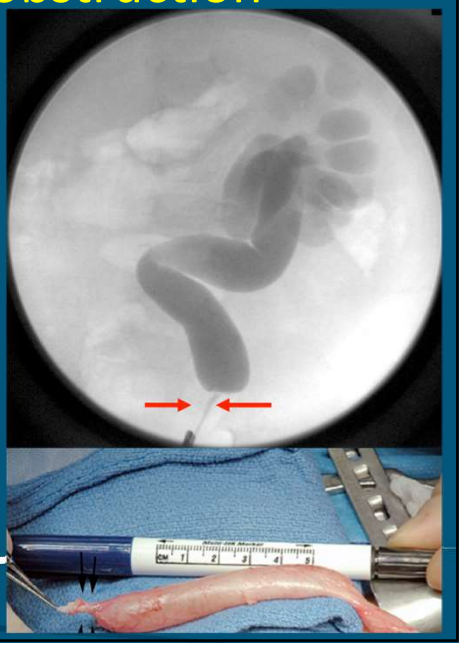
Primary Megaureter/UVJ obstruction

- ***Natural history is resolution with observation***
  - VCUG normal
  - MAG 3 scan – equal function, delayed drainage but radiotracer in ureter
- *Typically a distal narrow segment*
- Indications for treatment similar to UPJO
  - Ureteral reimplant most common, possible tapering
  - Ureteral dilation, prolonged ureteral stent, laser incision of distal narrow segment
  - Refluxing reimplant reported



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Vesicoureteral Reflux

- Embryology/theory
  - More distal ureteral bud -> more lateral and cranial UO in bladder
  - More lateral and cranial UO -> shorter intramural tunnel through detrusor
  - Shorter intramural tunnel -> lack of mucosal coaptation
  - Lack of mucosal coaptation -> VUR
- *Typically present with UTIs*
  - Incidental abnormal imaging, HTN, proteinuria less common
- Diagnosed by cystogram (VCUG, etc)
- Secondary reflux = bladder issues (neurogenic, dysfunctional voiding)

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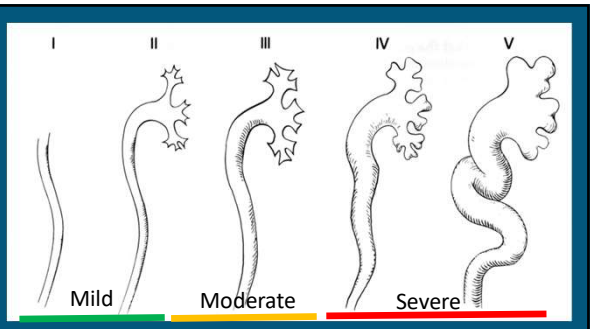
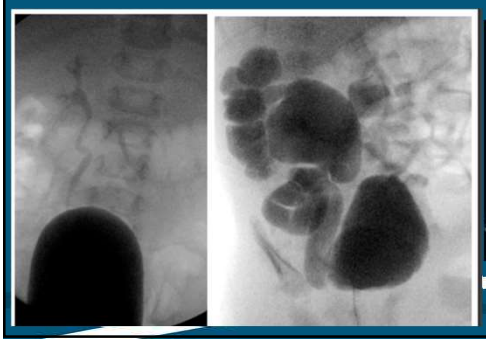
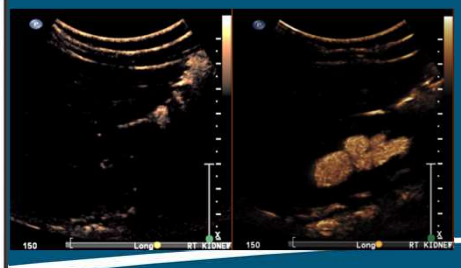
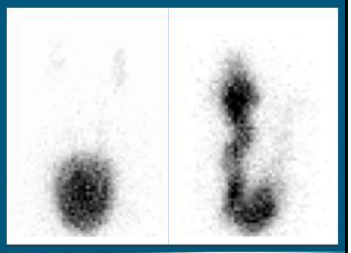
- VCUG
  - Grade 1-5[1]
- Nuclear medicine cystogram
  - mild, moderate, severe
- Contrast enhanced voiding cystourethrogram (CEVUS)
  - Grade 1-5









[1]Pediatrics. 1981 Mar;67(3):392-400.

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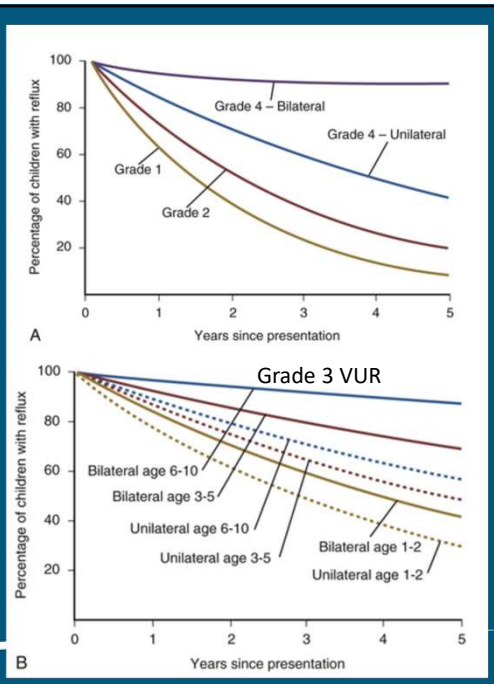
VUR: Natural history

- Lower grade more likely to resolve
- Younger children more likely to resolve
- Unilateral VUR is more likely to resolve
- ***Initial treatment is typically observation +/- prophylaxis***



J Urol. 1997;157[5]:1846–1851

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VUR and UTIs key points

- UTIs are one of the most frequent bacterial infections
- Prevalence of UTI in children 2-24 months with fever without apparent source ~ 5%[1]
- By age 8 yrs: 7-8% girls, 2% boys have a UTI[1]
- *VUR -> increased risk of subsequent UTI[2]*
- ~30-40% of children with febrile UTI have VUR[3]
  - 50-60% grade I-II, ~40% grade III, <5% grade IV or V

1.N Engl J Med, 2011;365:239-50
2. Pediatrics 2008;122:1064–1071
3. JAMA. 2007;298(2):179-186

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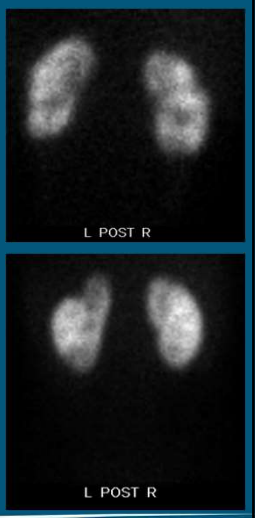
VUR and Renal Scarring Key Points

- 60% with febrile UTI < 24 mo have pyelo on DMSA
  - *initial normal DMSA = no renal scarring*
  - *initial abnormal DMSA = 15-30% risk renal scar*
- ~90% will have a normal renal ultrasound
- *VUR = increased risk of scar at 6 months*
  - *6% no VUR vs. 15% with VUR*



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N Engl J Med 2003; 348:195-202
Pediatrics 2008: 122:486-490

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VUR and Bowel Bladder Dysfunction

- VUR has strong association with bowel bladder dysfunction (BBD)
  - Holding behaviors, constipation, poor bladder emptying, etc
  - Leads to incontinence, urgency, recurrent UTIs (febrile vs nonfebrile)
  - Females have incontinence/UTIs, boys incontinence less UTIs
- *Important to treat BBD = urotherapy*
  - Timed voiding, double voiding, prevention of constipation, learning to relax sphincter with voiding (biofeedback)
- VUR may resolve with treatment of BBD
- *Failure to treat BBD can lead to treatment failure*
  - *And missing questions on ABU exams*

