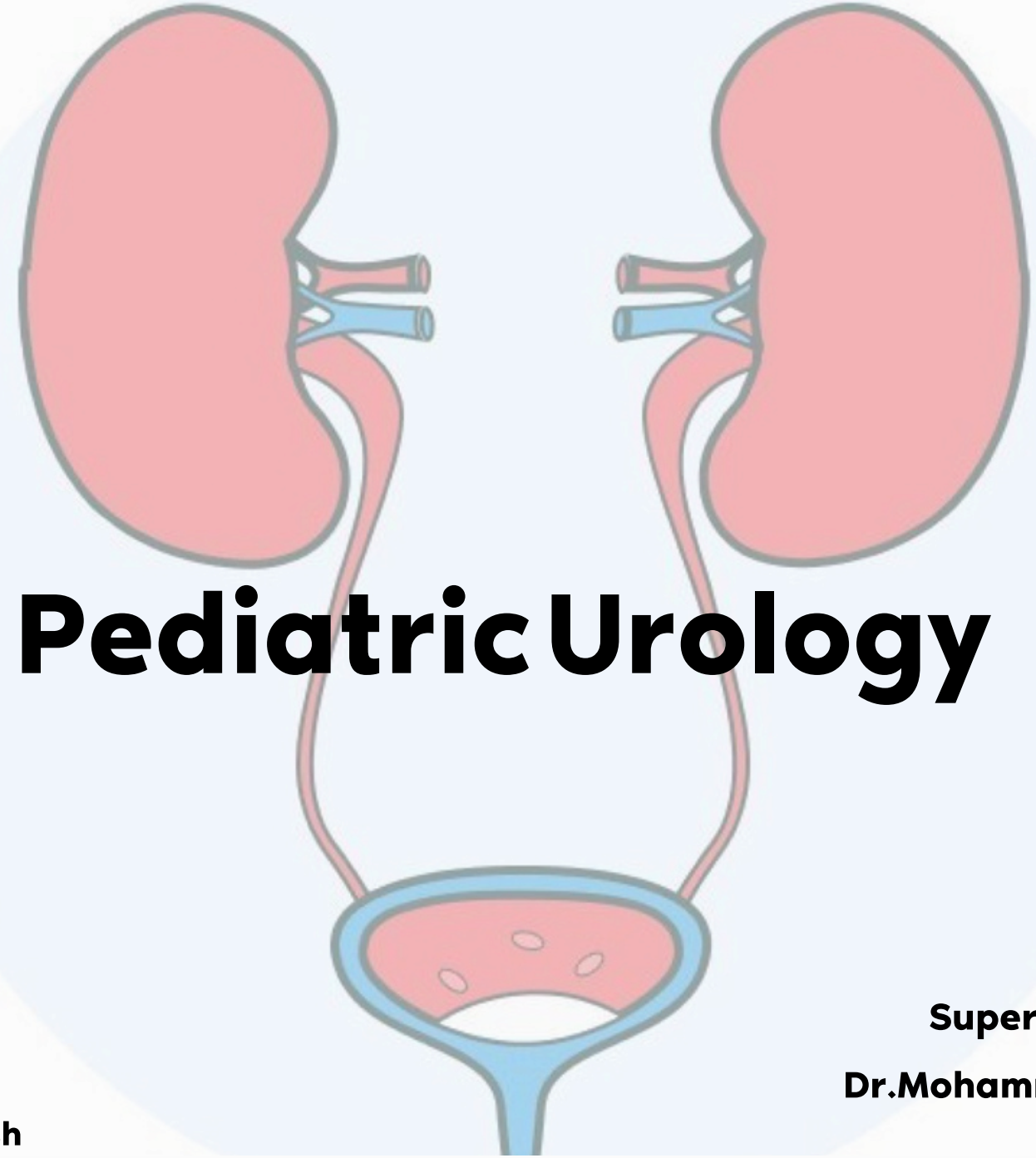

Pediatric Urology

Presented by:

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Supervised by :

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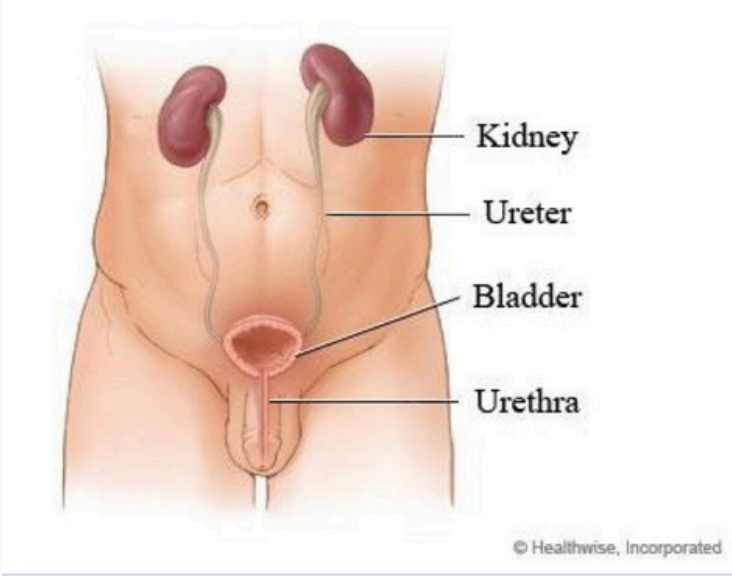
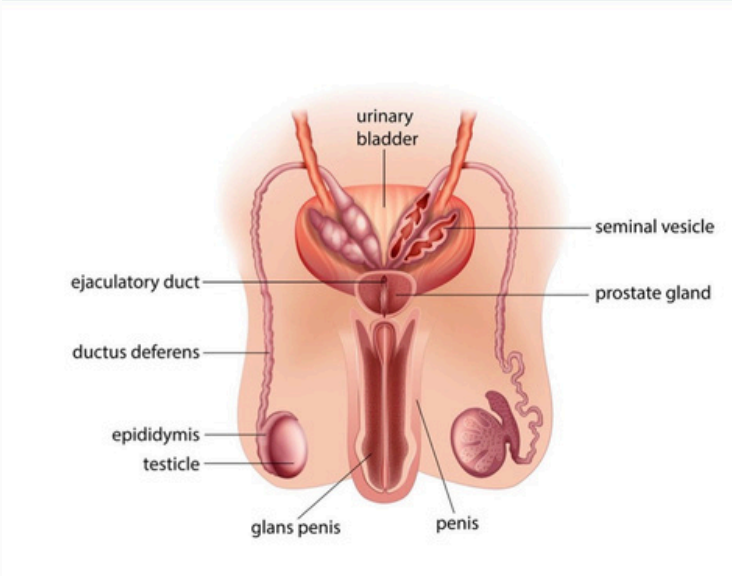
- Undescended testes
- Vesicouretericreflux (VUR)
- Hypospadias
- Nocturnal enuresis
- Posterior urethral valves (PUV)
- Extrophyof the Bladder
- Epispadias
- Phimosis
- Paraphimosis
- Disorder of sexual differentiations



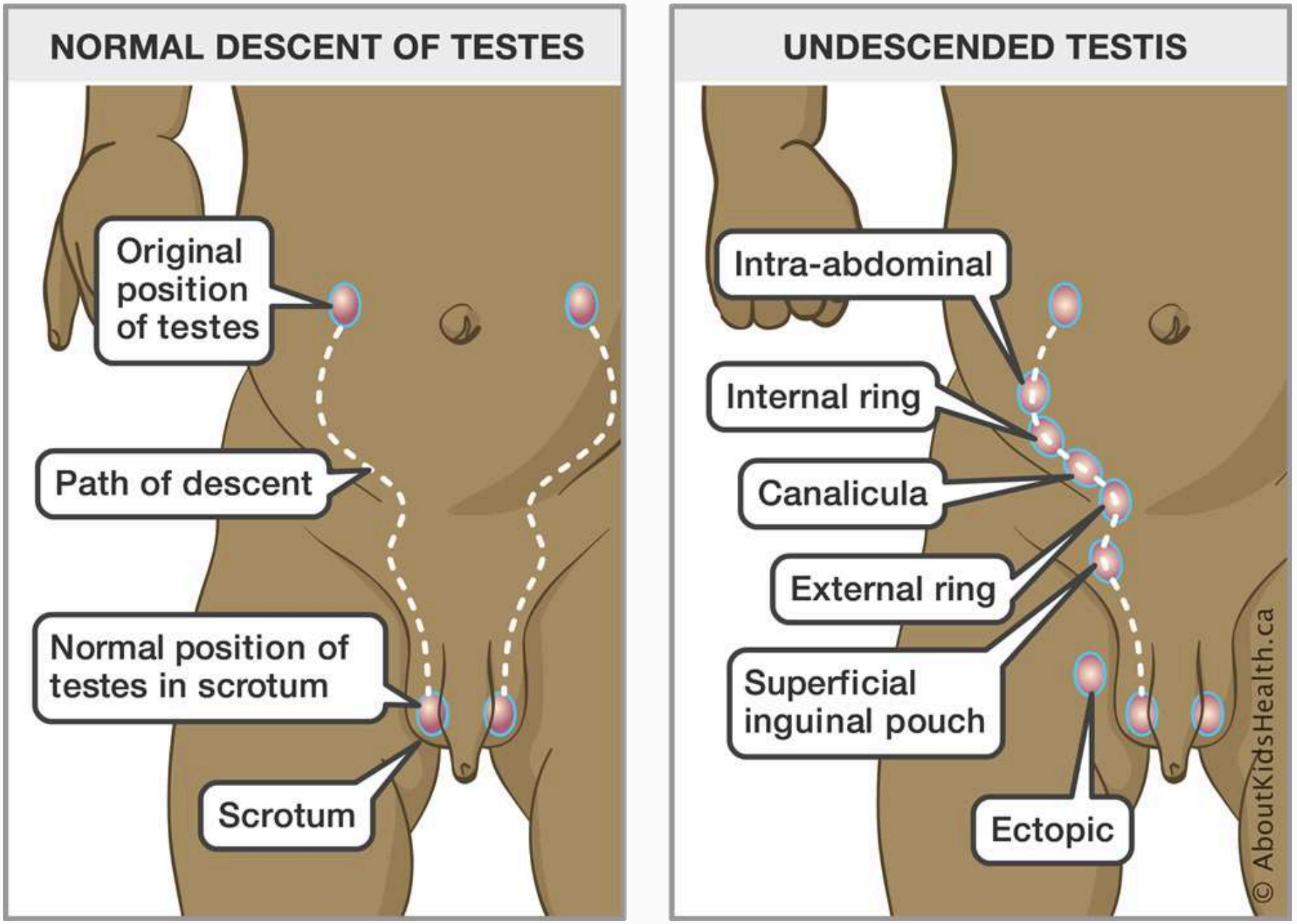


Undescended testes

The testes descend into thescrotum in the third trimester (passing through the inguinal canal at 24–28 weeks) , Failure of testicular descent results in cryptorchidism(or undescended testes).

- Incidence: Incidence is 3% at birth (unilateral > bilateral).Approximately 80% will spontaneously descend by 3months. The incidence at 1 year is 1%.

This is why we wait to fix this abnormality for >6 months of age.


NORMAL DESCENT OF TESTES
Original position of testes
Path of descent
Normal position of testes in scrotum
Scrotum
UNDESCENDED TESTIS
Intra-abdominal
Internal ring
Canalicula
External ring
Superficial inguinal pouch
Ectopic
© AboutKidsHealth.ca

Classification

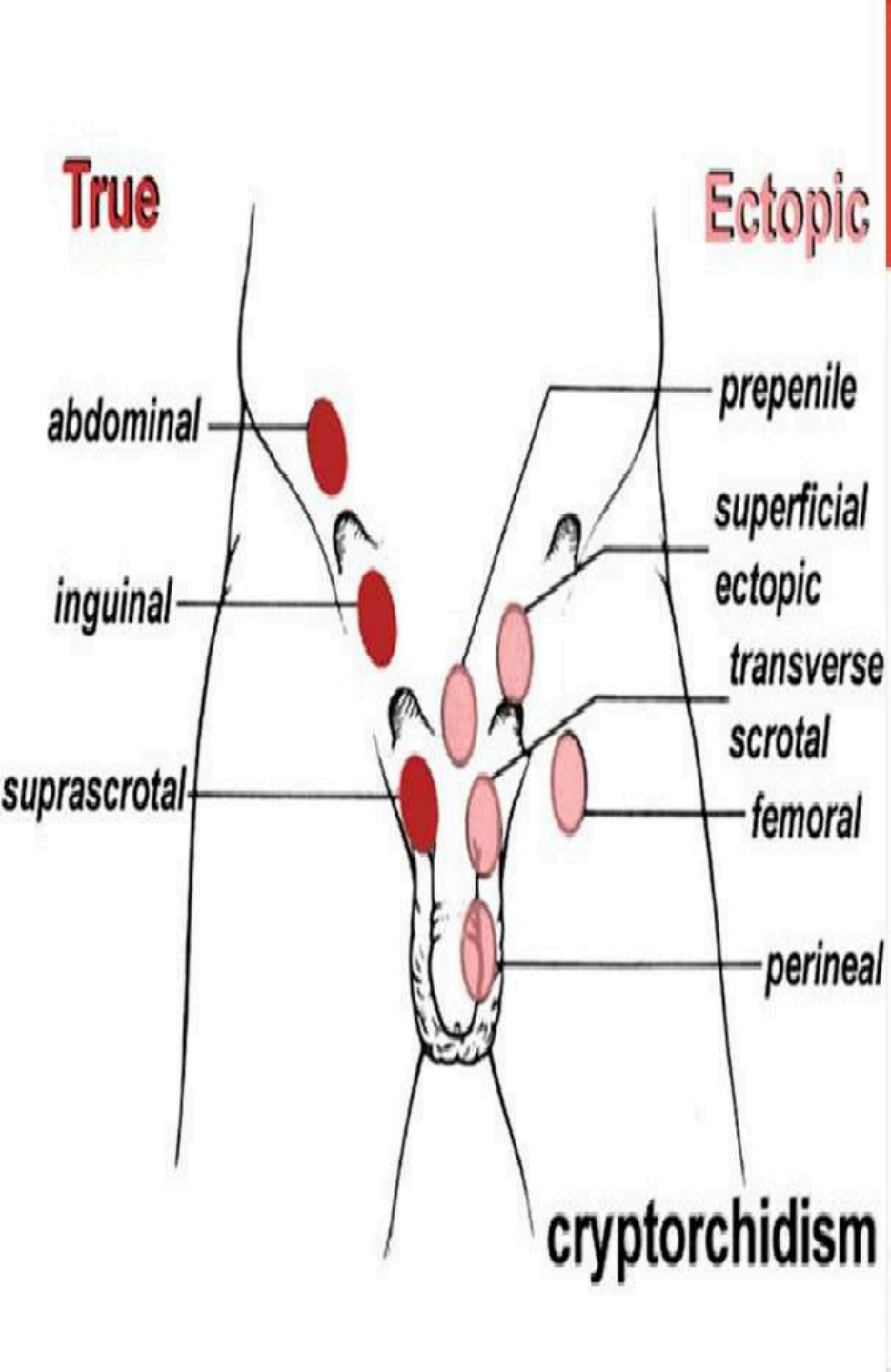
Retractile: an intermittent active cremasteric reflex causes the testis to retract up and out of the scrotum.

