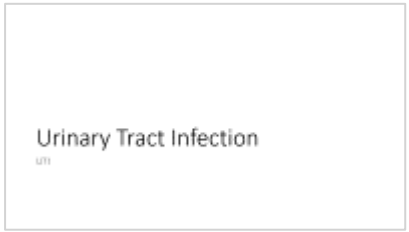
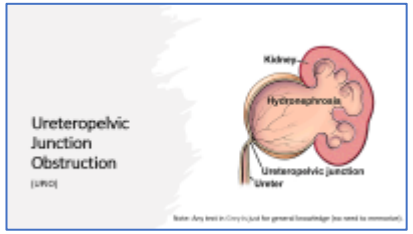
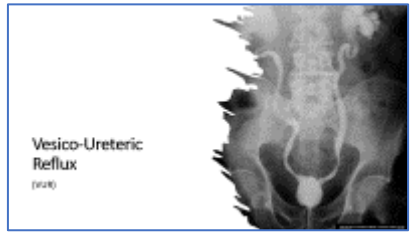
Principles of Pediatric Urology

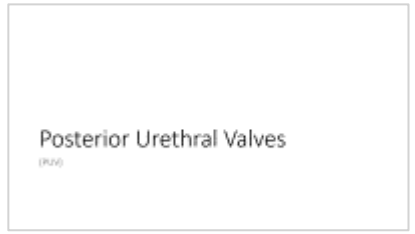
By Raed Al-Taher, M.D.

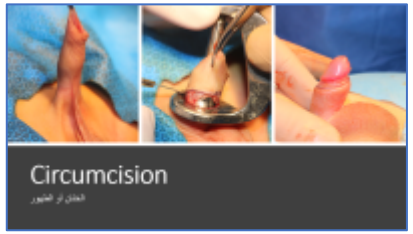
Topics in BLUE are the required ones

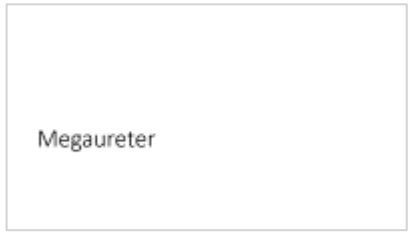


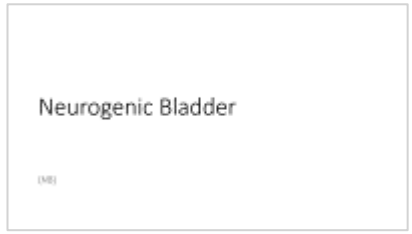


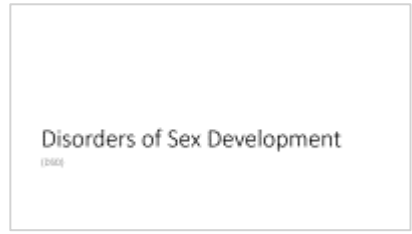












*Updated: Dec. 2025

Urinary Tract Infection

UTI

Urinary Tract Infection (UTI)

- The urinary tract is a common portal of infection in childhood.

- Diagnosis:
  - Suggestive symptoms
  - Urine microscopy
  - Urine quantitative culture of a properly collected sample

Urinary Tract Infection (UTI)

- F >> M (except in the first year of life when M>F).
- 8% of girls and 1–2% of all boys will get UTI during their childhood.

Urinary Tract Infection (UTI)

- Microbiology
  - E. coli (most common - 80%)
  - Other pathogens include:
    - Gram −ve (as Citrobacter, Proteus, Pseudomonas, and Serratia)
    - Gram +ve (as Staphylococcus spp., Enterococcus spp., and Hemophilus spp.)

- Route of infection:
  - Ascending (via the urethra)

    or
  - Hematogenous (more common in infants)

Risk Factors

- Antenatally diagnosed renal anomalies
- Family history of renal disease or VUR
- Bladder outflow obstruction (as urethral valves)
- Neurogenic bladder or dysfunctional voiding
- Urolithiasis
- Constipation
- Uncircumcised males

Clinical Features

- Symptoms (upon age of the child):
  - Older children and adolescents: fever, dysuria, urgency, frequency, and urinary incontinence.
  - Infants: poor feeding, failure to thrive, temperature instability, jaundice, and vomiting.

- PEx:
  - Renal angle, abdominal or suprapubic tenderness
  - Abdominal mass, or a palpable bladder
  - Dribbling, poor stream, or straining to void
  - Hypertension (suggests hydronephrosis or renal parenchyma disease)
  - Dehydration and sepsis (in severe cases)

Urinary Tract Infection (UTI)

- In general, UTI must be suspected and urine must be tested for:
  - any child with a fever above 38°C + no other cause is evident

Investigations

- Urinalysis
  - Clean-catch sample (adhesive urobags (convenient in infants) are not ideal because of contamination. In the male, retract foreskin to expose the meatus)
  - Catheter specimen or a suprapubic sample (under ultrasonic guidance).
- Urine dipstick:
  - for leukocyte esterase or nitrite positivity (recommended mainly for >3-year-old).
- Urine microscopy:
  - Mainly for <3-year-old, for WBCs (>5 per high-power field), RBCs, bacteria, casts, and skin contamination.
  - Bacteriuria (preferably with pyuria) is considered positive.
- Urine culture:
  - Clean-catch sample:
    - Bacterial counts of >105 are diagnostic of UTI.
    - Bacterial counts 103–5 □ repeat test.
    - Bacterial count <103 □ not significant.
  - Suprapubic sample: any organisms are diagnostic.
  - Multiple organisms:
    - Contamination with perineal flora OR Urinary tract abnormalities

Imaging

- Traditional approach
  - Birth to 1 year: US + DMSA scan + VCUG
  - 1–5 years: US + DMSA only
  - Five years: US only

- In UK, the National Institute of Clinical Excellence (NICE) has suggested modification to:
  - Birth to 6 months: US only. (DMSA and VCUG if there is abnormality on US, or severe, atypical, or recurrent UTI).
  - 6 months to 3 years: US and DMSA (+/- VCUG if abnormalities on US, family history of VUR, or a poor urinary stream).
  - 3 years: US and DMSA (no need of VCUG in most cases)

Imaging

- Renal and bladder US:
  - for size and shape of kidneys
  - presence or absence of ureter
  - urolithiasis, hydronephrosis, hydroureter, and ureterocele
  - bladder emptying (in toilet-trained children, for pre- & post-void volume measurement)
- VCUG:
  - to diagnose posterior urethral valves or VUR
  - Bladder anatomy, bladder diverticulae or features of neurogenic bladder
  - In girls, direct isotope cystogram (DIC) is preferred
- Radio-isotope renal studies:
  - Static scan (DMSA):
    - investigation of choice
    - shows the kidney outline and detects renal scars
  - Dynamic scan (MAG-3/DTPA):
    - preferred (to a DMSA scan) if there is hydronephrosis or if the ureters are dilated
    - can pick up VUR in the indirect cystogram phase in toilet-trained children

Management

- Parenteral antibiotics (e.g., ceftriaxone or cefotaxime):
  - All infants (<3 month) OR older patients who are sick
  - For ~10 days (if acute pyelonephritis is present)
- Oral antibiotic (e.g., cephalexin, co-amoxiclav, trimethoprim, or nitrofurantoin):
  - started once the patient is afebrile OR lower UTI/ cystitis in patients (>3 months) for 3 days only
  - provided patients are not sick AND good response within 48 h
  - up to 1/3 of community acquired UTIs are nowadays resistant to trimethoprim
- Routine antibiotic prophylaxis is not recommended (unless recurrent infections).
- Treat constipation (if present)
  - constipation and dysfunctional voiding are more common causes of recurrent UTI than diseases like VUR
- Asymptomatic bacteriuria:
  - does not require treatment
  - needs careful follow-up (if symptoms appear, urine tested again and treatment started)

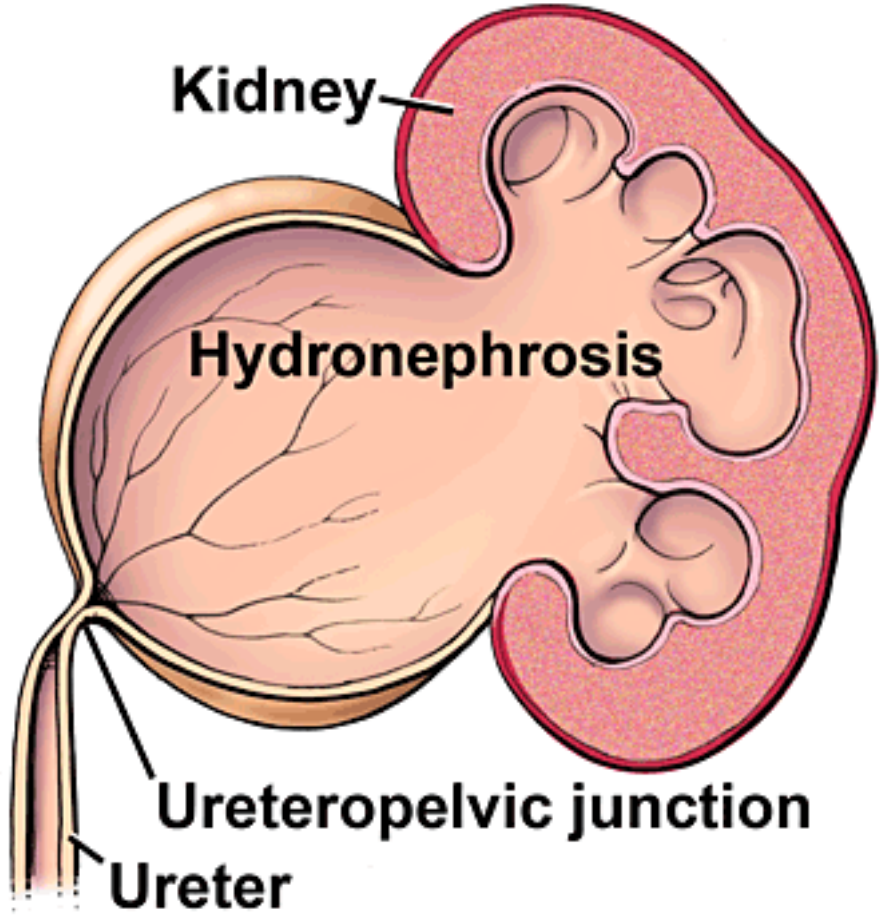
Outcome

- Most UTIs are uncomplicated and respond to outpatient antibiotics.

- Complicated UTI needs prolonged follow-up to prevent long-term sequelae, like:
  - Renal parenchymal scarring
  - Hypertension
  - Decreased renal function
  - Renal failure

Ureteropelvic Junction Obstruction

(UPJO)



Note: Any text in **Grey** is just for general knowledge (no need to memorize).

Ureteropelvic Junction Obstruction (UPJO)

- M:F 2:1 | Lt:Rt 3:2 | bilateral 10–40%
- Most present as **HYDRONEPHROSIS** (HN) that is detected by **antenatal US**

Antenatal US

- Society of Fetal Urology (SFU)
  - Grade 0 – normal kidney
  - Grade 1 – minimal pelvic dilation
  - Grade 2 – greater pelvic dilation without caliectasis
  - Grade 3 – caliectasis without cortical thinning
  - Grade 4 – HN with cortical thinning

Etiology

- **Intrinsic**
  - **Intrinsic narrowing (m.c.)**
  - Mucosal valves, polyps, and true ureteric strictures (rarely)

- **Extrinsic**
  - Aberrant renal vessel (~30% of UPJ, an artery directly enters the lower pole of the kidney).
  - Kinking as a result of severe vesicoureteral reflux (VUR).