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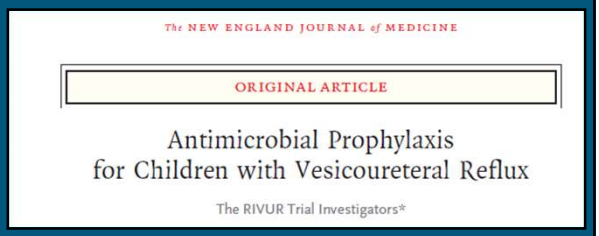
VUR: Antibiotic Prophylaxis/RIVER Trial

- Intervention: trimethoprim-sulfamethoxazole vs placebo
- Inclusion: 2 mo – 6 years with 1st febrile or symptomatic UTI
  - Grade I-IV VUR
- Outcomes
  - Recurrent febrile or symptomatic UTIs
  - Abnormal DMSA (renal scarring)
- 607 patients randomized
  - 10800 screened


The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Antimicrobial Prophylaxis for Children with Vesicoureteral Reflux

The RIVUR Trial Investigators*

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VUR: Antibiotic Prophylaxis (RIVUR)

- TMP-SMX lowers risk (p < 0.001, log rank).
  - *~50% risk reduction*
  - NNT = 8 for 2 years
- *No difference* in new renal scarring
  - 8.2% vs 8.4%
- Effect modifiers
  - Febrile UTI – more effect
  - *BBD – more effect*

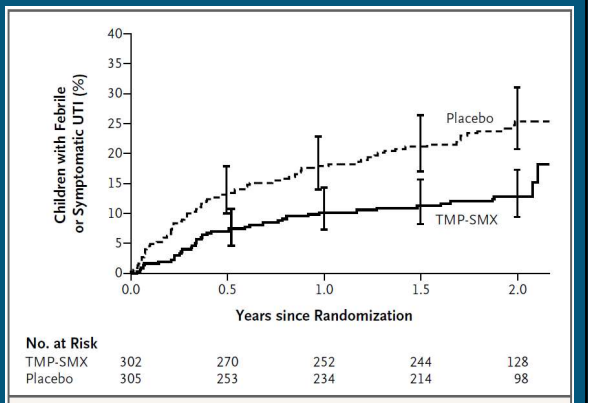

Figure 2. Time to First Recurrent Febrile or Symptomatic UTI.

Shown are Kaplan–Meier estimates of the cumulative percentage of children who had a recurrent febrile or symptomatic UTI according to study group. Fewer children assigned to TMP-SMX prophylaxis had a UTI than children assigned to placebo (P<0.001 by log-rank test). I bars indicate 95% confidence intervals.

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VUR: Surgical Treatment

- ***Indications for surgery***
  - ***Break through UTIs, renal scarring, family preference, high grade VUR that does not resolve***
- Open ureteral reimplant is gold standard
  - Cross trigonal most common technique
  - Complications rare: persistent VUR, ureteral obstruction
  - Typically overnight hospital stay
  - ***Decreases risk of febrile UTIs, decrease in renal scarring not proven***
- Success in meta-analysis
  - 99% grade I-II, 98.3% grade III, 98.5% grade IV, 80.7% grade V

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J Urol. 1997 May;157(5):1846-51. PMID: 9112544.

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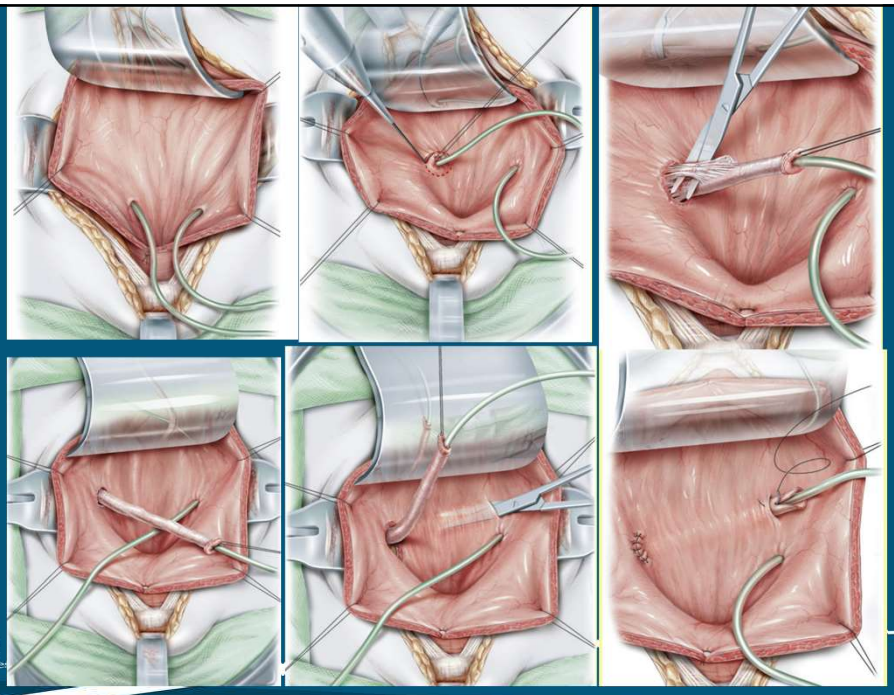

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PIERRE-YVES MURE and PIERRE D.E. MOURIQUAND. BJU INTERNATIONAL. 2004. 94, 679–698

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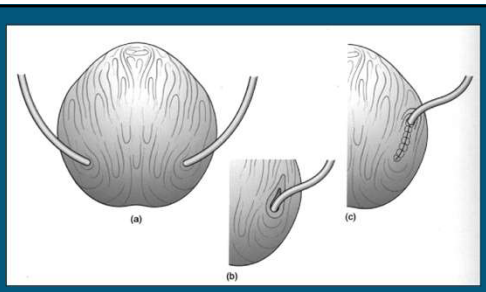
VUR: Robotic Reimplant



- Extravesical reimplant most common
- Perhaps easier recovery in older children
- Success rates initially lower than open but recent studies suggest similar results to open reimplant
- Some reports of ureteral complications (necrosis, urine leak)
- ***Special complication of bilateral extravesical ureteral reimplant***
  - ***Urinary retention. Usually transient, rarely permanent.***
  - ***Neuropraxia = temporary nerve injury***

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World J Urol. 2018 May;36(5):819-828.
J Pediatr Urol. 2018 Jun;14(3):262.e1-262.e6.

