Megaureter and beyond

VUR, POM, Duplex system-ectopic ureter-ureterocele

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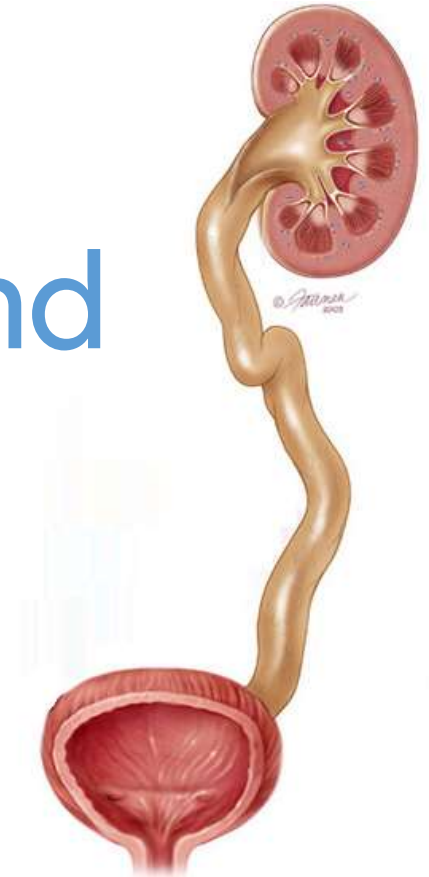


Introduction

- Megaureter refers to **dilated ureters >7mm**
- Causes include **reflux, obstruction** and **physiologic** (polyuria, infection)
- **Ultrasound** shows ureter dilatation more pronounced than pevicalyceal dilatation
- **VCUG** should be done
- **Renal scan** may help decide treatment (DMSA in refluxing, MAG3 in obstructing)
- Some patients may be **observed** while others require surgical intervention (urgent **decompression**, **endoscopic** procedures, **reimplantation with tapering**)

Outline

- **Embryology**

1. Refluxing megaureter

    1.1. Primary VUR (vesicoureteral reflux)

2. Obstructed megaureter

    2.1. **POM (primary obstructive megaureter)**

    2.2 **Duplex system-ectopic ureter-ureterocele**

3. Refluxing + obstructed megaureter

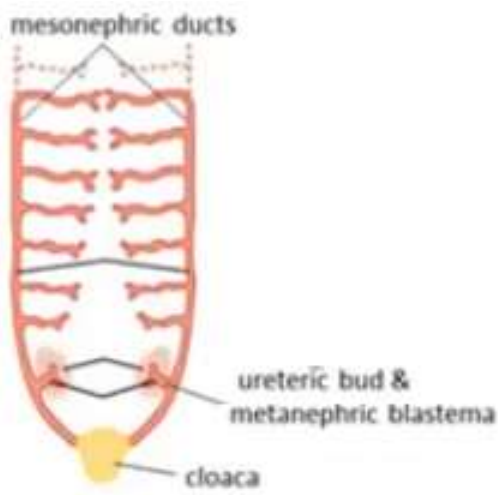
4. Non-refluxing + non-obstructed megaureter

Embryology:
ureteric development

Development of kidney and ureter

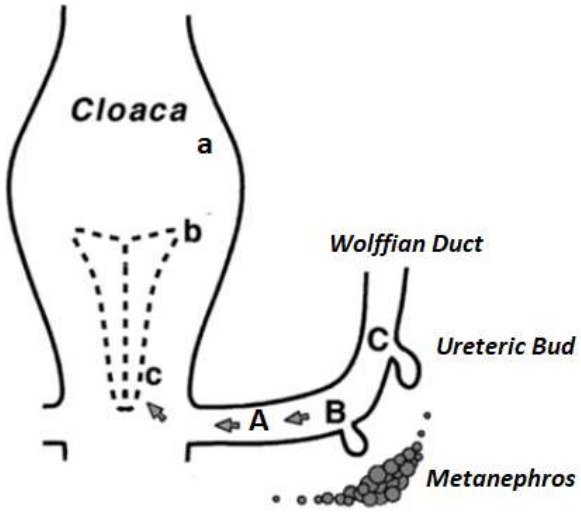
3 systems formed crania-caudal

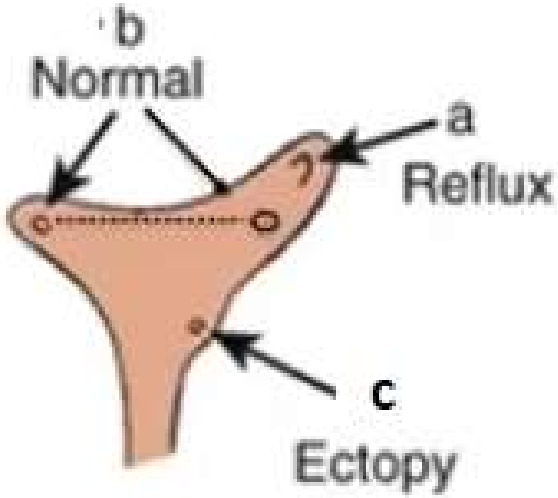
- **Pro**nephros (4th wk): non functional, regress completely (**rudimentary**)
- **Meso**nephros (4th wk) : thoracic, temporary kidney, regress but duct remains (**wolfian duct**) which gives **uretric bud,** seminal vesicle, gartner duct (female)
- **Meta**nephros (5th wk) : pelvic, permanent **kidney**
  - 6th wk = Uretric bud connect to metanephros (in middle) to induce each other
  - 9th wk = kidney ascend to adult position, rotate 90 degree, blood supply changed
  - 10th wk = Urine (but adequate enough for US at 16-20wk)
  - 36 wk = all nephrons formed (2 million/kidney)
  - Birth = still lobulated (disappear after 1yr)



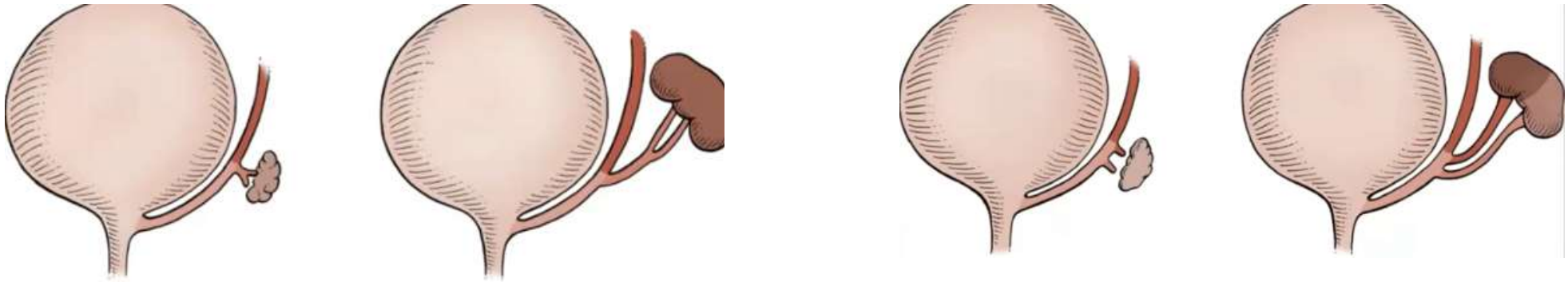
Theories of ureteric anomalies

- *Distal part of ureter last to develop (lack of smooth muscle) ----* **Primary UVJO**
- *Uretric bud anomalies (Renal dysplasia may occur b/c abn interaction to metanephric)*
  - *Early budding ---***Primary VUR**
  - *Late budding - --***Ectopic ureter**
  - *Early splitting of ureteric bud ----* **Incomplete duplication**
  - *Two uretric buds----* **Complete duplication**
- *Failure of rupture of chawalle's membrane ---* **Ureterocele**



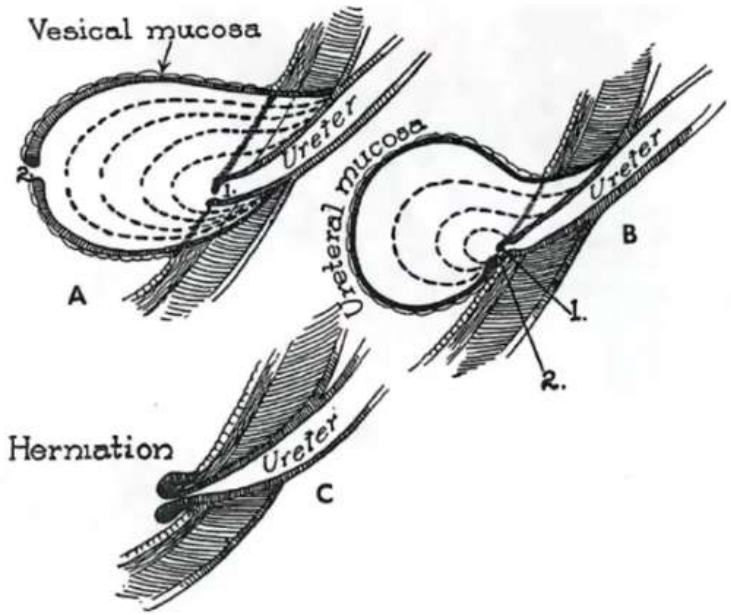


Theory of Ureteric bud misplacement– abnormal origin of uretric bud from mesonephric duct leads to abnormal ureteric insertion outside the trigone. The ureter may also not hit metanephros in the middle, making dysplasia more likely. Early budding leads to early incorporation to UGS making insertion lateral and high. Late budding leads to ureter being carried caudally to insert with mesonephric duct.



Theory of duplex systems: early splitting of uretric bud (incomplete duplication)
two uretric buds (complete duplication)

Wiegert-Meyer law of crossing ureters in a complete duplication: **Lower pole ureter** may be normal or bud early (cephalad and lateral ureter- **VUR, UPJO**) while **Upper pole ureter** buds late (caudal and medial ureter – **ectopic, ureterocele**). Ectopic ureter can obst & reflux if open at bladder neck or below

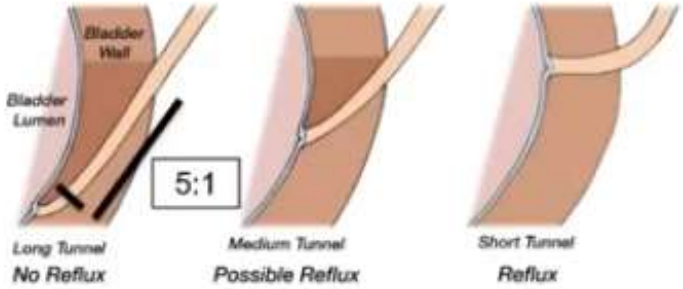


Chwalle's membrane: It was proposed that the a ureteral membrane exists that normally undergoes apoptosis. If the process of apoptosis is abnormal, it results in obstruction of the orifice. Ureter is ballooned out into the bladder creating a ureterocele. Some don't agree with this theory and suggest an **abnormal insertion of ureteric bud** is the cause. Histology shows deficiency in trigone muscle

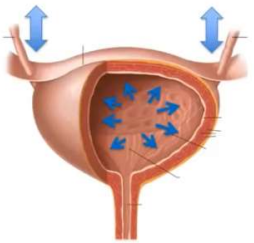
1 Vesicoureteral reflux (VUR)

Refluxing megaureter

- **Primary**
  - Congenital reflux- maldevelopment of UVJ anti-reflux mechanism (ratio of ureteric tunnel length to diameter <5:1)
    - *Additionally ureteric *orifice configuration*, UVJ *smooth muscle composition* and *neural function* may contribute



- **Secondary**
  - Posterior urethral valve
  - Neurogenic bladder (UDC, DSD)



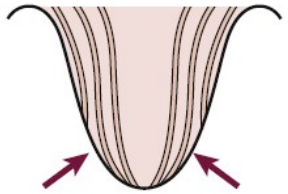
1.1 Primary VUR

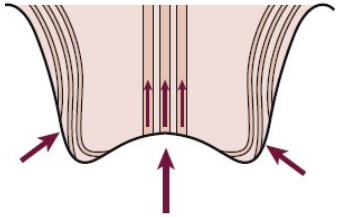
Epidemiology

- **1-2% of newborns**
- **Associated with UTI**
  - 30-40% in children with UTI (up to 70% in infants)
  - 17% without UTI
- **Decrease with age**
  - 70% in <1yr
  - 25% upto 4yr
  - 15% upto 12yr
- **Male preponderance if symptomatic**
  - 85% of VUR in females
  - Older male with UTI likely has reflux
  - 76% of VUR in infant in male
- **More in Caucasian** (?delayed maturation of anti-reflux mechanism)
  - 10x dec frequency in African Americans which equalizes after age of 10yr

Pathophysiology

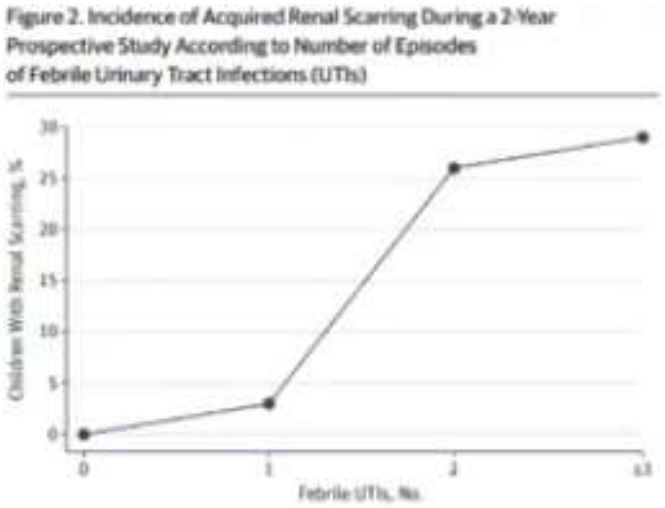
- Failure of UVJ passive valve mechanism >> Retrograde flow of urine
- **BBD** can cause 2ry reflux or aggravate primary reflux
- Sterile urine causes **back pressure** on kidney (water hammer effect)
- Reflux doesn't cause **UTI** but facilitates pyelonephritis
- **Scarring** occurs predominantly in upper/lower poles (location of compound papillae)
    - simple papillae have less intrarenal reflux b/c oblique insertion of collecting ducts .
    - Compound papillae facilitate IRR and scar with lower pressure even without UTI





Clinical implications

- Sterile reflux is unlikely to cause scarring
- **Pyelonephritis**
- **Renal Scarring (**focal/generalized -atrophy, failure of growth)
  - Greatest risk within 1st year
  - risk inc after 2nd febrile UTI
  - <4yr prone to develop scarring after single UTI
- **Reflux nephropathy**
  - HTN (3%)
  - ESRD (<2%)



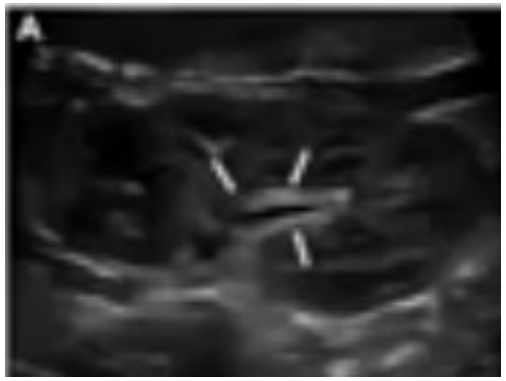
Screening

- **Genetic Risk – V**CUG if abnormal US or history of UTI
  - Siblings – 30%risk
  - Offspring – 66% risk
  - Identical twin - 100% risk
- **Febrile UTI** – VCUG if…
  - Atypical UTI (<2yr, male, septic, not responding, inc cr, non-Ecoli)
  - recurrent UTI
  - Abnormal US
- **Prenatal hydronephrosis** – VCUG if
  - Ureteric or bladder abnormalities
  - High grade hydronephrosis, cortical abnormalities
  - Duplex system, ureterocele