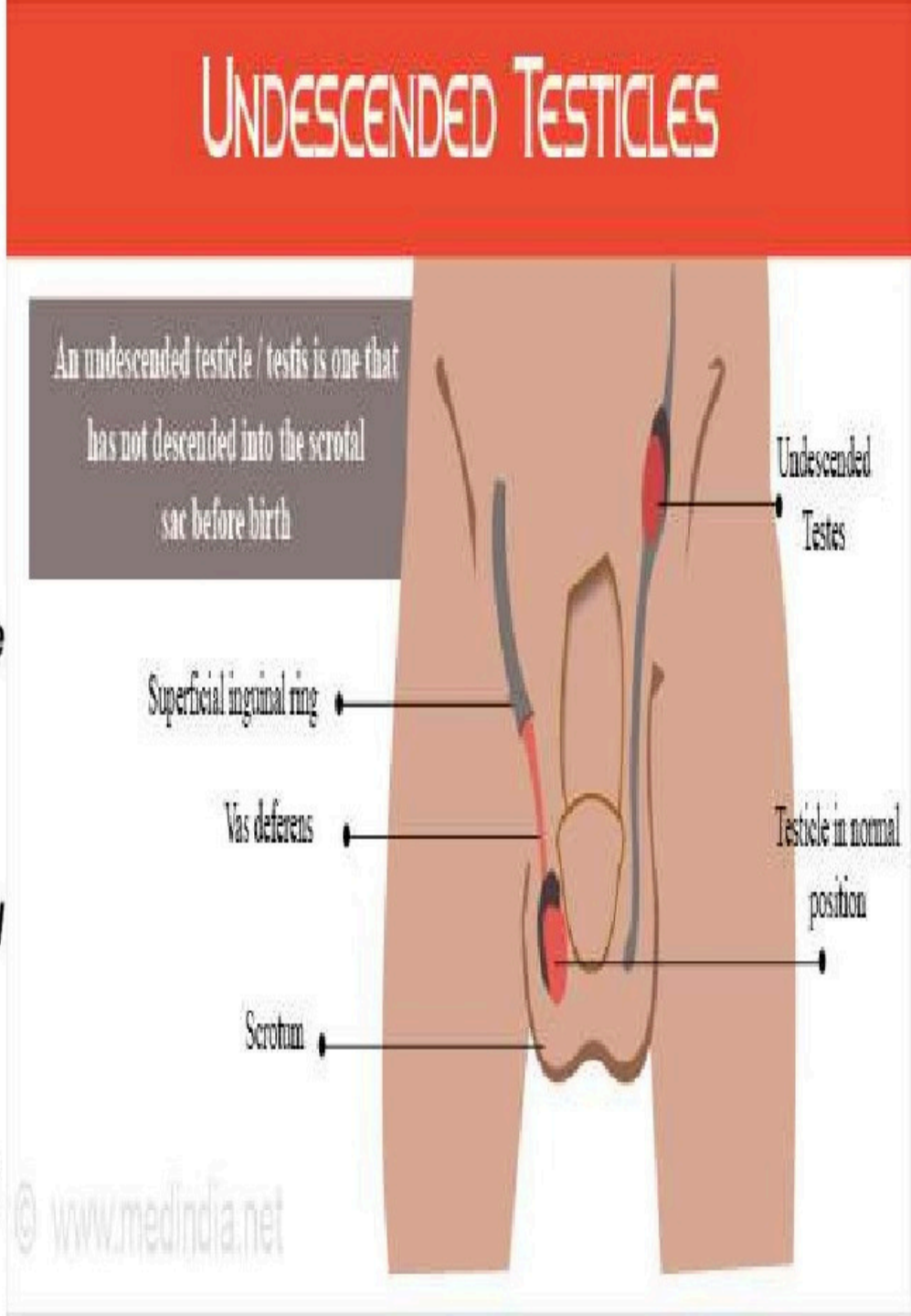
Ectopic (<5%): abnormal testis migration below the external ring of the inguinal canal (to perineum, base of penis, or femoral areas)

Incomplete descent (~95%): testis may be intra-abdominal, inguinal,orprescrotal

Atrophic/absent

No matter the type, our goal is to have the testes in the scrotum by 1 year of age.


True
Ectopic
abdominal
inguinal
suprascrotal
prepenile
superficial ectopic
transverse scrotal
femoral
perineal
cryptorchidism


UNDESCENDED TESTICLES
An undescended testicle / testis is one that has not descended into the scrotal sac before birth
Undescended Testes
Superficial inguinal ring
Vas deferens
Testicle in normal position
Scrotum
© www.medindia.net

Risk factors

These include preterm infants, low birth weight, small for gestational age, and twins.

Etiology

This includes :

1.Abnormal testis

2.Gubernaculum

3.Endocrine abnormalities (low level of androgens[hCG], (LH),calcitonin gene–related peptide)

4.Decreased intra abdominal pressure (prune-belly syndrome, gastroschisis).

Pathology

- There is degeneration of Sertoli cells, loss of Leydig cells, and atrophy and abnormal spermatogenesis.
- Long-term complications :

- Relative risk of cancer is 40-fold higher in the undescended testis.carcinoma in situ represents a small percentage (~2%).

- Reduced fertility (atrophy)
- Increased risk of testicular torsion
- Increased risk of direct inguinal hernia
- Increased risk for trauma

Diagnosis

- Full examination is required to elucidate if the testis is palpable and to identify location.
- Assess for associated congenital defects.
- If neither testis is palpable, consider chromosome analysis (to exclude an androgenized female) and hormone testing (high LH and FSH with a low testosterone indicates anorchia).

Management

- Treatment should be performed within the first year.
- Hormone therapy (hCG, LHRH) stimulates testosterone production.(currently studies have shown that hormonal therapy is useless)
- Surgery consists of inguinal exploration, mobilization of spermatic cord, ligation of processus vaginalis, and securing the testis into a dartospouch in the scrotal wall (***orchidopexy***; usually performed between 1-2 years of age).

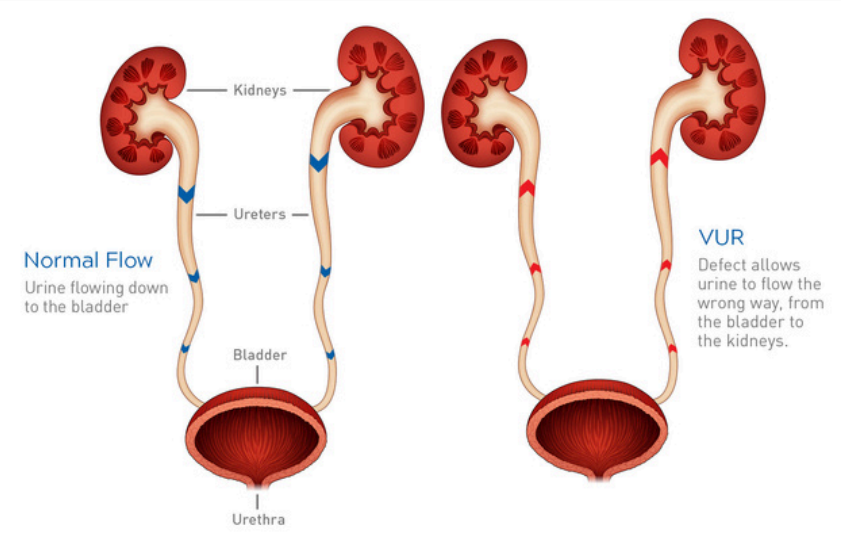
Why orchidopexy?

- To preserve fertility
- To preserve testicular growth
- To avoid malignancies, torsion &trauma
- For psychological issues for the growing child of having asingletesticle.

Vesicouretericreflux (VUR)

Definition

Results fromabnormal retrogradeflow of urinefrom the bladder into the upper urinary tract.



Epidemiology

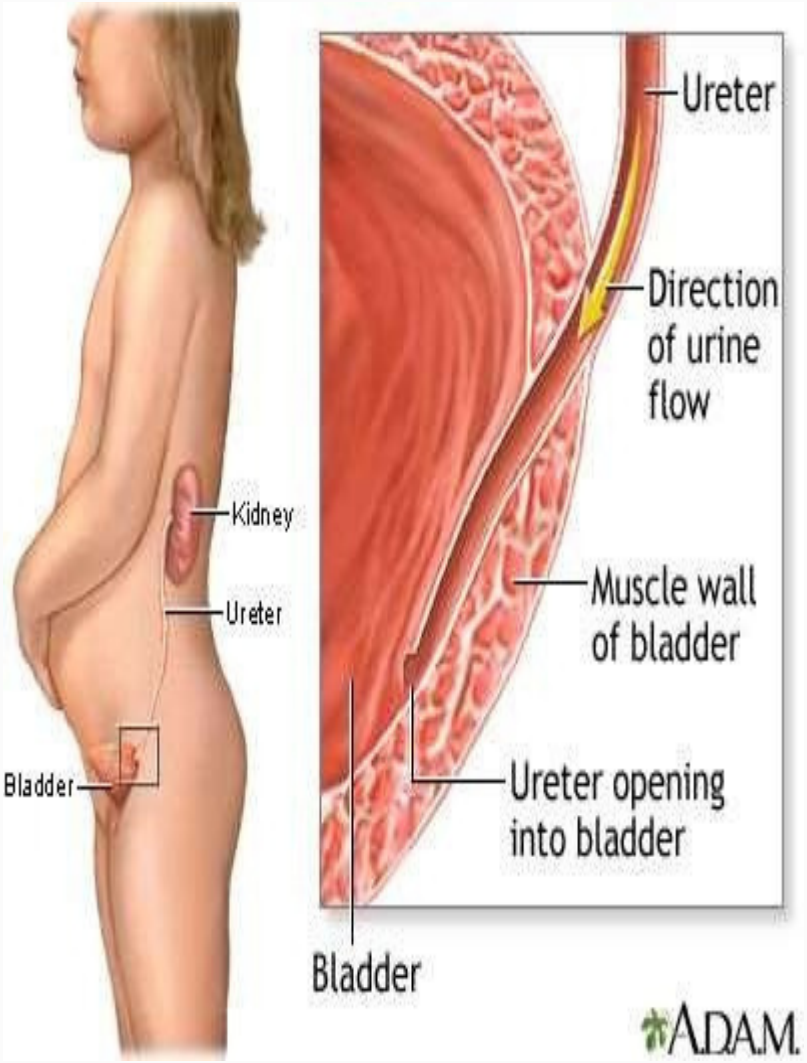
- incidence >10%;
- younger > older;
- girls > boys (5:1)
- Siblings of an affected child have

  40% risk of reflux

Pathogenesis

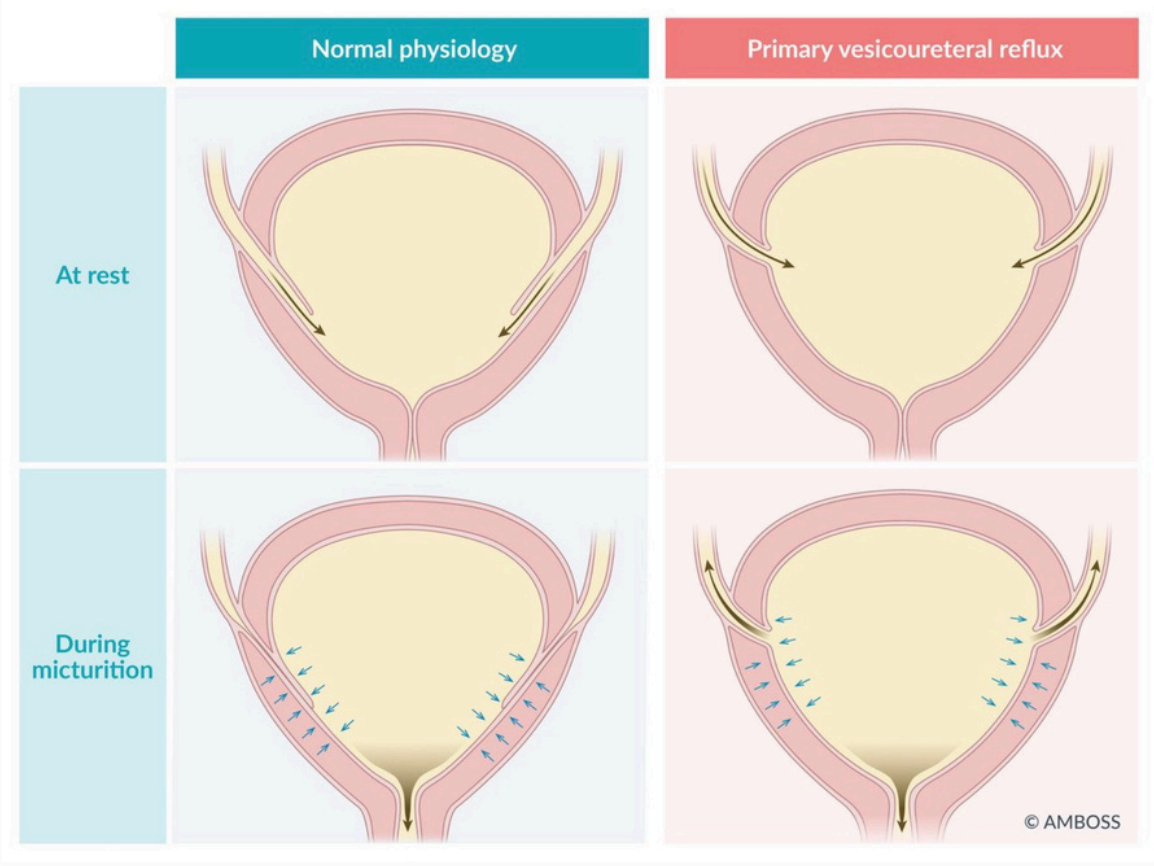
The ureter passes obliquely through the bladder wall (1–2 cm), where it is supported by muscular attachments that prevent urine reflux during bladder filling and voiding (physiological valve). The **normal ratio of intramural ureteric length to ureteric diameter is 5:1.**