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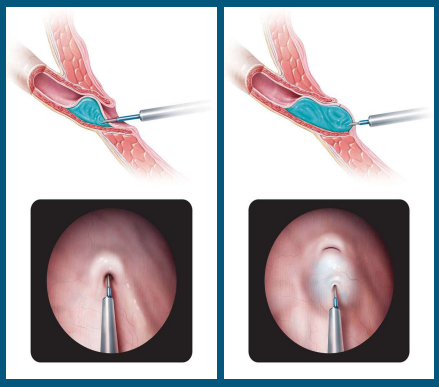
VUR: Endoscopic Injection = outpatient

- Hyaluronic Acid/Dextranomer (Deflux®)
- Systematic review with lower success
  - 89% grade I, 83% grade II, 71% grade III, 59% grade IV, 62% grade V
- ***Some risk of VUR recurrence over time***
  - ***BBD increases risk of recurrence***
- ***Can get calcified over years***
  - ***Confusion about possible ureteral stone***



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Kirsch. J Urology 2004
Lackgren and Kirsch, BJU Intl 2010
Pediatrics 2010;125:1010

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VUR: AUA Guidelines

AUA GUIDELINES

- Initial evaluation of child with known VUR
  - UA and renal US; serum Cr and DMSA optional
- ***< 1 year with febrile UTI or Grade III-V: antibiotic prophylaxis***
- > 1 year with BBD: antibiotic prophyaxis and treat BBD
- > 1 year without BBD: antibiotic prophylaxis optional
- Follow up
  - ***Renal US and UA annually for all***
  - Repeat VCUG in higher grades of VUR, optional for grade I-II
  - DMSA if renal US is abnormal and suggestive of scarring

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AUA Guideline: Management and Screening of Primary Vesicoureteral Reflux in Children (2017)

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VUR: AUA Guidelines

AUA GUIDELINES

- Recurrent or breakthrough UTI = ***Change management***
  - Start prophylaxis, change prophylaxis, consider surgery
- Surgery follow up
  - ***Renal US after any surgical treatment***
  - Cystogram after endoscopic injection, optional after open reimplant
- Long term follow up
  - If abnormal DMSA, annual blood pressure, weight, UA
- ***Screening for VUR in siblings not recommended***
- VCUG for prenatal hydro if SFU grade 3-4 or febrile UTI

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AUA Guideline: Management and Screening of Primary Vesicoureteral Reflux in Children (2017)

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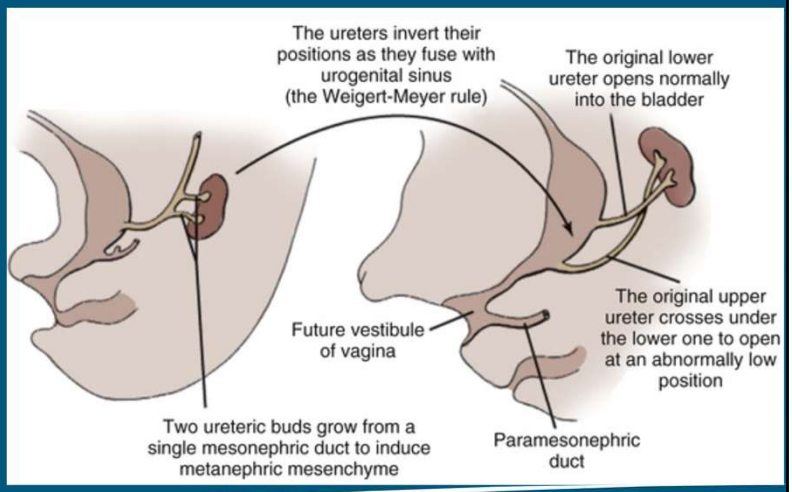
Duplicated kidney: Weigert-Meyer rule

- 2 ureteric buds -> duplex kidney
- Wolfian ducts fuse caudal to cranial to form trigone
- Caudal ureteric bud incorporated into trigone first and migrates cranial and lateral
- Cranial ureteric bud incorporated last and ends up caudal and medial



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Campbell's Urology 12th Edition

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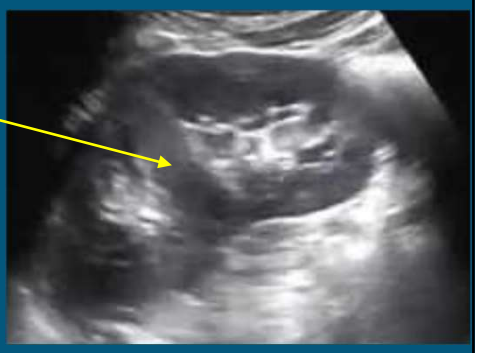
Duplicated kidney

- ***Normal variant***
  - 1-5%; Can be partial or complete
- Suggested by parenchymal bar on ultrasound
- Can have VUR or obstruction
  - Lower pole VUR
  - Upper pole (ectopic ureter, ureterocele)
  - Rare lower pole UPJ obstruction
- If no hydro or UTIs, no follow up or VCUG



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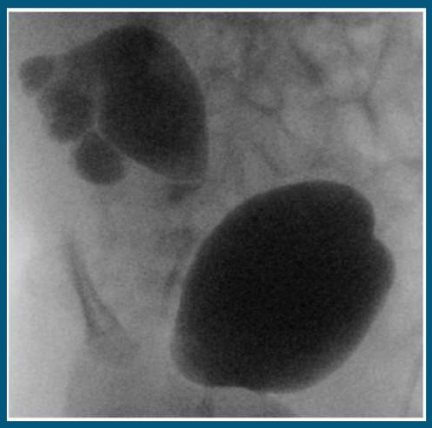
Duplicated Kidney and VUR



- Lower pole is more likely to have VUR
  - Lateral and cranial location
- Characteristic appearance = *"drooping lily"*
- ***Initial management similar***
  - ***Less likely to resolve spontaneously***
- Surgical treatment
  - Open double barrel ureteral reimplant
  - Endoscopic injection – less successful compared to non-duplicated?
  - Lower to upper pole uretero-ureterostomy –refluxing stump issues?

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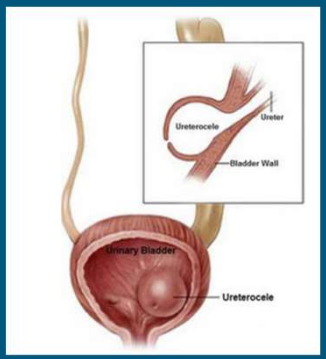
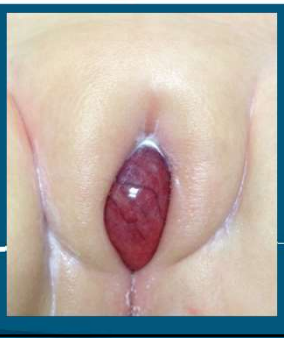
Ectopic Ureter and Ureterocele





- About 1/1000 births – *rare, but do not miss*
- Usually with duplicated kidneys (~80%)
  - Upper pole will be ectopic/ureterocele
- ***Continuous incontinence in female = ectopic ureter***
  - ***Can insert into mullerian structures, vagina, perineum***
- Males will not have incontinence with ectopic ureter
  - Always inserts proximal to sphincter
- Typically diagnosed on prenatal ultrasound
  - UTI, ureterocele prolapse, incontinence in females

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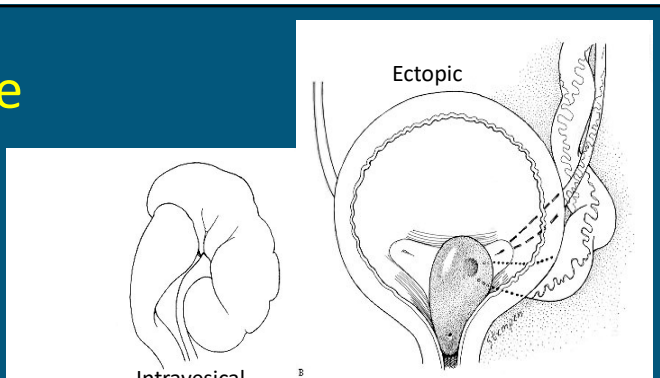
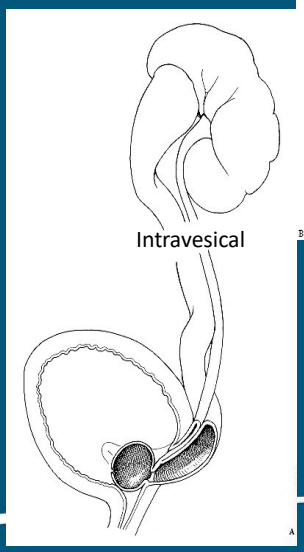
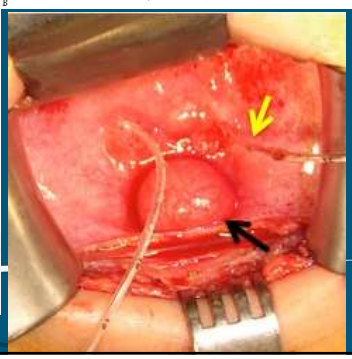
Ureterocele