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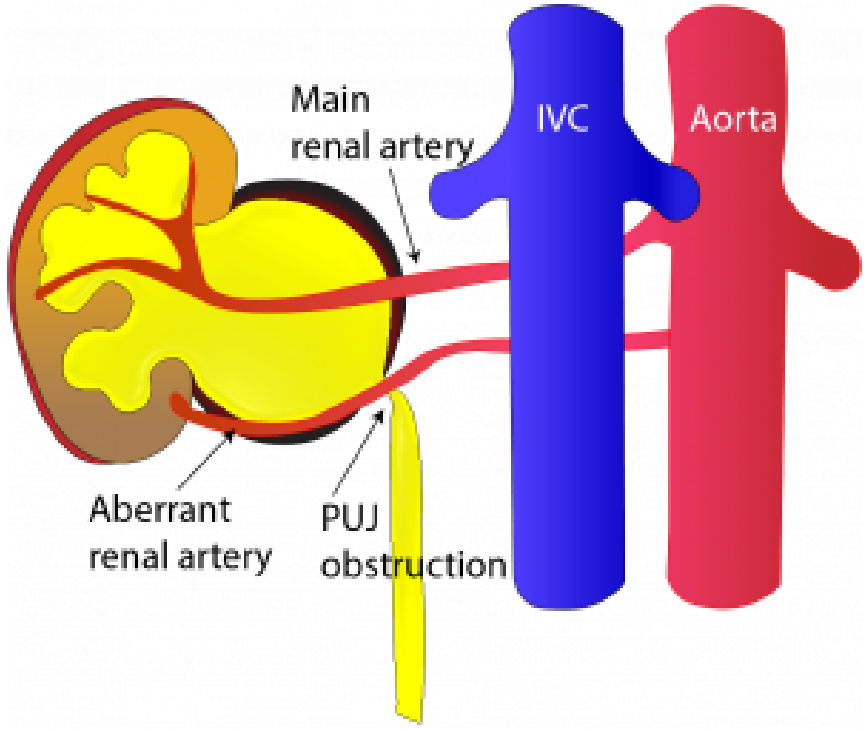

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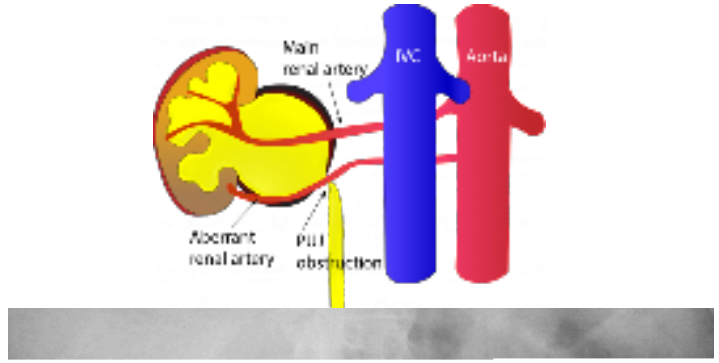

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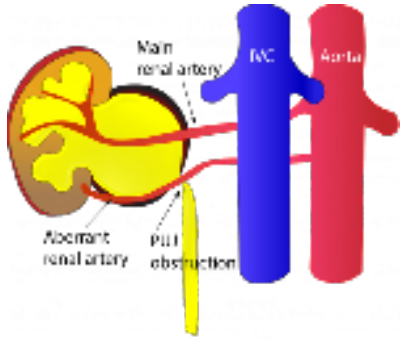

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Clinical Features

- Most are **asymptomatic** (detected via prenatal screening US)

- When symptomatic:
  - Flank or abdominal **pain** (~50%)
  - Palpable flank **mass** (~50%)
  - Hematuria
  - Recurrent UTIs (~30%)

Investigations

1 Postnatal US

- Assesses kidney **anatomy** & **AP diameter** of renal pelvis

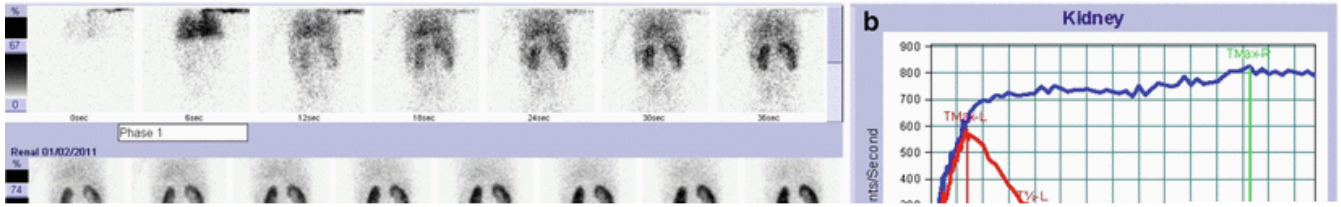
2 Renal radioisotope Scan

- **MAG3** is the scan of choice, & shows:
  - Differential renal function
    - normally 50%:50%
    - acceptable down to 40%
    - needs intervention when <40%
  - Pelvic drainage curve
    - shows pelvic emptying time (t½) after administration of furosemide
    - normally t½ <20 min
    - t½ >20 min = significant obstruction needs intervention

3 MCUG

- To rule out whether HN is due to **VUR**

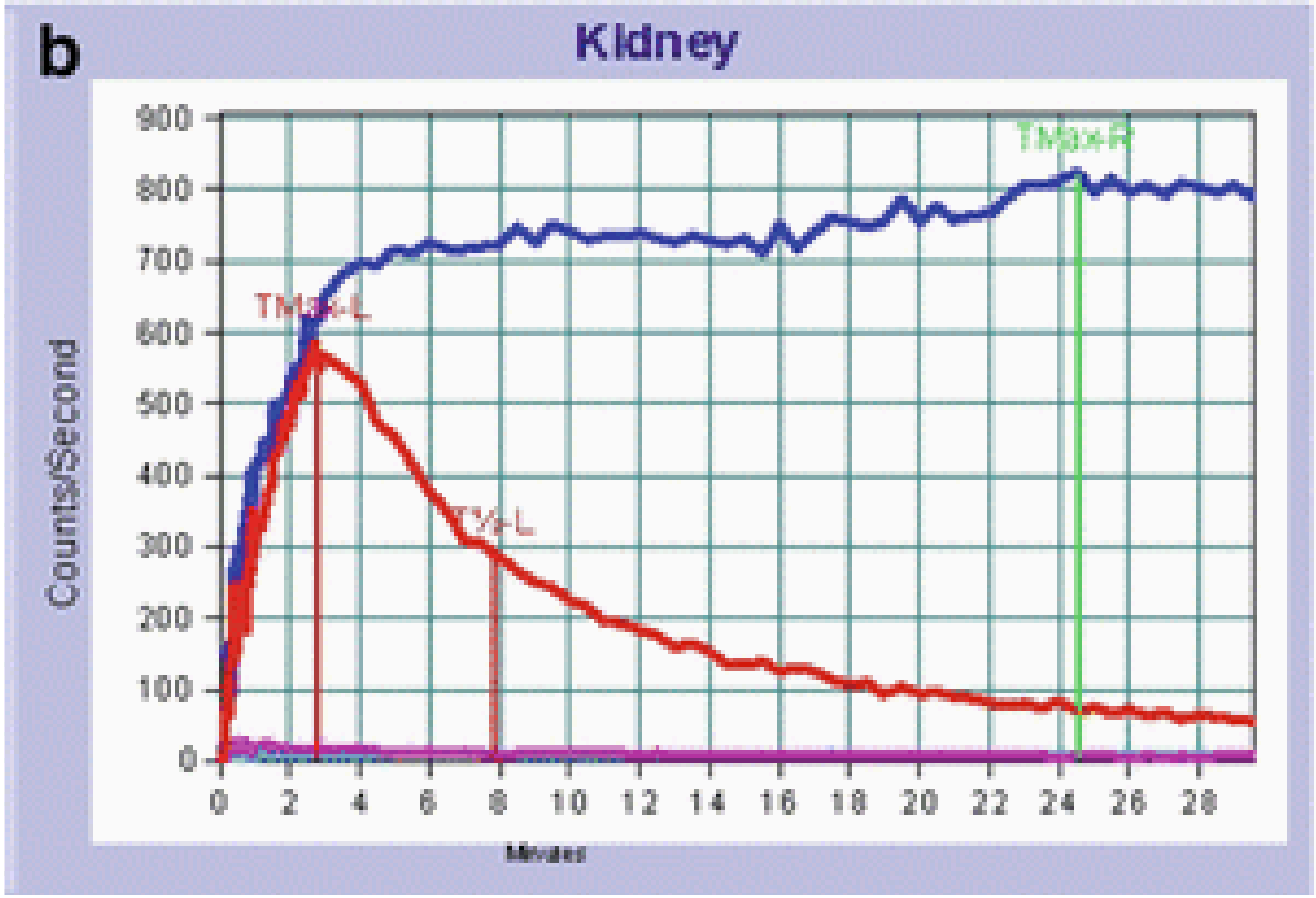
t½ = the time needed to drain half the urine amount from renal pelvis to the ureter



Obstructed kidney

Normal kidney

MAG-3 Dynamic Renal Nuclear Scan



Obstructed kidney

Normal kidney

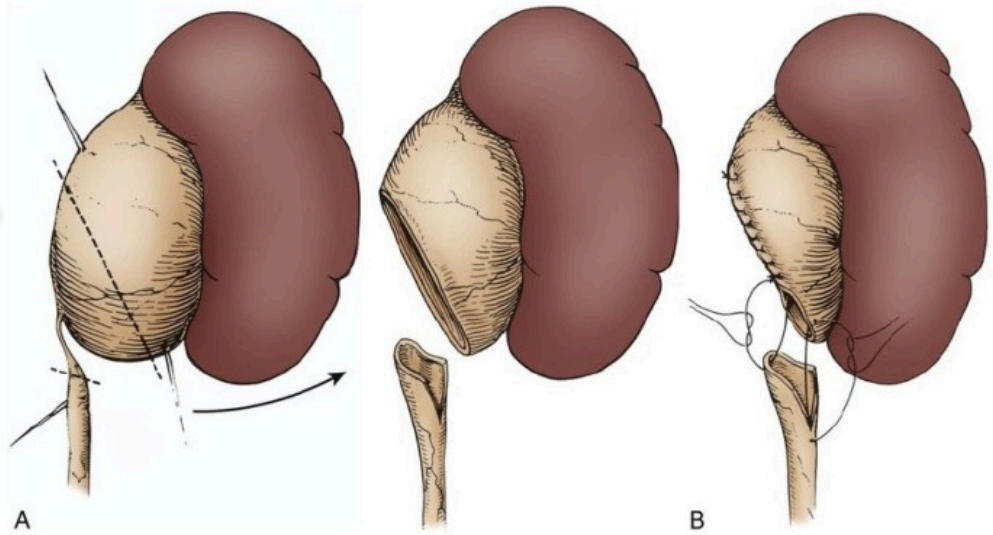
MAG-3 Dynamic Renal Nuclear Scan

Treatment

- **Antenatally detected HN**
  - Conservative management (follow up)
  - Surgery (needed in <50% of cases) when **any or all** of the following:
    - Functional deterioration (<40%)
    - $T_{1/2}$ > 20 minutes
    - Symptomatic

- **Surgery**
  - Open **OR** laparoscopic **pyeloplasty:**
    - Excision of the narrowed segment
    - Anastomosing ureter to the most dependent portion of the renal pelvis
    - Excision of redundant renal pelvis

- Endourological pyeloplasty
  - Use of balloon dilatations, percutaneous antegrade endopyelotomy, and retrograde ureteroscopic endopyelotomy.