Reflux occurs when the intramural length of ureter is **too short (ratio<5:1).**



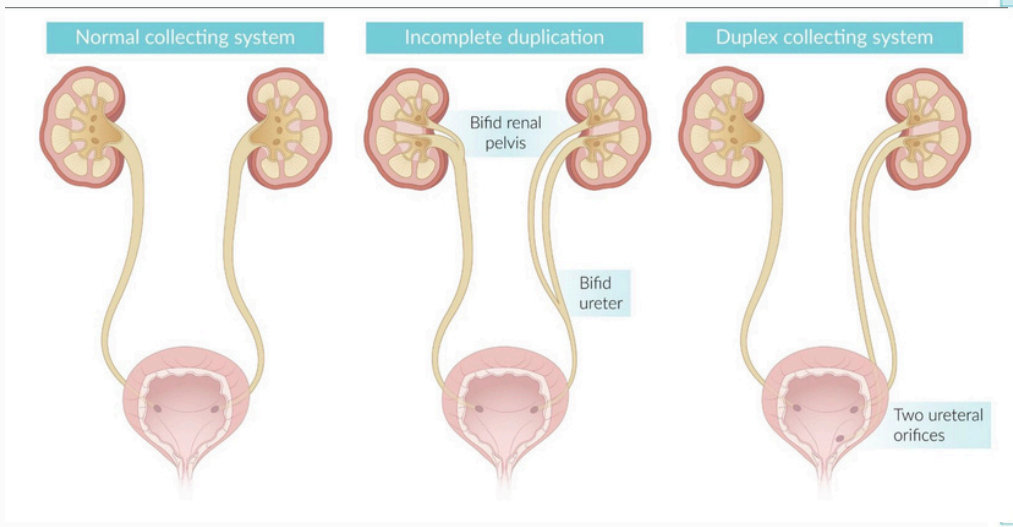
Classification:

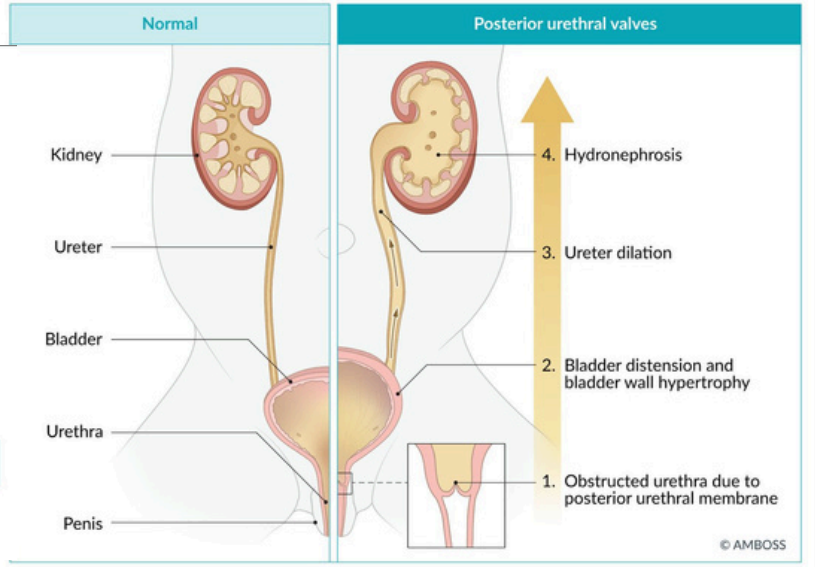
1. Primary reflux (1%) results from a congenital abnormality of the ureterovesical junction.



Classification:

2.Secondary reflux results from urinary tract dysfunction associated with elevated intravesical pressures. Causes include posterior urethral valves (reflux seen in 50%), urethral stenosis, neuropathic bladder, and detrusor sphincter dyssynergia (DSD: voiding against closed sphincter).





Complications

VUR associated with UTI can result in reflux nephropathy with hypertension and progressive renal failure.

Reflex induces Renal damage in 2 ways:

1. Secondary to reflex of infected urine and recurrent Pyelonephritis renal which induces scarring of the Renal cortex and reflex nephropathy & renal damage
2. Water hummer effect! The urine flows backwards with a high flow and causes injury in the renal collecting system.

Presentation

Patients have symptoms of UTI, fever, dysuria, suprapubicor abdominal pain, failure to thrive,vomiting, and diarrhea.

- Patients in the pediatric age group present with recurrent febrile UTIs we should rule out reflex

- Pediatrics with hydronephrosis we should rule out ut reflex

Investigation

- Urinalysis and culture to diagnose UTI
- Urinary tract ultrasound to assess condithion of the kidneysscan and VCUG/MCUG to diagnose(presence/absence) and grade
- Urodynamic assessment
- DMSA scan (nuclear scan) to detect and monitor associated renal cortical scarring

Kidney and bladder ultrasound grading

Grade I –reflux into non-dilated ureter

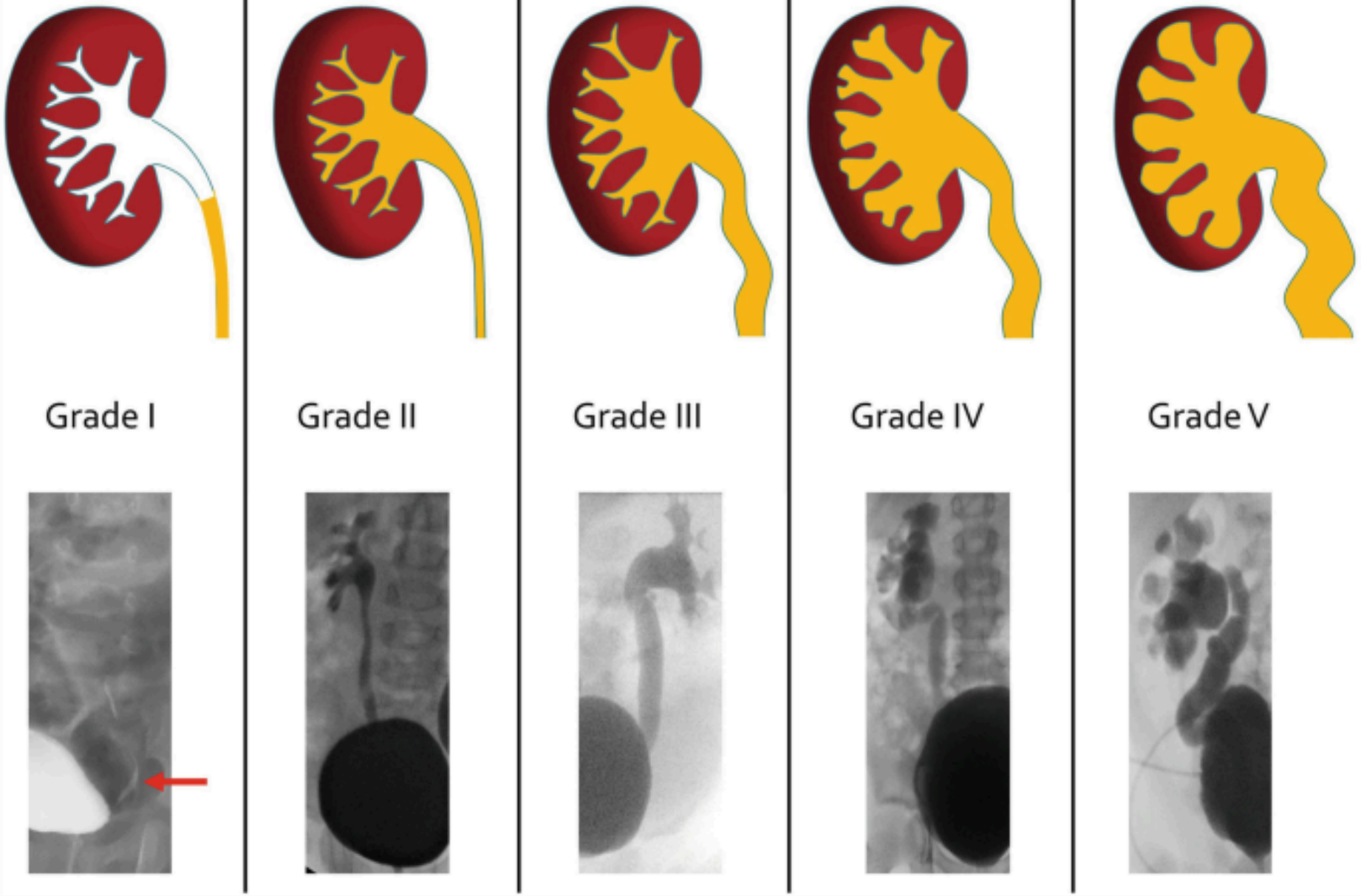
Grade II –reflux into the renal pelvis and calyces without dilatation

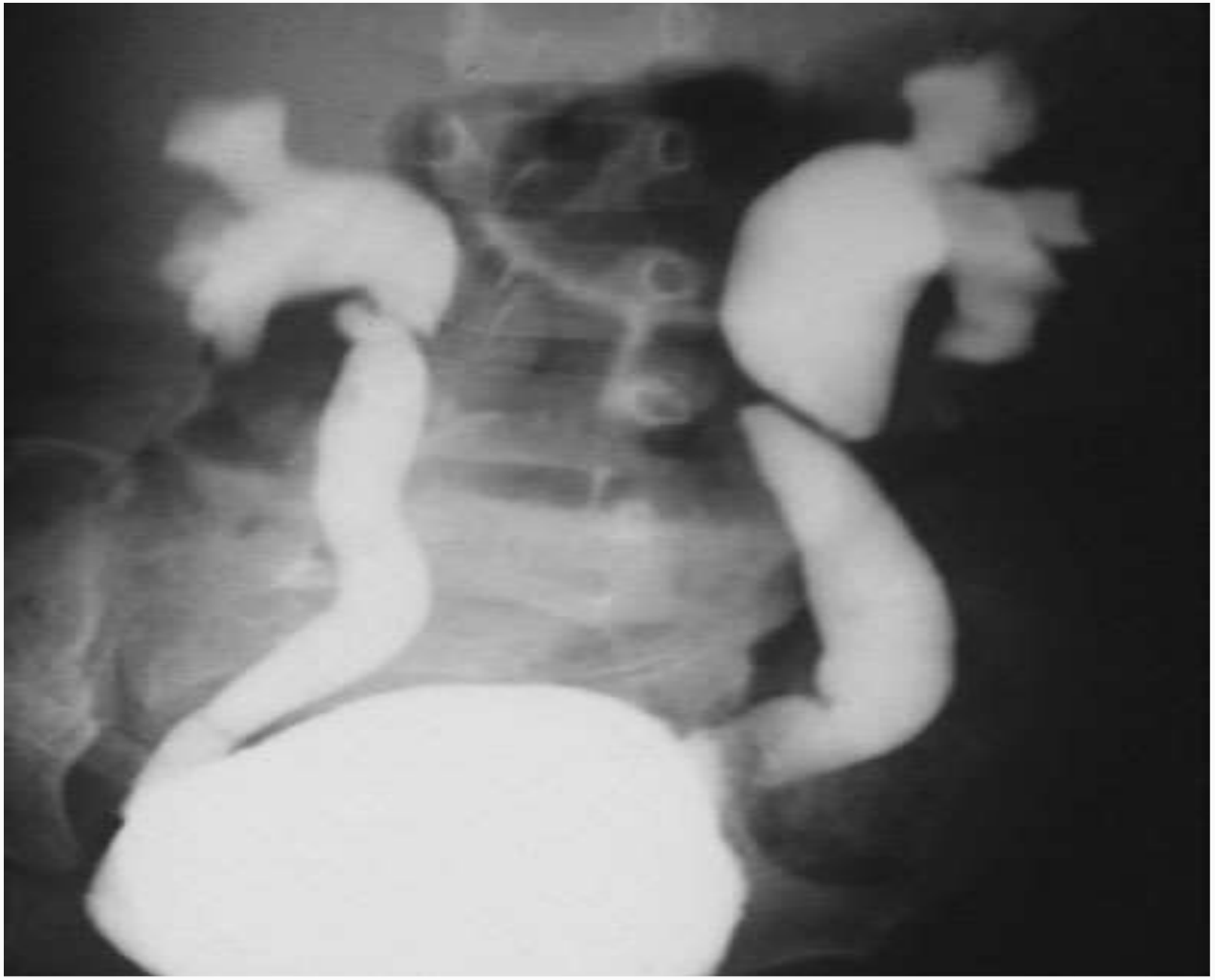
Grade III –mild/moderate dilatation of the ureter, renal pelvis and calyces with minimal blunting of the fornices

Grade IV –dilation of the renal pelvis and calyces with moderate ureteral tortuosity

Grade V –gross dilatation of the ureter, pelvis and calyces; ureteral tortuosity; loss of papillary impressions

- Grades 1,2,3 are low grade, 4 & 5 are high grade.


Grade I
Grade II
Grade III
Grade IV
Grade V



MCUG/VCUG: We inject contrast material via foley's catheter and take a series of x-rays: helpful in diagnosis & grading

Management

If we minimize the risk of infections & cystitis we reduce the risk of pyelonephritis & scarring.
If we reduce the intra-bladder pressure we lower therisk ofwater hammereffect andrenalinjury.

Correct problems contributing to secondary reflux. Most primary VUR grade I–II cases will resolve spontaneously (~85%), with 50% resolution in grade III. Observation and medical treatment are initially recommended.

Medical treatment

Conservative treatment:

Low-dose antibiotic prophylaxis should be given to keep the urine sterile and lower the risk of renal damage until reflux resolves.