- Cystic dilation of distal ureter
  - Associated hydronephrosis/hydroureter
  - Lower function in upper pole typical
- Ureteral orifice can be intravesical or ectopic near bladder neck or in urethra
- Prenatal diagnosis
- *Ureterocele prolapse, bladder outlet obstruction can occur*







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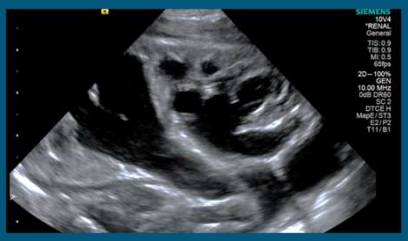
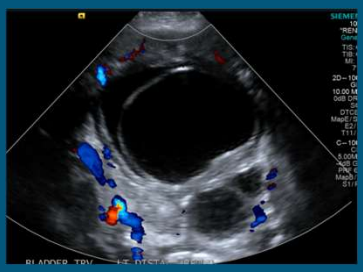
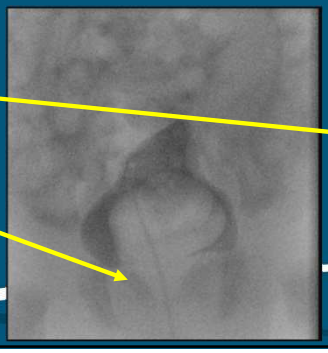
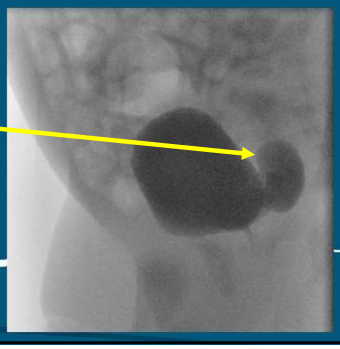
Ureterocele Imaging Findings

- Renal US
  - Duplex kidney
  - Dysplastic parenchyma
  - Upper pole hydronephrosis, can have lower as well
  - Cystic structure in bladder
- VCUG
  - Early filling defect
  - Lower pole VUR
  - Ureterocele eversion
  - Bladder outlet obstruction with ureterocele prolapsing into bladder neck









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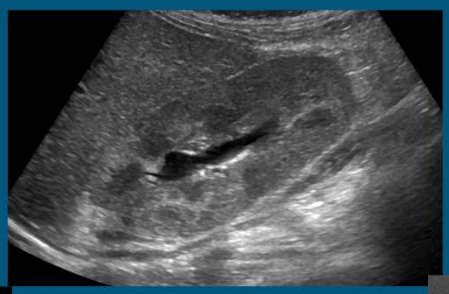
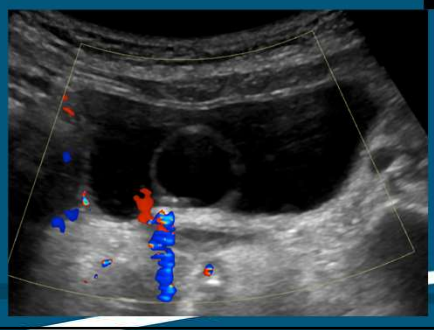
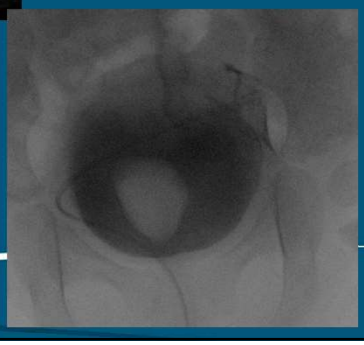
Ureterocele Imaging Findings - spectrum

- Single system
- Less hydronephrosis
- Healthy parenchyma
- Smaller ureterocele







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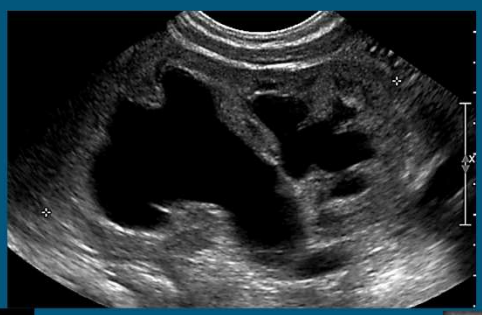
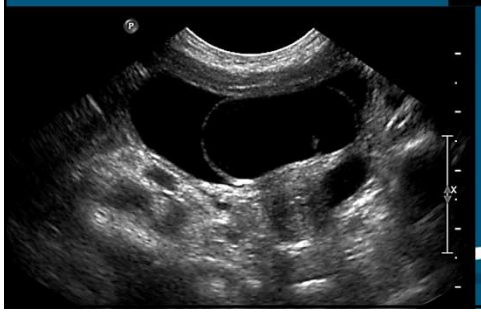
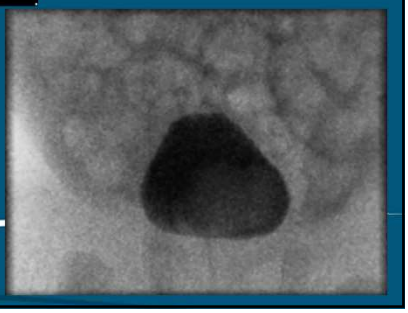
Ureterocele Imaging Findings - spectrum

- Duplex system
- Upper and lower pole hydronephrosis
- No ureterocele prolapse







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Ureterocele Management

- Infants used to present with sepsis
  - With prenatal ultrasounds, pre-emptive intervention began
- ***Renal US, VCUG soon after birth and antibiotic prophylaxis***
- *Endoscopic incision of ureterocele in infancy is common*
  - Selective observation (small, single system, male, mild hydro)
- Incision -> 50% risk ipsilateral VUR into upper pole ureter
- Ureterocele excision with duplex reimplant may be needed
  - Indications: recurrent UTIs, persistent high grade VUR

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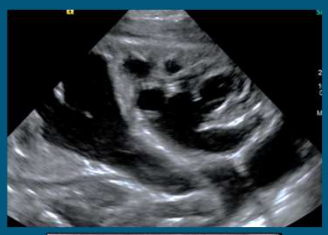
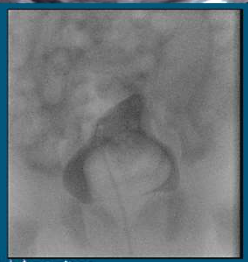
Post ureterocele incision VUR