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laparoscopic
pyeloplasty

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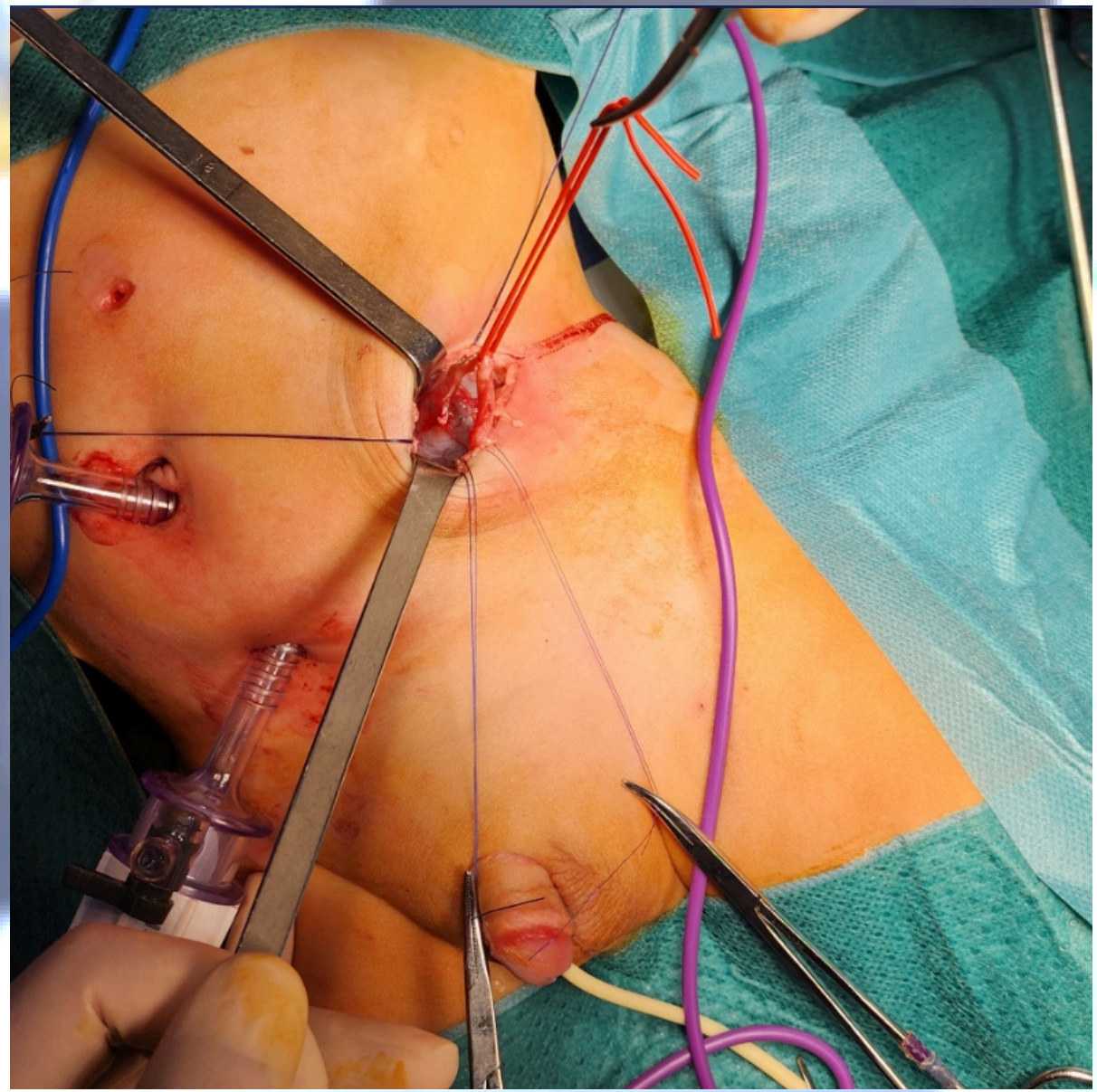
laparoscopic
pyeloplasty



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laparoscopic
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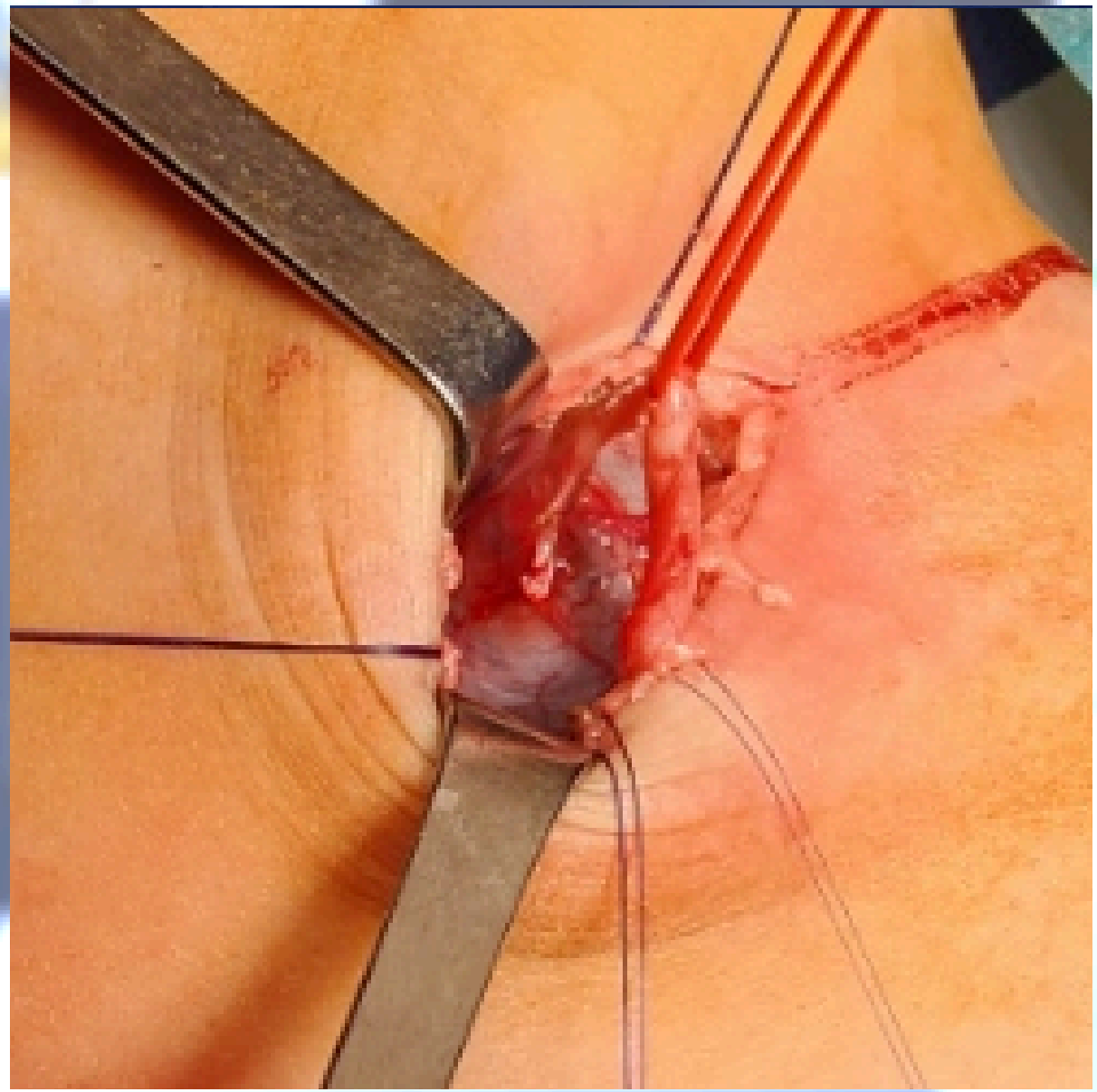

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Megaureter

Megaureter is classified as:

- Obstructed (M:F 4:1 | L > R | Bilateral 20%)
  - **Primary**: adynamic juxtavesical segment of the ureter.
  - **Secondary**: ↑ vesical pressures (due to PUV or a neurogenic bladder (NB)).

- Refluxing
  - **Primary**: severe VUR.
  - **Syndromic**: Megacystitis megaureter syndrome.
  - **Secondary**: ↑ vesical pressures (due to PUV or a neurogenic bladder (NB)).

- Obstructed/refluxing

- Nonobstructed/nonrefluxing

Clinical Features

- Increasing HUN
- Decrease in renal function
- UTI
- Recurrent flank pain

Investigations

- US HUN
- MAG3 scan degree of obstruction and differential renal function
- IVU to see anatomy (uncommonly used)
- VCUG r/o VUR

Management

- Ureteral reimplantation: when associated with severe VUR or obstruction.
  - Mobilize the megaureter via an intravesical, extravesical, or combined.
  - Reduce ureteral caliber.
  - Antireflux reimplant.

- Nephroureterectomy: when the function of the kidney drained by a megaureter is severely impaired.

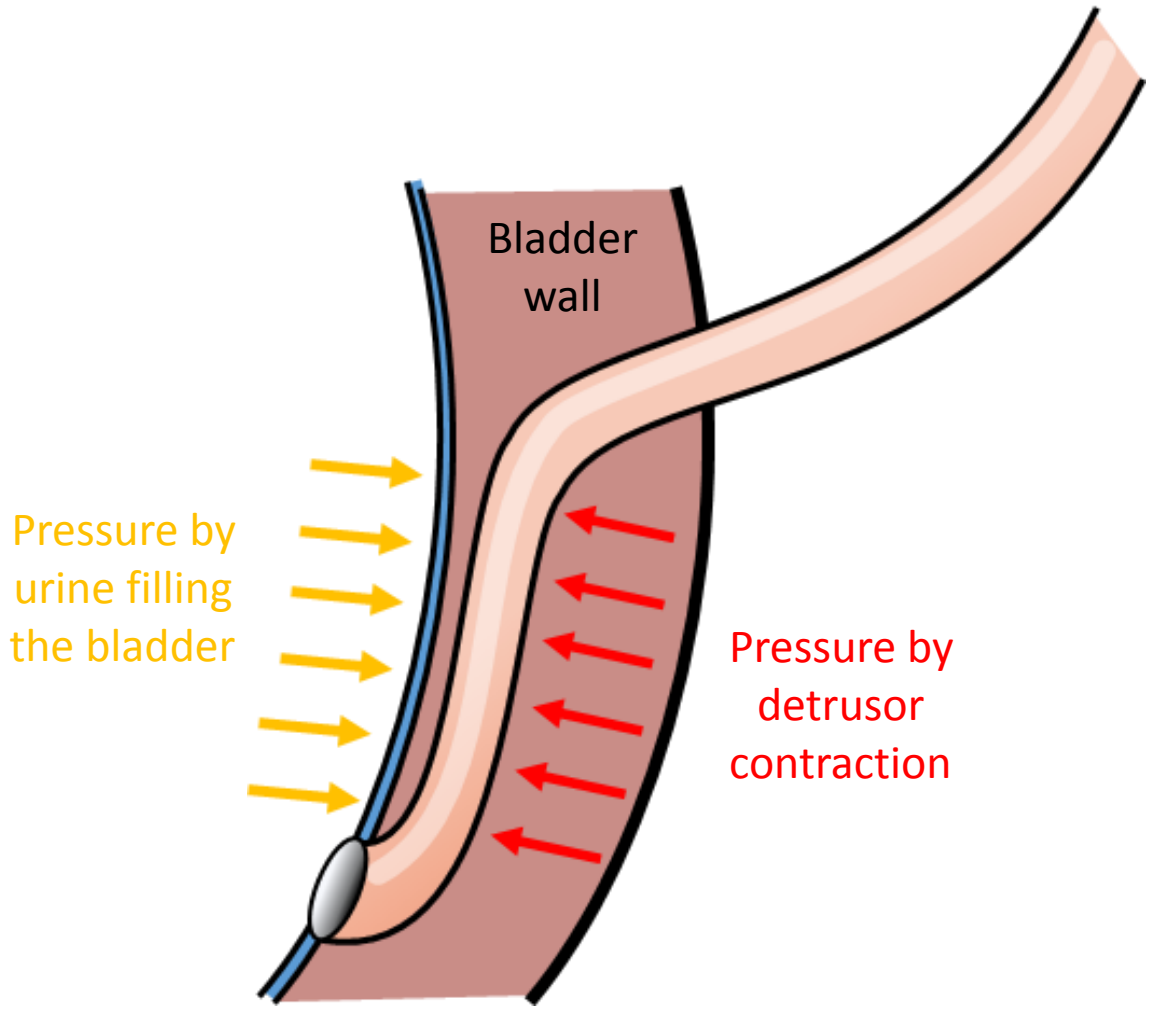
Vesico-Ureteric Reflux

(VUR)

Vesico-Ureteric Reflux (VUR)

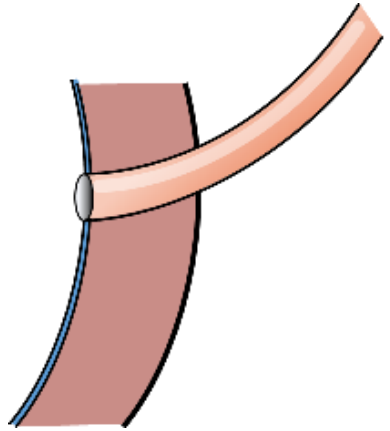
- Female predominant
- Peak incidence at 3 years
- Familial in 2–4%