Anticholinergic drugs are given to treat bladder overactivity

Surgical Management

- Our main goal is to maintain the:1 ratio!
- (ureteroneocystostomy± ureteroplasty (reimplanation surgeries)) or subureteralinjection

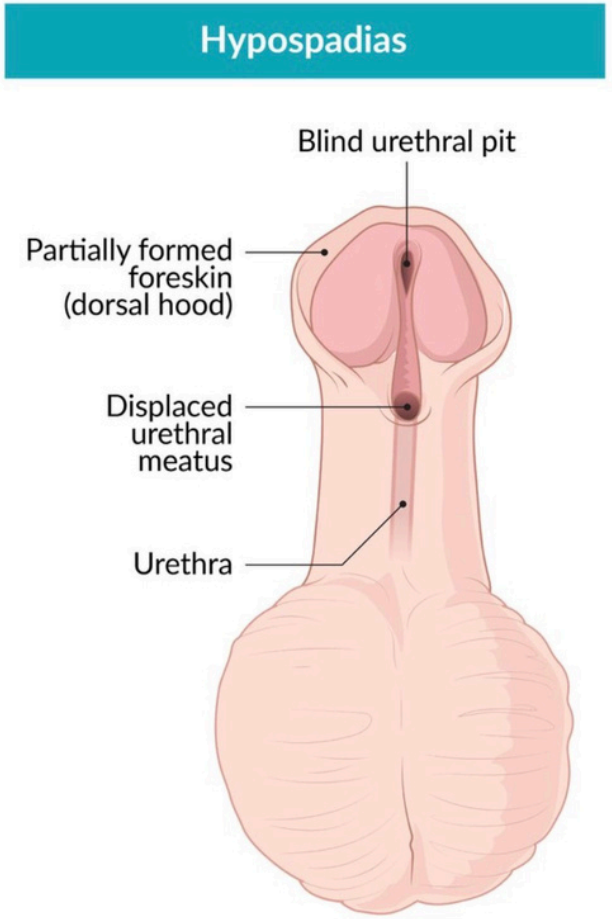
- Indication for surgery:
- If it is not possible to keep the urine sterile and reflux persists
- If acute pyelonephritis recurs despite a strict medical regimen and chronic suppressive antimicrobial therapy.
- If increased renal damage is demonstrated by serial excretory
- urogramsor nuclear scan.
- High grade reflux (grade IV or V -not an absolute indication)

If we fail to keep the pressure in the bladder low

Hypospadias

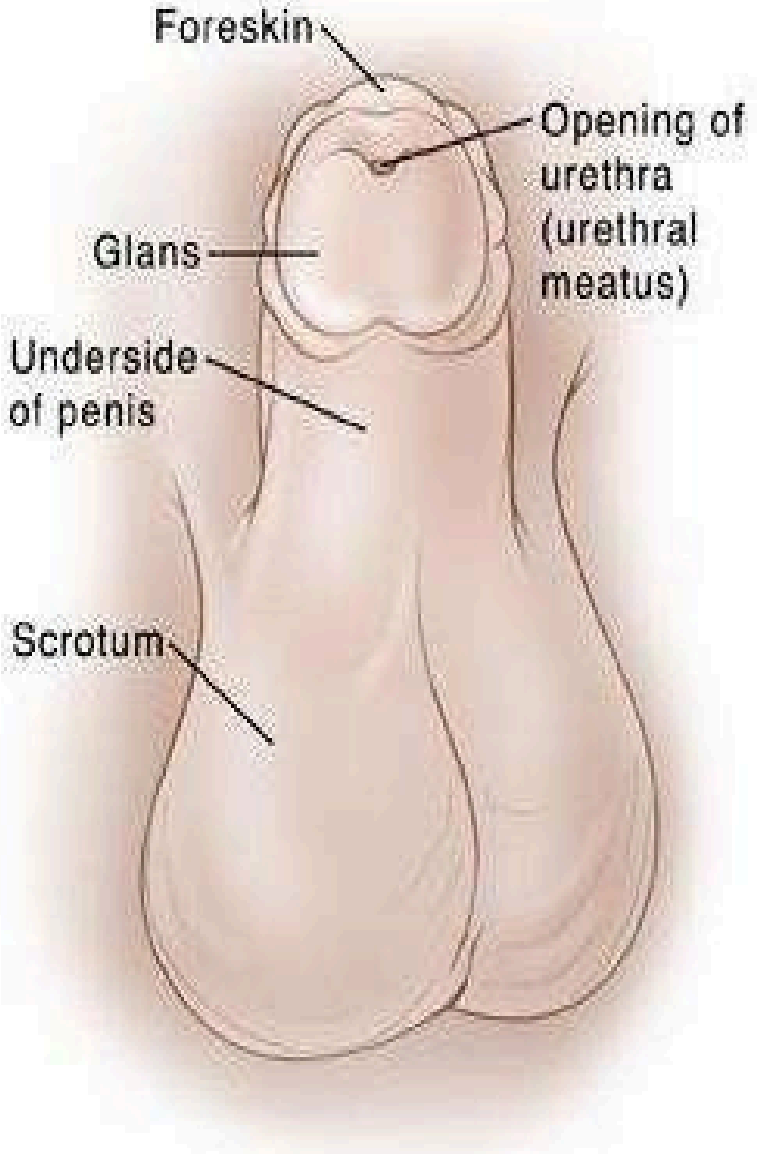
Hypospadias is a congenital deformity in which the opening of the urethra (the meatus) occurs on the underside (ventral) part of the penis, anywhere from the glans to the perineum. It is often associated with a **hooded foreskin** and **chordee** (ventral curvature of the penile shaft).

- It is the **most common congenital malformation of the urethra**. It occurs in 1 in 250 live male births

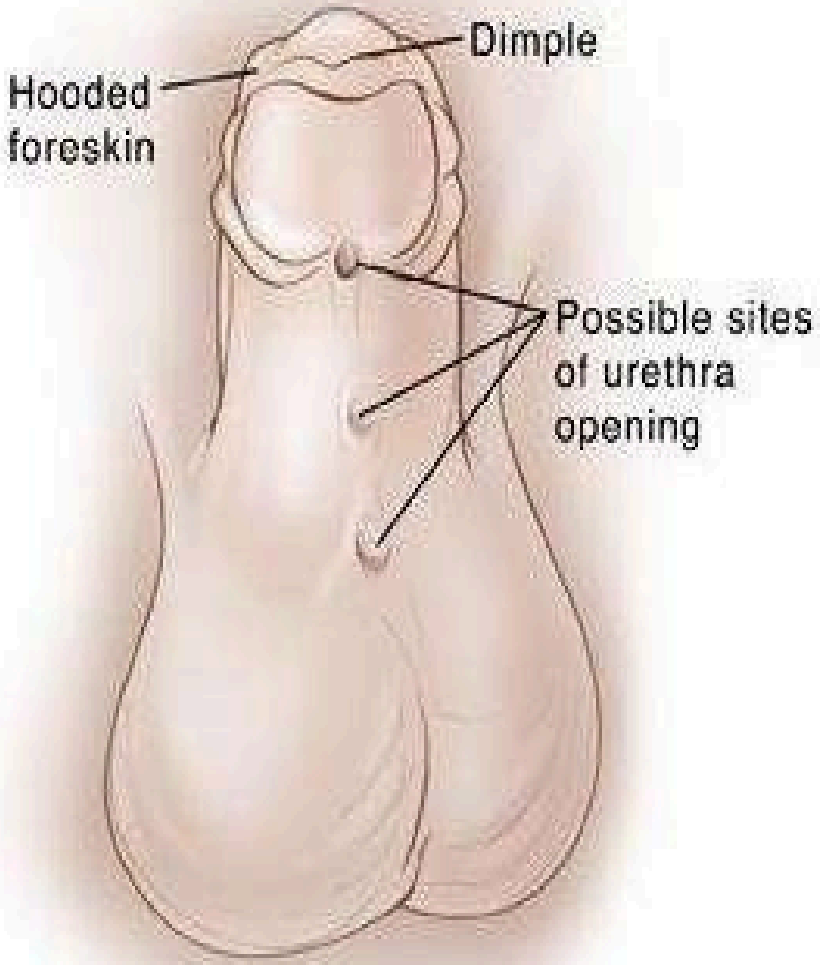


5 Associations with hypospidias:

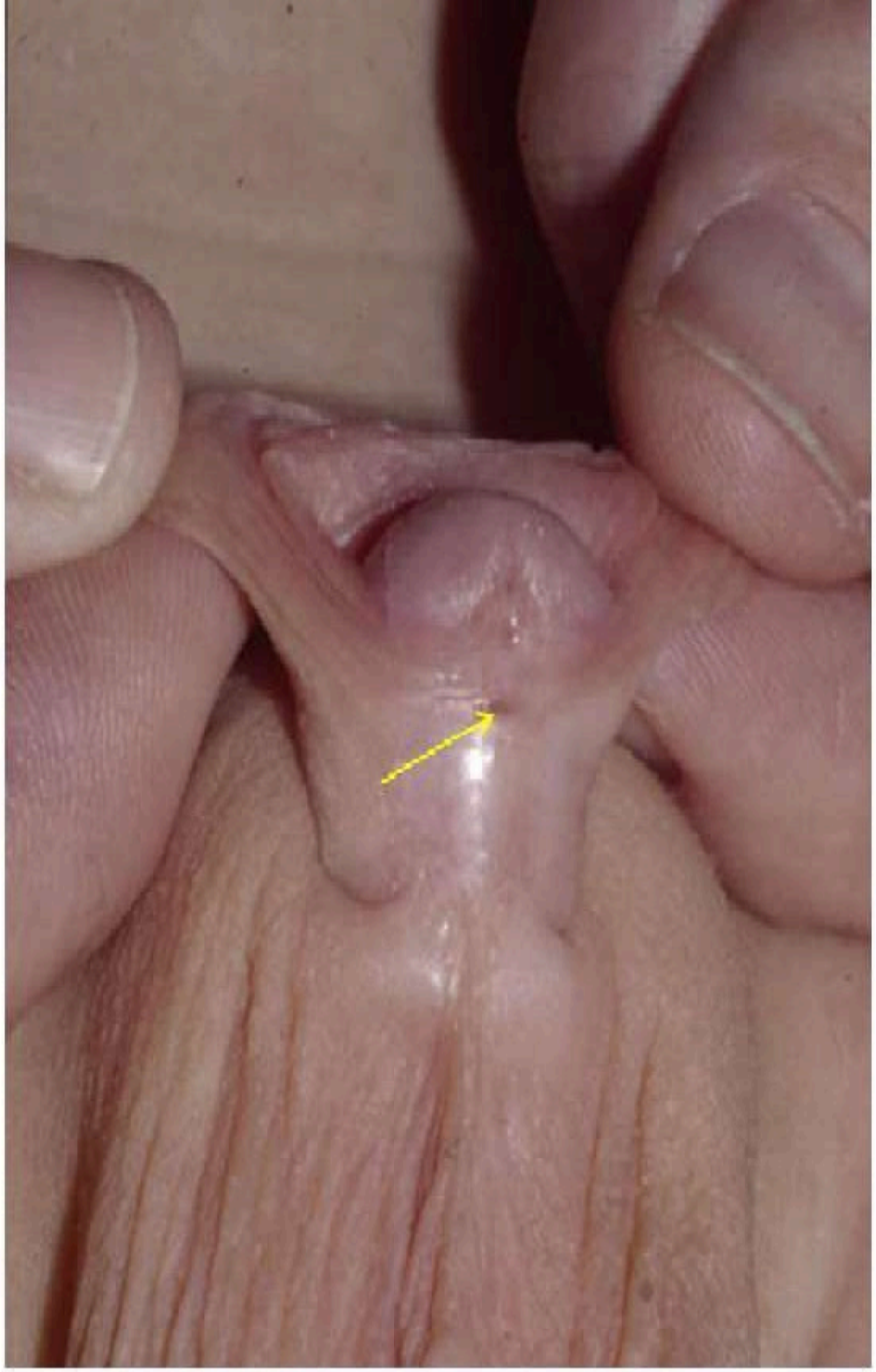
1. Ventrally located meatus
2. Absence of ventral foreskin
3. Penile chordee(ventral curvature of the penile shaft).
4. Absence of the penile ventral skin
5. Deviation of the median raphe to one side of the penis

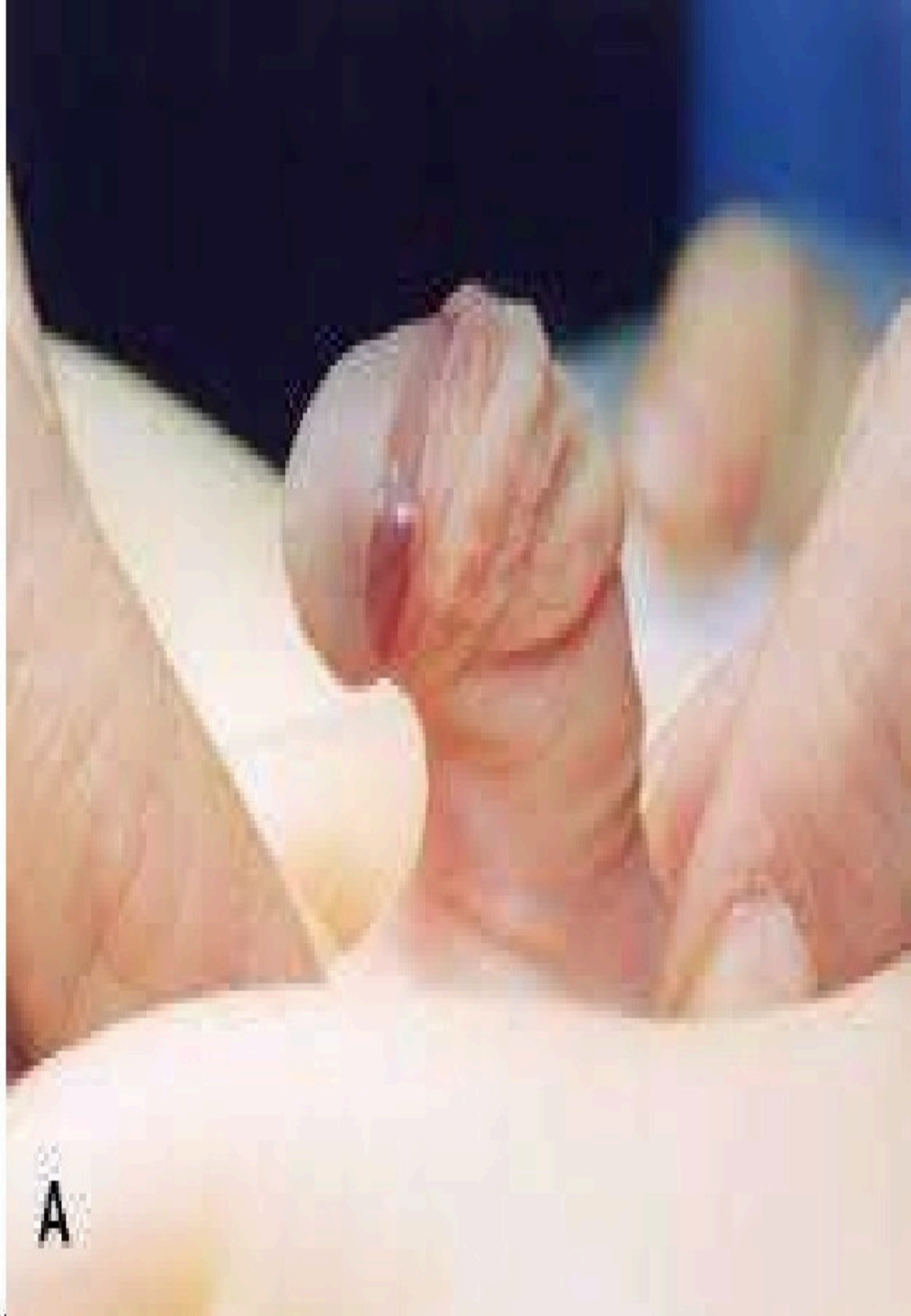


Normally, the opening of the urethral meatus is located at the tip of the penis.