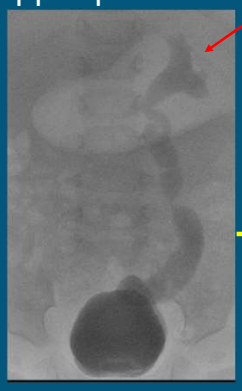




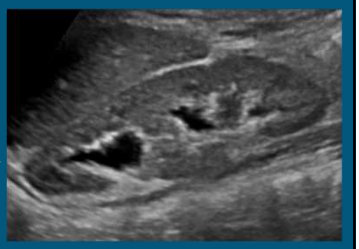
Ureterocele incision

Upper pole VUR

Not "drooping lily" but "hammer"



Duplex reimplant



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Ureterocele Management Options Summary

Figure 8: Options for Definitive Surgical Management, Ureterocele

	Ideal Indications	Advantages	Limitations
Transurethral incision	-Small infant -Large ureterocele with VUR	-Outpatient procedure* -Effective decompression -Occasionally definitive	-De-novo reflux into ureterocele segment necessitating subsequent lower tract reconstruction
Upper pole nephrectomy	**No lower moiety VUR** Nonfunctioning upper moiety	-May be definitive -Removes pathology -Avoids bladder surgery	-May still require lower tract reconstruction -Risk to lower moiety
UU/ureteropyelostomy	**No lower moiety VUR** Functional upper moiety	Drains obstructed segment with little risk for obstruction or UTI	Leaves ureterocele in bladder May develop VUR
Common sheath reimplant with ureterocele excision	**Associated lower moiety VUR** Functional upper moiety without significant dilation	Eliminates obstruction and VUR Removes ureterocele No renal risk	Complex surgery Risk to vagina and BN May require ureteral tapering

*unless patient is an infant requiring admission for oxygen monitoring

AUA core curriculum

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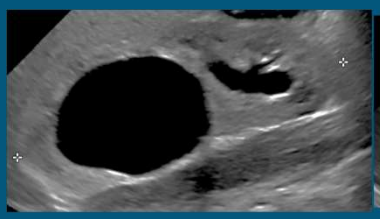
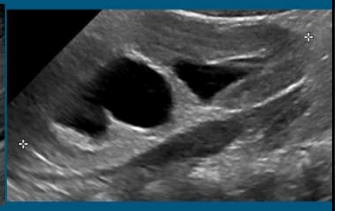
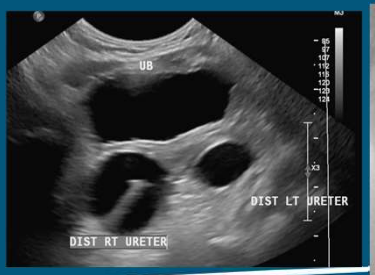
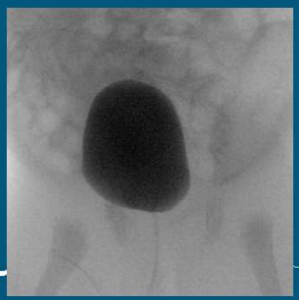
Ectopic Ureter

- Renal US, VCUG, prophylaxis
- Varying degrees of renal dysplasia
- Imaging findings
  - Duplex kidney (~80%)
  - Hydroureteronephrosis to bladder
  - Upper pole hydro and dysplasia
  - Possible lower pole VUR
  - "Pseudoureterocele" = large dilated ureter pushing into bladder, thick wall









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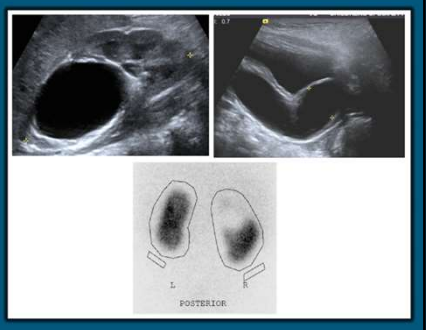
Ectopic Ureter Management



- *Will usually need surgery*
  - Very dysplastic kidney could be observed
- Uretero-ureterostomy (robotic vs open)
- Upper pole heminephrectomy (if no function)
- Ureteral reimplant
- Cutaneous ureterostomy followed by reimplant (if very large or sepsis)
- Nephrostomy tube followed by uretero-ureterostomy (if sepsis)
- Endoscopic incision ***NOT*** an option
  - Fistula from bladder to ectopic ureter -> incontinence in females

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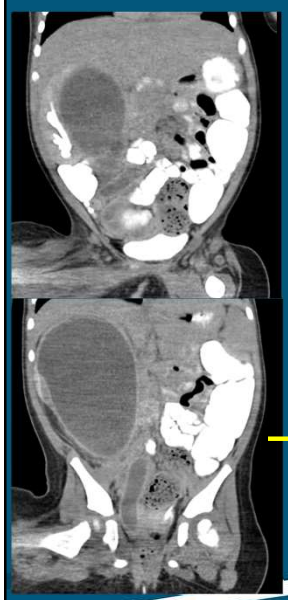
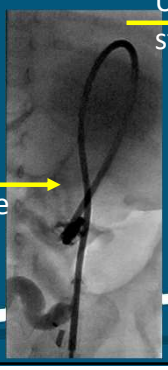
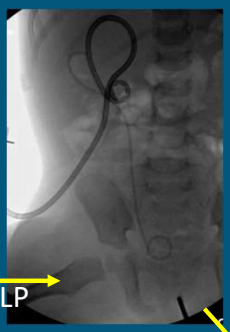
Ectopic ureter 14 month old with sepsis



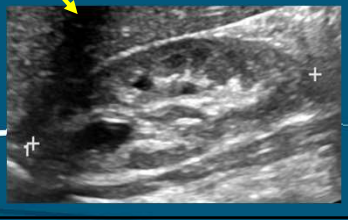
PCN to upper pole



UU with stent in LP



F/U RUS



62

CAKUT - Categories

- Renal parenchymal anomalies
  - Renal dysplasia, MCDK, ADPKD, ARPKD, renal agenesis
- Collecting system anomalies
  - Hydronephrosis, UPJ obstruction, megaureter, megacalycosis, ectopic ureter, ureterocele, duplicated kidney, VUR
- Renal ascent and fusion anomalies
  - Ectopic kidney, horseshoe kidney, crossed renal ectopia (fused/unfused)
- Lower urinary tract anomalies
  - Posterior urethral valves, anterior urethral valves, urethral atresia, megalourethra, prune belly syndrome, urachal remnants

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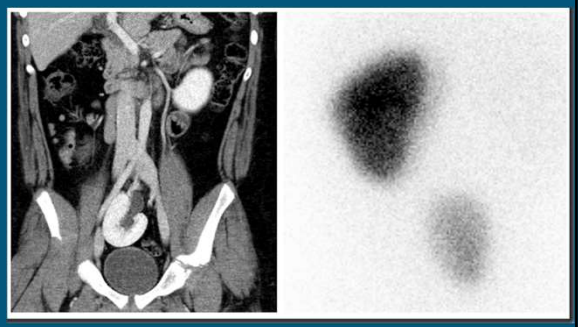
Renal Ascent and Fusion Anomalies

- Ectopic pelvic kidney
  - Most common ectopic kidney, ~1/1000
  - Increased risk of VUR, UPJ obstruction, lower differential function
  - If no UTIs or hydronephrosis, limited follow up necessary
  - Blood supply is anomalous
  - Renal pelvis often anteriorly rotated
- Other locations rare (Thoracic)