Normal ureteral submucosal tunnel



tunnel length : ureteral diameter ⍰ should be about 5:1 to prevent VUR

Pathology



Primary VUR:

- Due to a **short ureteral submucosal tunnel** in the bladder wall

Secondary VUR:

- Distal obstruction due to either:
  - Posterior urethral valve (PUV) in males
  - Neurogenic bladder (NB)
  - Anterior urethral valves
  - Ureteroceles
  - Bladder diverticula
  - Ectopic ureters associated with duplex system

Clinical Features

- Symptoms of UTI | **recurrent UTIs (m.c)**
- **Renal scarring** (due to previous pyelonephritis – upper UTI)
- Renal dysfunction
- Hypertension
- Reduced somatic growth

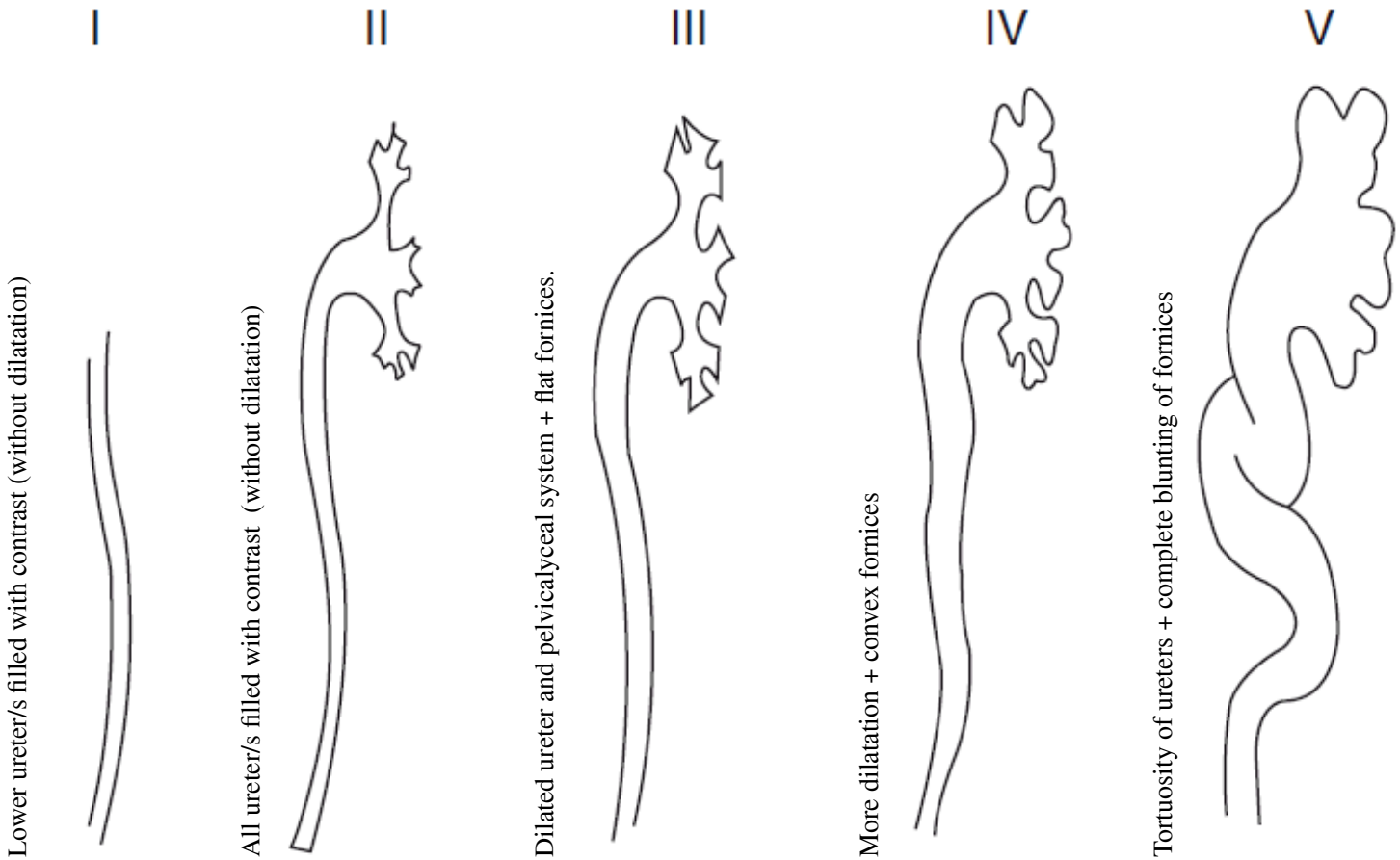
Investigations

- Urine analysis → r/o infection

1. **US** → HUN (hydro-uretero-nephrosis)
2. **DMSA nuclear scan** → renal scars and differential renal function
3. **MCUG** → confirm Dx & grade of VUR

- Direct isotope cystography (DIC) → for follow-up scans

MCUG | grading

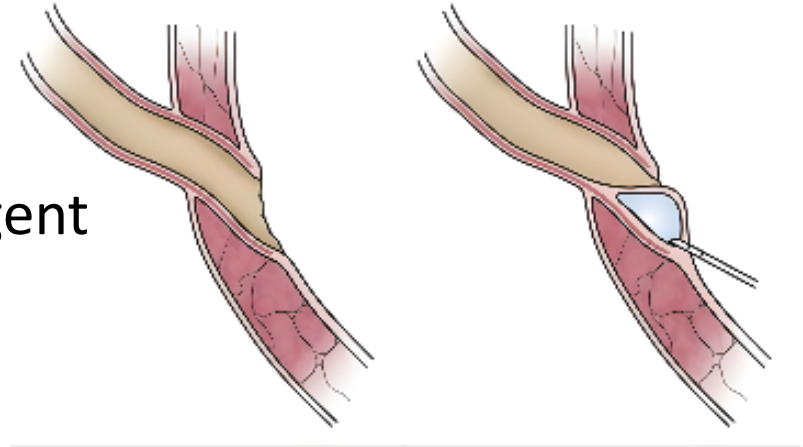


Treatment

Low-grade reflux (I, II, & III):

- More likely to resolve spontaneously with age
- Antibiotic prophylaxis

- +/- Subureteral submucosal injection of bulking agent
  - Results better for lower grades of reflux (>80% success)
  - Less successful in children with neurogenic bladder (NB)

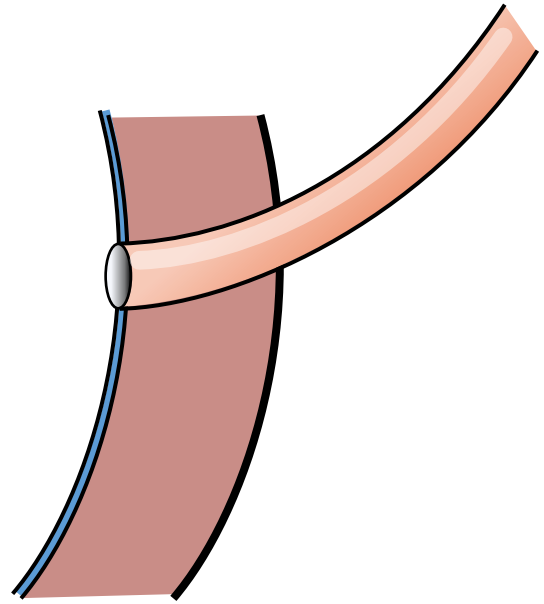


Surgical treatment

- **Indications:**
  - Failure of chemoprophylaxis and/or submucosal injection therapy
  - Deterioration of renal function &/or appearance of new scars
  - Secondary VUR (due to ureterocele, duplex ureter, PUV, or neurogenic bladder)
  - Higher grades VUR (IV, V)
  - Hypertension
  - Single kidney with higher grade of VUR
  - Decrease in renal growth or somatic growth

Surgical treatment

- **Reimplantation of ureters**
  - Transtrigonal ureteric (Cohen) reimplantation [most common]
  - Intravesical technique (Leadbetter–Politano)
  - Extravesical detrusorraphy technique (Lich and Gregoir)
  - ± Ureteric tapering or plication



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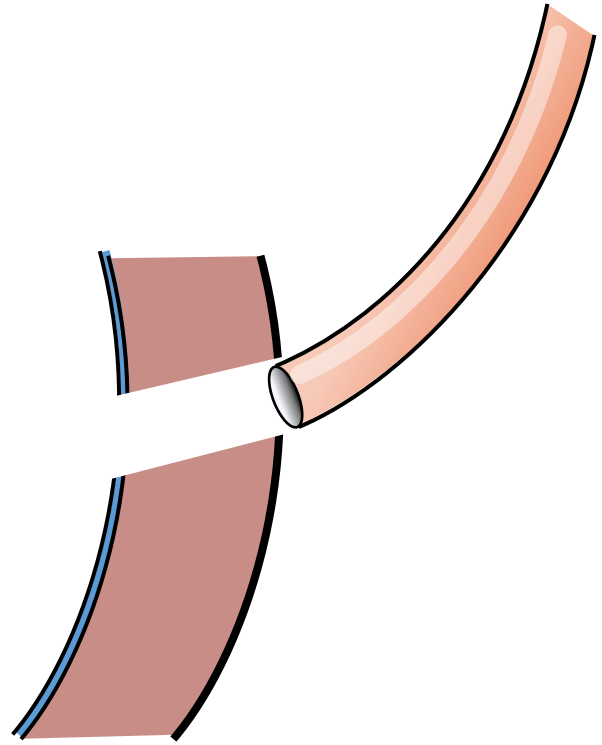

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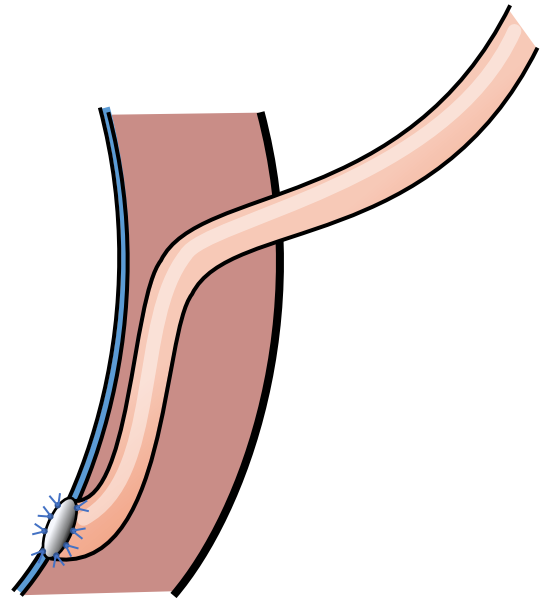


Surgical treatment

Postoperative Management

- Antibiotics for ~3 months
- Follow-up VCUG or DIC

Complications

- Persistent reflux
- Ureteric obstruction (devascularisation, kinking or torsion)
- Intravesical calculi
- Injury to the bowel, fallopian tubes and/or vas deferens

Surgical treatment | Outcome

- Success rate up to 98%
- Corrects VUR.. however..
  - does not reverse kidney scarring nor parenchymal damage

Neurogenic Bladder

(NB)

Neurogenic Bladder (NB)

Ideal bladder function:

- Bladder should have the ability to fill to capacity while maintaining low pressures.
- Sensation to void when full bladder and be under voluntary control.
- Bladder should be empty at the end of voiding without residue.

Neurogenic Bladder (NB)

Neurology

- Parasympathetic (predominant)
  - S2–S4 spinal segments, via pelvic nerves, to the detrusor muscle.
- Sympathetic
  - T9–L1 spinal segments, via sympathetic chain and hypogastric plexus, to the bladder neck.
- Somatic innervation
  - S2–S4 segments, via pudendal nerve, to the external sphincter.

Neurogenic Bladder (NB)

Etiology

Congenital	Acquired
Myleomeningocele	Trauma
Spina bifida occulta (including tethered cord, lipoma of cord)	Tumors
Sacral agenesis	Infarction
	Transverse myelitis

Neurogenic Bladder (NB)

Etiology

- Almost always due to spinal cord anomalies (congenital and acquired) (myelomeningocele most common).

