Pediatric Urology

Undescended testes

Vesicouretericreflux (VUR)

Hypospadias

Nocturnal enuresis

Posterior urethral valves (PUV)

Extrophyof the Bladder

Epispadias

Phimosis

Paraphimosis

Disorder of sexual differentiations

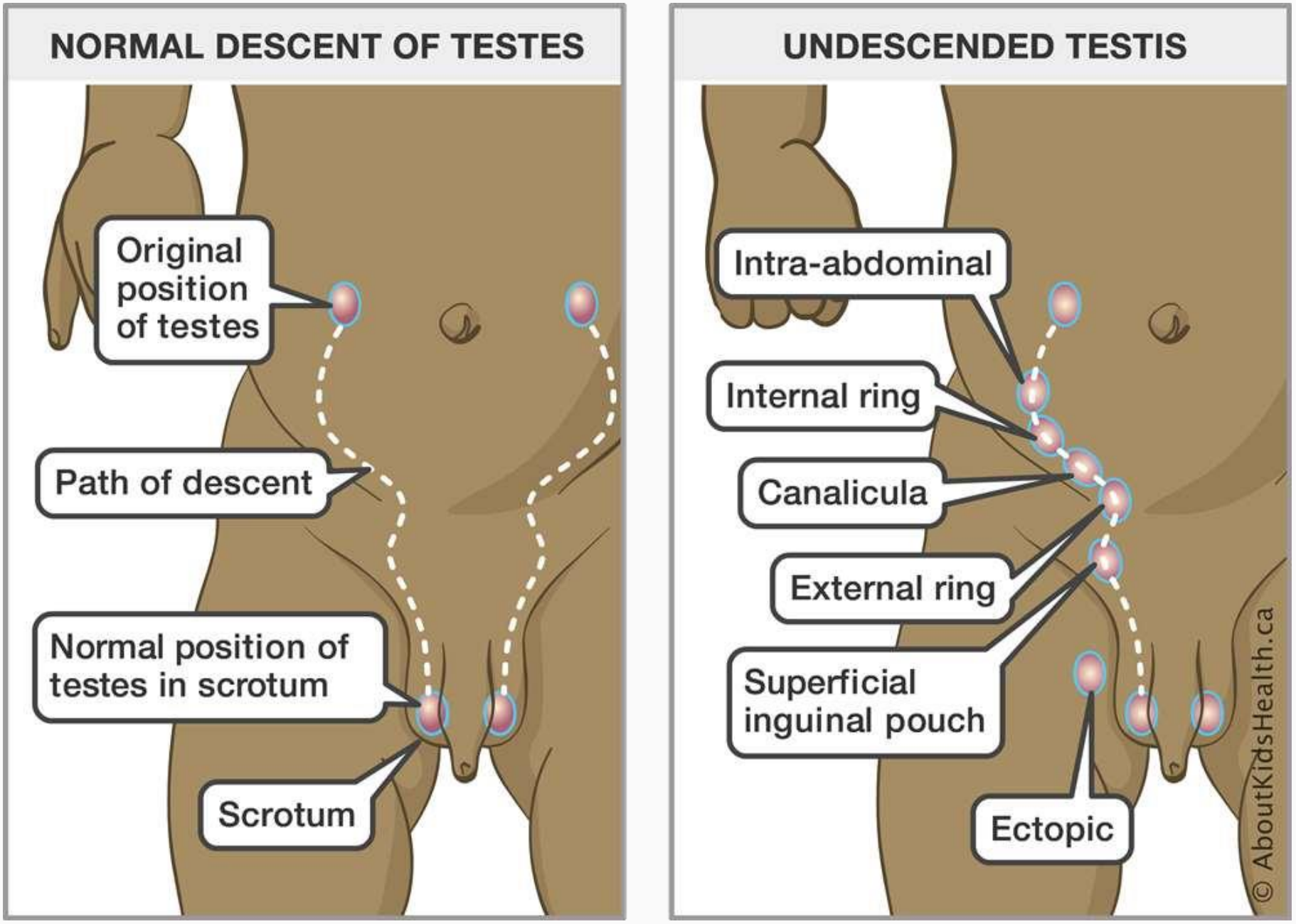
Undescended testes

- The testesdescendintothescrotuminthe third trimester (passingthroughtheinguinalcanalat 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 3 months. 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
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- Classification