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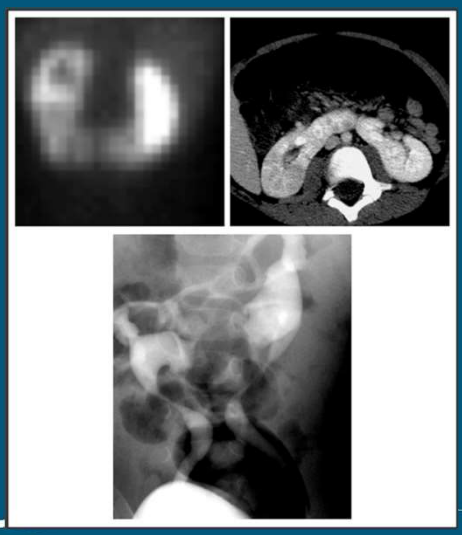
Renal Ascent and Fusion Anomalies

- Horseshoe kidney
  - Most common fusion anomaly (~1/600)
  - ***Isthmus below the inferior mesenteric artery***
  - Anomalous blood supply -> crossing vessels
  - Increased risk of VUR, UPJ obstruction
  - If no UTIs or hydronephrosis, limited follow up
  - ***Medially oriented calyces on VCUG***
- Crossed/fused and crossed/unfused ectopia
  - Less common, similar management



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CAKUT - Categories

- Renal parenchymal anomalies
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- Collecting system anomalies
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- Lower urinary tract anomalies
  - Posterior urethral valves, anterior urethral valves, urethral atresia, prune belly syndrome, urachal remnants

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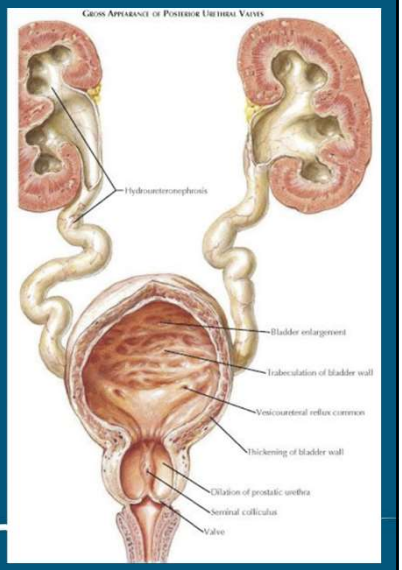
Posterior Urethral Valves

- ~1/5000-8000
- Membranous folds in posterior urethra
- Presentations
  - Prenatal hydronephrosis (most common)
  - UTIs, incontinence, renal failure
- Wide spectrum of severity
  - Anhydramios with fetal demise to mild symptoms with late presentation
- ***An important diagnosis to not miss***



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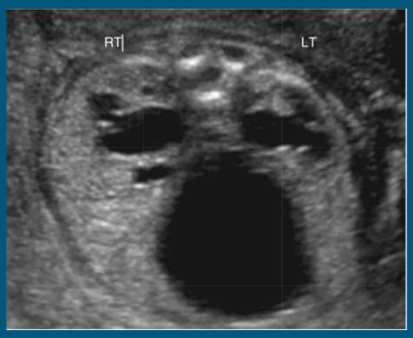
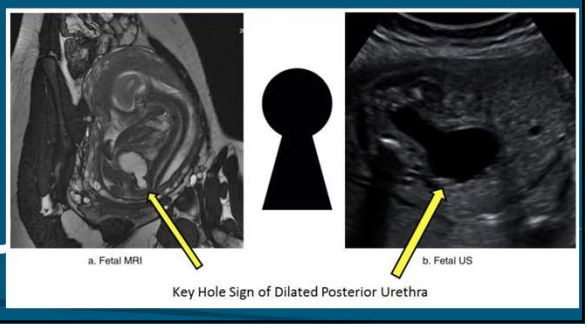
PUV: prenatal imaging findings

- Hydronephrosis and hydroureter, usually bilateral
- Renal dysplasia
- ***Distended bladder***
  - ***"Keyhole" sign***
- Amniotic fluid levels
  - Can be normal to anhydramnios





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LUTO: Prenatal Intervention

- PUV is most common cause of Lower Urinary Tract Obstruction (LUTO)
- Normal amniotic fluid levels = better postnatal renal function
- Severe oligohydramnios -> Fetal death due to pulmonary hypoplasia
- Fetal urine begins at ~10 weeks, oligohydramnios seen by 16 weeks
- ***Goals of prenatal intervention***
  - ***Improve lung function by increasing amniotic fluid***
  - ***Improve renal function by relieving obstruction***
- Fetal urine should be dilute (concentrated urine = bad outcomes)
  - Urine osmolality >200 mOsm/L, Urine sodium >100 mEq/L; predicts poor outcomes

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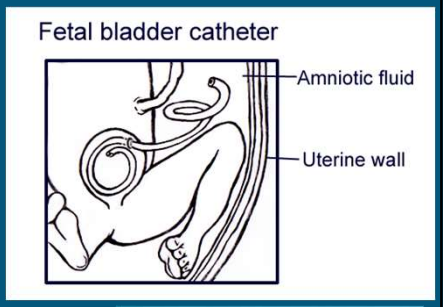
Vesicoamniotic Shunt for LUTO

- 2013 randomized trial (PLUTO trial)
  - Recruited 31 patients (goal 150)
  - *Intention to treat, no difference in survival*
  - *As treated analysis, survival benefit*
    - *2 years 8/16 vs 2/15 (p=0.02)*
  - *No difference in renal outcomes*
- 2016 Meta-analysis of 6 available studies
  - <200 patients
  - *Early survival benefit, but not at 6, 12, 24 mo*
  - *No significant renal benefit, no good predictors*



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Lancet. 2013 Nov 2;382(9903):1496-506.
Ultrasound Obstet Gynecol. 2017 Jun;49(6):696-703.

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Vesicoamniotic Shunt for LUTO

- ***Possible survival benefit due to improved lung function***
- ***Unproven renal benefit***
- Complications common
  - Preterm labor, fetal loss, shunt dislodging, wrong diagnosis
- Fetuses most likely to benefit
  - Oligohydramnios/anhydramios
  - Severe bilateral hydronephrosis but no cystic dysplasia
  - Favorable urine parameters on amniocentesis (dilute urine)
- Overall, limited outcome data. Controversial.

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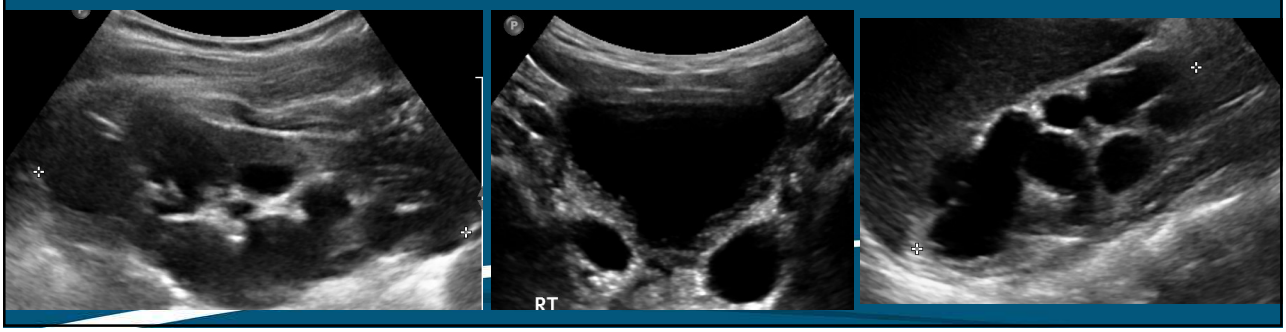
PUV: Management after birth

- Renal US and VCUG
- Drain bladder
- Monitor renal and pulmonary function
- Antibiotic prophylaxis typical