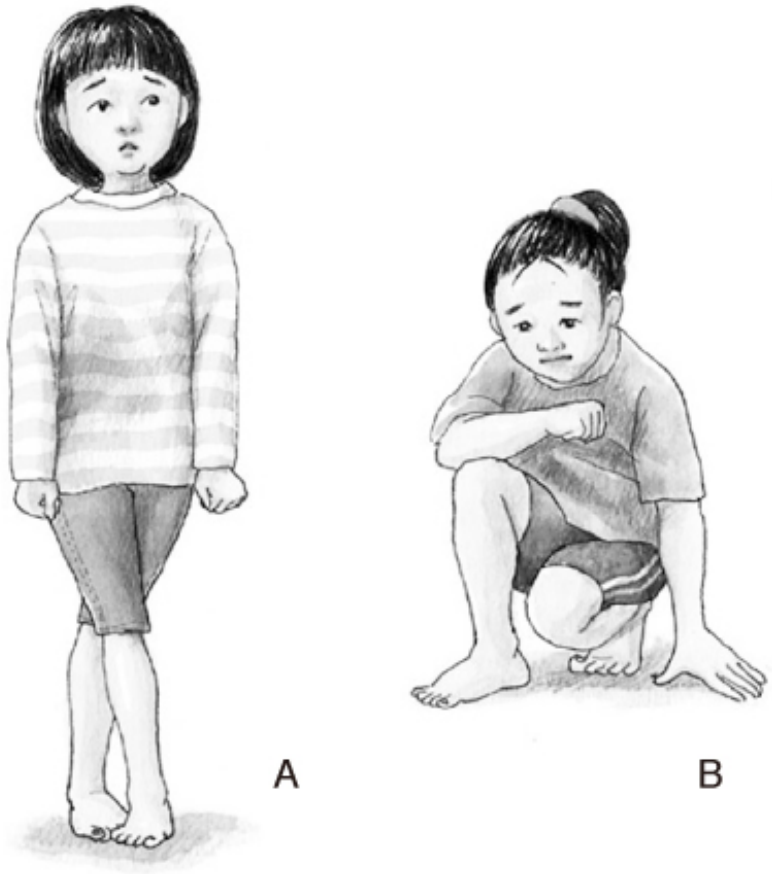
- Nonneurogenic neuropathic bladder (Hinman–Allen syndrome) psychological overlay in addition to bladder dysfunction.

Neurogenic Bladder (NB)

Clinical Features

- Urge incontinence (overactive detrusor)
- Stress incontinence (underactive sphincter)
- Vincent's curtsy postures to inhibit voiding

(cross-legged "A" or squat with heel pressing perineum "B")



- Assess the back for evidence of:
  - spina bifida occulta (hairy patch or lumbosacral lipoma)
  - sacral agenesis
- Full neurological assessment to elicit the integrity of segments S2–4

Neurogenic Bladder (NB)

Concept

- “Unsafe” bladder inability to empty distended bladder
- “Safe” bladder leaks easily under pressure or usually empty on examination

Neurogenic Bladder (NB)

Investigations

- US:
  - upper tract dilatation
  - postvoiding bladder residual volume

- Cystogram:
  - r/o VUR, bladder trabeculation or diverticular formation

- Urodynamic Studies [next slide]

Neurogenic Bladder (NB)

Basic Video Urodynamics

- Normal bladder capacity (mL)
  - Infants capacity = weight (in kg) × 7
  - 1–12 years capacity = age (in years) × 30 + 30

- Detrusor hyperreflexia (significant if pressure is >30–40 cm H_2O)

- Compliance (at expected bladder capacity, detrusor pressure should be <30–40 cm H_2O)

- Sphincteric incompetence (opens abnormally at low pressures)

- Leak-point pressure (at which external sphincter opens)
  - <40 cm H_2O □ “safe” bladder
  - >40 cm H_2O □ potential for upper tracts deterioration

- Detrusor sphincter dyssynergia (detrusor contraction and external sphincter non-relaxation □ ↑ intravesical pressures)

Neurogenic Bladder (NB)

Patterns of Neurogenic Bladder

- Contractile bladder (usually the result of suprasacral cord lesions)
  - Innervation of the detrusor and external urethral sphincter are intact
  - Intravesical pressures are usually high (ultimately lead to secondary upper renal tract complications)

- Acontractile bladder
  - Innervation of both detrusor and external sphincter is destroyed
  - Voiding occurs either by overflow or by raising intraabdominal pressure

- Intermediate bladder dysfunction
  - Combination of anomalies: various degrees of detrusor noncompliance, detrusor hyperreflexia, and sphincteric incontinence

Neurogenic Bladder (NB)

Management

- Natural history: progressive deterioration by the age of 3 years (in up to 60% of all children)
- Early institution of CIC can prevent both renal damage and secondary bladder wall changes improving long-term outcomes
- Both detrusor hyperreflexia and detrusor noncompliance can be treated:
  - medically with anticholinergics (e.g., oxybutanin and tolerodine)
  - +/- surgically (to increase the capacity and outlet resistance)

Neurogenic Bladder (NB)

Clean intermittent catheterization (CIC)

+

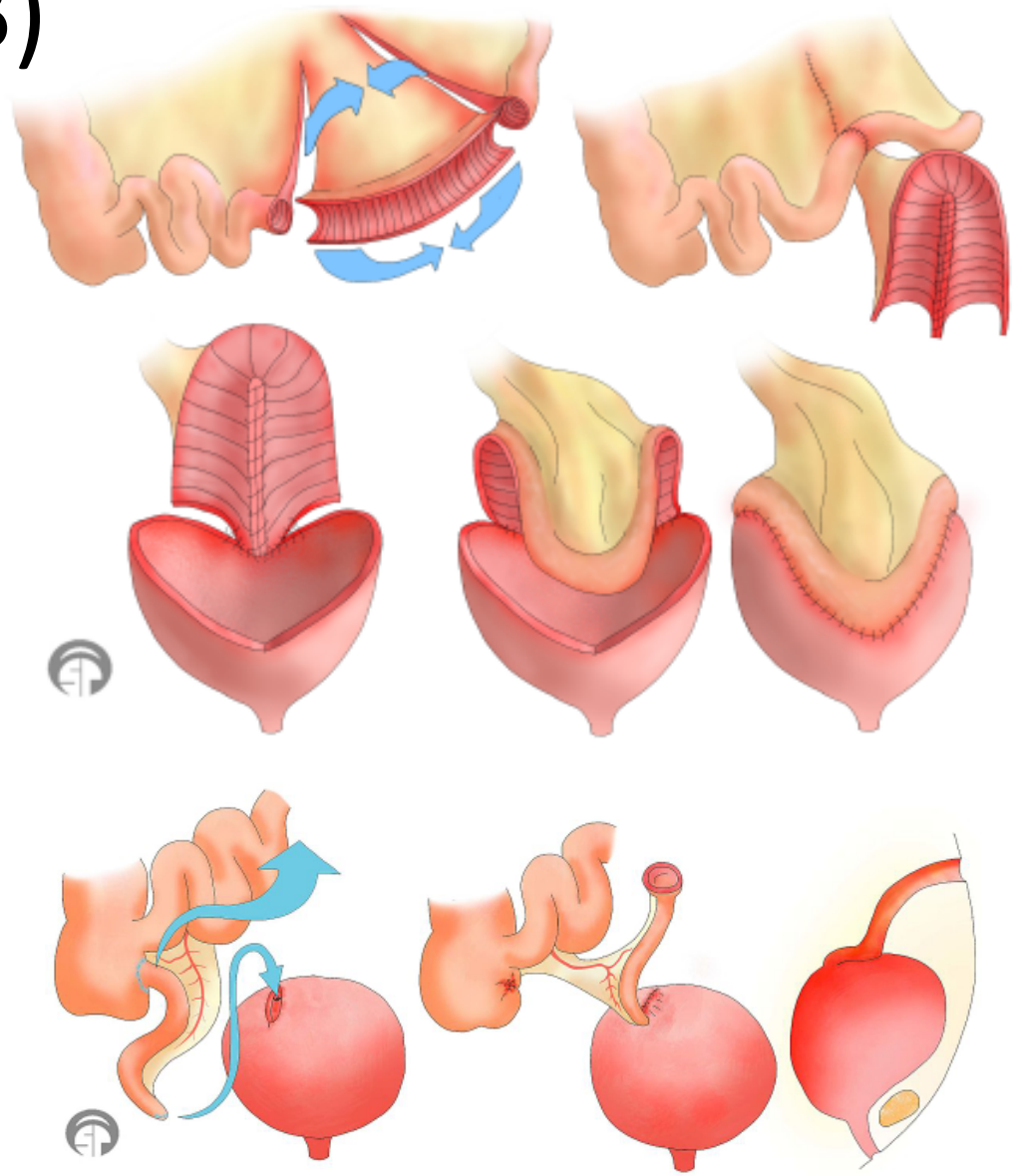
Anticholinergic (oral or intravesical)

is the standard therapy
for neurogenic bladder

Neurogenic Bladder (NB)

Management (surgical)

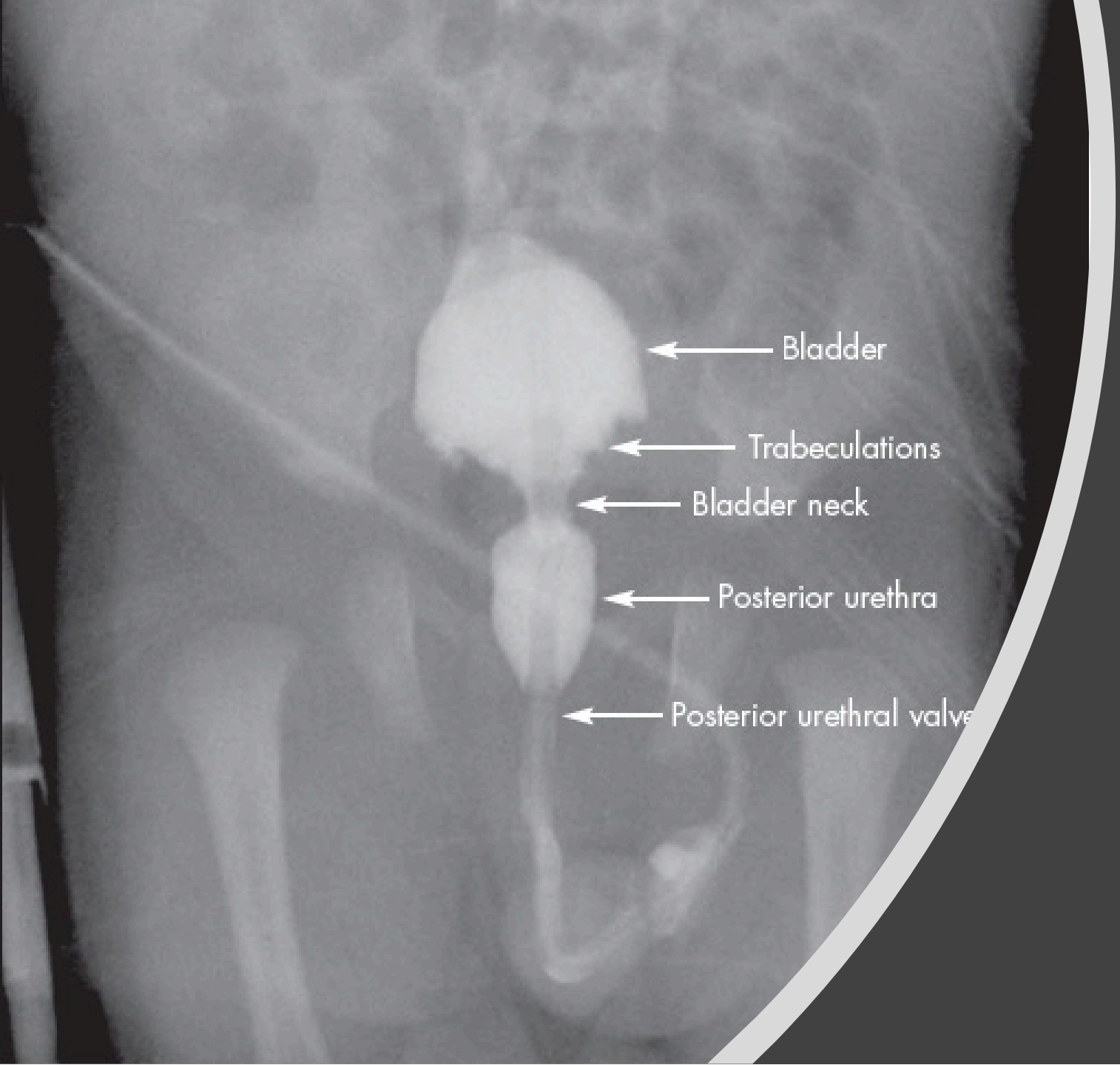
- Augmentation cystoplasty ± catheterizable conduit
  (continent diversion of urine)
  - A cystoplasty using ileum is most commonly performed.
  - Conduits: appendix (most common), ileocecum, sigmoid colon, or stomach.