With hypospadias, the opening of the urethra is located on the underside of the penis or near the scrotum.

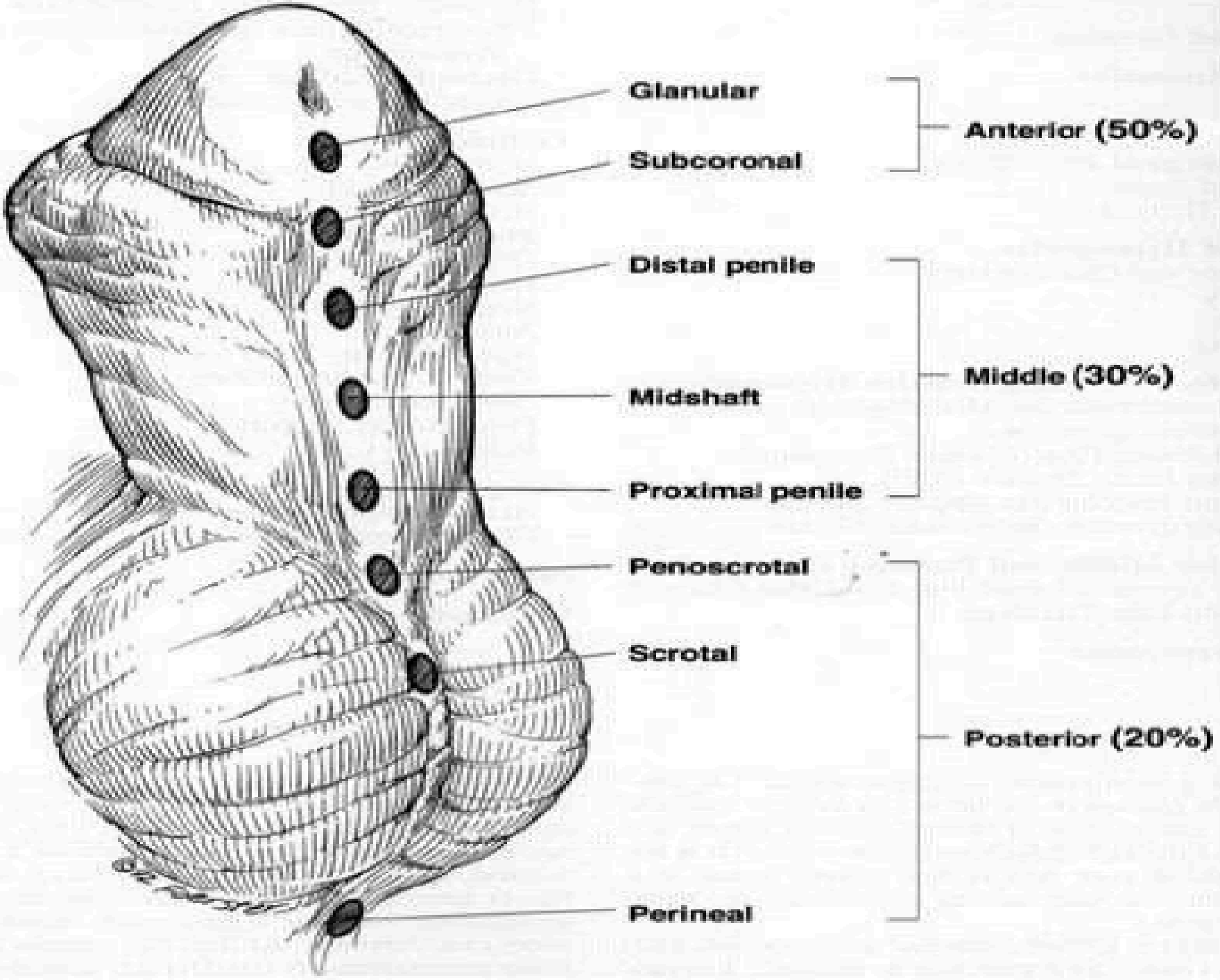


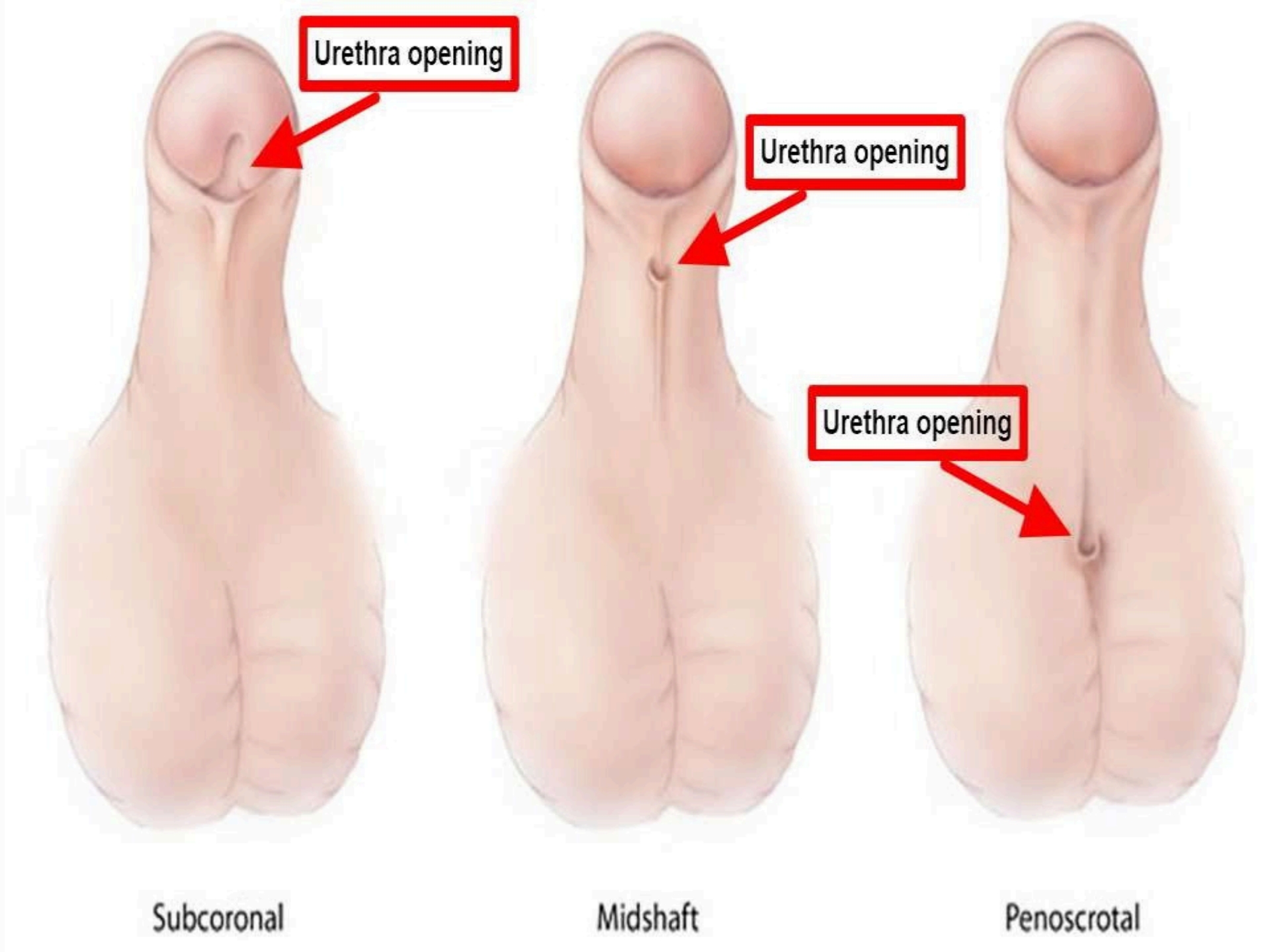

A

Classification

- Hypospadias can be classified according to the anatomical location of the urethral meatus
- Anterior (or distal)—glanular, coronal, and subcoronal(~50%) mild, does not affect fertility
- Middle—distal penile, midshaft, and proximal penile (~30%) severe
- Posterior (or proximal)—penoscrotal, scrotal, and perineal (~20%) severe, affects fertility, seminal ejection won't be in the vagina

The more proximal, the more severe.

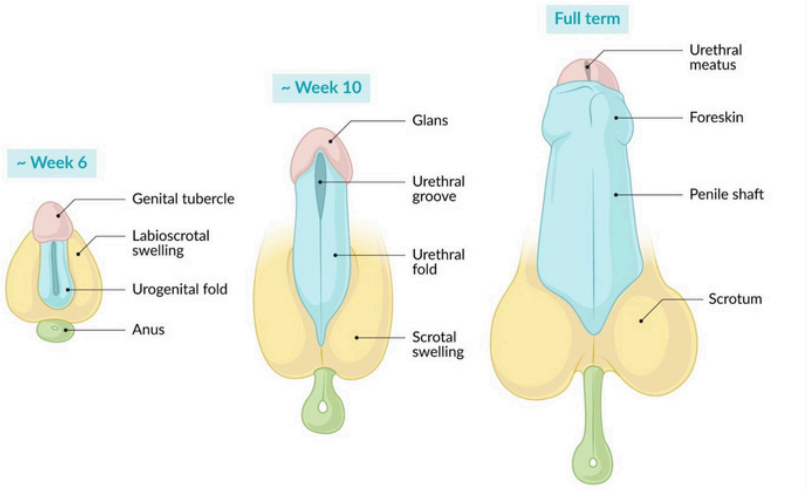

Glanular
Subcoronal
Distal penile
Midshaft
Proximal penile
Penoscrotal
Scrotal
Perineal
Anterior (50%)
Middle (30%)
Posterior (20%)


Urethra opening
Urethra opening
Urethra opening
Subcoronal
Midshaft
Penoscrotal

Etiology

- Hypospadias results from incomplete closure of urethral folds on the underside of the penis during embryological development.

- This is related to a defect in production or metabolism of fetal androgens, or the number and sensitivity of androgen receptors in the tissues.



Diagnosis

- A full clinical examination will make the diagnosis.

  However,itis also important to seek out associated abnormalities that will need treatment (undescended testes, inguinal hernias, and hydroceles).
- Patients with absent testes and severe hypospadias should undergo chromosomal and endocrine investigation to exclude intersex conditions

- After diagnosis you have to inform the family that:in the case of distal or mild; circumcision should bed one at the time-of the the hypospidias surgery which is done at 1 year of age, this condition will not increase the incidence of Uris and will not affect the fertility.
- In proximal hypospidias; it is rare! And you should refer the patient to a pediatric urologist.

Treatment

- No matter the severity surgery is the mode of treatment (cosmetically & functionally).

- Surgery is indicated where deformity is severe, interferes with voiding, OR is predicted to interfere with sexual function. Surgery is now performed between 6 and 12 months of age.

- Local application of testosterone for 1 months preoperatively can help increase tissue size.

- The aim of surgery is to correct penile curvature (orthoplasty), reconstruct a new urethra, and bring the new meatus to the tip of the glans using urethroplasy, glanuloplasty, and meatoplasty techniques.

Complications

- These include bleeding, infection, urethral strictures, meatal stenosis,urethrocutaneous fistula,urethraldiverticulum,andfailed procedures requiring reoperation

Nocturnal enuresis

- **Enuresis :** is the involuntary, intermittent voiding of urine, occurring at an inappropriate time or place, in the absence of organic disease.

- **Nocturnal enuresis (bed wetting) :** repeated voiding of urine during sleep in an individual with a developmental age $\geqslant$ 5 years.

- **Prevalence of Nocturnal Enuresis :**
  - **At 5 years: ~ 15–20%**
  - **At 10 years: ~ 5%**
  - **At 15 years: ~ 1–2%**

• What are the types of nocturnal enuresis?

- There are two main types of bedwetting:

- Primary nocturnal enuresis occurs when a person has never remained dry throughout the night for six months in a row or longer.

- Secondary nocturnal enuresis occurs when a person wets the bed again after not wetting the bed for six months or more.
- Secondary enuresis is usually the result of a medical or psychological condition.

• Etiology:

- Familial
- Delay in functional bladder maturation
- Altered antidiuretic hormone (ADH) secretion; abnormal decrease in ADH levels at night causes increased urine production (nocturnal polyuria)
- Altered sleep/arousal mechanism
- Psychological factors
- UTI (1% of cases)

Evaluation of Nocturnal Enuresis

1. History:

- Frequency of episodes
- Daytime symptoms
- New or recurrent bed-wetting
- Family history of enuresis
- Urinary tract infections (UTIs)
- Bowel problems / constipation
- Psychosocial history (stress, trauma, school issues)

2. Physical Examination

- Focus on excluding organic causes
- Look for neurological disease, spinal anomalies, or signs of systemic illness

3. Investigations

- Urinalysis
  - Detect infection
  - Check specific gravity (reduced in nocturnal polyuria)
  - Glucose / protein
- Voiding diary
  - Track urine output and frequency

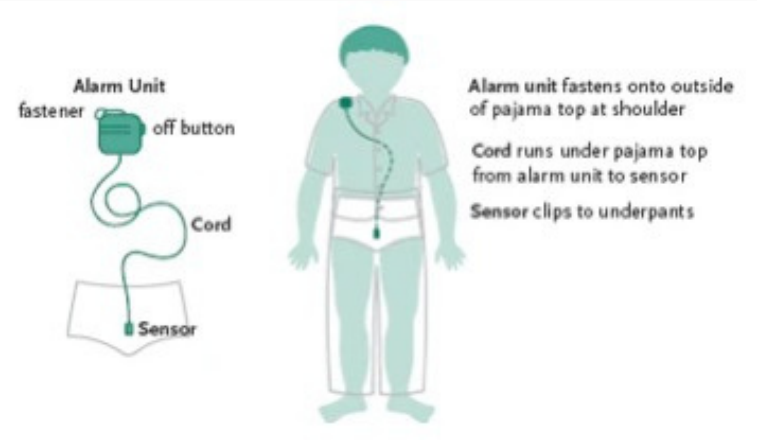
Management and Treatment

- **How is bedwetting (nocturnal enuresis) treated?**

- Treatment for nocturnal enuresis varies based on the cause. Treatment options could include:

- **Behavioral changes :**

- Provide reassurance
- Bladder training
- Motivational techniques to improve self-esteem
- Conditioning therapy (alarm connected to underwear, which is triggered with the first few drops of urine).