Ultrasound

unable to diagnose VUR

- **Findings**
  - Hydroureter
  - Rena dysmorphia
  - Duplication
- **Improving sensitivity**
  - Uroepithelial thickening: predictor of high grade reflux



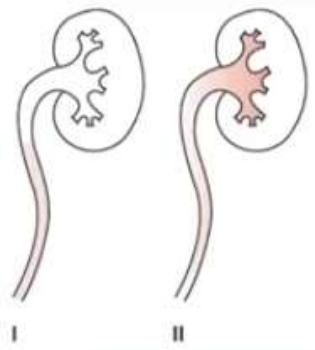
VCUG

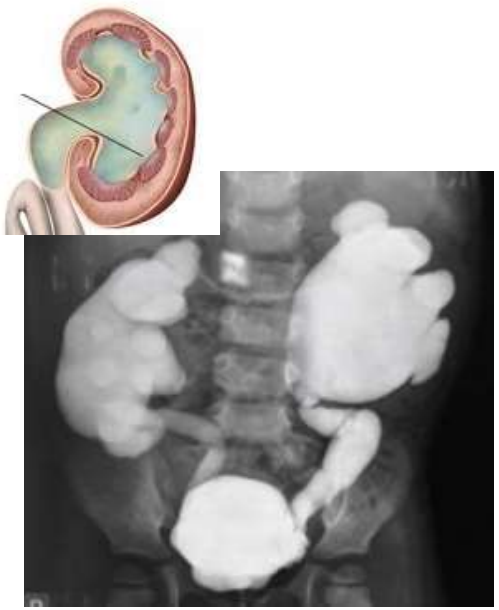
Gold standard

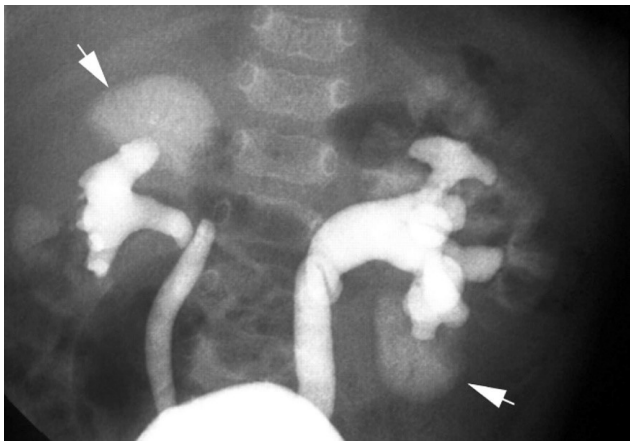
- Timing
  - Urine becomes sterile after 24 hr of successful antibiotic (can go for VCUG early)
- Preparation
  - Prophylactic antibiotic dose is warranted for VCUG
- Technique
  - **Cyclic VCUG** (min 2 voiding cycle) to identify intermittent VUR and ectopic ureter
  - **Delayed films** to identify simultaneous obstruction (UPJO or UVJO)
    - *Obstruction is also indicated by dilution of contrast

Classification

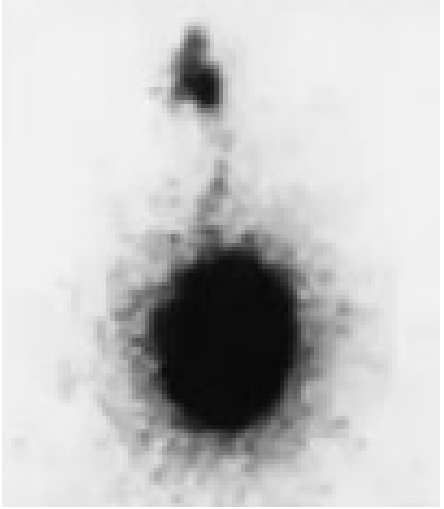
- **Ureteric Anomalies**
  - Simple
  - Complex (duplex system, ureterocele, UVJO, PUJO)
- **Pressure (timing)**
  - Low pressure - only during voiding
  - High pressure – reflux on filling phase
- **Volume**
  - high volume - occur>75% of predicted bladder capacity
  - low volume - occur at <75% of predicted capacity
- **Dilatation (grade)**
  - Non-dilating reflux Grade 1-2 (1- flow to ureter, 2- pelvis-calyces),
  - Dilating reflux - Grade 3-4 (3- mild dilatation & calyceal blunting, 4- ureteric & complete blunting, 5- massive tortuosity, loss of papillary impressions, IRR)

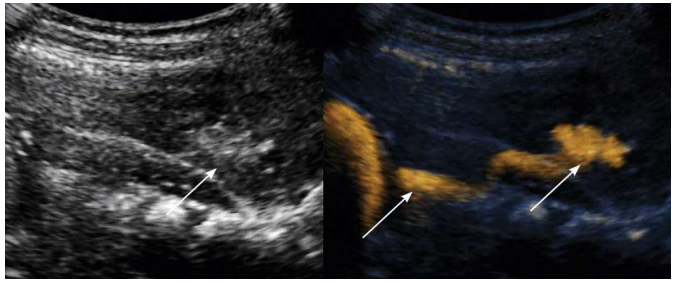






VCUG– blunting of fornices (flattened but maintain papillary impression), loss of papillary impression, intrarenal reflux.



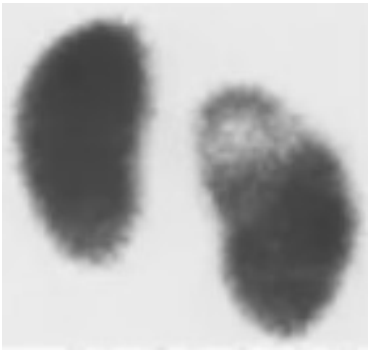


Alternatives to traditional VCUG – VCUG via nuclear cystogram. ceVUS (contrast enhanced voiding urosonography) PIC cystogram (positional instillation of contrast with cystoscopy- for those with negative VCUG),

DMSA

Difficult study to obtain

- **Diagnose renal scarring** (order 4-6 months after febrile UTI)
    - some obtain it for UTI before VCUG (top down approach)
    - Some don't recommend
- **Differentiating renal dysplasia from renal scarring maybe difficult



Goals of treatment

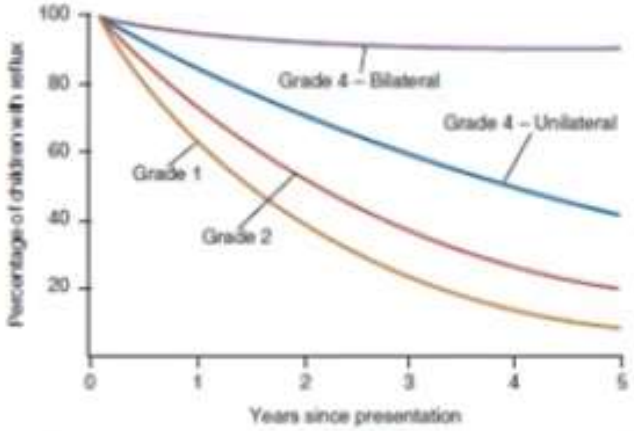
Engage family (shared decision making)

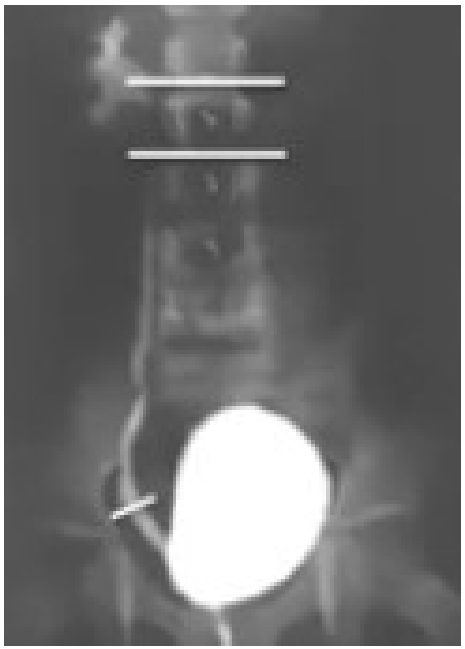
- **Prevent UTI**
- **Prevent renal scarring**
- **Minimize morbidity of treatment**

Natural history

may resolve overtime, sterile reflux won't cause renal damage

- **By grade** - chance of resolution…
  - Grade 1-2 – 60-90%
  - Grade 3 – 50%
  - Grade 4-5 – 10-25%
- **By age** – <1yr can resolve with remodeling of UVJ (**upto 5 yr**)
  - Less likely to resolve if diagnosed at older age (no consensus on how long to observe older school age children)
  - Annual resolution rate (G1-5) -45%, 20%, 17%, 15%, 9%
- **Other factors**
  - Ureteric anomaly
  - Laterality
  - Mode of diagnosis – screening/prenatal hydronephorisis





Variables	Score	Minimum	Maximum
Gender	0-1	0	1
VUR Timing	1-3	1	3
Early-mid Filling	3		
Late Filling	2		
Voiding	1		
Ureteral Anomalies	0-1	0	1
High-grade VUR	0-1	0	1
VURx	1-6	1	6

Improving predictors of prognosis on VCUG: **distal ureteral diameter ratio** (largest ureteral diameter in pelvis/bottom L1 to top L3)**, VUR index** (1-6 point scale)….poor prognosis VUR has more UTI and less chance of spontaneous resolution.

Surveillance

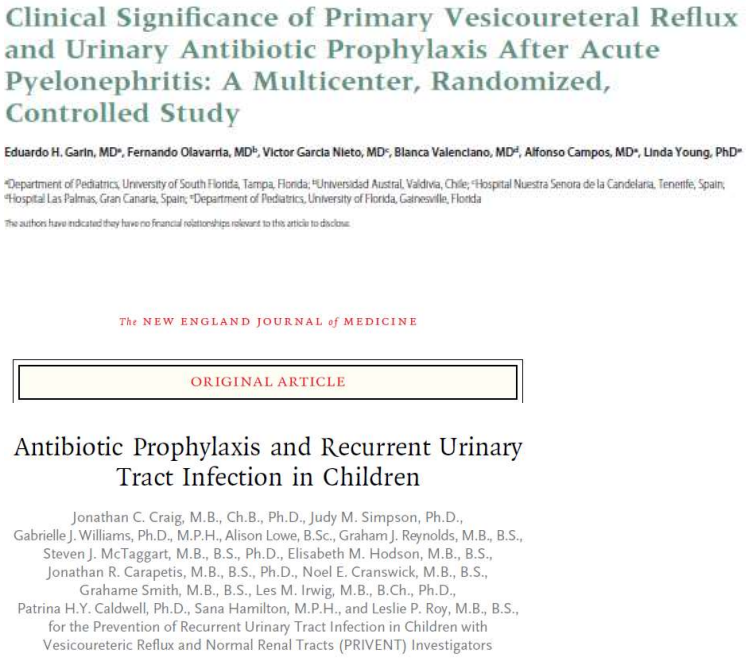
- Renal imaging – (U/S or DMSA) – every 6-12mo
- VCUG- not more than once a year
- Other factors – growth, renal function, blood pressure

Continuous antibiotic prophylaxis (CAP)

- Use is controversial
  - Benefit – reduce febrile UTI & scarring…
  - Pitfall- adherence, resistance, cost, allergies,
- recommended
  - high grade reflux and UTI,
  - BBD
- Optional
  - Circumcised (dec UTI in 1st 6 mo by 10 fold, some say more effective than CAP)
  - low grade without UTI, no BBD. Normal US
- Breakthrough UTI
  - Not resistant - Non compliance, low dose
  - Resistance – high bladder volume, high dose
- When to stop? (until toilet trained)

Age	Drugs	Dose
Neonates	Amoxicillin	12.5mg/kg daily
	cephalexin	5-10mg/kg daily
>2mo	Cotrimoxazole	2/10 mg/kg daily
	nitrofurantoin	2mg/kg daily

CAP: should be 1/4th of therapeutic dose, once daily, preferably at night (physiologic retention)



Clinical Significance of Primary Vesicoureteral Reflux and Urinary Antibiotic Prophylaxis After Acute Pyelonephritis: A Multicenter, Randomized, Controlled Study

Eduardo H. Garin, MD[a], Fernando Olavarria, MD[b], Victor Garcia Nieto, MD[c], Blanca Valenciano, MD[d], Alfonso Campos, MD[e], Linda Young, PhD[a]

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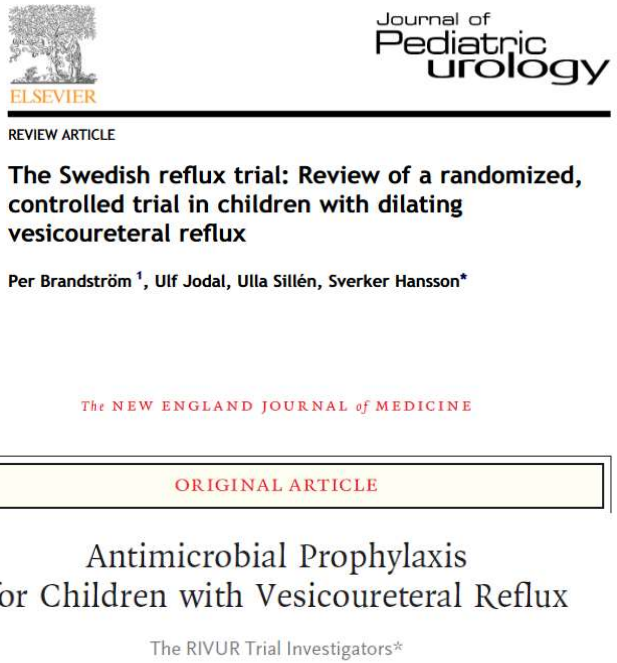
The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Antibiotic Prophylaxis and Recurrent Urinary Tract Infection in Children

Jonathan C. Craig, M.B., Ch.B., Ph.D., Judy M. Simpson, Ph.D., Gabrielle J. Williams, Ph.D., M.P.H., Alison Lowe, B.Sc., Graham J. Reynolds, M.B., B.S., Steven J. McTaggart, M.B., B.S., Ph.D., Elisabeth M. Hodson, M.B., B.S., Jonathan R. Carapetis, M.B., B.S., Ph.D., Noel E. Cranswick, M.B., B.S., Grahame Smith, M.B., B.S., Les M. Irwig, M.B., B.Ch., Ph.D., Patrina H.Y. Caldwell, Ph.D., Sana Hamilton, M.P.H., and Leslie P. Roy, M.B., B.S., for the Prevention of Recurrent Urinary Tract Infection in Children with Vesicoureteric Reflux and Normal Renal Tracts (PRIVENT) Investigators



ELSEVIER

Journal of Pediatric urology

REVIEW ARTICLE

The Swedish reflux trial: Review of a randomized, controlled trial in children with dilating vesicoureteral reflux

Per Brandström [1], Ulf Jodal, Ulla Sillén, Sverker Hansson*

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Antimicrobial Prophylaxis for Children with Vesicoureteral Reflux