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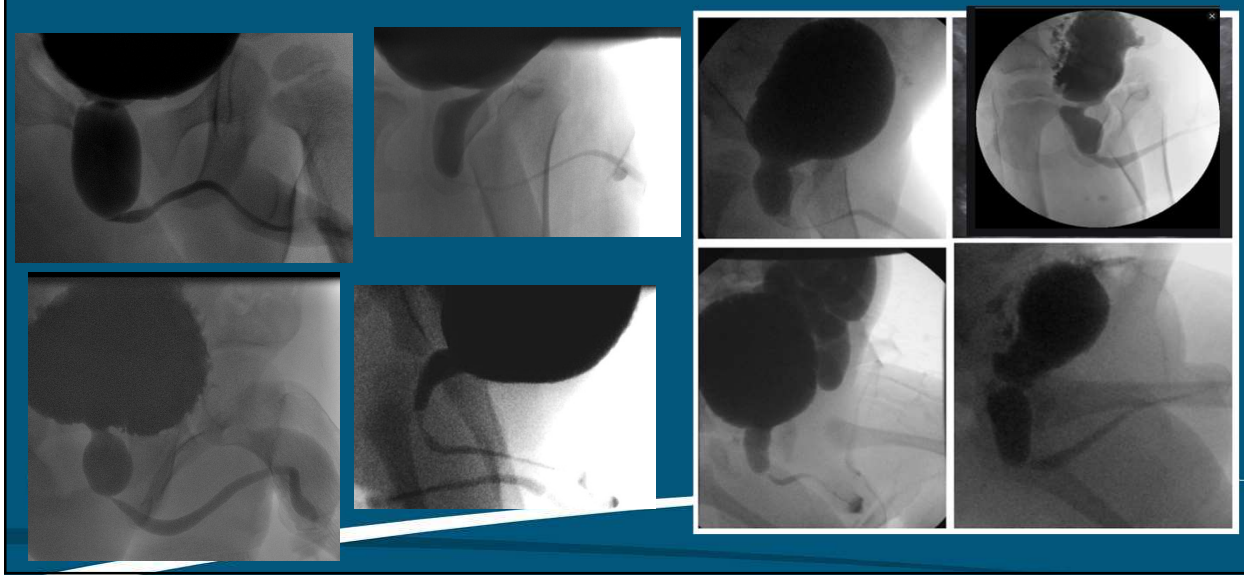
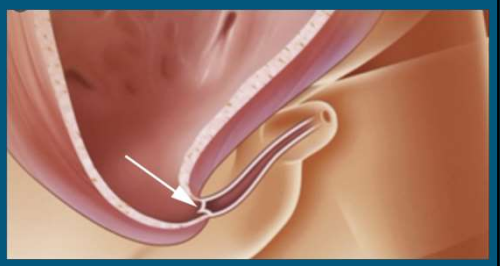
PUV: VCUG variable but patterns exist



73

PUV: VCUG findings

- Dilated posterior urethra
- Narrow segment at membranous urethra
- VUR (up to 75%)
- Bladder trabeculations and elongation
  - Can have smooth bladder



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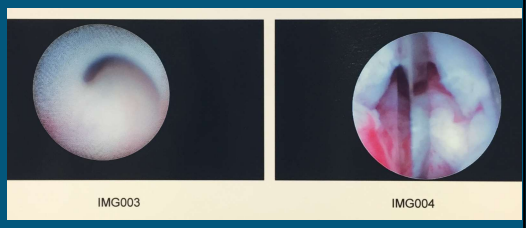
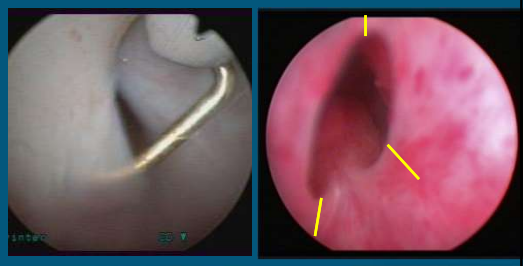
PUV: Cystoscopy and Endoscopic Incision

- Endoscopic incision for most infants
  - If too small, can leave catheter and dilate urethra
  - Vesicostomy rarely needed before valve ablation
- Incision at 5, 7, +/- 12 o'clock
- Cut current, cold knife, laser
- Complications: stricture, incomplete resection
- Catheter for 1-3 days
- Circumcision associated with lower UTI risk[1]
  - 3% vs 20% first 2 years, 2022 randomized trial





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[1]Eur Urol. 2022 Jan;81(1):64-72. d

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PUV: Follow Up After Incision

- ***Repeat VCUG after resection***
- ***Hydronephrosis: monitor***
  - Majority improve/resolve
- VUR: 2/3 will resolve & more improve
  - Rarely surgery necessary
- Toilet training
  - On average delayed about 2 years (age 5-6)
  - Double voiding, timed voiding can be helpful
  - Poor renal function associated with more delay
  - Overnight catheter drainage for select cases

Table 2. Spontaneous vesicoureteral reflux resolution on conservative management in group 1

Variable	Group 1: Conservative Management				
	First Year No (%)	Second Year No (%)	Third Year No (%)	Fourth Year No (%)	Fifth Year No (%)
VUR resolution*					
Grade I	0	10 (33.3)	24 (80.0)	25 (83.3)	25 (83.3)
Grade II	0	2 (8.7)	9 (39.1)	15 (65.2)	18 (78.2)
Grade III	0	0	1 (20.0)	1 (20.0)	2 (40.0)
Grade IV	0	0	0	2 (22.2)	2 (22.2)
Grade V	0	0	0	0	0
VUR improvement*					
Grade I	0	10 (33.3)	24 (80.0)	25 (83.3)	25 (83.3)
Grade II	0	2 (8.7)	9 (39.1)	15 (65.2)	18 (78.2)
Grade III	0	0	1 (20.0)	3 (60.0)	4 (80.0)
Grade IV	0	0	1 (11.1)	2 (22.2)	4 (44.4)
Grade V	0	0	0	1 (25.0)	1 (25)

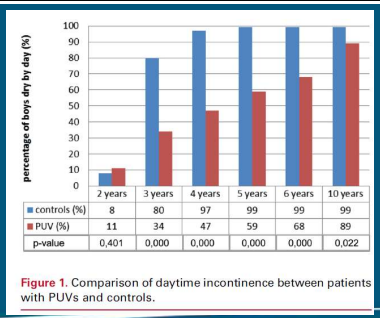

Figure 1. Comparison of daytime incontinence between patients with PUVs and controls.

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Urology. 2014 Jan;83(1):199-205.
J Urol. 2009 Oct;182(4):1555-60.
J Urol. 2016 Jul;196(1):213-8.