- **Pharmacological:**

- **Imipramine**
  - Tricyclic antidepressant with anticholinergic & antispasmodic properties
  - Not commonly used

- **DDAVP (Desmopressin)**
  - Synthetic analogue of ADH
  - Administered intranasally or orally

- Prognosis
- 15% of patients have spontaneous resolution of symptoms per year.

Posterior urethral valves (PUV)

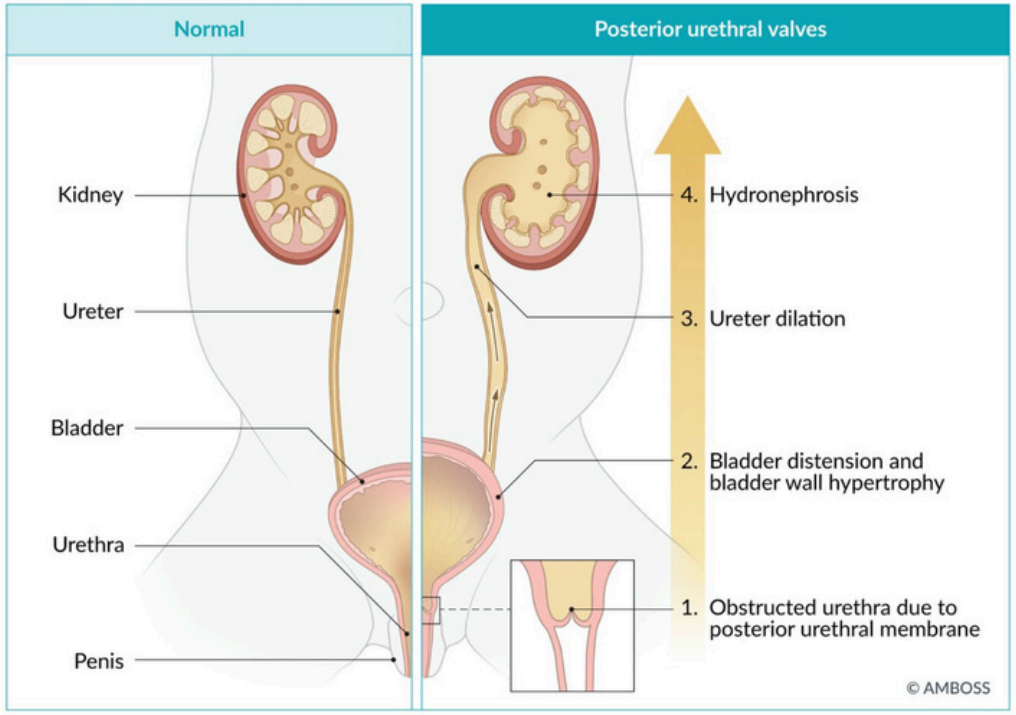
Definition:

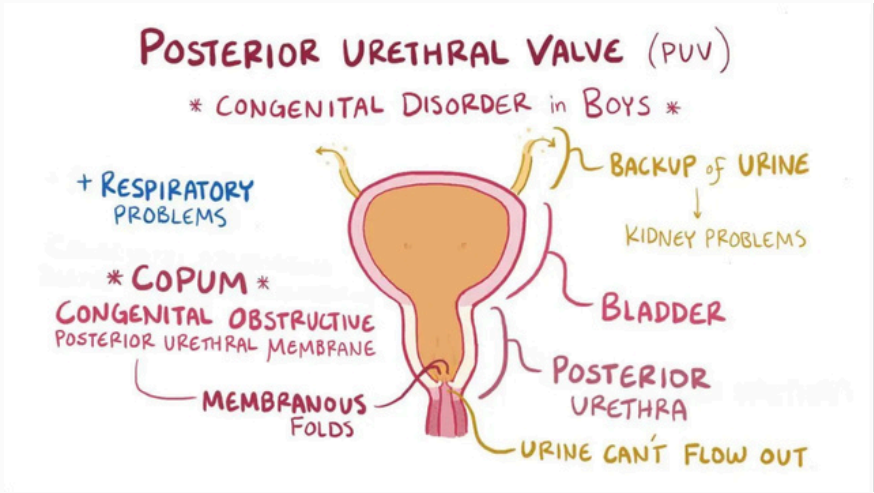
- congenital malformation in males where membranous folds of the urogenital membrane obstruct the membranous and prostatic urethra (posterior urethra)
- Occurs only in males
- Most common cause of lower urinary tract obstruction in newborns

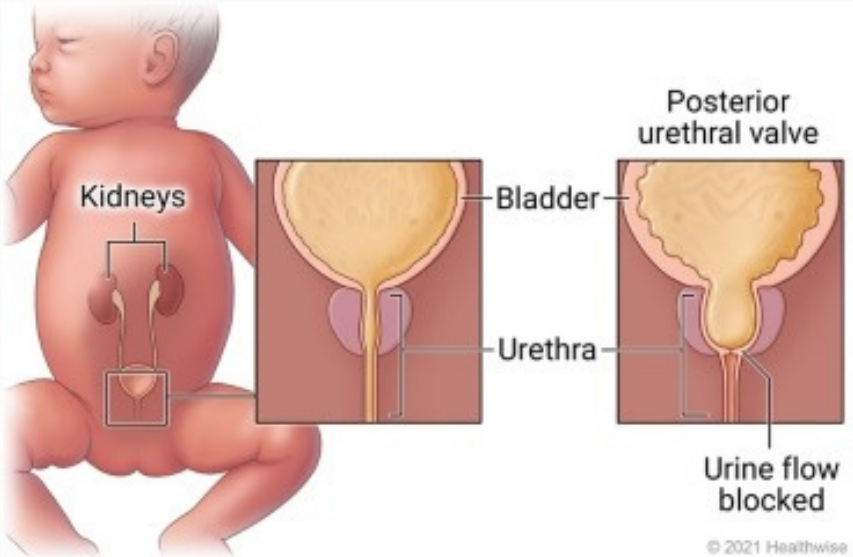
Causes:

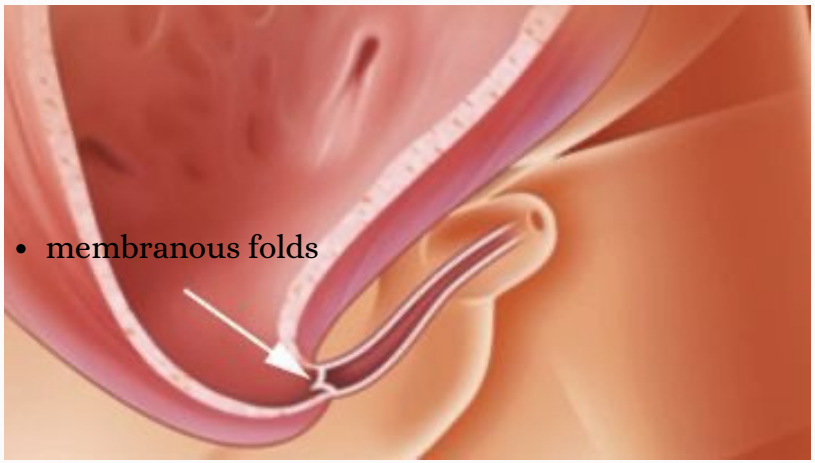
- **Normal anatomy:**
- Male urethra has small paired lateral folds (plicae colliculi)
- Located between the lateral distal edge of verumontanum and lateral urethral wall

- **PUV mechanism:**
- Congenital overgrowth of these folds
- Due to abnormal insertion of Wolffian ducts into the posterior urethra during fetal development


Normal
Posterior urethral valves
Kidney
Ureter
Bladder
Urethra
Penis
4. Hydronephrosis
3. Ureter dilation
2. Bladder distension and bladder wall hypertrophy
1. Obstructed urethra due to posterior urethral membrane
© AMBOSS


POSTERIOR URETHRAL VALVE (PUV)
* CONGENITAL DISORDER in BOYS *
+ RESPIRATORY PROBLEMS
BACKUP of URINE
KIDNEY PROBLEMS
* COPUM *
CONGENITAL OBSTRUCTIVE POSTERIOR URETHRAL MEMBRANE
BLADDER
POSTERIOR URETHRA
MEMBRANOUS FOLDS
URINE CAN'T FLOW OUT


Kidneys
Bladder
Urethra
Posterior urethral valve
Urine flow blocked
© 2021 Healthwise

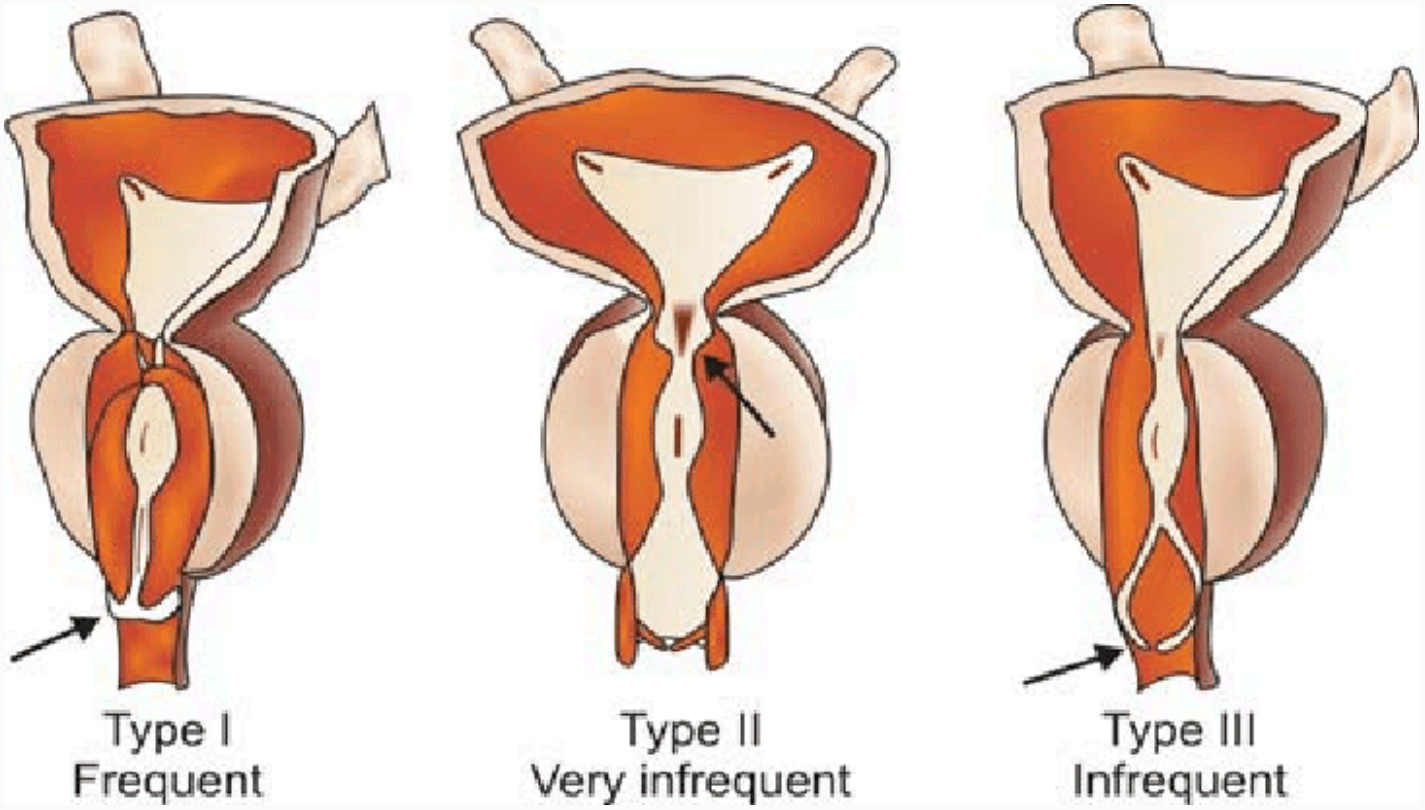

• membranous folds

Classification of Posterior Urethral Valves (PUV):

- Landmark: Verumontanum ("mountain ridge") in the prostatic urethra

Types:

- **Type I (Most common)**
  - Anterior fusion of plicae colliculi
  - Mucosal fins extend from the bottom of verumontanum distally along prostatic and membranous urethra
- **Type II (Least common)**
  - Vertical / longitudinal folds between verumontanum and proximal prostatic urethra / bladder neck
- **Type III (Less common)**
  - Disc of tissue distal to verumontanum
  - Theorized as developmental anomaly of congenital urogenital remnants in bulbar urethra


Type I
Frequent
Type II
Very infrequent
Type III
Infrequent

Presentation of Posterior Urethral Valve (PUV)

Prenatal (Ultrasound features)

- Bilateral hydroureteronephrosis
- Dilated bladder with elongated ectatic posterior urethra
- Thick-walled bladder
- Oligohydramnios (↓ amniotic fluid)
- Renal dysplasia

Early severe findings → poor prognosis

Newborns & Infants

- Respiratory distress (pulmonary hypoplasia)
- Palpable abdominal mass (hydronephrotic kidney / distended bladder)
- Ascites
- Recurrent UTI
- Electrolyte abnormalities
- Failure to thrive

Older Children (milder cases)

- Recurrent UTI
- Poor urinary stream
- Incomplete bladder emptying
- Poor growth
- Incontinence

- <u>Risk of:</u>
  - Renal failure
  - Vesicoureteric reflux (VUR)
  - Voiding dysfunction
    - Overactive or underactive bladder
    - Valve bladder syndrome