The RIVUR Trial Investigators*

RCTs on CAP: **2006 (multicenter trial)** - no benefit of antibiotic; **2009, Australia (PRIVENT trial)-** no benefit, cause Bactrim resistance; **2011, (Swedish reflux trial)** – dec UTI in females but not boys; **2013, USA (RIVUR trial)**- dec UTI but didn't affect scarring

Treat UTI

Early treatment of febrile UTI limits renal scarring (postponing 2-3 days significantly inc scarring)

- **Symptoms**
  - Neonate = sepsis
  - Infant = fever, GI symptoms
  - Children = LUTS
- **Urinalysis** = microscope (>5WBC/HPF), dipstick (leukocyte and nitrate positive)
- **Urine culture** = can't be interpreted without urinalysis and overall clinical picture
  - Suprapubic aspirate – any organism
  - Catheterized – 50,000 CFU/ml
  - Clean catch – 100,000 CFU/ml (Midstream void for toilet trained)
  - Bag specimen – least reliable (only no growth result is useful)
- **Antibiotic** - 7-14d, type based on sensitivity

Clean catch urine in babies not toilet trained: The method of urine collection and the presence of pyuria are of utmost importance in the diagnosis of UTI to avoid false-positive culture results. Quickwee trial (2016) found **rubbing suprapubic area by wet gauze results in spontaneous voiding** <5 minutes

Treat Bladder and Bowel Dysfunction

Close link between BBD (especially constipation) with VUR and recurrent UTI

- **Epidemiology :** Identified in 43% of patients with primary VUR
- **Symptoms:** Urinary urgency, frequency, prolonged voiding intervals, daytime wetting

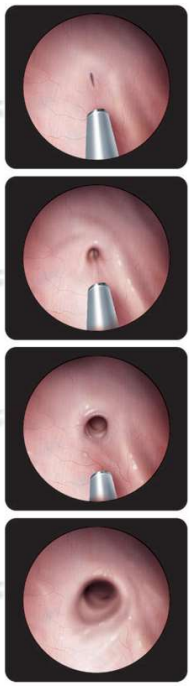
  : Constipation, incontinence, holding maneuvers, Perineal and penile pain
- **Implication:** Constipation leads to sympathetic, concomitant contraction of both anal & urinary sphincters
  - Should be recognized and eliminated for spontaneous resolution of reflux
  - Leads to poor surgical outcome (persistence/recurrence of reflux, breakthrough infection)
- **Treatment**
  - Timed voiding (every 2 hours), good hydration
  - Treat constipation, perineal hygiene

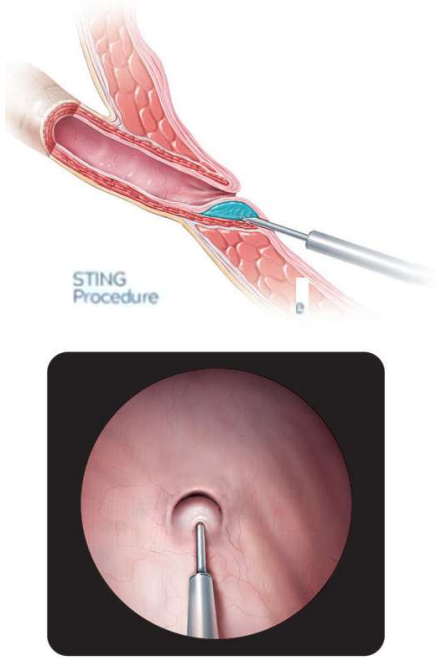
Surgery: Indications

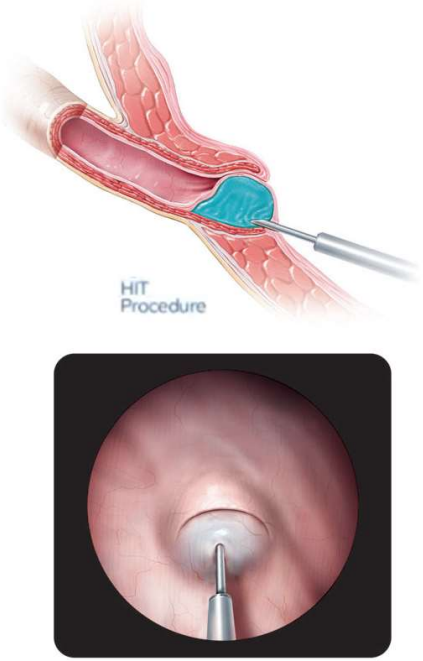
- **UTI**
  - Recurrent breakthrough infection
  - Intolerance or non-compliance to medical therapy
- **Renal status**
  - Renal scarring
  - Progressive renal injury
  - Failure of renal growth
- **Relative indications**
  - **Persistent** high grade VUR
  - Parental **preference**

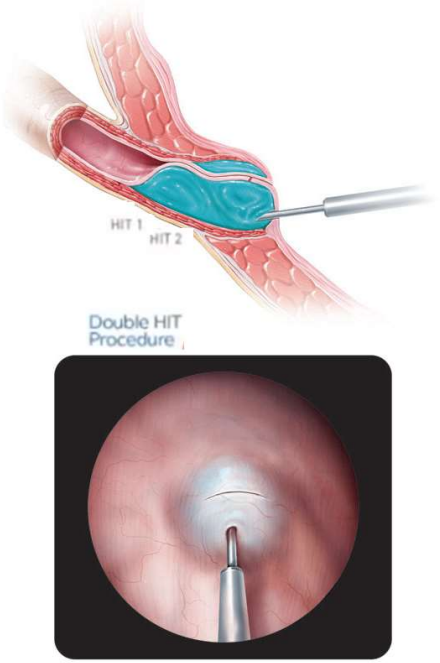
Endoscopic injection of bulking agents

- Candidates – grade 1-4 VUR
- Agents used- biocompatible (no fibrosis/inflammation), persist minimum of 5 yrs
  - Teflon - 1984
  - Deflux - 2001(degradable, no immunogenic/malignant)
- Risks – obstruction (0.6%), implant migration, dissolution, disruption (mucosal breach)
- Success - 70-80%
  - Follow up VCUG is mandatory (at 3mo)
  - If fails deflux can be removed. Then repeat deflux may be tried 1 or 2 times.
  - *NB. Implant may be come dense overtime and mistaken for urolithiasis*









Deflux injection techniques: **STING** (*subureteric teflon injection*- on bladder wall under orfice), **HIT** (*hydrostatic implantation technique*- intra-ureteric, improved success, less migration), **Double HIT** (most commonly used)

Hydrodistention is performed with tip of scope at ureteral orfice. Injection is made 2–3 mm below the refluxing orifice at a 6o'clock position and continued until there is a prominent bulge on the ureteral orifice. Once a volcano appearance, with the ureteral meatus on top of the mound, is achieved, additional volume is injected until the ureteric orifice ebecomes crescent or slit shaped. Lack of hydrostatic distention post injection confirms success.

Reimplantation

- Cystoscopy at beginning
  - exclude inflammation
  - confirm position& number of ureteral orifices
- **Intravesical reimplantation-** bladder open, ureteral anastomosis *(risks = obstruction, hematuria, spasm)*
  - Polatino-leadbetter (1958)
  - Glen aderson (1967)
  - Cohen cross triagonal (1975)
- **Extravesical reimplantation-** bladder never opened, detrusor closure *(risks = diverticula, nerve injury)*
  - Lich-gregoir (1961, 1964)
  - Robotic surgery (2004)
- Success - 96% regardless of technique
  - Continue CAP until U/S (6wk post op)
  - Follow up VCUG is optional b/c good success
  - Consider VCUG if suspect obst or reflux (hydronephrosis, UTI)

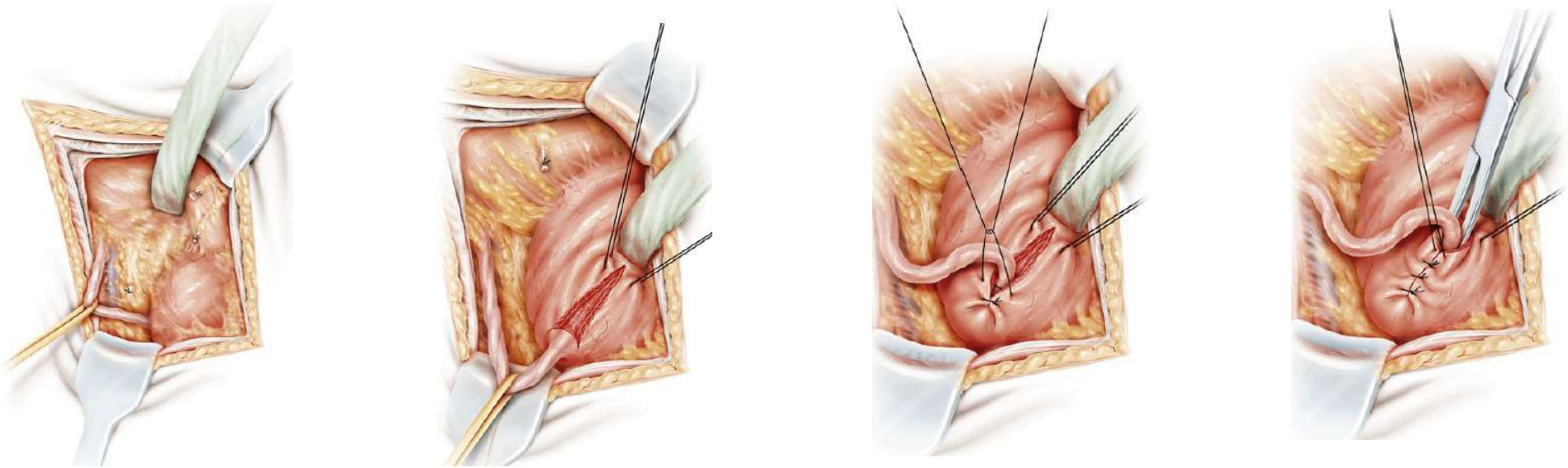
Principles

Adequate bladder size (1yr), **Exclude 2ry causes**

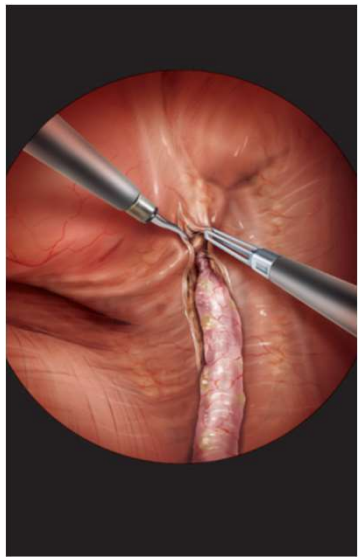
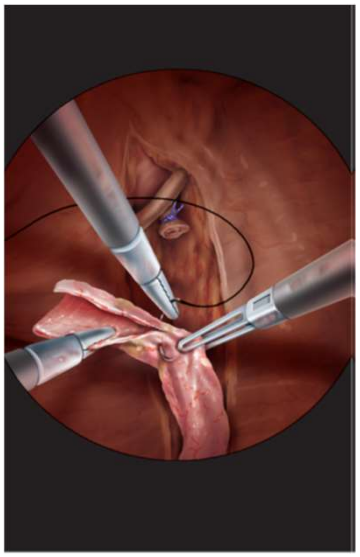
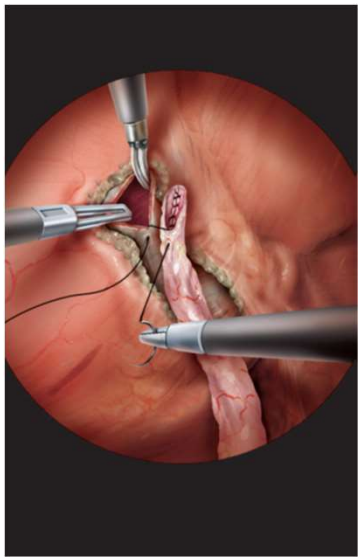
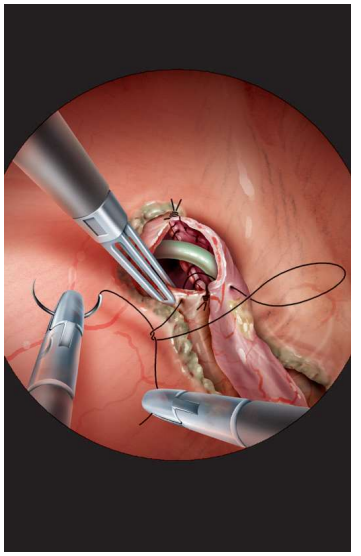
- Adequate exposure and mobilization
- Gentle tissue handling, preserving blood supply
- submucosal tunnel with 5:1 ratio (some argue a full 5:1 ratio may not be absolutely required)
- Ensure ureter is straight (no twist or angulation)
- Tension free anastomosis (mobile ureter with adequate blood supply)
- Patency of anastomosis
- Intravesical reimplantation - gentle handling of bladder (to minimize spasm)
- Extravesical reimplantation - mobilize detrusor flaps, make sure there is space (don't cause obst)

Extravesical Reimplantation

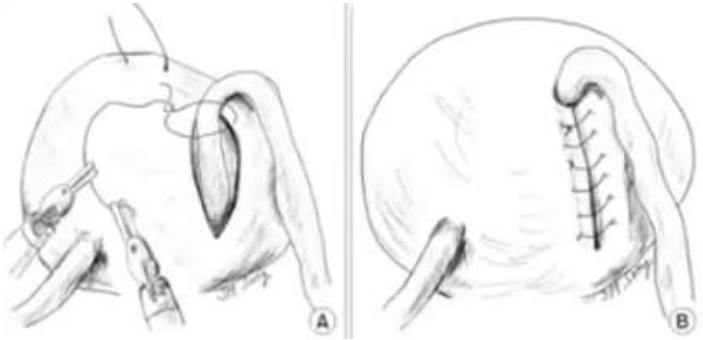
- Popular in Europe
- Maintain anatomy
  - no hematuria, minimal spasm, no stent/drain
  - can discharge early (remove catheter 1d for unilateral, 2d for bilateral).
- 4% Risk of temporary urinary retention (sometimes permanent) if done for bilateral
  - potential injury to pelvic nerves and resultant bladder emptying neurogenicity
  - Risk appear to be infants and girls with large, thin-walled bladders and preexisting retentive voiding
  - one at a time for bilateral, child needs to be toilet trained or need to leave suprapubic tube).
- **Robotic**
  - Better cosmesis, shorter hospital stay, less pain.
  - Higher cost, port site hernia.



Lich-Gregoir (Detrusserorraphy) ureter mobilization stops at UVJ, where dissection should stop to avoid injuring the small nerves that circumscribe the ureter and branch out to the detrusor lateral and distal to the UVJ. Plane between the detrusor and bladder mucosa dissected ureter must be anchored inferiorly to stabilize the UVJ. The ureter is then laid into the trough created by opening the detrusor and the detrusor is brought together over it. Periodically a right angle clamp should be placed anterior to the intramural ureter to be certain that the tunnel is not too tight. Anchoring suture as it enters the tunnel to prevent it from everting during bladder filling.