Neurogenic Bladder (NB)

Augmentation cystoplasty ± catheterizable conduit

- Complications:
  - Mucus production leading to catheter blockage, infection, and bladder stones.
  - Metabolic changes (hyperchloremic alkalosis, electrolyte disturbance, systemic alkalosis (gastrocystoplasty).
  - Spontaneous perforation.
  - Metaplasia/malignancy.
  - Bowel problems (diarrhea, vitamin B12 deficiency).
  - Dysuria and hematuria (gastrocystoplasty).



Posterior Urethral Valves

(PUV)

Posterior Urethral Valves (PUV)

- Posterior urethral valves remain the most common reasons for renal failure and renal transplantation in children.

Posterior Urethral Valves (PUV)

Anatomy

The male urethra is divided into four segments:

- Posterior:
  - Prostatic urethra – from the bladder neck to the site of “urogenital diaphragm.”
  - Membranous urethra – “urogenital diaphragm”.

- Anterior:
  - Bulbar urethra – from the distal margin of the urogenital diaphragm to penoscrotal junction.
  - Penile urethra – urethra that traverses the penile shaft including the glans.

Posterior Urethral Valves (PUV)

Table 5.5.1 Young's Classification[1]

	Description	Result
Type I (~95%)	Bicuspid valve from posterior edge of the verumontanum extending distally and anteriorly and fusing in the midline	Obstructive
Type II	Prominent longitudinal folds extending from verumontanum toward the bladder neck	Nonobstructive
Type III (5–10%)	Circumferential ring distal to the verumontanum at the level of the membranous urethra	Obstructive

Don't memorize this table

Posterior Urethral Valves (PUV)

Clinical Features

- Antenatal (66%):
  - dilated posterior urethra and bladder (“key-hole” sign)
  - ± hydroureteronephrosis
  - ± high renal echogenicity
  - ± oligohydramnios (possible cause of hypoplastic lungs)

- Postnatal (33%):
  - Poor stream ± palpable bladder
  - UTI
  - Renal failure with poor somatic growth and lethargy
  - Diurnal incontinence

Posterior Urethral Valves (PUV)

Investigation

- Micturating Cysto-Urethrogram (MCUG)
  - Wide posterior urethra and prominent bladder neck
  - Partial filling of anterior urethra
  - Posterior urethral bulging forward over the bulbous urethra
  - Valve leaflet lucencies (occasionally)

Posterior Urethral Valves (PUV)
Worse Prognostic Factors

- Antenatal factors
  - Gestation at detection (<24 weeks).
  - US appearance (renal cystic changes imply renal dysplasia).
  - Oligohydraminos.
  - Fetal urine analysis (usually hypotonic with Na<100 mmol/L and Cl<90 mmol/L.)
  - ↑ Na >100, Cl >90, and b-2-microglobulin >40, urine osmolality >210 mOsm, and urine output <2 mL/h
- Postnatal factors
  - Lowest serum creatinine >1 mg/dL at 1 year of age.
  - US appearance: lack of corticomedullary differentiation suggests poor function.
  - Incontinence – inability to achieve diurnal continence signifies bladder dysfunction and this significantly worsens long-term renal function.
  - Lack of a protective “pop-off” mechanism such as gross unilateral reflux or urinary ascites.
  - Presence of severe reflux.

Posterior Urethral Valves (PUV)

Management

- Drain the bladder
  - Infant “feeding” tube is ideal (avoid Foley --> stimulate bladder spasms in the already hypertrophic bladder + can cause secondary ureteric obstruction)
  - Suprapubic catheter (if urethral route is unsuccessful)
- Prevent infection (Prophylactic antibiotics)
- Preserve renal function (intensive IV fluids and electrolyte balance)

Posterior Urethral Valves (PUV)

Management (surgical)

- PUV Ablation
  - Transurethral diathermy ablation and division of the valve leaflets at the 5 and 7 o’clock (Nd YAG laser used occasionally).

- Temporary Urinary Diversion
  - If ablation not feasible (small infant or lack of scopes)
  - Includes vesicostomy, ureterostomy, or pyelostomy

Posterior Urethral Valves (PUV)

Associated clinical features

- Urinary Ascites (5–10%)
  - Peritoneal absorption of urine can lead to uremia.
  - Protect the kidneys from the deleterious effects of constant high back pressure from bladder.

- Vesicoureteric Reflux (50%)
  - Subsides with effective valve ablation (~30%).
  - Persistent reflux (~30%).

Posterior Urethral Valves (PUV)

Bladder Dysfunction and the “Valve Bladder”

- Voiding dysfunction is extremely common in children with PUV.
- Manifests as incontinence and persistence of upper tract dilatation.
- Treatment:
  - Treatment is difficult.
  - ADH treatment is not usually successful.
  - Timed voiding in older cooperative children and overnight free drainage in children with a continent catheterisable channel can protect upper tracts.

Posterior Urethral Valves (PUV)

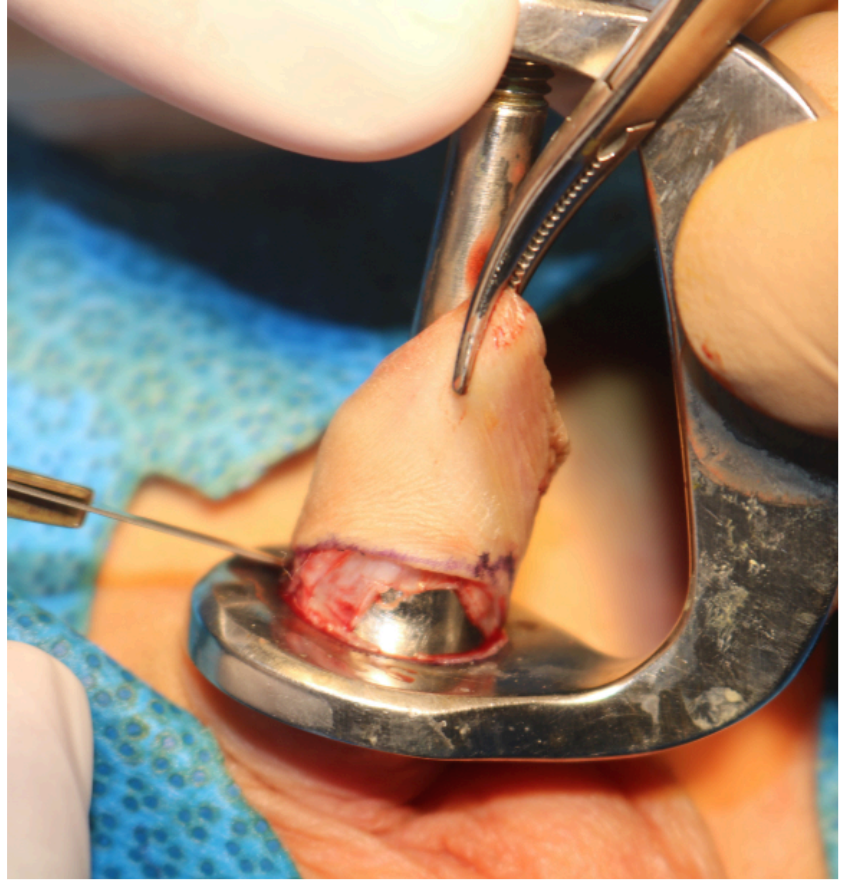
Renal Transplantation

- Up to 30% will ultimately require renal replacement therapy.
- Persistent bladder dysfunction decreases graft survival post transplant.

Posterior Urethral Valves (PUV)

Fertility Issues

- Diminished fertility possibly due to:
  - ↑ Posterior urethral pressure in utero may affect prostate development.
  - ↑ Incidence of UDT.
  - Semen analysis has shown a thick ejaculate with decreased sperm motility.
  - Voiding dysfunction and retrograde ejaculation.


Circumcision

الختان أو الطهور

Natural History of Foreskin Separation

- **At birth:**

  Foreskin (prepuce) is adherent to the glans (non-retractable) (physiological phimosis)

- **At 2–4 years:**

  dissolution of adhesions foreskin can retract

- **At 5 years:**

  most boys should have normal foreskin retraction

History

- An act of faith in both Jewish and Muslim religions:
  - Jewish: timing is important (8th day of life)
  - Muslims: timing is less important (tends to 1st 1-2 weeks of life)

- Not a necessary part of Christian faiths, though lots still encourage it.

Indications

- **Religious or ritual preference** (m.c. indication in our region)
- **Pathological phimosis** (m.c. medical indication in general)
- Paraphimosis
- Recurrent UTI with no known underlying cause

Phimosis

- Foreskin is unable to be retracted to expose the glans.

Types:

- Physiological phimosis (normal state in first years of life)
- Pathological phimosis:
  - **Primary**: true congenital phimosis with pin-hole meatus.
  - **Secondary to**:
    - Bacterial infection:
      - **balanitis** (inflammation of the glans)
      - **posthitis** (inflammation of the foreskin)
    - Balanitis xerotica obliterans (BXO)

Phimosis | Management

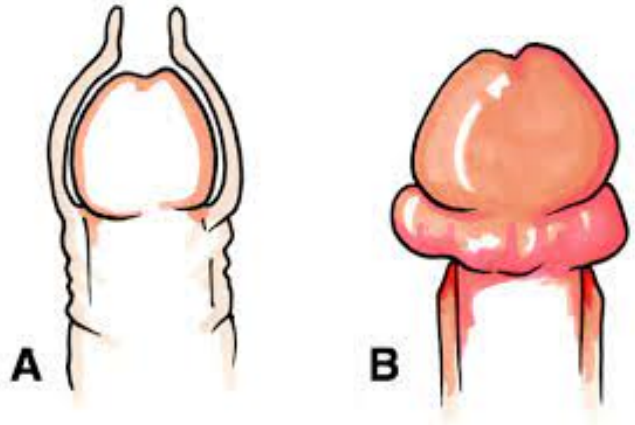
- **Conservative**:
  - Reassure, gentle self-retraction
  - Topical steroids (e.g., betamethasone 0.1%)