Investigations of Posterior Urethral Valve (PUV)

- **Ultrasound (KUB)**
  - Kidneys and bladder
  - Detects hydronephrosis, thick-walled bladder
- **VCUG (Voiding Cystourethrogram)** ★
  - Shows distended, elongated posterior urethra
  - Partially filled anterior urethra
  - Bladder neck hypertrophy
  - Gold standard for diagnosing PUV
- **Isotope Renal Scan (MAG-3, DMSA)**
  - Assesses renal function and drainage
- **Video Urodynamics**
  - Diagnoses associated voiding dysfunction
  - Assesses urethra and bladder neck

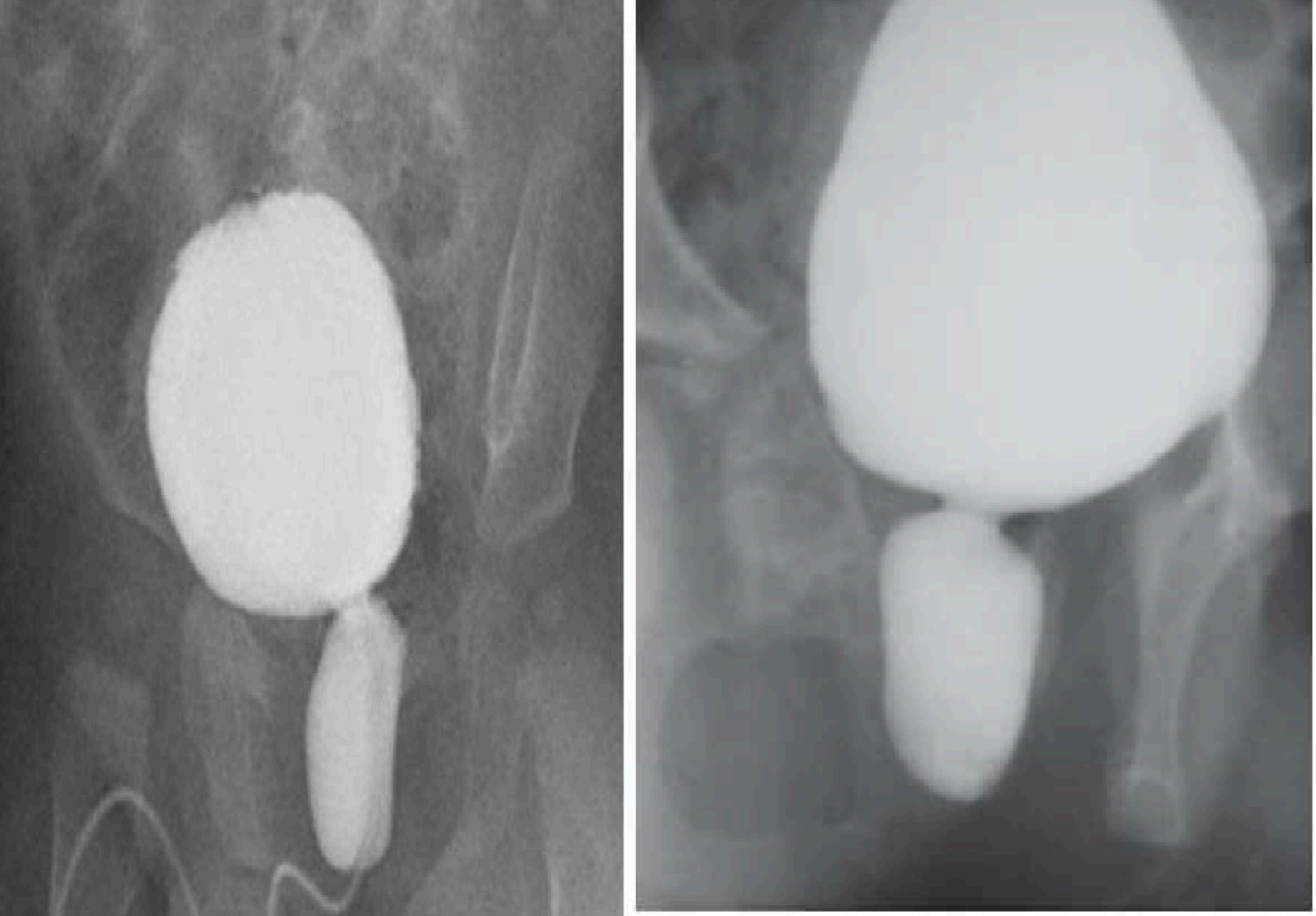


Figure 15.9 VCUG in infant with posterior urethral valves shows dilated elongated prostatic urethra and thickened bladder neck.

Treatment of Posterior Urethral Valve (PUV)

Initial Management

- Start prophylactic antibiotics
- Check serum electrolytes
- Bladder drainage with pediatric feeding tube

Definitive Management (if improves)

- Cystoscopy + transurethral valve ablation
  - Incisions at 5 & 7 o'clock using electrocautery
  - Possible complication: urethral stricture

If No Improvement After Drainage

(Upper tracts still dilated + raised creatinine)

- Temporary cutaneous vesicostomy
  - Stoma between bladder dome & suprapubic abdominal wall
  - Allows free urine drainage
- Alternative: Ureterostomy drainage

Later

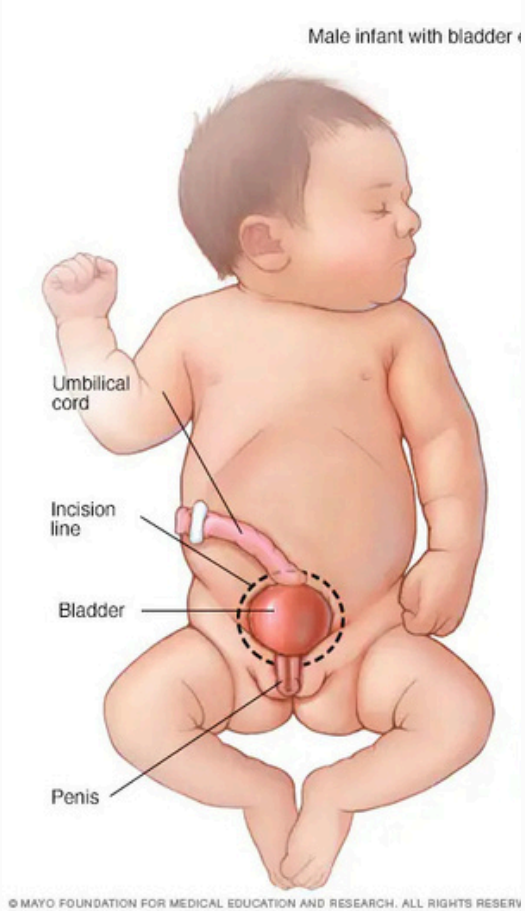
- Definitive valve ablation once stable

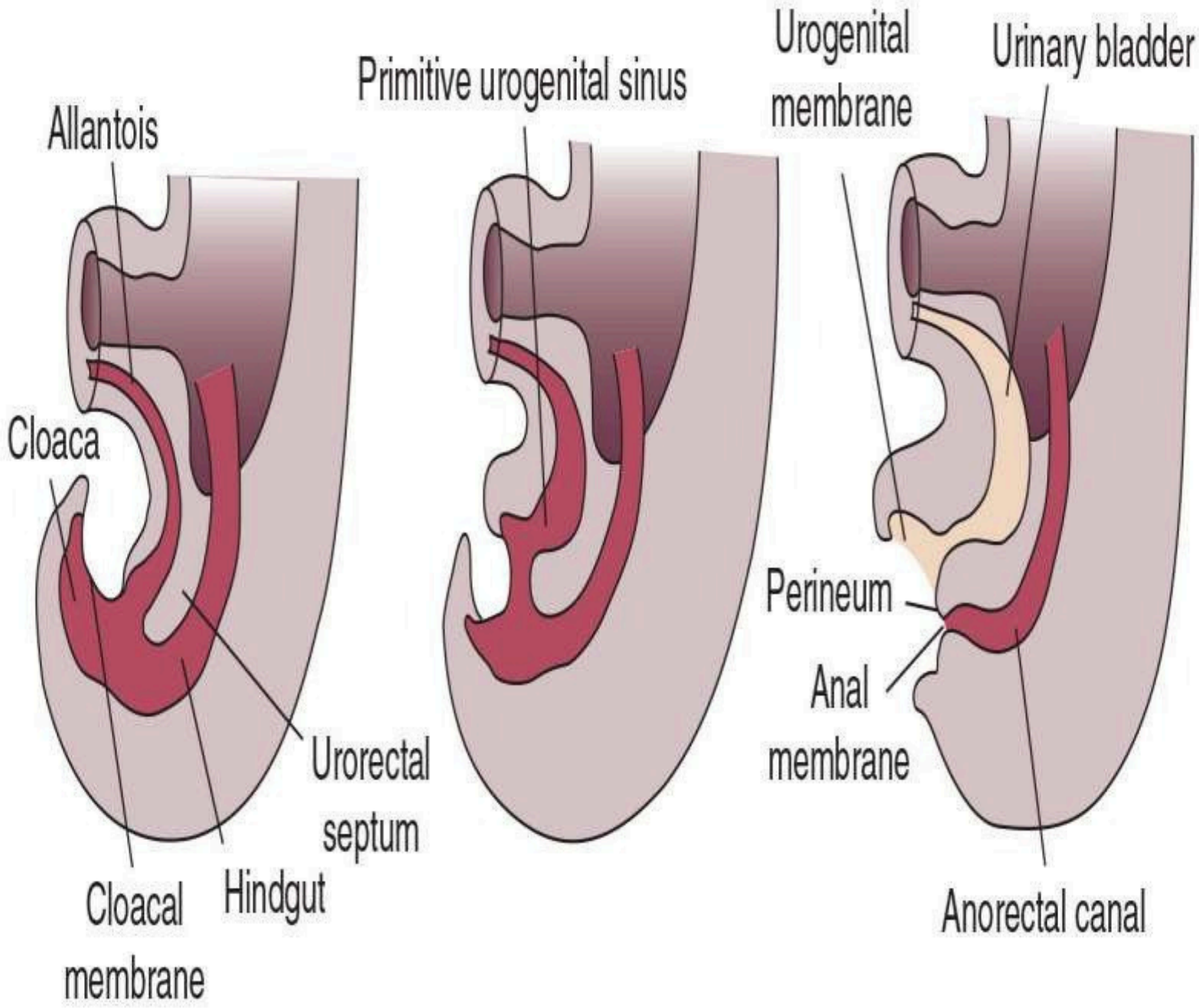
Exstrophy of the Bladder

- Definition:
  - Defective development of anterior bladder and lower abdominal walls
  - Posterior bladder wall exposed on the abdomen

- Epidemiology:
  - Male > Female (>2:1)

- Risk factors:
  - Positive family history
  - Younger maternal age
  - Increased parity




Allantois
Cloaca
Urorectal septum
Cloacal membrane
Hindgut
Primitive urogenital sinus
Urogenital membrane
Urinary bladder
Perineum
Anal membrane
Anorectal canal

- **Embryology of Bladder Exstrophy**

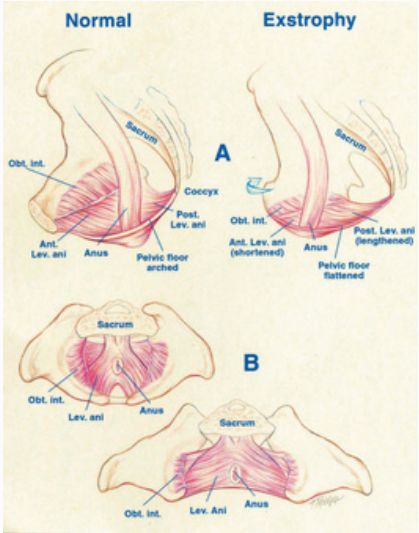
- Cause:

- Embryological malformation → abnormal overdevelopment of cloacal membrane
- Prevents in-growth of lower abdominal tissues

- Normal: Cloacal membrane perforates → forms urogenital and anal openings

- In Exstrophy: Premature rupture → triangular defect below umbilicus

- Timing of rupture determines type of defect:
  - Bladder exstrophy
  - Cloacal exstrophy
  - Epispadias



Associated Anomalies in Bladder Exstrophy

- Urinary tract defects:
  - Vesicoureteral reflux (VUR)
- Bone defects:
  - Widening of the pubic symphysis

- Genital defects:
  - Epispadias

- Musculofascial defects:
  - Inguinal hernia
  - Femoral hernia

- Investigation – Bladder Exstrophy (Prenatal US)

- Typical prenatal ultrasound features:

- Lower abdominal wall mass
- Absent bladder filling
- Low-set umbilicus
- Small genitalia
- Abnormal widening of iliac crests

Management of Bladder Exstrophy

At Birth

- Cover exposed bladder with plastic film
- Regular irrigation with sterile saline
- Urgent referral to a tertiary center

Chronic mucosal exposure → squamous metaplasia, cystitis cystica, adenocarcinoma, SCC

Definitive Surgical Repair

- Selected cases: one-stage repair
- Most patients: three-stage reconstruction

Three-Stage Repair

1) Newborn

- Pelvic osteotomy + external fixation
- Closure of bladder, abdominal wall & posterior urethra

2) 6–12 months

- Epispadias repair

3) 4–5 years

- Bladder neck reconstruction (Young–Dees–Leadbetter)
- Anti-reflux surgery (ureteric reimplantation)
- Performed when bladder capacity is adequate and child can void

If Bladder Capacity is Inadequate

- Bladder augmentation
- Or urinary diversion

Prognosis of Bladder Exstrophy

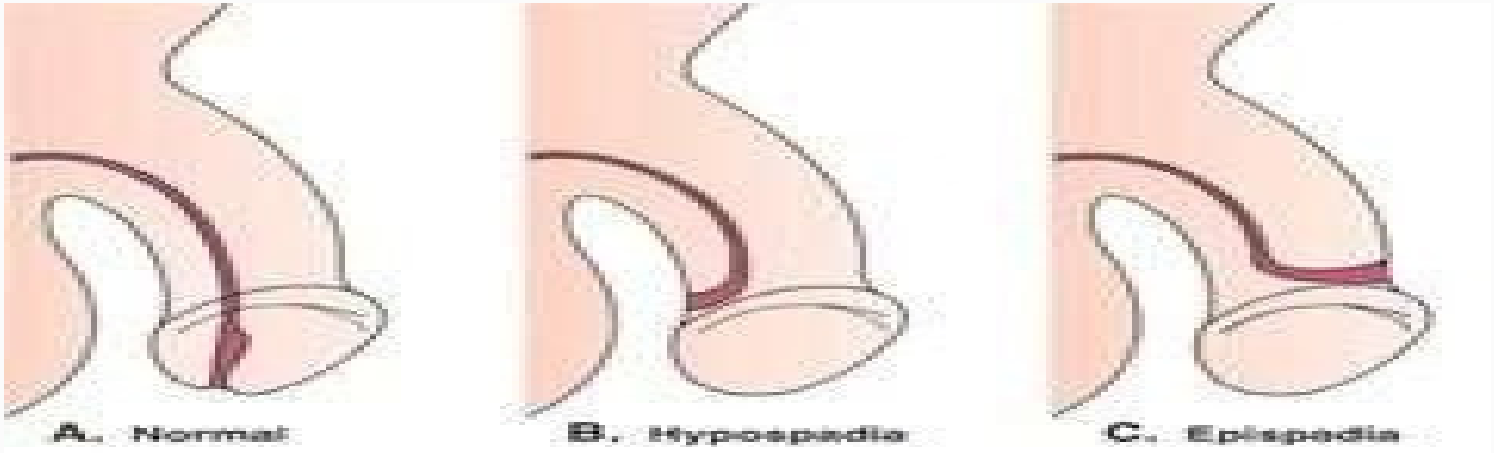
- **Even after successful surgery, long-term complications may include:**
  - Incontinence
  - Urinary reflux
  - Recurrent UTIs
  - Bladder adenocarcinoma
  - Colonic adenocarcinoma
  - Uterine prolapse
- Sexual function and libido are usually normal in exstrophy patients

Epispadias

- The urethraopeninthedorsalsurfaceof the penis any wherefromthe glans,penile shaftormostcommonlythe penopubicregion . Extremely rare.
- Etiology represents failure of closure of the cloacal membrane, resulting in the bladder and urethra opening directly through the abdominal wall

•M:F … 5:1

•High morbidity -+ incontinence, infertility, reflux.