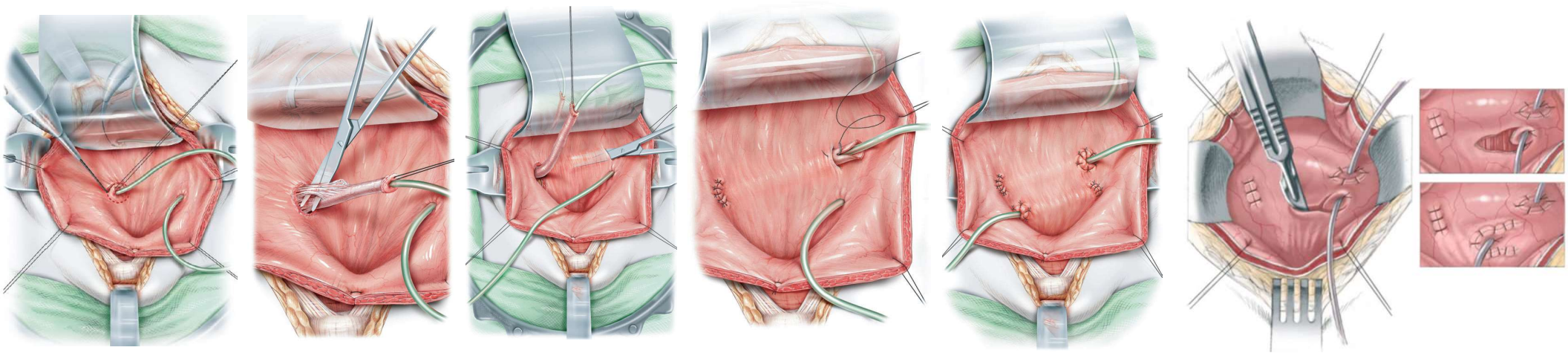
Lich-Gregoir (with ureteroneocystostomy) UVJO ***r****esection, tapering and opening bladder mucosa equal to distal ureter for ureteroneocystostomy.*



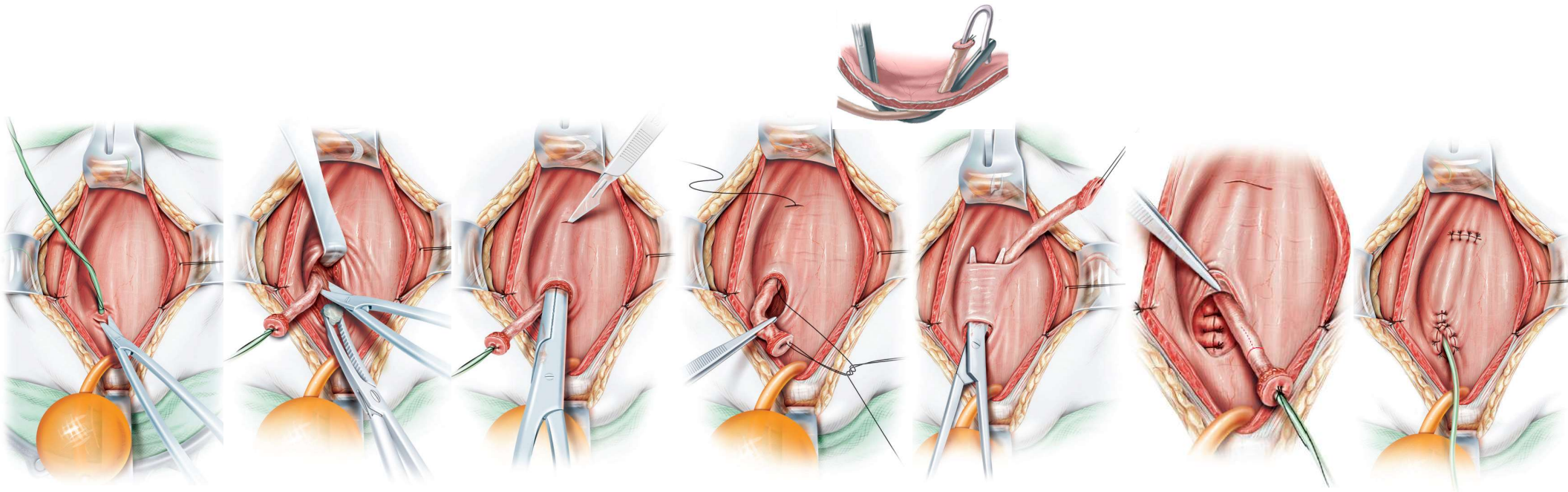
Robotic Reimplant : for adolescent, obese.

Intravesical Reimplantation

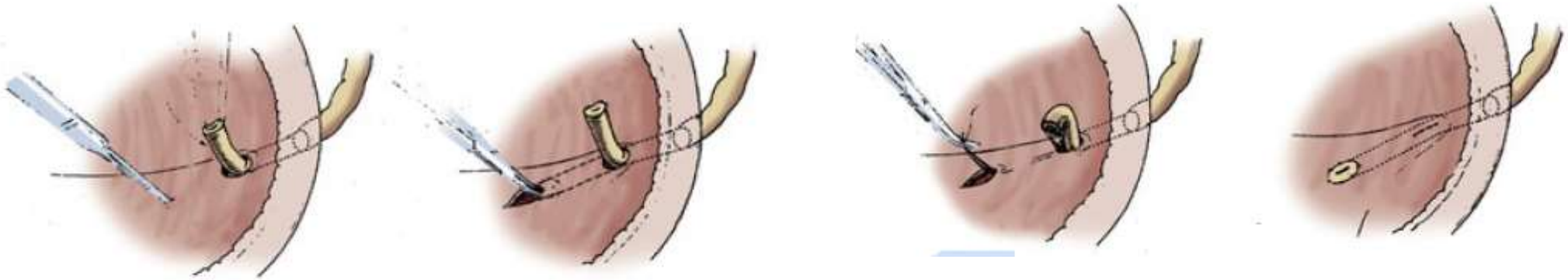
- **Cohen cross triagonal**
  - Most common.
  - Difficult to sent (if develop stone disease in the future)
- **Polatino-leadbetter**
  - Orthotopic reimplantation (easily catheterizable neo-orifice) and good option for bilateral.
  - More complex technique. Concern for kinking.
- **Glen-Aderson**
  - avoidance of angulation and kininkg.
  - tunnel limited by bladder neck (may not be enough space)



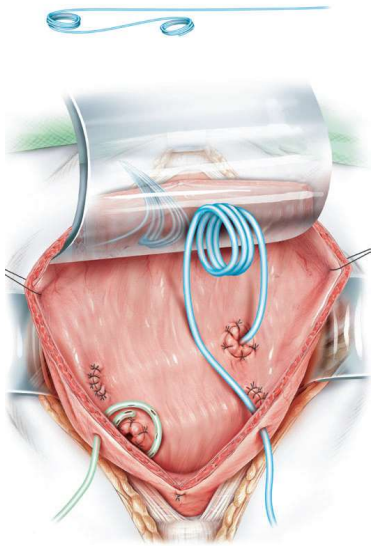
Cohen (cross trigonal advancement): *Uretric orfice circumcised with diathermy and dissection continues to mobilize ureter until peritoneum. uretral orfice may be narrowed to prevent diverticula. Tunnel created horizontally just above trigone. Distal 2cm is excised and spatulated. Ureter is anchored to bladder muscles and ureterovesicostomy done. Same can be done to other side. tunnel for the more laterally displaced ureter is directed superior to the contralateral orifice. If procedure is done for unilateral there is a risk of contralateral reflux in 10-50% which may be prevented Gil-Vernet ureteral reimplant (contralateral ureteral meatal advancement- medially with Y shaped incision)*

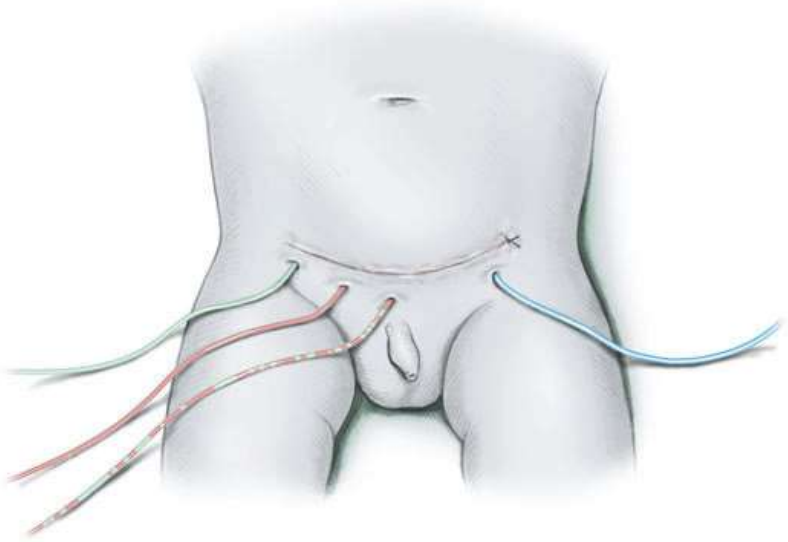


Polatino-leadbetter (creation of new hiatus): dissect distal mesoureter and transvesical mobilization of the adherent peritoneum. Neohiatus created 3cm above old orfice. Transvesical pullthrough of moblizied ureter using free suture as guide rail. Ureter is then transposed into submucosal tunnel prepared from old to new hiatus. Extravesical dissection may be combined with intravesical to avoid injury to bowel/peritoneum. Orfice is fixed at initial position after resection of dysplastic ureteric segment.



Glen Anderson (advancement of hiatus) : same hiatus with ureteral advancement distally.

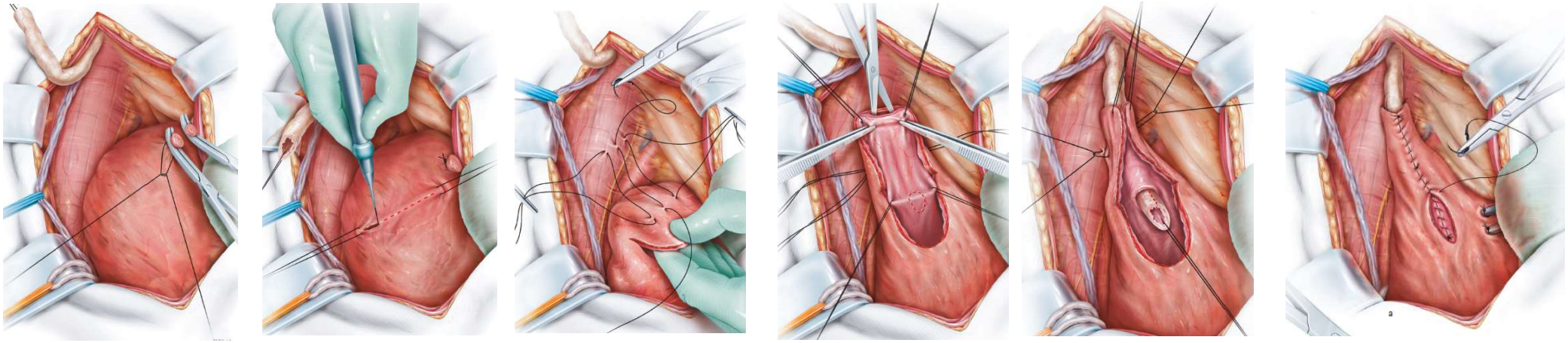




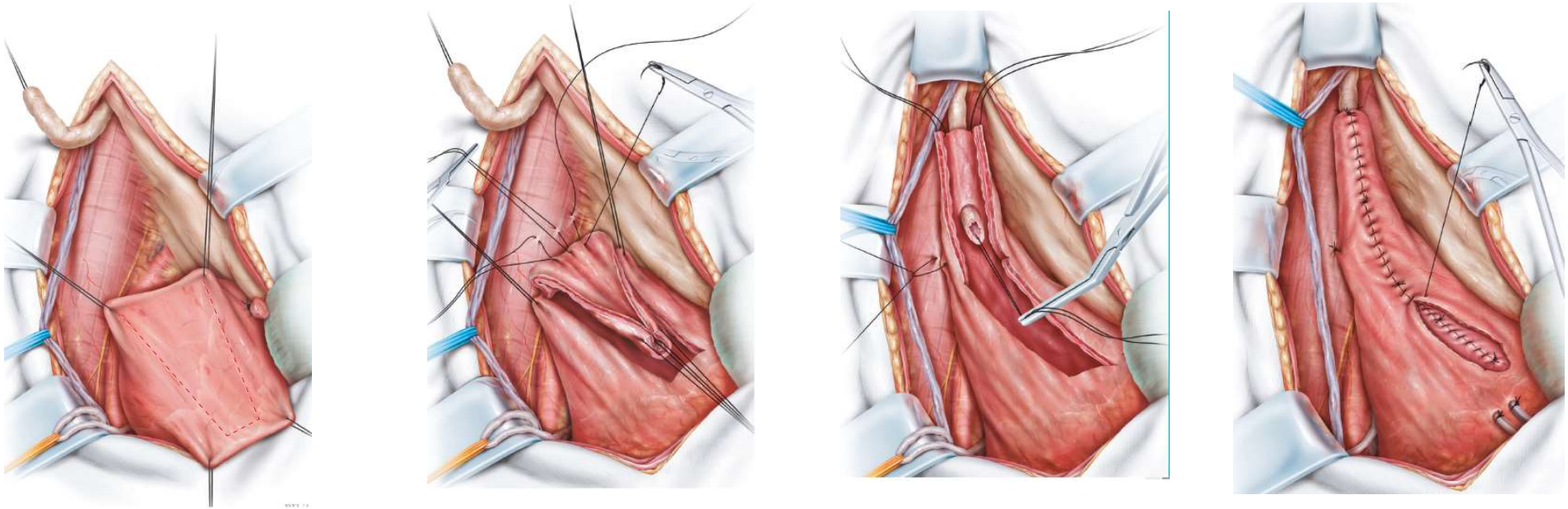
Drainage : There is no consensus on the efficacy of drainage in routine reimplantation. Ureteral stent should be considered in reoperative cases and ureteral tailoring. Bladder spasms are common and oral oxybutynin can be useful to reduce the discomfort. **Wound drain** - Remove on day 3, **transurethral catheter** when urine is clear (day 3–5), **exteriorized uretric stent** (4 or 6fr feeding tube) - 2 days upto 10 days (if the ureter has been re-modelled) **cystostomy** 9d

Redo-reimplantation

- technically more challenging.
  - preferable to create a new hiatus and tunnel.
  - excise ischemic segments until free bleeding and peristalsis (intra & extravesical mobilization as needed)
- The ureter may be short
  - Unilateral - **psoas hitch** .
  - Bilateral - psoas hitch on one side with a **transureteroureterostomy** for the other
  - higher defects (>5-8 cm above ureteric orfice) - **Boari flap**
  - Very significant shortening - a reconfigured segment of bowel similar to the **Monti procedure**.



Psoas Hitch ureteroneocystostomy: Mobilization of the bladder to allow a tension-free fixation of the bladder at the psoas muscle at least 2–3 cm above the common iliac vessel. Implantation of the ureter into an immobilized part of the bladder to prevent kinking during filling and emptying of the bladder.



Boari Flap ureteroneocystostomy: If bladder hitch doesn't guarantee a tension-free ureteric anastomosis, additional bladder flap may be used to bridge lesions of distal ureter.