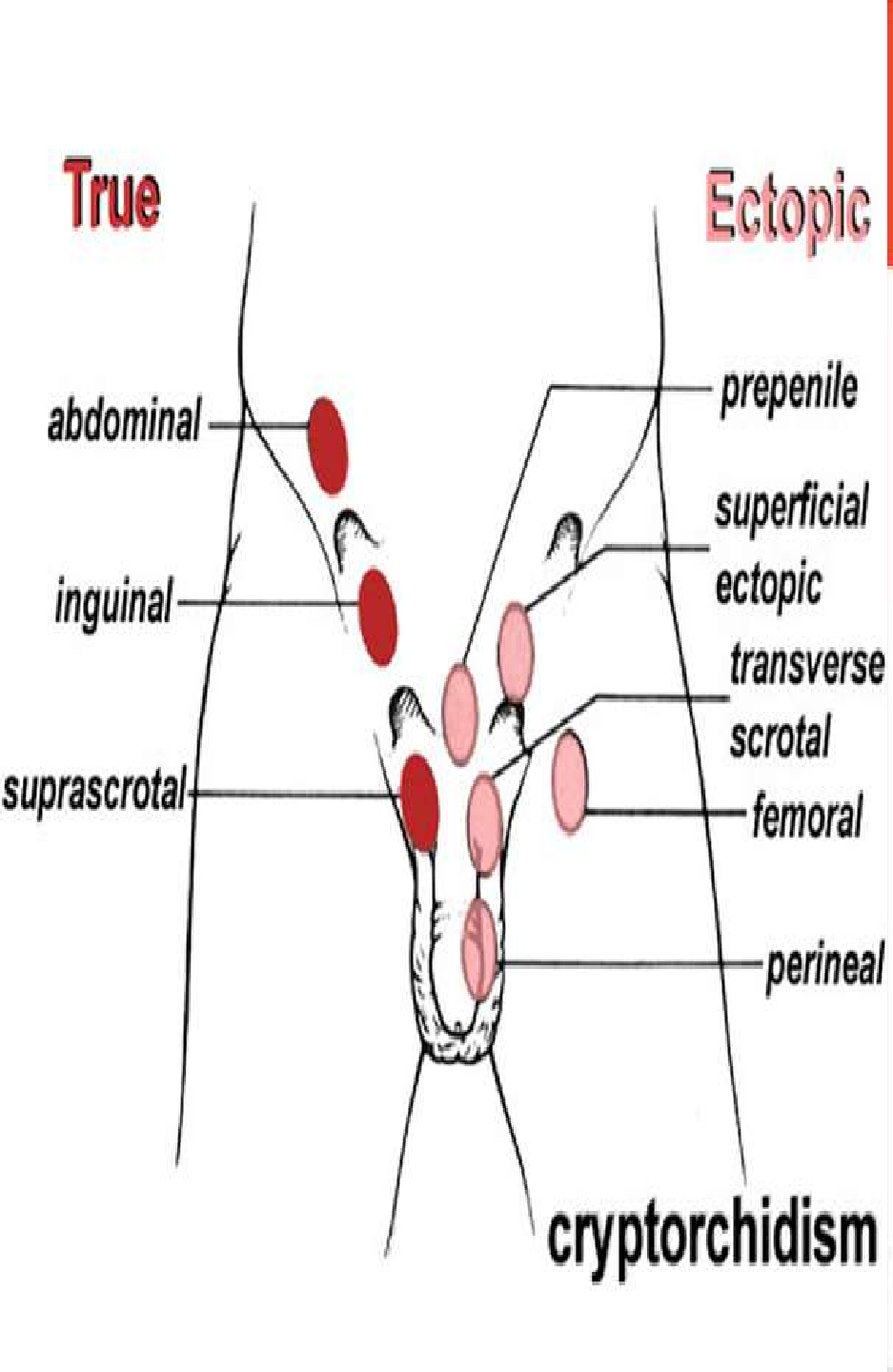
Retractile: anintermittent active cremastericreflex causes the testis to retract up and out of the scrotum.