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PUV: Urodynamic patterns

- ***Classic teachings***
  1. Infants/toddlers: high pressure, detrusor overactivity
  2. Children: improved compliance, detrusor overactivity
  3. Older children: large capacity, myogenic failure
     - "end stage valve bladder". Higher risk with worse CKD and high UOP. ***Possible overnight drainage.***
- ***Most will toilet train, but delayed on average***
  - ***Timed voiding, double voiding common***
- Routine UDS, α-blockers, anticholinergics of unclear utility.
- ***Follow clinically ,check renal UA, UA, labs, PVRs annually***

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PUV: Long term outcomes

- Vesicostomy or cutaneous ureterostomy
  - Unclear indications/benefit. Some are more aggressive with diversion.
- *20-50% will have ESRD in lifetime -> nephrology f/u*
- < 20% will have incontinence in adulthood
- Paternity possible, less likely if ESRD
- Possible CIC or overnight drainage for myogenic failure/high UOP
  - APV can facilitate CIC for rare case that needs it (sensate urethra).
- Primary urology involvement: do a good valve ablation, circumcision (or resolve phimosis) can lower UTI risk, manage voiding expectations

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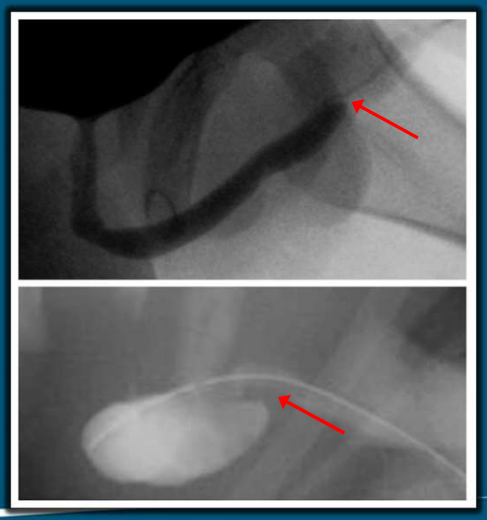
Anterior Urethral Valves and Urethral Atresia

- Very rare causes of LUTO in males
- Severe bilateral hydronephrosis and distended bladder
- Anterior Valves
  - VCUG: obstructing ventral web +/- diverticulum
  - Endoscopic incision, +/- urethral reconstruction
- Urethral atresia
  - If no patent urachus, then survival unlikely
  - If patent urachus, then vesicostomy followed by urethral reconstruction/diversion



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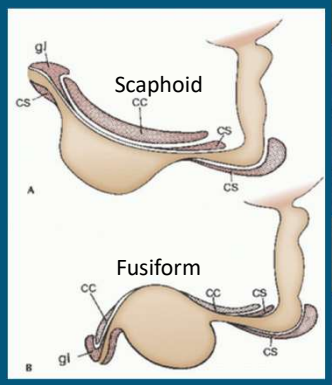
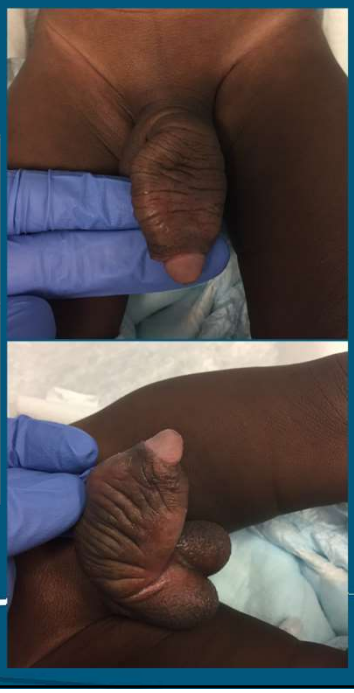
Megalourethra

- Rare congenital anomaly
- Scaphoid:
  - Deficiency of corpus spongiosum
- Fusiform
  - Deficiency of corpus cavernosum & spongiosum, ? abnormal glans
  - Upper tract anomalies
- Prune belly syndrome, VACTERL
- Scaphoid variant easier to reconstruct and function





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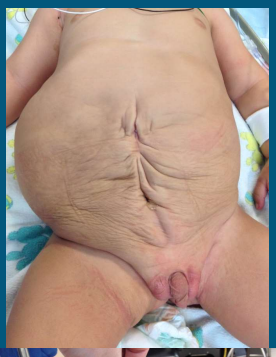
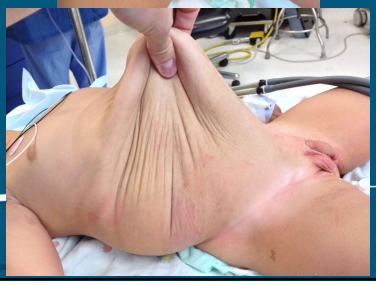
Prune Belly Syndrome/Eagle Barret Syndrome

- 3-4 in 100,000 births; >95% males
- ***Constellation of findings (classic triad)***
  - ***Abdominal wall laxity***
  - ***Urinary tract dilation: more severe distally***
  - ***Bilateral intra-abdominal UDT***
- Associated finding
  - Renal dysplasia
  - Chronic lung disease
  - Prostate hypoplasia
- Can be misdiagnosed with PUV prenatally





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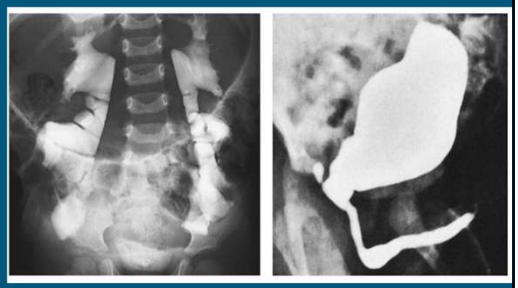
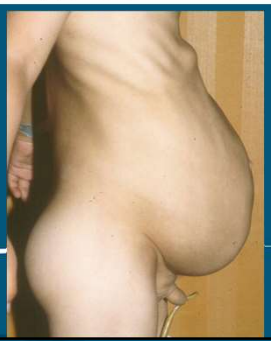
Prune Belly Syndrome Management

- Prevent UTIs
  - Circumcision vs steroid cream for phimosis
  - Antibiotic prophylaxis
  - Avoid unnecessary catheterization
- Bilateral orchiopexy
- +/- abdominal wall reconstruction
  - Cosmetic, ? improve bladder emptying & pulmonary function
- +/- urinary tract reconstruction
  - Individualized management, most do not need reconstruction
- Voiding – timed voiding/double voiding
- CKD management – long term follow up with nephrology/urology





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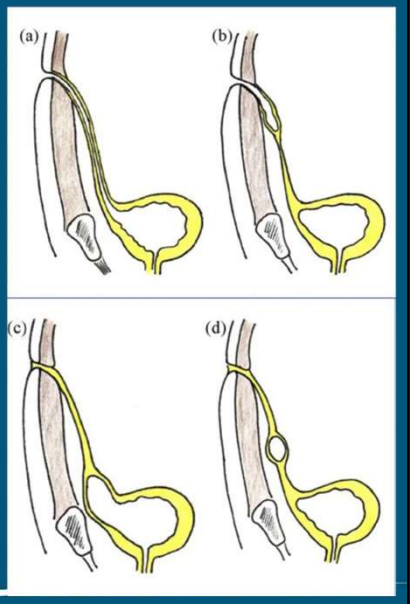
Urachal Remnants

- Connection between bladder and allantois
  - Median umbilical ligament
- Types: Patent urachus, urachal cyst, urachal diverticulum, urachal sinus
- Historically: malignancy concern -> surgery
  - >5000 surgeries to prevent 1 cancer[1]
- Incidental -> non-operative/observational[2]
- Patent urachus, infected cyst -> excision[2]
- Patent urachus -> obstruction?



From: Tazi, F., Ahsaini, M., Khalouk, A. et al. Abscess of urachal remnants presenting with acute abdomen: a case series. J Med Case Reports 6, 226 (2012).

1. Gleason JM, Bowlin PR, Bagli DJ, et al. A comprehensive review of pediatric urachal anomalies and predictive analysis for adult urachal adenocarcinoma. J Urol. 2015 Feb;193(2):692-6.
2. Gelikman DG, Ibanez KR, Ghattas YS, et al. Management of urachal anomalies in pediatric patients: A comparison of treatment strategies between pediatric urology and general surgery. J Pediatr Urol. 2024 Feb;20(1):75.e1-75.e8.

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Thank you!

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