Follow-up

- **Restrictions**
  - stay away from school for 2 weeks after surgery
  - avoid sport for 1 month.
- **Ultrasonography** (ensure that there is no increased dilatation)
  - at 2 weeks, 3 months, 12 months, and 24 months
- **VCUG** is not typically obtained in an asymptomatic patient

Complications

- **Bladder spasm** (intravesical approach, for one to 2 weeks post-operatively)
  - dysuria, urgency, and hematuria
  - Oxybutynin chloride TID for 10 to 14 days
- **Contralateral reflux**
  - distortion of the contralateral hemitrigone, missed preop
  - 10%, but 80% resolve spontaneously
- **Persistent reflux** (Inadequate tunnel, BOO)
  - majority of low grade
  - postoperative reflux on VCUG at 3-month followup disappeared spontaneously
  - Even with successful surgical correction, 10% will develop a febrile UTI over the following 10 years.
- **Diverticula**

Obstruction

- Causes
  - Early obst = Twisting, intramural blood clots. submucosal hematoma or edema
  - at neohiatus – high (angulation), stricture, devascularization
  - Tunnel = tight
  - kinking at obliterated umbilical artery
- **Acute postoperative obstruction**
  - Nephrostomy >> change to stent as early as possible>> antegrade contrast study when stable
- **Late obstruction**
  - balloon dilation and stenting or unroofing the distal ureter
  - If fail, reimplant

Special scenarios

- **Bladder diverticula**
  - Paraureteral diverticula – not indication for intervention
  - Ureter refluxing into diverticula – reflux not expected to resolve
- **Renal agenesis**
  - associated with higher contralateral anomalies
  - Reflux less likely to spontaneously resolve as compared to MCDK
- **Megacystitis-megaureter association**
  - Massive bilateral VUR leading to gradual bladder dilatation
  - Important to d/t from PUV
  - Bladder may normalize if rehabilitated after VUR is corrected
  - Vesicostomy may be done to temporize until reimplantation done
- **Reflux persisting through puberty in girls**
  - Some recommend surgery b/c reflux cause problems during pregnancy
  - Some prefer discontinuing CAP and monitoring

Concomitant UPJO

- **Cause**
  - Primary UPJO - with incidental reflux
  - Secondary UPJO - Kinging, severe diltation, repeated inflamation
- **Epidemiology**
  - Upto 3%
  - 5x more common in higher degree reflux
- **Signs**
  - Severely dilated ureter with little or no filling of pelvis may indicate kinking.
  - contrast entering pelvis may be poorly visualized b/c diluted by urine
  - Delayed films show retention of contrast in dilated pelvis
- **Treatment** - Peyloplasty required if drainage not good on postvoid images or on scintigraphy
  - Correction of UPJO takes precedence over VUR (secondary UPJO cant be corrected by correcting reflux)
  - May intervene VUR via endoscopy during pyeloplasty (Simultaneous reimplantation risks ureteral devascularization)

2 Ureterovesical junction (UVJ) obstruction

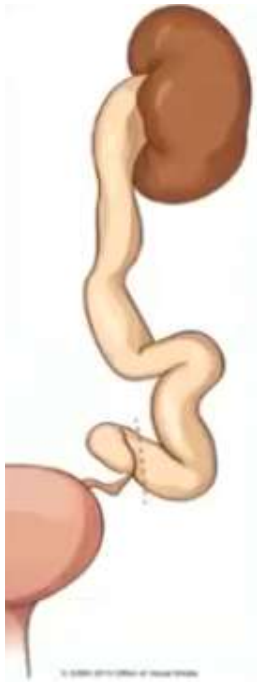
Obstructive megaureter

- **Primary**
  - Adynamic distal ureter (primariy obstructive megaureter)
- **Secondary**
  - Duplex system- Ectopic ureter-ureterocele
  - Ureteral valves
  - Extrinsic mass or tumor

2.1 Primary obstructive megaureter (POM)

Pathophysiology

- Adynamic distal ureter
  - disruption of muscle continuity/peristalsis) but stent can pass
  - UVJ last to mature (craniocaudal development)



Epidemiology

- 10-20% of antenatal HN
- M:F = 4:1
- Lt > Rt
- Bilateral in 25% (more likely diagnosed in infants)
- Contralateral dysplasia or obst in 10-15%

Evaluation

- Presentation- prenatal, Abd mass, pain, vomiting, hematuria
- Ultrasound
- VCUG
- MAG 3 –**Indication**: no VUR, ureter dilatation >1 cm, associated parenchymal thinning

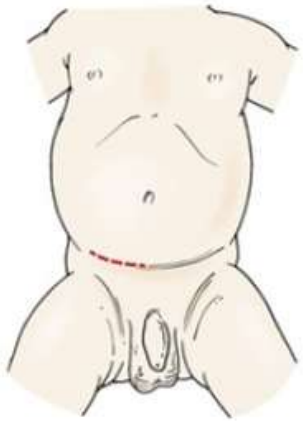
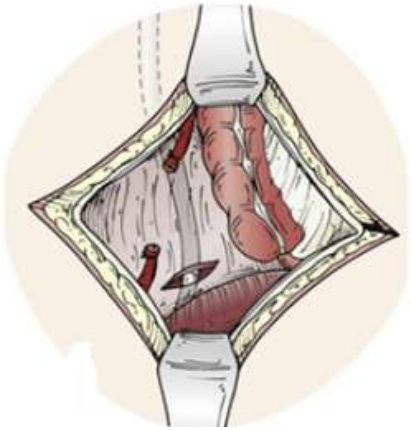
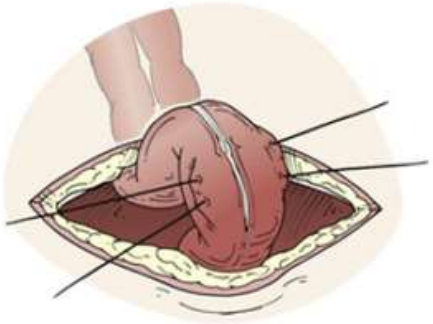
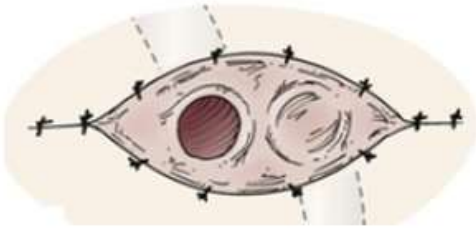
Observation

Neonatal megaureter with obstruction but preserved function can be observed

- majority resolve with time – 75-85%
  - Spontaneous resolution inversely related to size (>1.5 cm less likely to resolve).
  - However some stable megaureter may worsen overtime (puberty)
- Prophylactic antibiotic first 6-12mo of life (controversial)
- Surgery for 10-25% by age 7
  - recurrent UTI
  - Increased hydronephrosis
  - Renal scan (low function<40%, dec function >5%)
  - older age

Temporary diversion

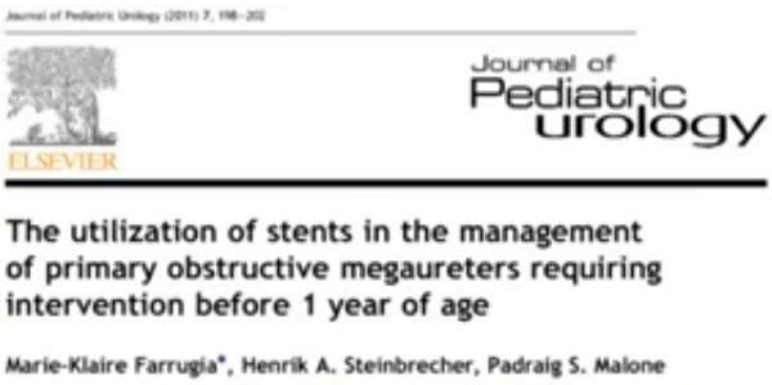
- infant in whom the chance of successful reimplantation is diminished (bulky ureter into a small bladder.)
- Urgent drainage – for severe obstruction/ infection
- External diversion
  - **Nephrostomy**
  - **end-cutaneous ureterostomy**
- Internal diversion
  - **Stenting**
  - **refluxing end-to-side anastomosis of the ureter to the bladder**

Cutaneous ureterostomy (CU): small inguinal incision, split muscle, identify ureter, split umbilical artery if necessary, ureter brought out as a loop and fixed to fascia, and skin. No resection of redundant ureter is performed because this will shorten in time and some length will be needed to perform the reimplantation. *Problematic complication is severe stenosis. Not preferred in bilateral cases b/c it will defunctionalize bladder.*

Dilatation and stenting

- Success - 70-80%
- **Technique**
  - ureteral orifice dilated (distal 3cm) by a ureteral catheter or high-pressure balloon under fluoroscopy until the indentation or waisting on the balloon disappears.
  - Thereafter, a double pigtail catheter is left for 2 to 6 months .
- **Complications**
  - VUR
  - Stent migration - 30%
  - ureteral injuries - requiring emergent reimplantation
  - Persistent dilatation - may respond to re-stenting or require reimplantation



Journal of Pediatric urology

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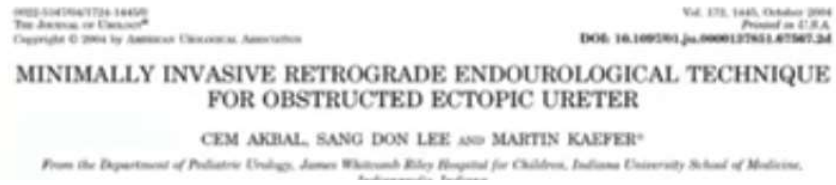
The utilization of stents in the management of primary obstructive megaureters requiring intervention before 1 year of age

Marie-Klaire Farrugia*, Henrik A. Steinbrecher, Padraig S. Malone



MINIMALLY INVASIVE RETROGRADE ENDOUROLOGICAL TECHNIQUE FOR OBSTRUCTED ECTOPIC URETER

CEM AKBAL, SANG DON LEE AND MARTIN KAEFER*

Balloon dilation and stenting : Described primarily in older children with a success rate >90% (second diltation or endoureterotomy may be needed to achieve this), onset VUR occurred in 27% of ureters, one-third of which spontaneously resolved on follow-up. In infants stenting is difficult b/c tortuous (require fluoroscopy) and has higher comp (31%). May also make definitive surgery difficult b/c inflammation.

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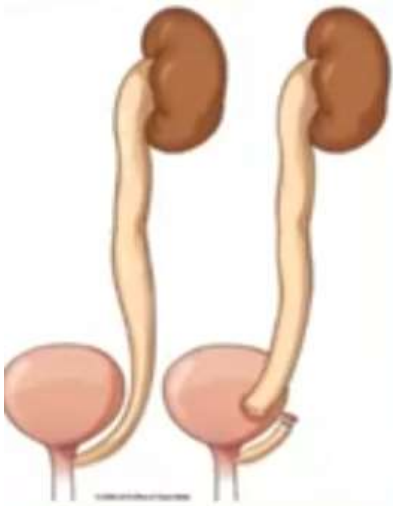
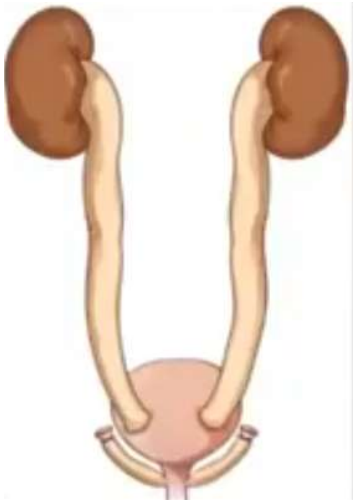
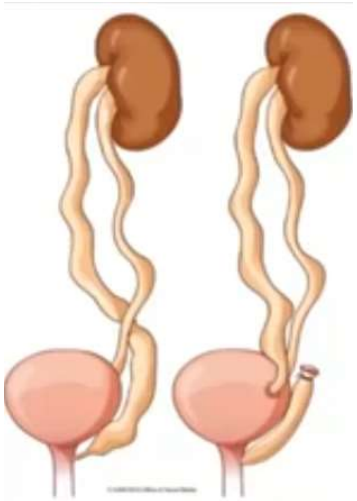
REFLUXING URETERAL REIMPLANT AS TEMPORARY TREATMENT OF OBSTRUCTIVE MEGAURETER IN NEONATE AND INFANT

SANG DON LEE, CEM AKBAL AND MARTIN KAEFER*

From the Department of Urology, Riley Hospital for Children, Indiana University, Indiana, and College of Medicine, Pusan National University, Busan, Korea (SDL)



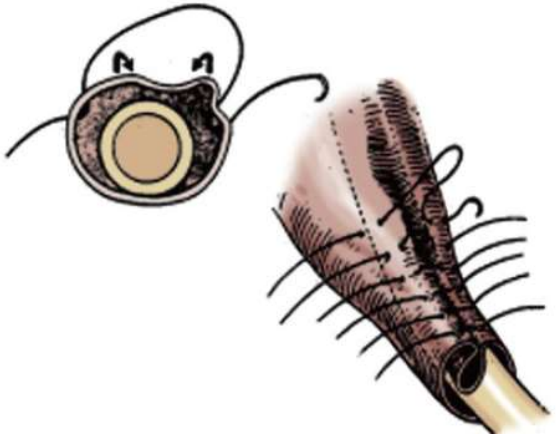


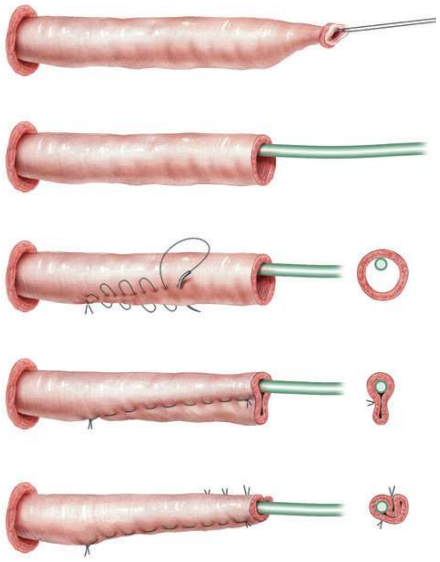


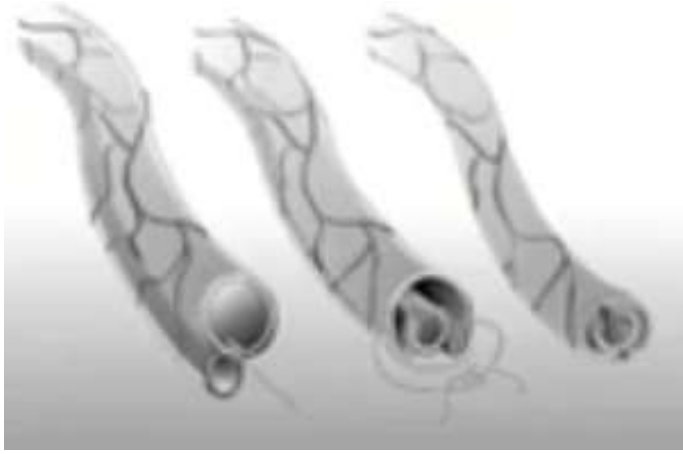
Temporary Refluxing ureteral reimplant (TRUR): single or bilateral 3cm incision, ureteric division proximal to obstruction and end-side anastomosis to bladder in freely refluxing fashion (trade obstruction for reflux). Definitive surgery done through same incision at 1 year. Good option for obstructed megaureter in small infants, especially if bilateral. *Advantage of avoiding stoma, and stenosis while maintaining bladder cycling in bilateral cases. However may have more febrile UTI and greater need for tapering.*

