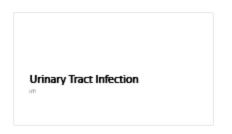
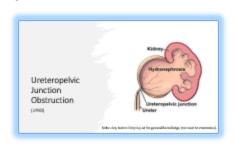
# Principles of Pediatric Urology

By Raed Al-Taher, M.D.

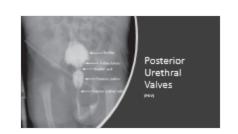


#### Topics in BLUE are the required ones



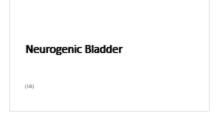










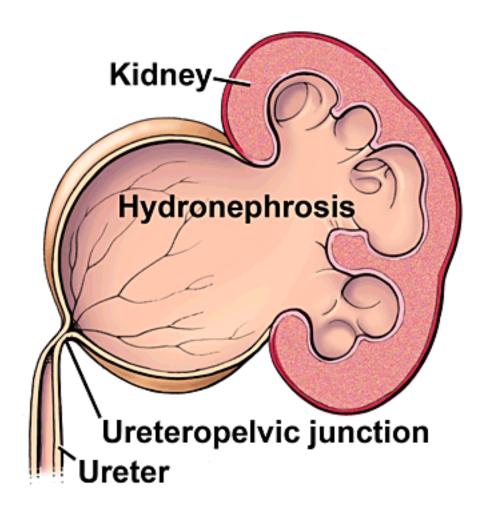




Disorders of Sex Development

Edited by: Haya Khader

# 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
- Rarely: mucosal valves, polyps, and true ureteric strictures

#### 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).

## 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

#### Postnatal US

- The primary investigation tool for HN
- Assesses kidney anatomy & AP diameter of renal pelvis

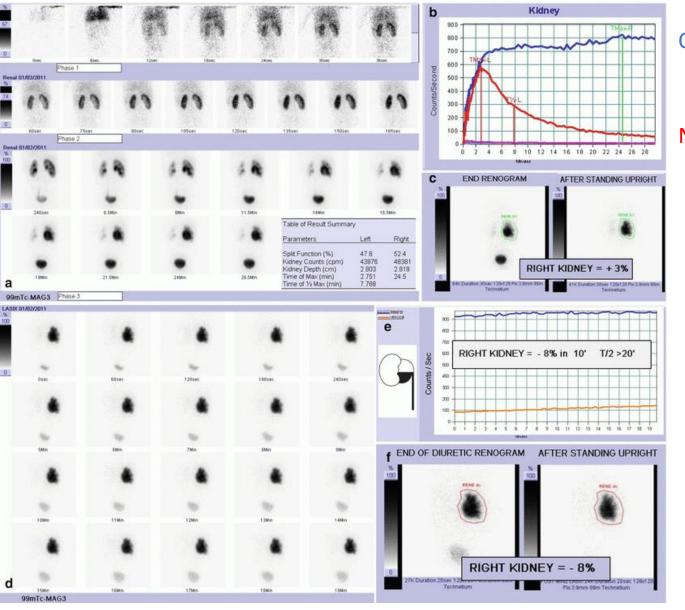
#### Renal radioisotope Scan

- 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 (t½) after administration of furosemide
    - normally t½ <20 min</li>
    - t½ >20 min = significant obstruction → needs intervention

#### 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

#### Treatment

#### Antenatally detected HN

- Conservative management (follow up)
- Surgery (needed in <50% of cases) when any or all of the following:
  - Functional deterioration (<40%)</li>
  - $T_{1/2} > 20 \text{ minutes}$
  - 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.

# Vesico-Ureteric Reflux

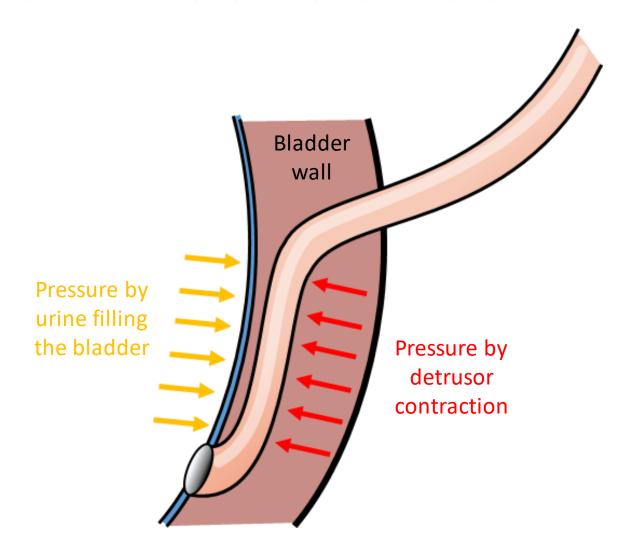
(VUR)



# Vesico-Ureteric Reflux (VUR)

- Female predominant
- Peak incidence at 3 years
- Familial incidence is 2–4% of all cases