- **Surgical options**:
  - Circumcision
  - Preputial plasty

Paraphimosis

- Foreskin is able to be retracted but becomes stuck in that position

 resulting in distal congestion and edema of glans



Paraphimosis | Management

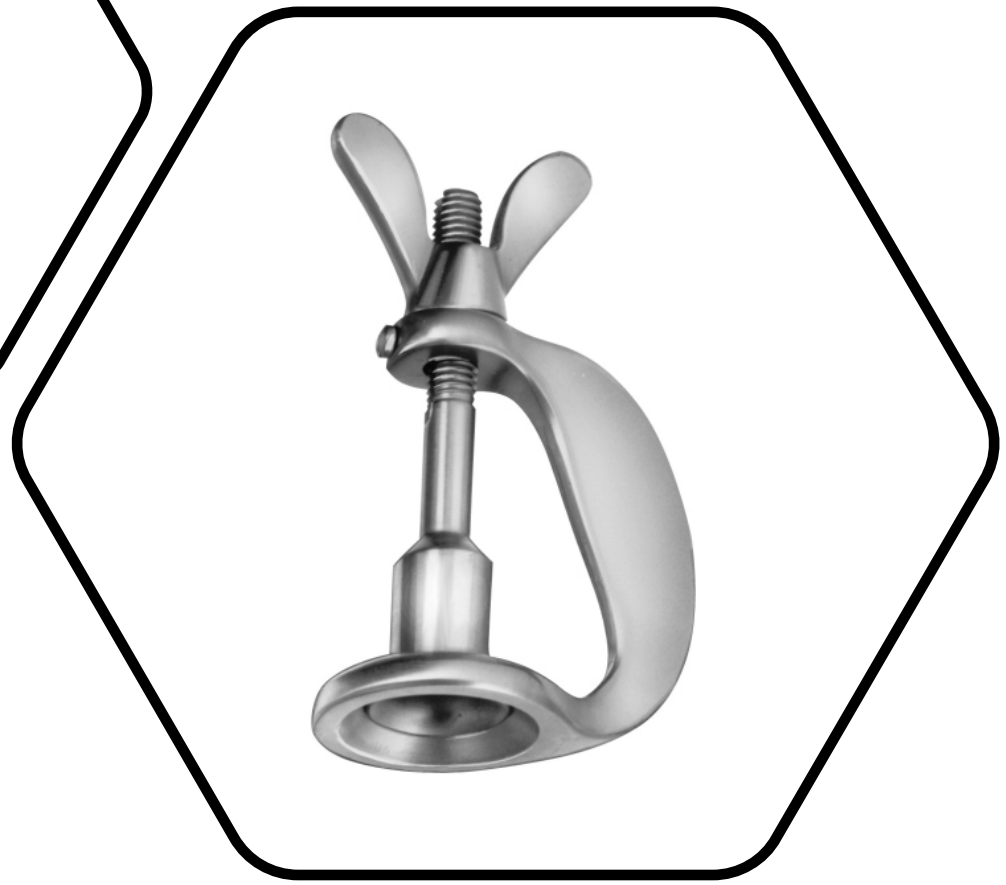
- **Surgical emergency**

- **Manoeuvres can be done in ER:**
  - Compresses with ice or sugar (to reduce the swelling and allow protraction)
  - Multiple needle punctures (to allow fluid to be squeezed out)

- if failed send to OR

  dorsal slit of the tight band +/- circumcision (under GA)

Circumcision



Surgical Freehand

Surgical using specific clamps

- Plastibell® | Gomko clamp® | Winkelmann clamp® | Mogen clamp

Circumcision

Contraindications

- **Absolute:**
  - Family history of bleeding disorders
  - Newborn with known bleeding tendency/disorder
  - Newborn with pathological jaundice

- **Relative:**
  - Hypospadias (as foreskin helps in the surgical repair of hypospadias)

Circumcision | Complications

- **Bleeding**
- Infection
- Meatal stenosis
- Insufficient/excessive foreskin removed
- Adhesions, skin bridges, or inclusion cysts
- Entrapped penis or secondary phimosis
- Urethral injury (iatrogenic hypospadias)
- Necrosis of the penis (injudicious use of electrocautery to control bleeding)
- Amputation of the glans (partial or complete)
- **Death** (mainly due to unnoticed bleeding)

Hypospadias

الإحليل التحتي أو الطهور الملائكي

Definition

A complex of..

- abnormal ventral urethral meatus
- dorsal hooded foreskin
- glans defect
- underdeveloped corpus spongiosum
- +/- phallic torsion
- +/- phallic ventral curvature (chordee)

Incidence

- Usually isolated
- Can be part of the DSD spectrum

- **One in 300 live-births**

- Associated with:
  - ↑ parity
  - ↑ maternal age
  - ↓ birth Wt
  - +ve family history
  - inguinal hernia & hydrocele (10%)
  - undescended testes (8%)

Etiology

Genetic Factors

- Exact mode of inheritance is unknown
- Monozygotic twins (x8)
- +ve family history (8% fathers | 14% brothers)

Endocrine Factors

- Deficient androgenic stimulation
  (production, conversion, or sensitivity)
- Increased maternal progesterone exposure (x5)

Environmental Factors

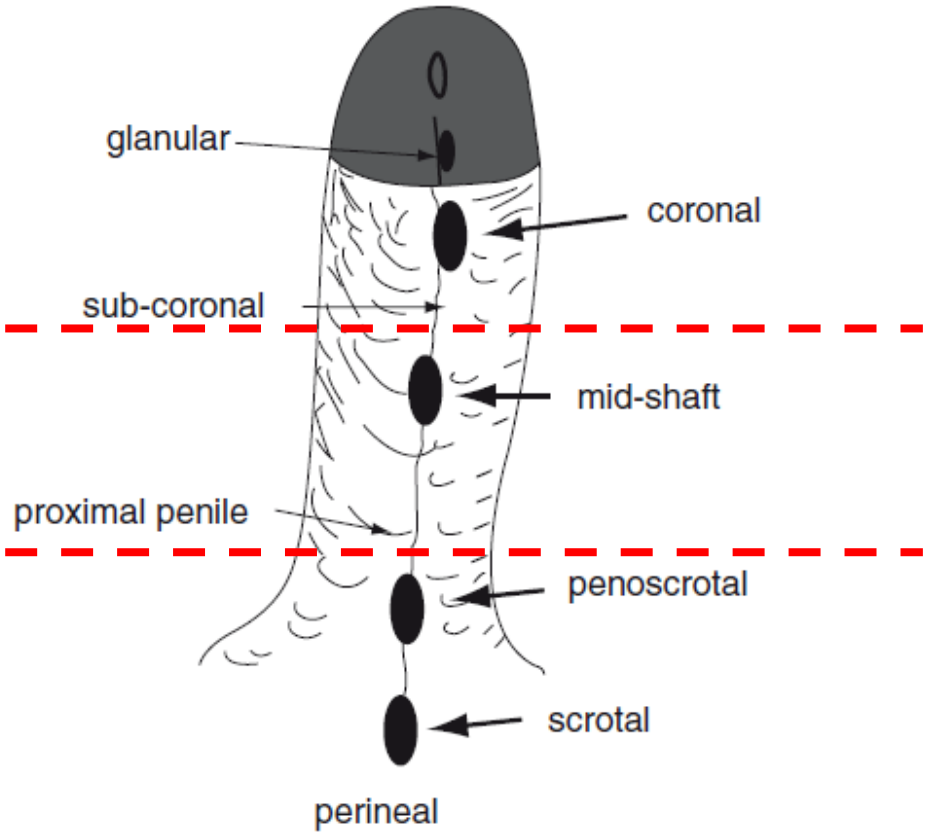
- Maternal exposure to estrogenic substances
  (in pesticides, milk, and pharmaceuticals)

Classification

Distal (50%)

Middle (30%)

Proximal (20%)



Surgery

Best timing recommendation:

- before 18 months of age (minimizes psychological impact of genital surgery)

Hormone manipulation preoperative:

- penile size can be increased by..
  - weekly IM testosterone or hCG
  - or topical testosterone or DHT

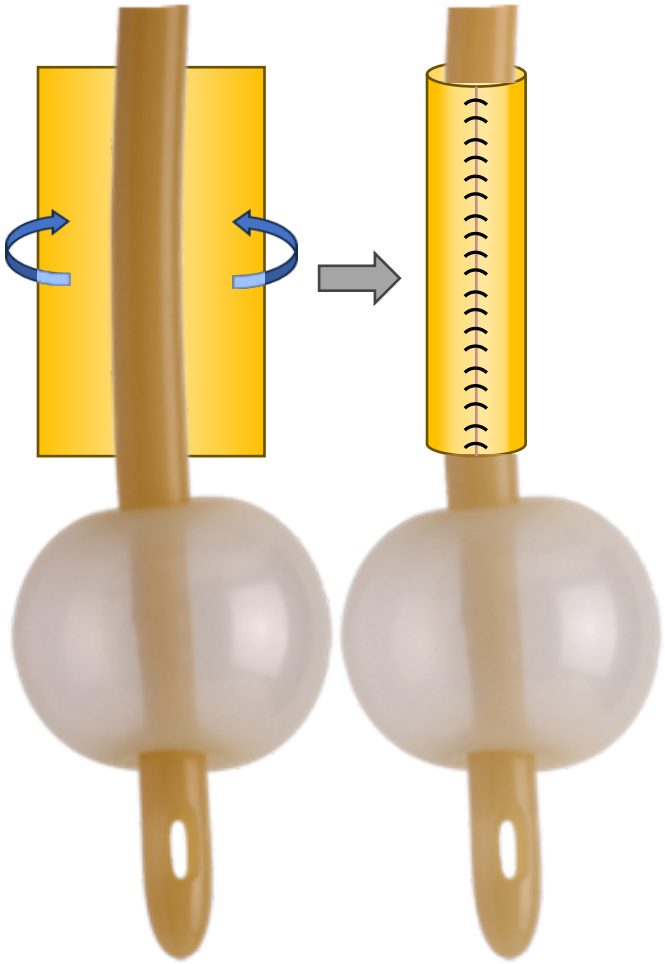
Surgery

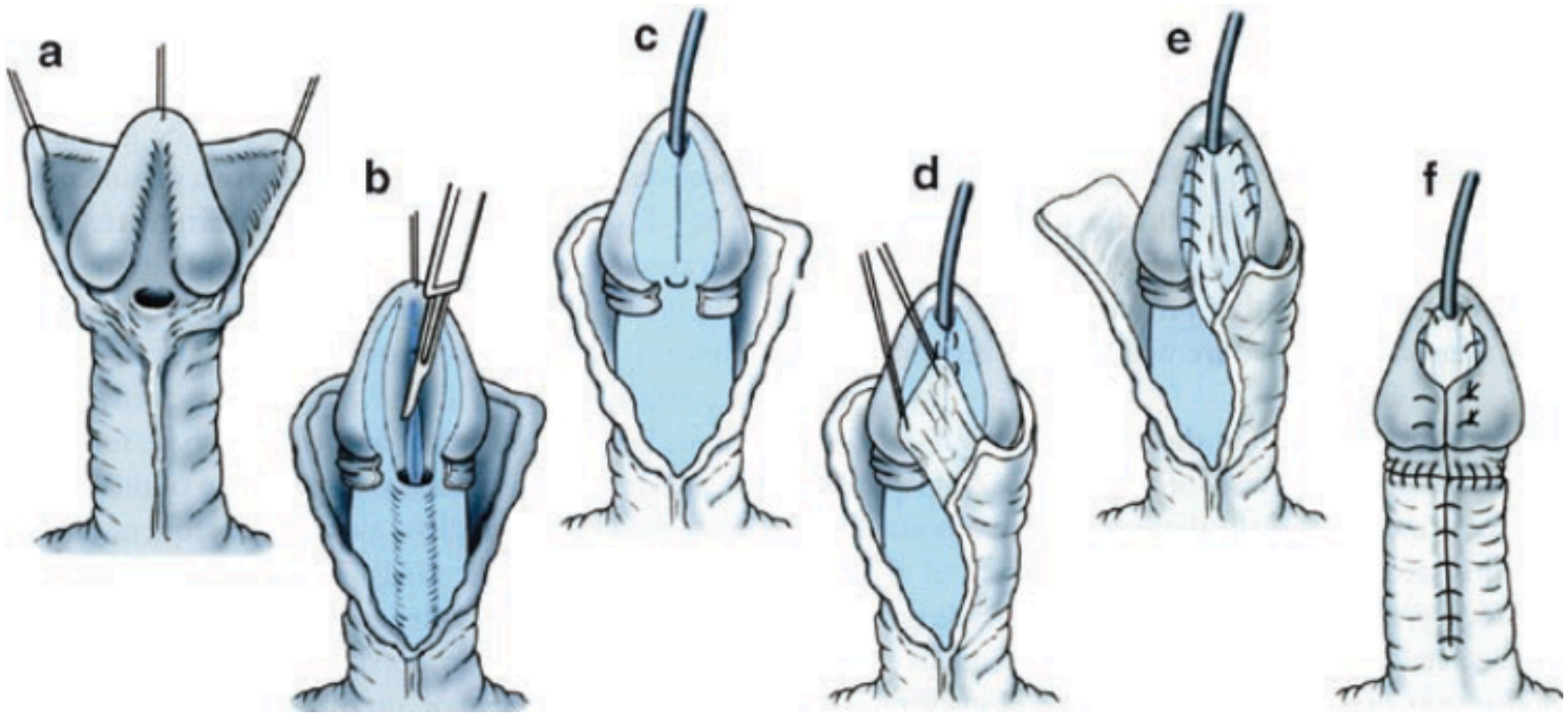
Tube urethroplasty

urethral plate is tubularized to neourethra **[main step]**

+

- **Corpus spongioplasty:** deficient corpus spongiosum is compensated by fascia (preputial or dartos fascia) to support the neourethra
- **Straightening phalloplasty:** chordee is released
- **Glansplasty:** glans defect is corrected
- **Circumcision:** dorsal foreskin is removed
- **Phalloplasty:** phallic torsion is corrected