Ureteral Remodeling

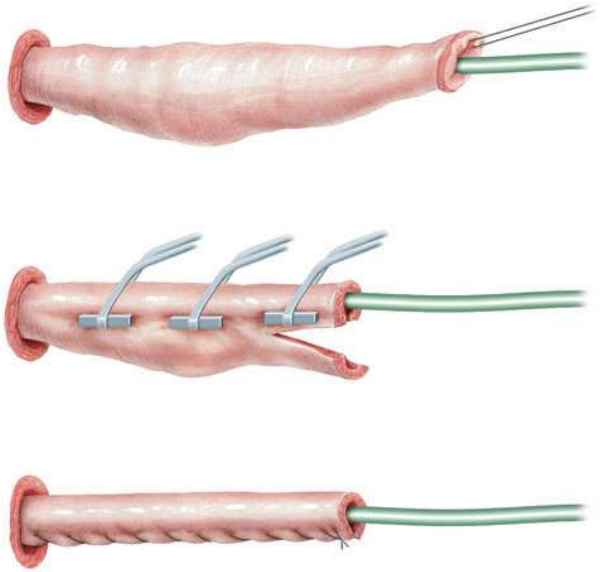
- **Timing** –9-12mo of age (avoid reimplantation of large ureter into infant bladder)
- **Technique**
  - A longitudinal segment of ureter is remodeled over a 8-10 Fr catheter.
  - suture line is positioned against the detrusor during reimplantation to decrease the chance of fistula.
- **Ureteral folding/ Plication** – suitable for <1.75 cm, 90-95% sucess
  - theoretically preserve ureteral blood supply (decrease the risk of ischemic injury)
  - increased bulk from these folding techniques can be technically challenging in infant bladders.
- **Excisional tapering -** for massive dilatation
  - Technically challenging in newborn-small bladder size, risk injuring blood supply redundant tissue is excised,
  - risk of stenosis and ischemia (excise lateral, less vascular ureter)







Plication (folding techniques): **Starr plication-** multiple interrupted lambert sutures.
Kalicinski's technique- continuous sutures to defuctionalize redundant aspect which is folded back and fixed along lumen.
Wrap plication of megaureter around normal ureter in duplex system reimplantation



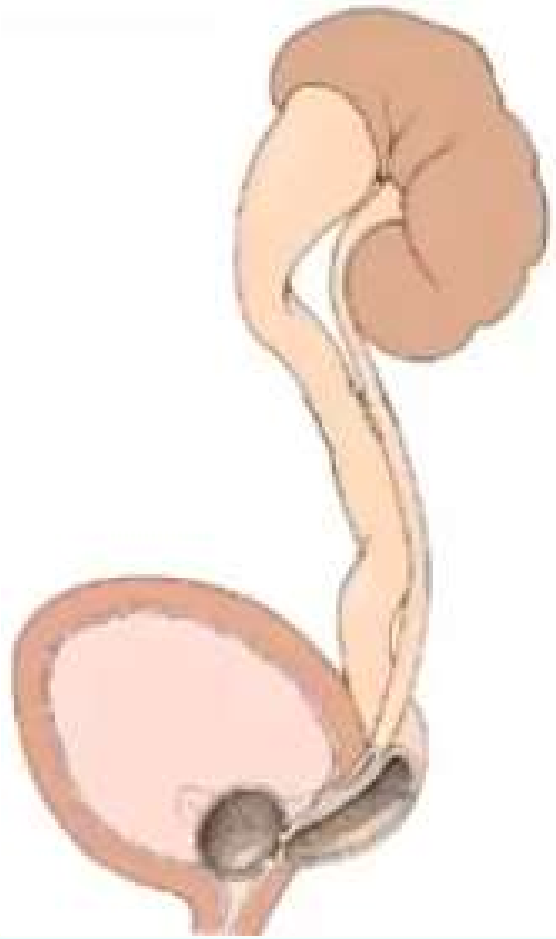
Excisional tapering (henderen's technique): with gradual tapering so as not to cause abrupt change. and closed with continuous locking surutre in proximal 2/3 and interrupted dsutures in distal 1/3

Complications

- **A persistent obstruction/ dilatation**
  - **Postop edema** - stenting
  - **ischemic structuring** - redo reimplantation
- **VUR**
  - reflux has a tendency to resolve spontaneously in many cases.
  - Redo-reimplantation sub ureteric injections fail, especially in cases of recurrent febrile UTIs
- **Urinary Extravasation**

2.2

Duplex system-
Ectopic ureter-
Ureterocele

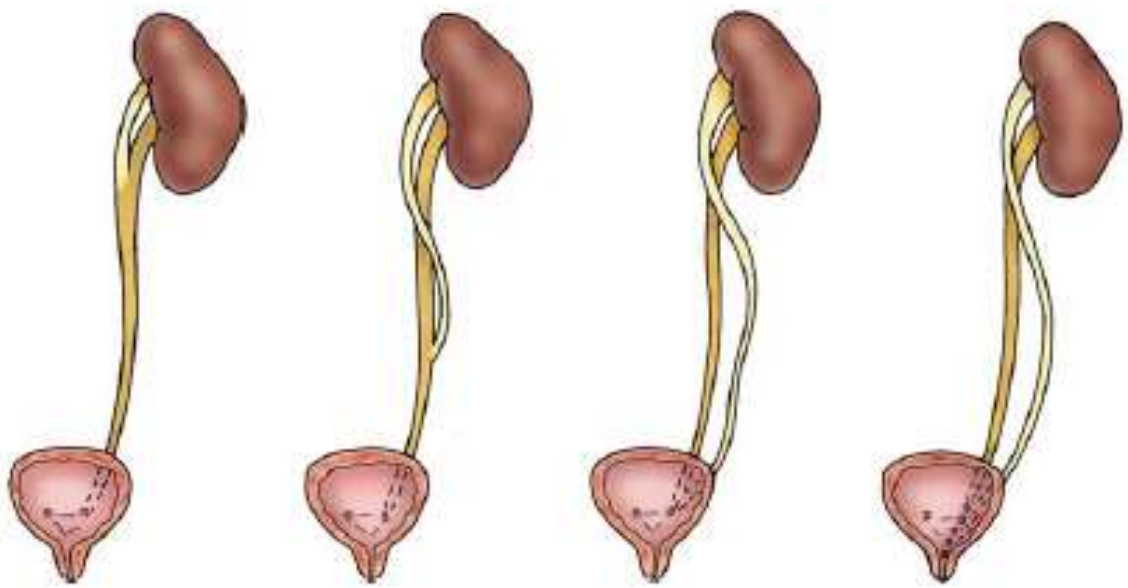


Epidemiology

- The most common anomaly of upper urinary tract (2-4%)
- Both sides equally affected
- F>M = 2:1

Pathophysiology

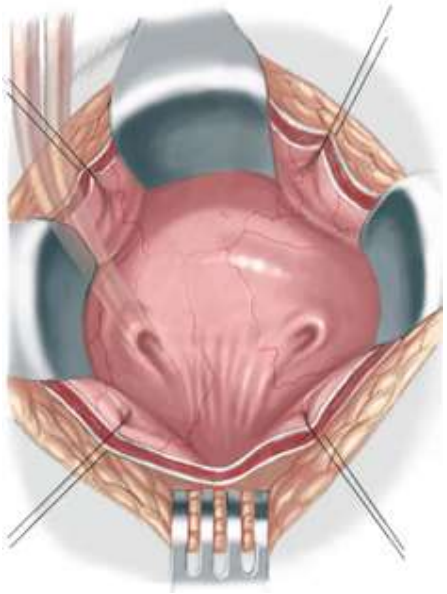
- Can be complete or partial
- Upper pole moiety is usually dysplastic (ass. With ectopic ureter & ureterocele)
- Reflux of lower pole is most common cause of disease (ass. With VUR)
    - Chance of resolution is same as single system
    - Common sheath reimplantation should be done (common blood supply)

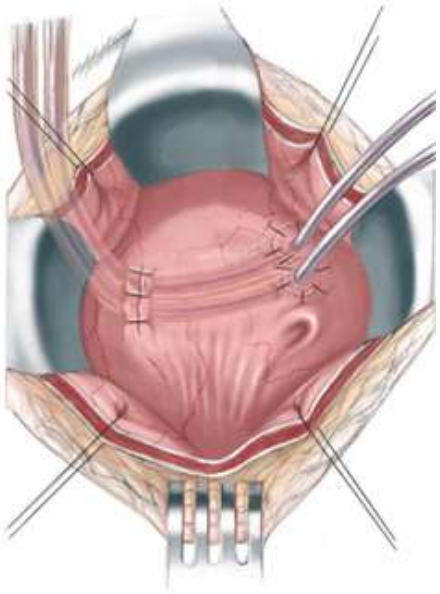


Types of duplication: can be incomplete (bifid pelvis, Y ureter or V ureter) or complete. Rarely ureteral triplication and quadruplication can occur.

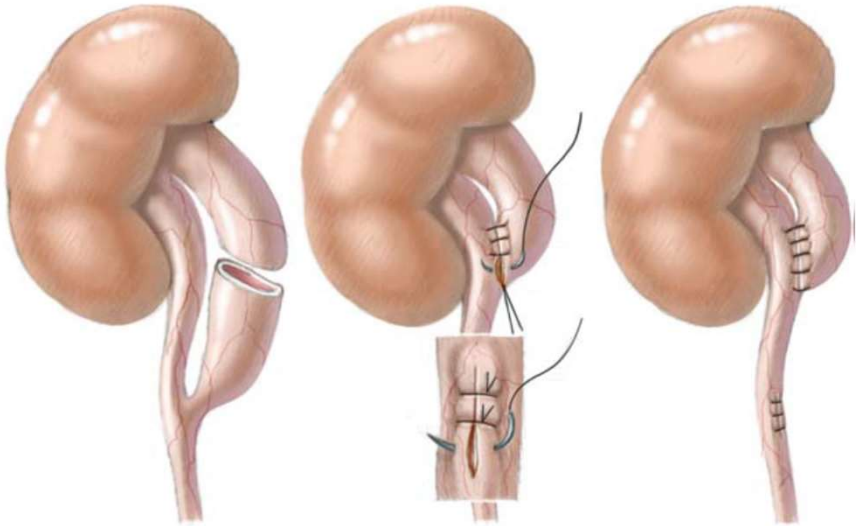
Procedure	Ideal candidate	Advantage	Limitation
Transurethral incision	Infant with large uretherocele + VUR	Simple Occasionally definitive	-May produce VUR -windsock effect (in ectopic urethrocele)
Common sheath reimplantation	Older with VUR	definitive	Complex
Upper tract surgery (heminephrectomy	Older child with no VUR, intravesical ureterocele + nonfunctional	Avoid bladder surgery	VUR Risk to lower pole May not be definitive
Uretero-uretrostomy/ uretero-pyelostomy	Older child with no VUR, intravesical ureterocele + functional	Avoid bladder surgery	May develop reflux Leaves urethrocele

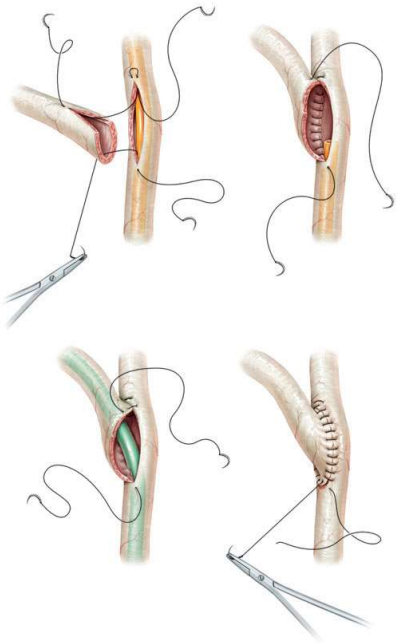
- Decision Making: For ectopic ureter decision is easier (weather to maintain kidney or not). Reflux may indicate lower tract surgery. For urethrocele availability of cystoscopy incision creates more options. Even if half of these patients may need subsequent surgery, it can be safely deferred until the child is older



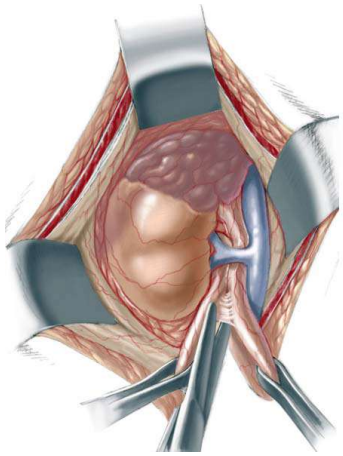
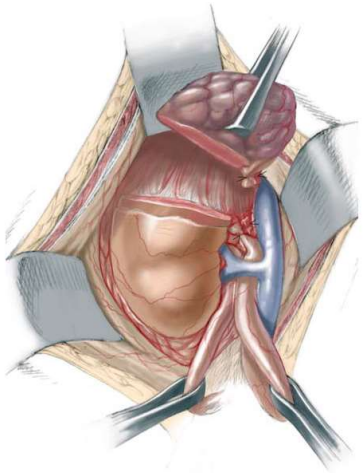
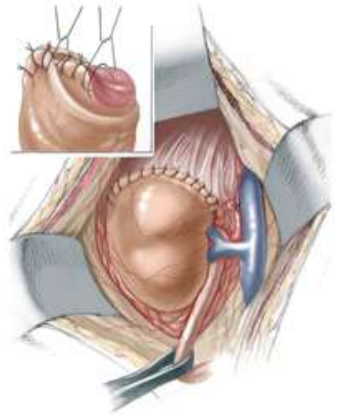


Common sheath reimplantation: separation of ureters is discouraged to avoid injury to common blood supply running longitudinally between the two ureters. The two ureters are dissected as a unit, the upper pole ureter is tapered as needed, and both ureters are reimplanted submucosally. The distal portion of the ureterocele is dissected. Bladder mucosal flaps are raised to cover the removed area. Detrusor muscle is placated if it is attenuated. Persisting reflux can be an issue in 5% to 10% of patients.

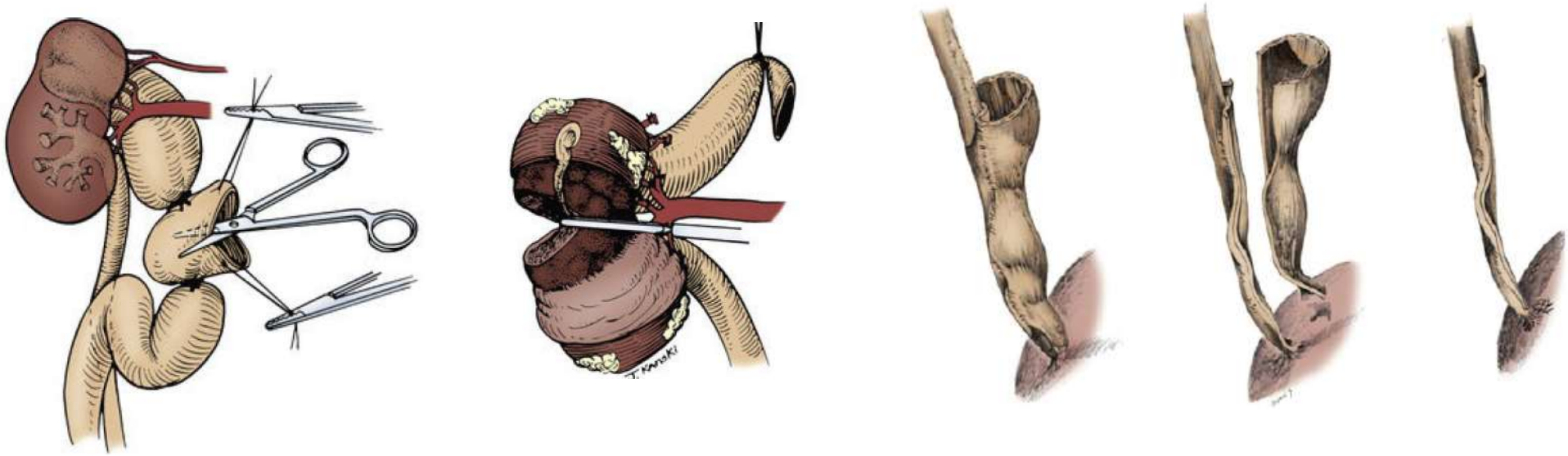




Ureteroureterostomy/pyelostomy: Whether the “yo-yo” phenomenon is clinically significant remains uncertain, but some prefer a proximal anastomosis when there is a significant mismatch in ureteral size. Make a large end to side anastomosis and the distal portion of the upper pole ureter should be resected as far inferiorly as possible, with care taken to stay directly on this ureter's wall and avoid the vasculature of the adjacent lower pole ureter. If there is no reflux, the resection is taken as distally as possible and the remnant lower portion of ureter may be left open.