#### Normal ureteral submucosal tunnel

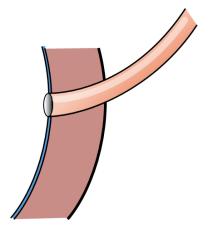


tunnel length: ureteral diameter → should be about 5:1 to prevent VUR

# Pathology

#### Primary VUR:

• due to a short ureteral submucosal tunnel in the bladder wall



#### Secondary VUR:

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

## Clinical Features

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

# Investigations

• Urine analysis  $\rightarrow$  r/o infection

- US → HUN (hydro-uretero-nephrosis)
  DMSA nuclear scan → for renal scars and differential renal function
  MCUG → for degree of VUR

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

# MCUG | grading

Ш IV complex calyx dialated ureter tortous ureter Dilated ureter and pelvicalyceal system + flat fornices. Lower ureter/s filled with contrast (without dilatation) Tortuosity of ureters + complete blunting of fornices All ureter/s filled with contrast (without dilatation) More dilatation + convex fornices

#### 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

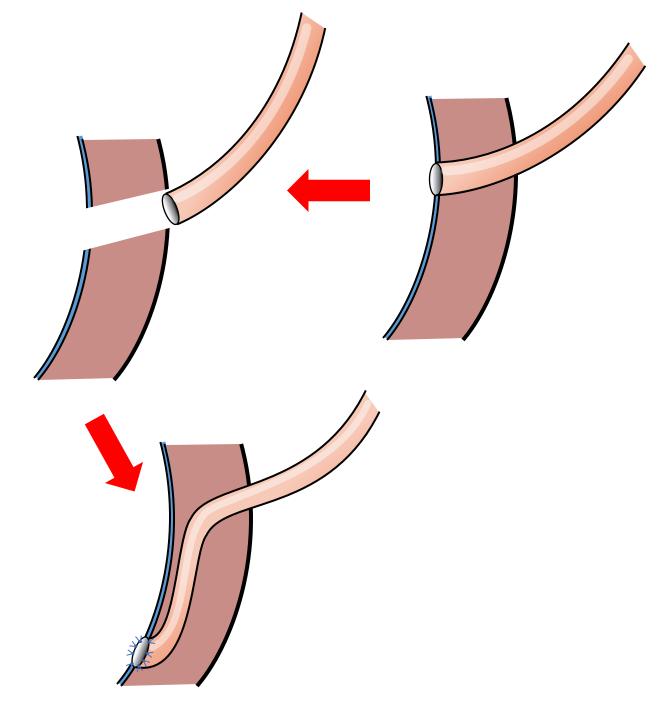
#### Indications:

- 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



# Surgical treatment

#### **Postoperative Management**

- Antibiotics for ~3 months
- Follow-up VCUG or DIC Direct isotope cystography

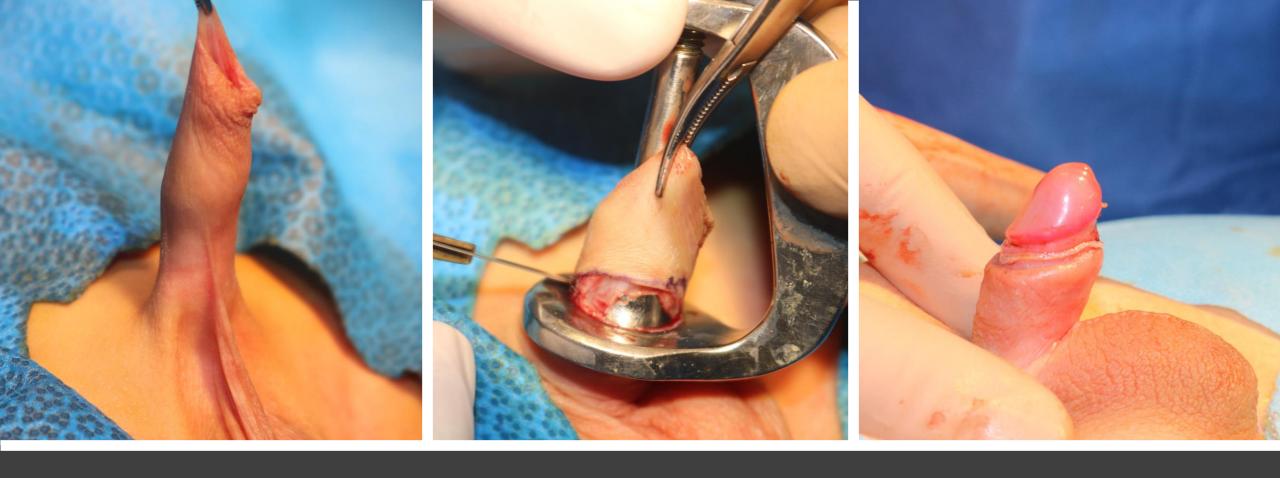
#### **Complications**

- 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 scarring nor parenchymal damage



# 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  $\rightarrow$  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 (8<sup>th</sup> 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
- Pathological phimosis
- 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 "stretch" or -plasty

# **Paraphimosis**

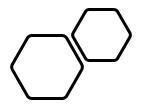
 Foreskin is able to be retracted but becomes stuck in that position