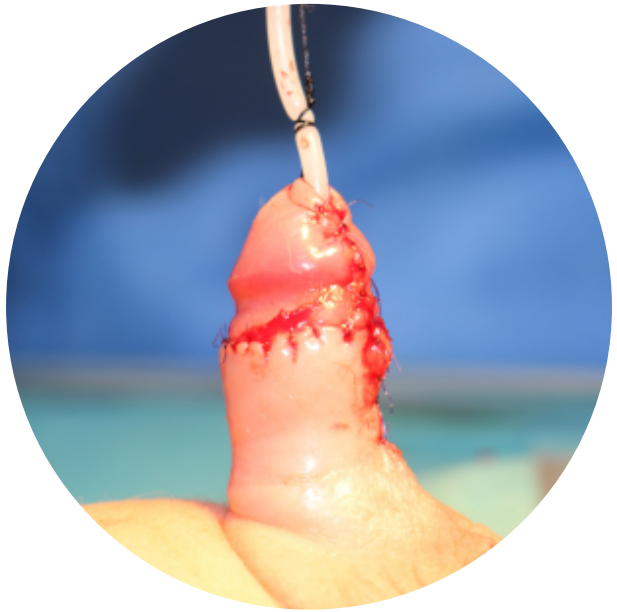


Don't memorize this slide

Incised-plate tube urethroplasty "Snodgrass repair"

Adapted from Belman et al. (Belman AB, King LR, Kramer SA (eds) (2002) Clinical pediatric urology, 4th edn. Martin Dunitz, London, p 1077)

Postoperative care



- Neourethra is protected for 1 week with a “stent”
- Simple analgesics
- Oral antibiotic

- **Early complications:**
  - Bleeding
  - Hematoma
  - Infection
  - Breakdown of repair

- **Late complications:**
  - Meatal stenosis
  - Urethrocutaneous fistula (UCF)
  - Persistent chordee
  - Urethral stricture
  - Urethral diverticulum

Disorders of Sex Development

(DSD)

Disorders of Sex Development (DSD)

- Rarely, genital appearance at birth may be ambiguous or even contrary to genetic sex.
- Should be regarded as a medical emergency to achieve the best gender outcome for the baby.

Disorders of Sex Development (DSD)

- Ince 2006, the phrase disorders of sexual development (DSD) has been advocated, and appears preferable to the older terms:
    - ambiguous genitalia
    - Hermaphroditism
    - Pseudohermaphroditism
    - intersex

Disorders of Sex Development (DSD)

- Embryology

- From 6 weeks, early bipotential gonads develop within the genital ridge from primordial germ cells.

Disorders of Sex Development (DSD)

- Clinical Features
- The presentation tends to be either that of an under-virilized male, or an over-virilized female (there are exceptions).
- The birth of an infant with ambiguous genitalia will be a cause of much anxiety for the parents and in some cases (congenital adrenal hyperplasia), the sequelae may be life-threatening.
- This event should, therefore, be regarded as medical emergency and subsequent management should proceed in a logical and timely manner in a specialized center with multidisciplinary input.
- It is important to avoid guessing the sex and using pronouns such as “he” or “she.” Phrases such as “your baby” are more appropriate.

Disorders of Sex Development (DSD)

- **Clinical Features**
- Particular points to be noted when examining include:
  - Presence of a palpable gonad on either side.
  - Appearance of phallus (length, width, and chordee).
  - Appearance of scrotum/labioscrotal folds, degree of skin rugosity/pigmentation.
  - Location of external urethral opening and number of orifices present on the perineum.
- A proportion of children with DSD will only present later in life, such as those with Swyer syndrome who look unequivocally female (XY, complete gonadal dysgenesis), but who are unable to develop secondary sexual characteristics or menstruate.

Disorders of Sex Development (DSD)

- **Symmetrical** genital appearance imply a biochemical etiology (e.g., congenital adrenal hyperplasia (CAH))
- **Asymmetrical** appearance implies a chromosomal abnormality (e.g., mixed gonadal dysgenesis)

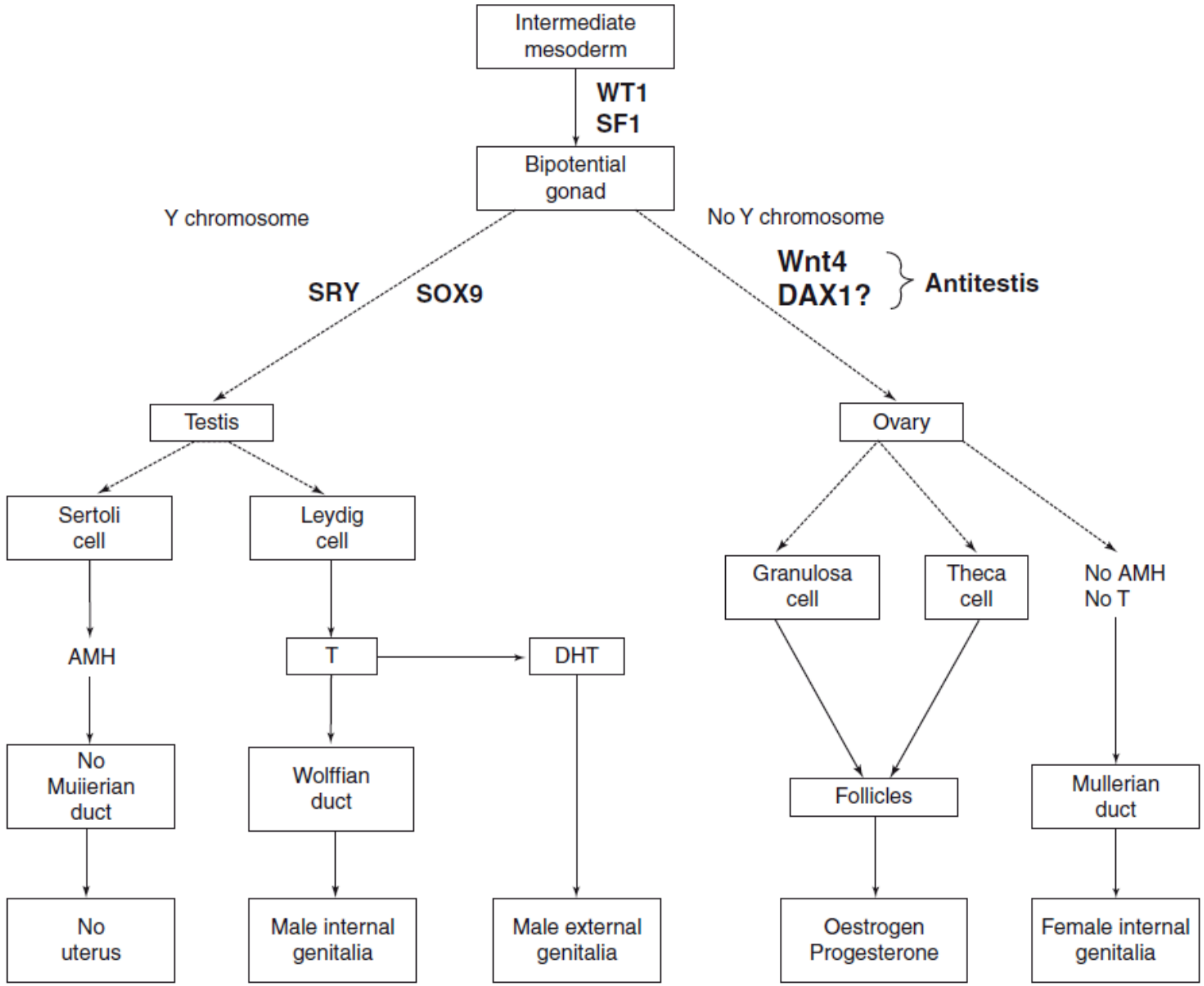


Fig. 5.8.1 Embryology of sexual development

Embryology of sexual development

Table 5.8.1 Classification of DSD (after Lee PA, Houk CP, Hughes IA, Ahmed SF, Houk C, et al. Consensus statement on management of intersex disorders. Pediatrics 2006; 118:e488-e500)

	Variant	Notes
46XY DSD (undervirilized male)	Severe hypospadias	
	Androgen insensitivity syndromes (complete and partial)	Female phenotype, *AR* mutations
	5 α-Reductase deficiency	Failure to produce more active DHT
	Complete gonadal dysgenesis (Swyer syndrome (Swyer GI (1955) Male pseudohermaphroditism: a hitherto undescribed form. Br Med J 2:709–712); 46XY sex reversal)	Female phenotype, *SRY* mutations (~20%), usually detected at puberty
	Leydig cell hypoplasia	Autosomal recessive
	Disorders of anti-Mullerian hormone (AMH) production and receptor	*AMR* and *AMHR-II* mutations
46XX DSD (overvirilized female)	Congenital adrenal hyperplasia	*Vide infra*
	Fetoplacental aromatase deficiency	
	Exogenous androgen exposure	
	Ovotesticular DSD	
Sex chromosome DSD	45XO (Turner[2] syndrome)	Short, webbed neck, female phenotype, and aortic coarctation (60%)
	45XO/46XY (mixed gonadal dysgenesis)	
	46XX/46XY (chimeric ovotesticular DSD)	

Disorders of Sex Development (DSD)

- **Investigations**
  - Genetic – karyotype and specific gene arrays
  - Endocrine – blood and urine biochemistry, hormone assays
  - Imaging – ultrasound (renal/pelvic), contrast studies (cystogram/genitogram), and MRI
  - Surgical – cystovaginoscopy, laparoscopy, and skin/gonadal biopsies
- It is important that the birth is not registered until the final decision has been made.

Disorders of Sex Development (DSD)

- Wherever possible, gender is assigned according to karyotype, rather than being reassigned to suit the initial appearances of the external genitalia, although this may not always be possible.

Disorders of Sex Development (DSD)

- **Principles**
  - Once a karyotype has been obtained, subsequent management should focus on:
    - Hormone replacement and electrolyte balance if indicated
    - Delineation of anatomy (US, cystovaginoscopy, MRI, and laparoscopy)
    - Need for and timing of reconstruction
    - Management of gonads
      - ↑ Risk of malignancy, streak ovaries → gonadoblastoma
    - Genetic counseling regarding future pregnancies
    - Psychological support for family and patient

Disorders of Sex Development (DSD)

- **Congenital Adrenal Hyperplasia**
  - Usually 21-Hydroxylase Deficiency
  - Appear as infants with ambiguous genitalia and bilateral impalpable gonads.
  - Can become life-threatening due to salt-losing nature (75%) with ↓ aldosterone.
  - Workup:
    - Urgent karyotype – XX.
    - Steroid profile – ↑ 17-hydroxy progesterone ↑ androstenedione levels.

46,XY DSD (severe hypospadias)

46,XX DSD (congenital adrenal hyperplasia)

Disorders of Sex Development (DSD)

- Surgery

- 46,XY DSD (Undervirilized Male)
- The main issues for this group include:
    - Hypospadias – cosmetic/functional aspects
    - Undescended testes/impalpable gonads (?malignant potential).
    - Streak gonads or intra-abdominal gonads at high risk of malignancy (e.g., mixed gonadal dysgenesis) should be removed.
    - Assessment and management of persistent Müllerian structures. Often asymptomatic and if so do not require excision. Recurrent infection or postmicturition dribbling may necessitate their removal.

Disorders of Sex Development (DSD)

- Surgery
- 46,XX DSD (Overvirilized Female)
- The main issues are to improve the external cosmetic appearance, preserve clitoral function, and separation of the urethra and vagina.
- Feminizing genitoplasty may include the following:
  - Clitoroplasty including recession, concealment, or reduction.
  - Vaginoplasty – to separate vagina and urethra.
  - Total or partial urogenital mobilization, vaginal pull-through, or replacement.
  - Introitoplasty to produce a more feminized appearance.


thank
you