Upper pole heminephrectomy: the atrophied and dysplastic upper pole is often clearly demarcated and dissecting clevage plane is preferred (less vascular) . The renal capsule on the upper pole is incised to be used later to suture over the renal parenchyma after suturing. Upper pole vessels (usually 2 or 3) are sequentially ligated with care not to make lower pole ischemic. Demarcation of upper pole becomes apparent. Nephropexy to posterior wall to avoid rotation around the long stretched pedicle.. Urinoma in 20% (mostly in series in which there is no formal closure of the polar defect). Whether these urinomas are caused by injury to the lower pole or remnant upper pole is unclear. Asymptomatic urinomas can be observed if not expanding



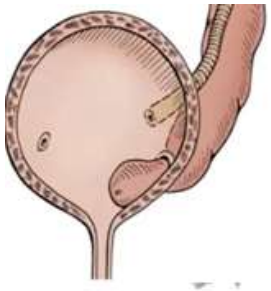
Refluxing stump. If in addition to obstruction there is concomitant reflux into the ureter, some recommend a second incision (Gibson incision) to resect the ureter in its entirety. Laparascopic approaches avoid a second incision that is often needed for the distal ureterectomy. upper pole ureter is usually dilated and tortuous and can be identified readily. maintain the dissection immediately on the wall of the upper pole ureter as much as possible to preserve the blood supply. Backwall of the upper pole ureter can be left attached. Resection is carried out to the level of the bladder, where several sutures are placed to close the upper pole ureteral hiatus.

2.2 .....Ureterocele

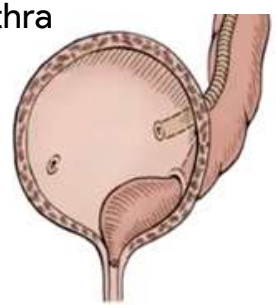
Pathophysiology

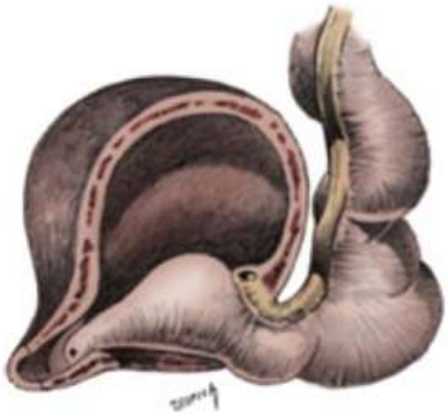
- Cystic dilatation of intravesical ureter (ass with duplex 80%, ectopic 60%)

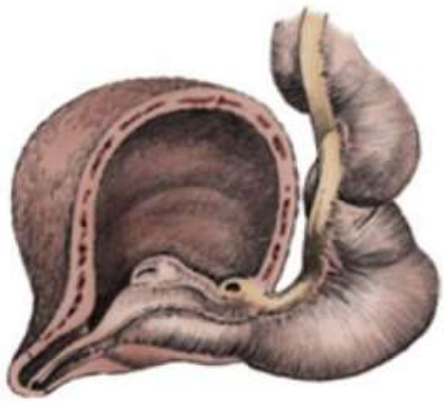
- **Intravesical ureterocele**– above bladder neck (more likely with single system)
  - May prolapse to urethra with voiding



- **Ectopic ureterocele** – portion of ureterocele at bladder neck or urethra permanently (obstruction & risk of damage during surgery)







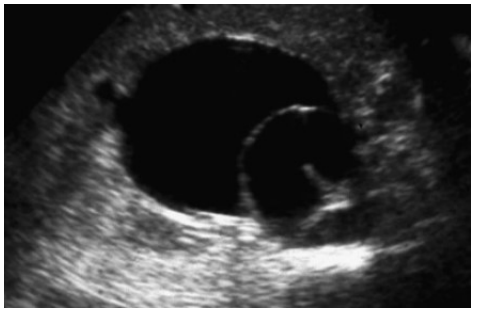
Ectopic ureterocele : can be **sphincterostetoic:** opening in bladder neck**, Ceco uretrocele:** large incompetent orfice in bladder with lumen extending beneath into the urethral submucosa as a long tongue (reach urethra but opening in bladder) **Prolapsed uretrocele:** is visible externally. DDx include other causes of interoital mass (urethral prolapse, RMS, imperforate hymen. Gartner duct cyst)

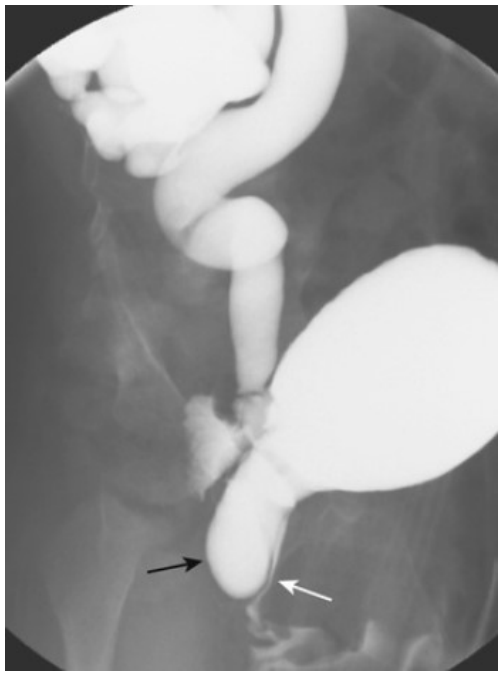
Presentation

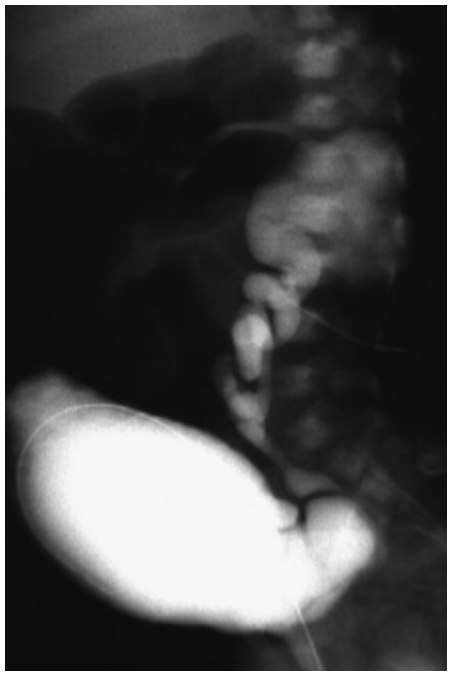
- Purulent discharge (infection)
- BOO
  - prolapse in girls
  - Ectopic urethrocele – rarely obstruct b/c decompress on voiding

Imaging

- **US** - Thin walled cystic dilatation, acute angle, intravesical
    - *A full bladder with effaced ureter may mimic ectopic ureter
- **CUG** – see ureterocele, ipsilateral or contralateral reflux
- **DMSA** – function of upper pole/ectopic moiety
- **MAG3** – replaces DMSA if conservative approach is considered
- **MRI** – complex anatomy, function
- **Cystoscopy** – examine when full and empty b/c may be missed in full bladder if compressible







Ureterocele imaging: urethrocele is seen on early filling of VCUG. Must d/t ectopic ureterocele from intravesical by noting prolapse during voiding. Must also note eversion which indicates a weak trigonal floor that may require surgical repair.

Urgent drainage

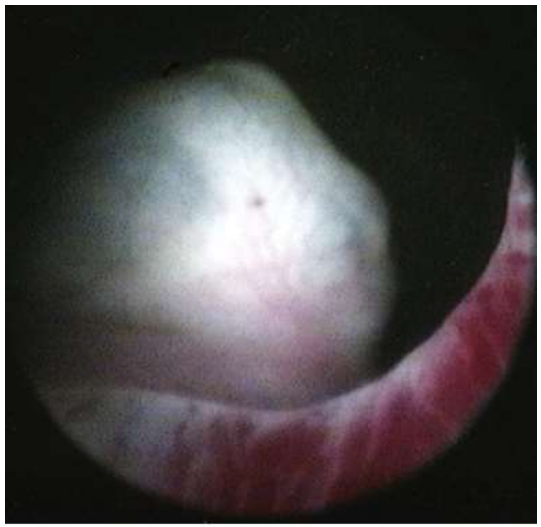
- **Indications**
  - infection (not responding to antibiotic),
  - BOO (prolapsed urethrocele),
  - bilateral ureterocle
- **Options**
  - cystoscopic puncture
  - Ureterostomy or Refluxing implant
  - PCN

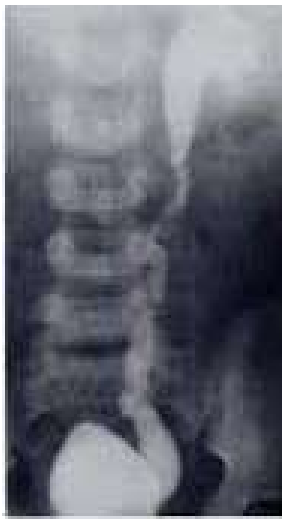
Observation

- **10-15% of prenatally identified** can be observed
    - multicystic dysplasia, with no hydroureter and mild VUR (dysplastic moiety involutes)
- **Single system urethroceles in older children** may be observed
    - If asymptomatic and good renal status
- Antibiotics are needed significant hydroureteronephrosis (assume significant obstruction)

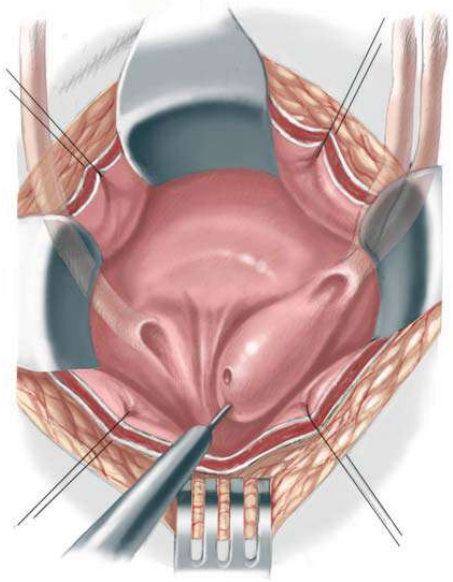
Ureterocele intervention

- **Cystoscopic incision "unroofing" =** definitive in 35% (if no pre-existing VUR)
  - 80-97% successful decompression, initial procedure in neonates
  - Cure in >90% intravesical , 10-50% of ectopic urethrocele, 55% single, 15% duplex
  - Decompressing dilated ureters prepares patient for open surgery if needed.
- **Simplified Upper tract approach =** definitive in 85%
  - Ureteroureterostomy
  - Upper pole heminephroureterectomy - 40% to 50% require second surgery
- **Ureterocele excision with reimplantation** - required in 50% of ectopic ureteroceles
  - Marsupialization - Incomplete excision can result in obstructing flap
  - Complete excision and reconstruction of bladder base – complete excision risks damaging BN
- **Simultaneous upper and bladder surgery** - rarely required (decompressed distal ureter may not cause symptom)

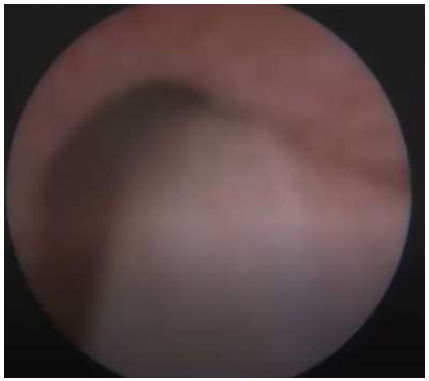
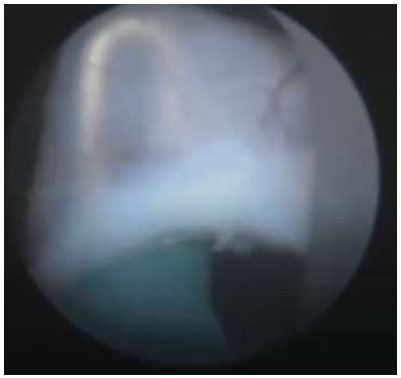


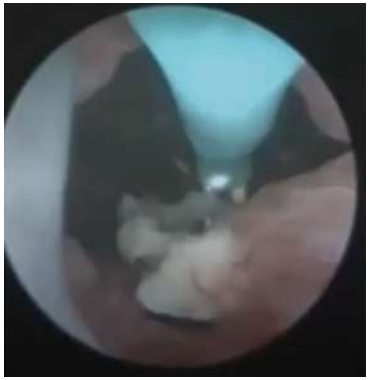


Cystoscopic evaluation: urethrocele seen just inside bladder neck. The appearance of the ureterocele will vary with bladder filling, and it is best to start with little filling and slowly increase bladder volume. The ureterocele will be seen to slowly flatten. Its true limits are best appreciated with limited bladder filling; this will expose the lowest portion, which is probably the best site for incision. Retrograde contrast injection can confirm the presence of ureterocele disproportion (non dilated ureter), and if there are unusual connection to genital ducts.