> resulting in distal congestion and edema of glans

# Paraphimosis | Management

A 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<sup>®</sup> | Gomko clamp<sup>®</sup> | Winkelmann clamp<sup>®</sup> | Mogen clamp



## Circumcision

#### **Contraindications**

#### Absolute:

- Family history of bleeding disorders
- The newborn has known bleeding tendency/disorder, or pathological jaundice

#### Relative:

• Hypospadias (as foreskin helps in the surgical repair of hypospadias)

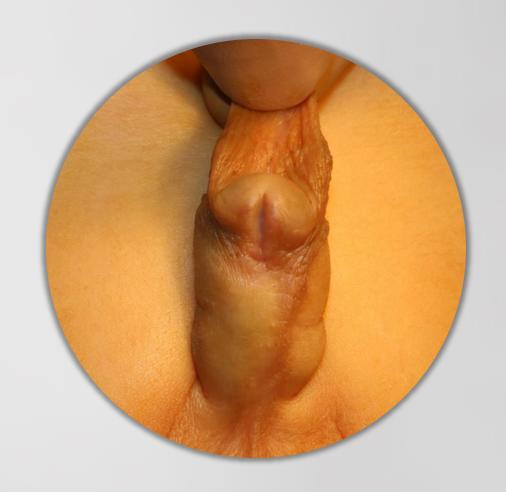
a birth defect where the location of the opening of the urethra is not at the tip of the penis

# **Circumcision** | Complications

- Bleeding
- Infection
- Meatal stenosis particular to neonatal circumcision
- 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:

- 个 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)

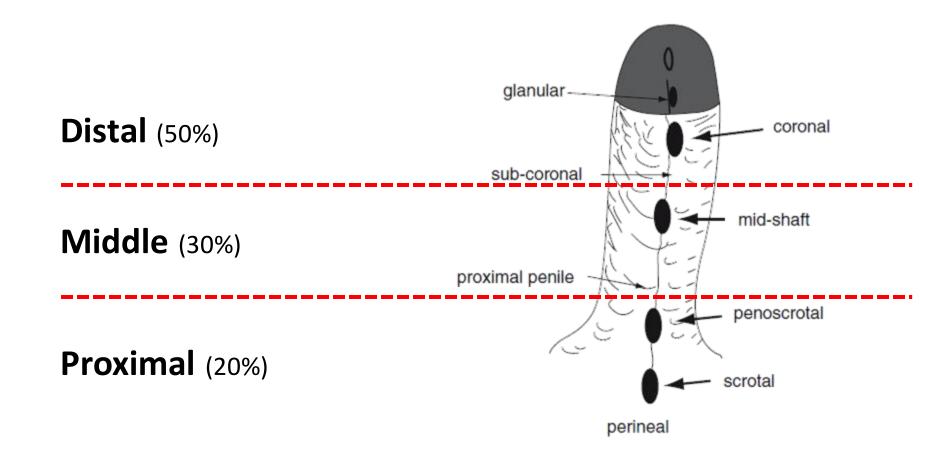
# Endocrine Factors

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

# Environmental Factors

• Maternal exposure to <u>estrogenic</u> substances (in pesticides, milk, plastic linings of metal cans, and pharmaceuticals)

# Classification



# 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

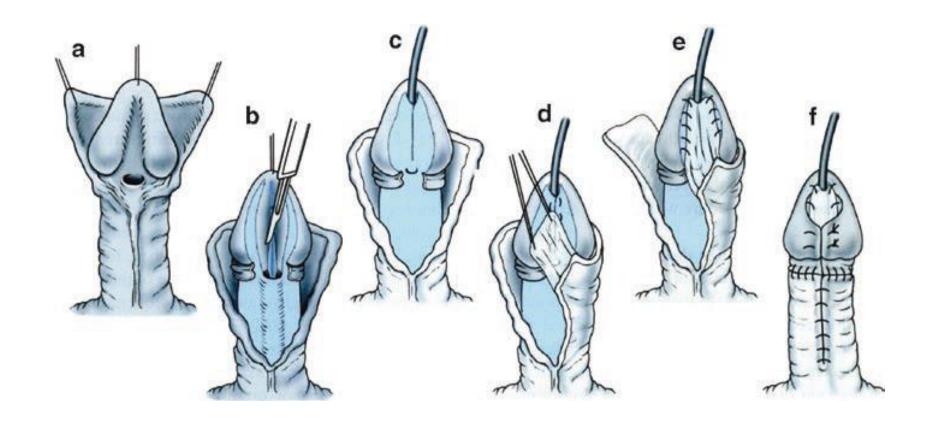
# Surgery

## **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

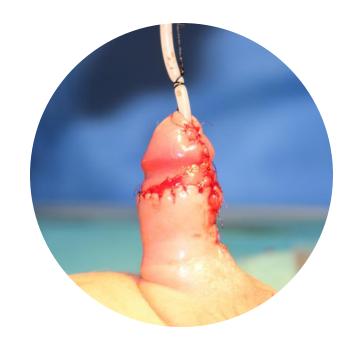


#### Incised-plate tube urethroplasty "Snodgrass repair"

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

# Thank You