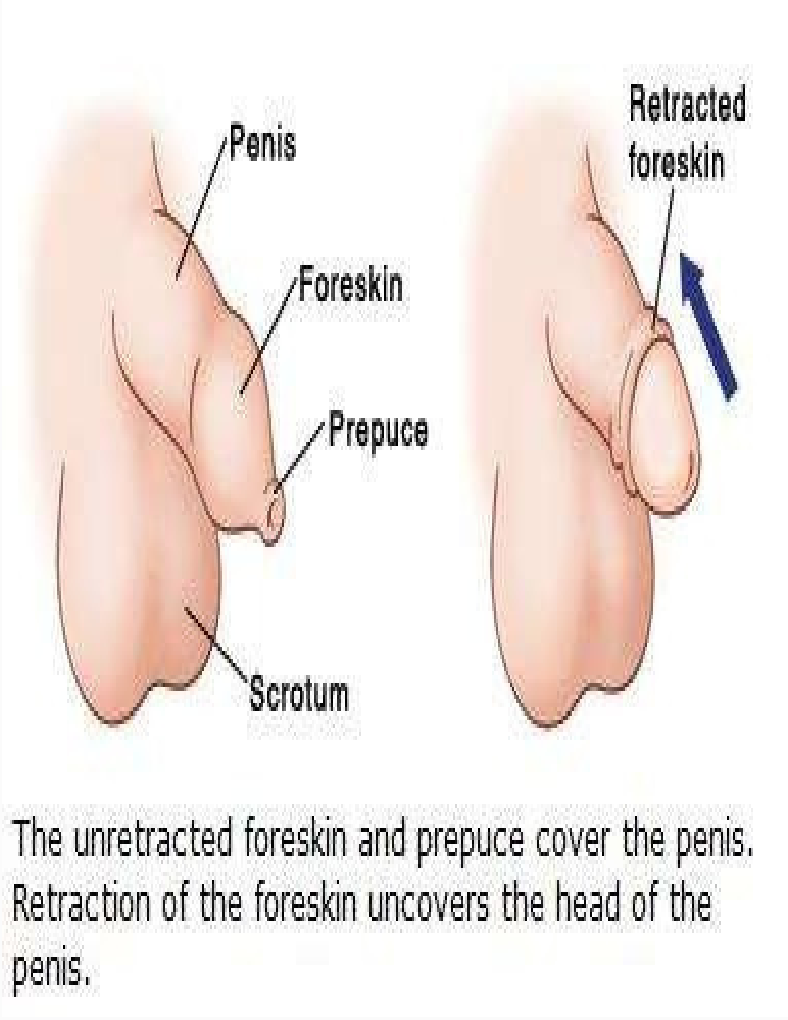
Management same as hypospidias

- at6–12months >>Thisinvolvesurethroplastywith functional and cosmetic reconstruction of the external genitalia (penile lengthening and correction of chordee) .
- The modified Cantwell–Ransleytechnique is commonly used in males. It describes mobilizing the urethra to the ventral aspect of the penis, with advancement of the urethral meatus onto the glans with a reverse MAGPI (meatal advancement-glanuloplasty) .
- From age 4–5 years>> when children can be toilet trained, bladder neck reconstruction can be performed (Young–Dees–Leadbetterprocedure). This achieves continence, and any bladder residuals may then be emptied by urethral catheterization.

Phimosis

Is when the foreskin cannot be retractedbehind the glans. Which can lead to recurrentsUTIs due to collection of bacteria underneath the foreskin, and ballooning of the foreskin during voiding.

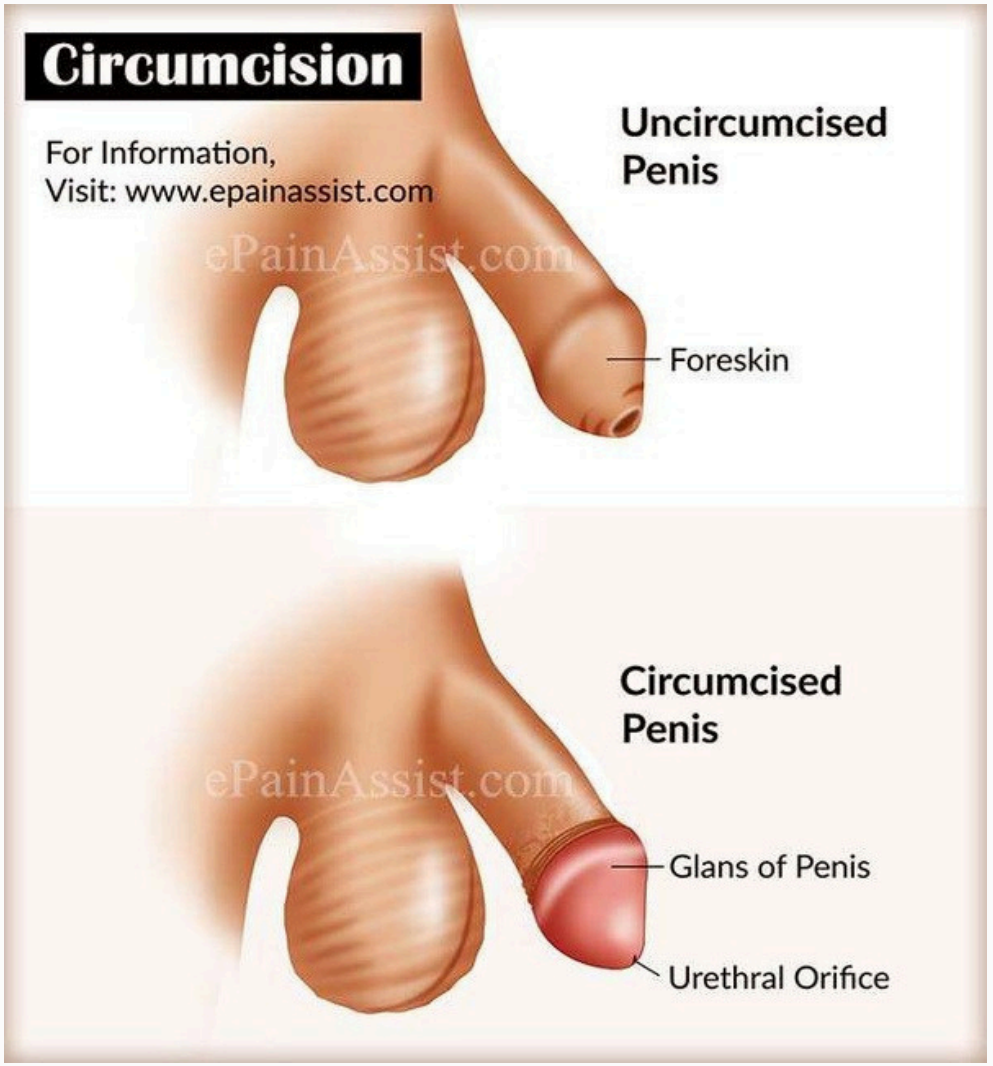
- A physiological phimosisis present at birth due to adhesions between the foreskin and glans.
- As the penis develops, epithelial debris (smegma) accumulates under the foreskin, causing gradual separation.
- 90% of foreskins are retractile at age 3 years , (<1% of phimosis at age 17)
- Resolves with time or with corticosteroid creams, if not : do circumsicion.



The unretracted foreskin and prepuce cover the penis. Retraction of the foreskin uncovers the head of the penis.

Pathological phimosis

- Recurrent balanitis in uncircumcised males can cause new phimosis due to scarring



Treatment

- Olderc hildrenwith phimosis, suffering recurrent infection (balanitis), can betreatedwitha6-week course of topical 0.1% dexamethasonecream, which acts to soften the phimosisand allow foreskin retraction (avoid circumcision where possible).
- Adultsmay require a dorsal slit or circumcision for recurrent balanitis, voiding obstruction, or difficulties with sexual intercourse.

Paraphimosis

- Paraphimosisis when the uncircumcised foreskin is retracted under the glans penis and the foreskin becomes edematous, and cannot be pulled back over the glans into its normal anatomical position. Which leads to gangrene & necrosis of the glans.

phimosis

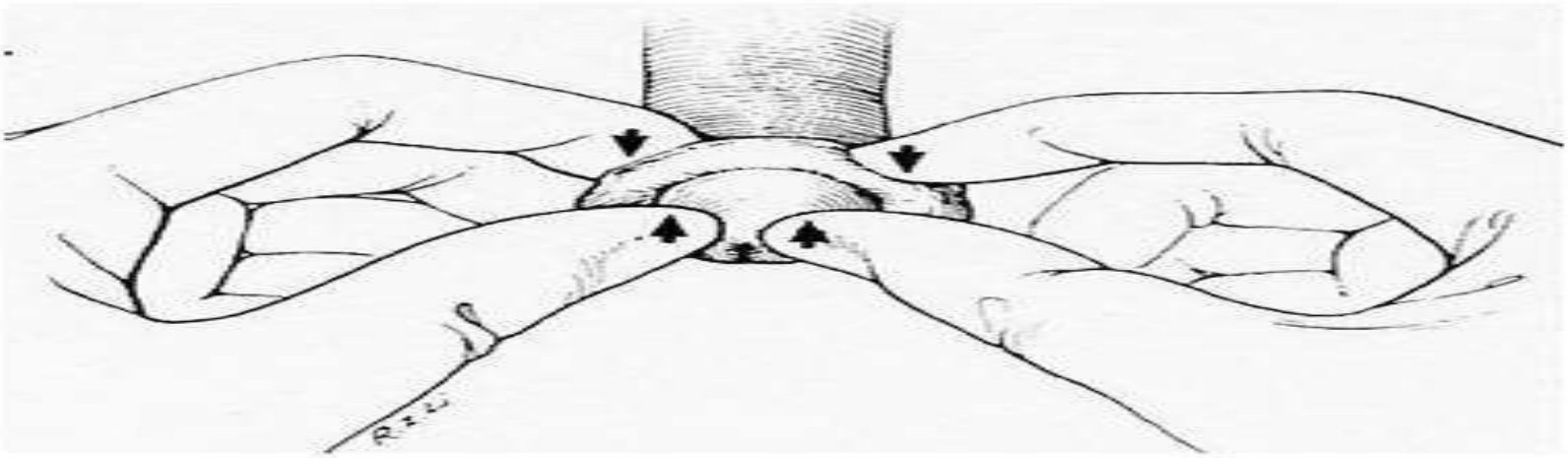
paraphimosis

- It occurs most commonly in teenagers or youngmenand alsoinelderlymen(whohave had the foreskin retracted during catheterization, but where it has not been returned to its normal position).
- Paraphimosisis usually painful. The foreskin is edematous and a small area of ulceration of the foreskin may have developed.

Treatment :

- Manualreductionispreferred using ice packs, elastic compression, and topical anesthetic such as 2% lidocainegel.
- Operative dorsal slit may be required in refractory cases.
- Elective circumcision for definitive treatment (paraphimosis tends to recur).

Figure 5. Paraphimosis Reduction



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Disorder of sexual differentiations:

are defined as congenital conditions in which the development of chromosomal, gonadal, or anatomical sex is atypical. They are estimated to affect 1 in 4500 births.

DSD are subdivided into:

*Seminiferous tubule dysg enesis(klinfelter syndrome XXY,46XXmale).

*Turner syndrome45xo.

*True hermaphrodites 46xx,xy with both ovarian and testicular tissue.

*Mixed gonadal dysgenesis (streak gonads and ambiguous genitalia).

*Pure gonads dysgenesis (female with streak gonads).

Diagnosis:

*Detailed history and maternal history especially drugs used during the pregnancy as steroid and contraceptivepills

*General ex may show associated syndrome ,evidence of dehydration, position of urethral meatus ,carful palpation may show the presence of testes and the presence of female psudohermaphrodites.

Chromosomal analysis confirm karyotype.

*serum electrolyte ,testosterone ,DHT for salt wasting in CAH.

*17 Hydroxyprogesterone done after 3 days can diagnose 21 hydroxylase deficiency.

*Hcgstimulation test can diagnose androgen resistance and 5a reductase

deficiency.

Treatment

A multidisciplinary approach is required with full parental input. Gender assignment of ambiguous genitalia is guided by the functional potential of gonadal tissue, reproductive tracts, and genitalia, with the aim of optimizing

psychosocial well-being and producing a stable gender

identity.

Patients have a higher risk of gonadal malignancy, which

requires surveillance and/or removal of gonadal tissues and hormone replacement.


Thank
you!