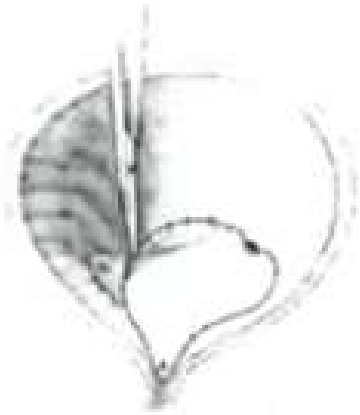


Intravesical ureterocele incision: 2-3 mm low transverse incison (puncture with bugbee). ueterocele should be incised deeply because ureteroceles may be thick walled. Adequacy of ureterocele incision is confirmed either by the escape of a jet of urine from the ureterocele or by viewing the urothelium on the inside of the ureterocele. Making the incision as distally on the ureterocele and as close to the bladder floor as possible lessens the chance of postoperative reflux into the ureterocele. Another technique “watering can technique” (use of laser to make multiple holes) also dec the risk of VUR)

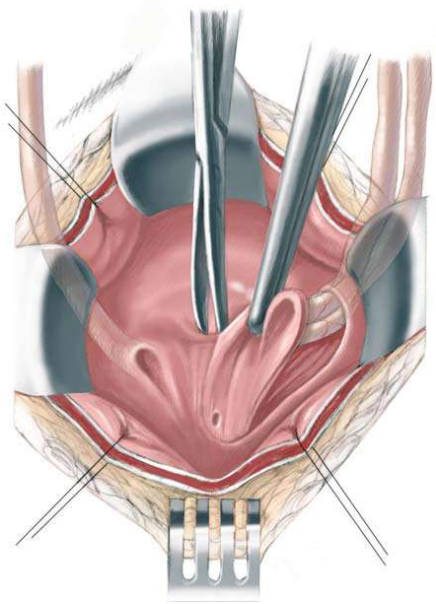



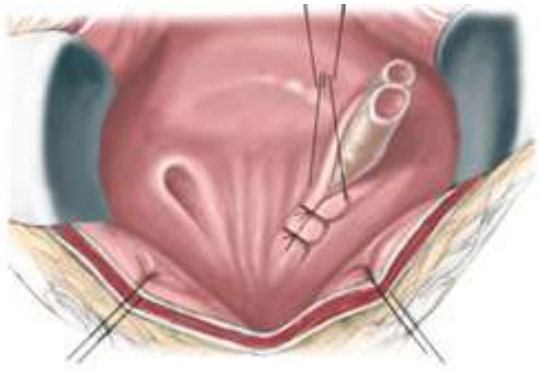



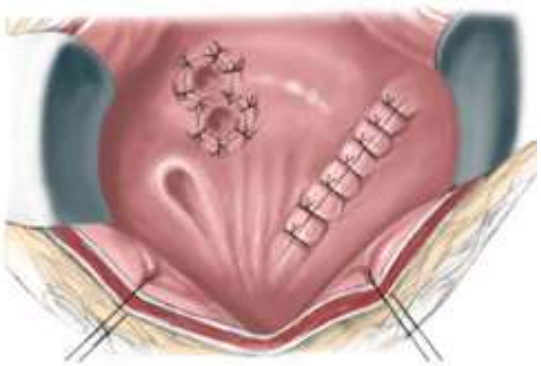
Extravesical ureterocele incision: needs either a longitudinal incision that extends down from the intravesical portion into the urethral portion or two separate punctures, one in the intravesical portion of the ureterocele and one in the urethral portion of the ureterocele. The additional puncture in cecourethrocele is in the uretera to prevent windsock. The additional puncture in sphincteric is in bladder to prevent obstruction during bladder neck closure.

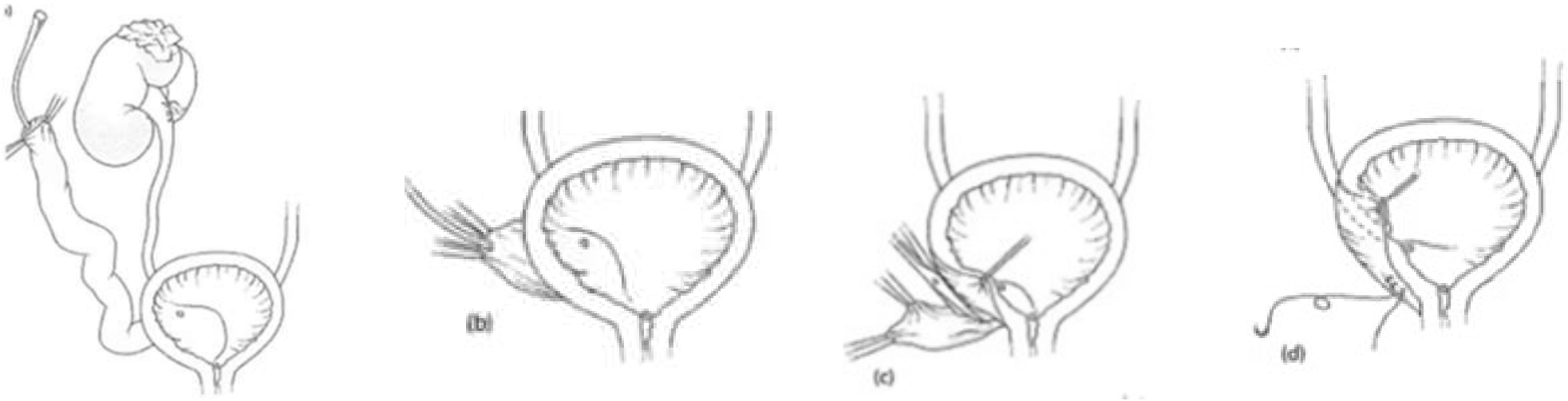

Marsupialization: excise anterior wall of uretherocele (intravesical aspect) and anastomose the mucosal defect (edge is sutured) then followed by reimplantation. No attempt is made to reinforce the back wall. Defect may not be definitively corrected but of eliminates the need to dissect out the ureterocele and avoids injury to the underlying vagina.







Intravesical excision-reconstruction: circumferential incision to excise anterior wall of uretherocele, find hole in bladder, close bladder, then followed by reimplantation. The distal portion of the ureterocele may extend below the bladder neck. Extreme care must be taken in this part of the dissection to avoid injury to the sphincter mechanisms. **Cecoureteroceles present a unique challenge**. Resect by gentle retraction of the ureterocele if it is not very large, closure of the opening with two layers of tissue, or fulguration of the lumen to cause collapse and closure. Careful postop assessment to identify if remnant causes obstructive voiding.



Extravesical excision - reconstruction: if approached upper tract the ureter can be followed down to trigone and reconstructed extravesically as done in diverticulectomy.

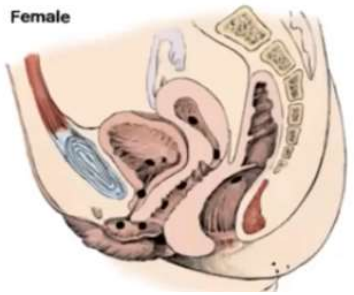
Follow-up

- **VCUG after 6-12mo if bladder level surgery done**
- Dilatation, recurrent infections, incontinence - consider possibility of inadequate trigonal support
  - ballooning of the trigone posterior to the bladder neck and an obstructive process
- Bladder dysfunction – management depend on cause
  - bladder and trigonal reconstruction
  - bladder neck repair,
  - intermittent catheterization,
  - endoscopic injection of bulking agents.

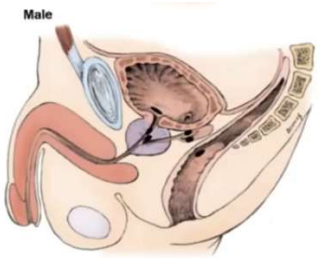
2.2 ....Ectopic ureter

Pathophysiology

- Insertion caudal to trigone (bladder neck or distal),
- girls – below continence mechanism (insert with gartner's duct /mesonephric remnant)
  - bladder neck (1/3),
  - vestibule/urethra (1/3),
  - vagina/uterus (1/3),
  - Rectum



- Boys (15%) – above external sphincter (insert with Wolffian derivatives)
  - posterior urethra,
  - seminal vesicle
  - ejaculatory duct/vas



Epidemiology

- Incidence 1 in 2,000
- 80% associated with duplex system.
    - Single system likely in boys
- Bilateral in 20%
    - if occur in a single system leads to hypoplastic bladder (needs reimplantation, augmentation, BNR)

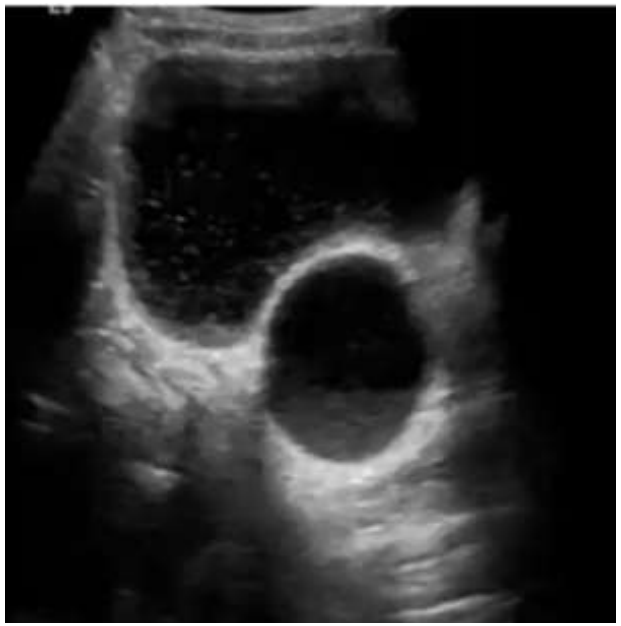
Presentation

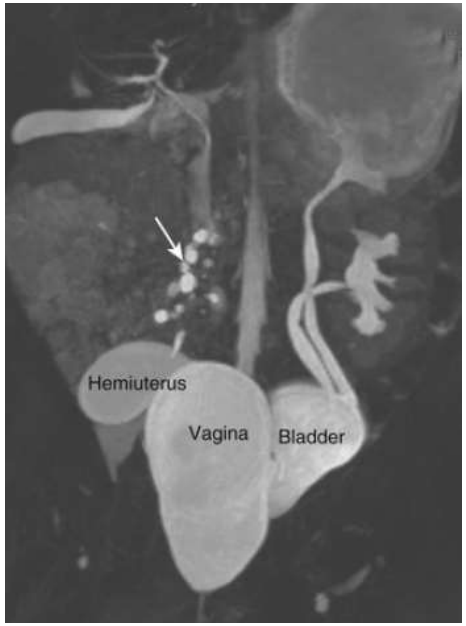
- Girls
  - **Incontinence** with normal voiding pattern
  - Foul smelling vaginal discharge,
  - Sometimes uretric orfice may be seen

- Boys (*infection and pain; no incontinence)*
  - Epididymitis, postvoid dribbling (pooling of urine in the prostatic urethra)
  - May be asymptomatic until onset of sexual activity
  - prostatitis, infected seminal vesical cyst
  - Zinner syndrome – ectopic ureter, ipsilateral renal agenesis, seminal vesicle cyst

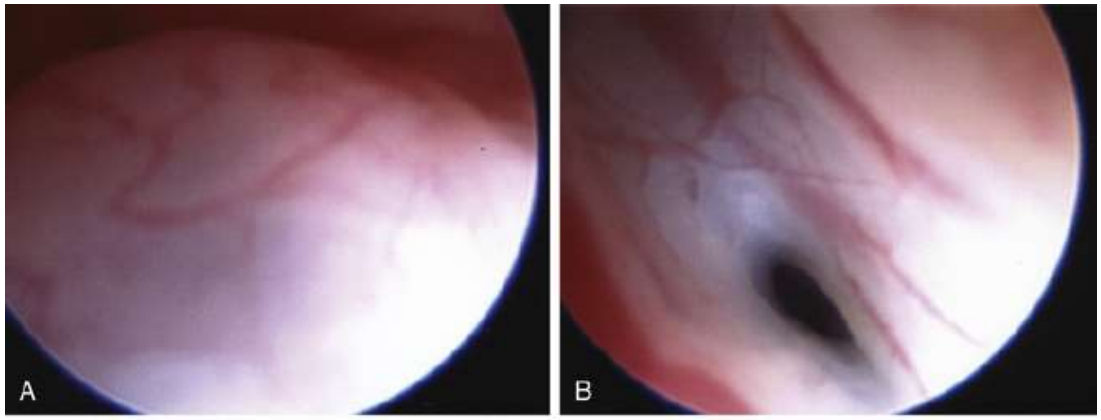
Imaging

- **CUG** – to exclude reflux
- **US** – Thick walled ,obtuse angle, extravesical component
- **DMSA** – can localize small upper pole/ectopic moiety
- **MAG3**– value is limited as dilated ectopic ureter is rarely suitable for observation
- **IVP** – functional information (qualitative) with anatomy
- **MRU** – most precise for making diagnosis
- *Diagnosis confirmed with*
  - **Physical exam** - filling bladder with dyes to d/t clear urine from ectopic
  - **Cystoscopy/vaginoscopy**





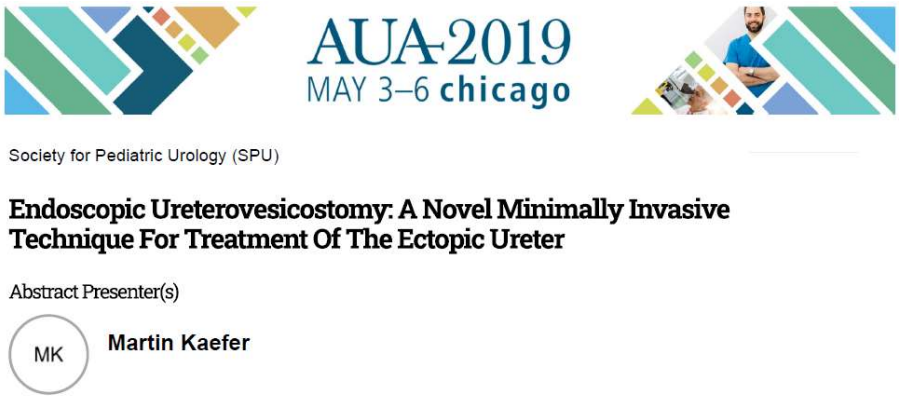
Imaging of ectopic ureters: large ectopic ureter pushing posterior wall of bladder may confuse on US (pseudoureterocele). Ectopic ureter to bladder neck/sphincter may have concomitant VUR. MRI best to delineate anatomy



Cystoscopic evaluation: ectopic ureters that appear to have a very thin layer of tissue separating them from the urethra may confuse with ureterocele. The anatomic connections of the ectopic ureter may be demonstrated using retrograde contrast studies. Ureteral orifice may be very difficult to find endoscopically. If it is in the bladder neck, it is often patulous but do not always reflux. These may be associated with defects in the bladder neck and incontinence. Based on endoscopic appearance, more extensive removal of patulous segments with reconstruction of the bladder neck may be considered.

Management options

- **Temporary end ureterostomy**
  - Infection not responding to antibiotics
  - If the degree of function is ambiguous (permit later assessment)
- **Ureteroureterostomy** (proximal/distal) – concern size discrepancy
- **nephroureterectomy**–usually successful as definitive b/c the residual stump is not problematic.
  - Resection of ureteral stump - to avoid BN injury, keep close to ureter or use transvesical/vaginal approach
- **Common sheath reimplantation with BNR** - fix lower pole VUR at the same time,



Novel techniques: **Simple laparoscopic ligation** of the ectopic ureter has been proposed. Long term results are pending. Caution in chronically infected systems. More recently **endoscopic ureterovesicostomy** has been described in a few patients). Ureter is cannulated, balloon inserted and area of bulge of balloon is punctured under cystoscopy to create fistula. Make sure there is no proximal kink otherwise won't work. Fistula may also narrow and stop draining.

3 Refluxing & obstructed megaureter

Refluxing + Obstructed megaureter

- **Primary**
  - Kink superior to reflux
  - Ectopic ureter to bladder neck or sphincter
- **Secondary**

4 Non-refluxing, Non-obstructed megaureter

Non-Refluxing Non-obstructed megaureter

Diagnosis of exclusion

- **Primary**
  - Idiopathic
  - Physiologically insignificant adynamic segment
- **Secondary**
  - Polyuria (SIADH)- volume of urine exceeds emptying capacity
  - Infection- bacterial endotoxin can cause diltation that resolves after treatment of UTI
  - Postoperative residual diltation

References

- **Hollcomb and Aschcraft pediatric surgery** 7th E (2020)
- **Campbell Walsh Urology** 12th E (2020)
- **Springer Pediatric Surgery Atlas** (2006)