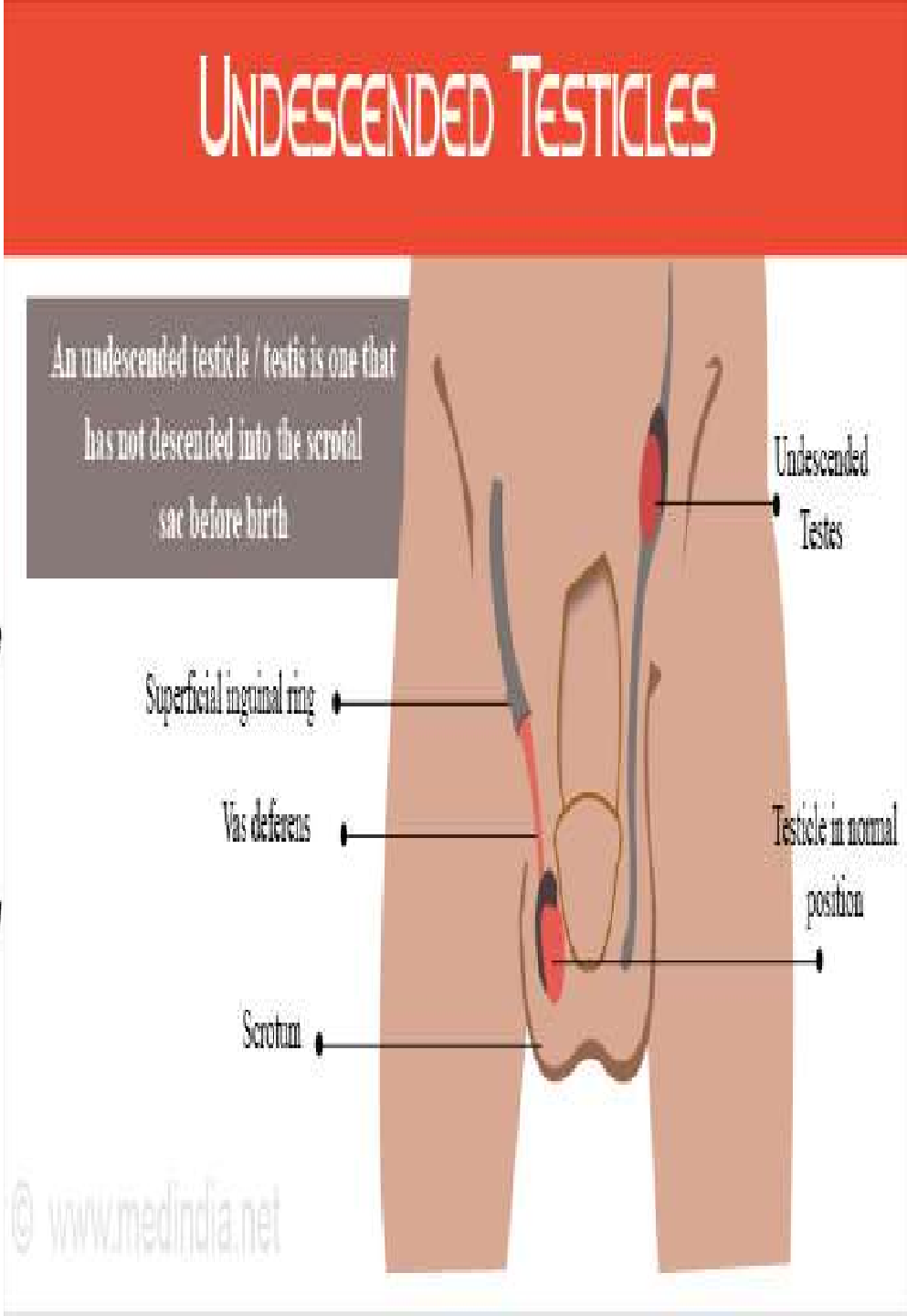
Ectopic (<5%): abnormal testis migration below the external ring of the inguinal canal (to perineum, base of penis, or femoral areas)

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

Theseincludepreterminfants, low birth weight, small for gestational age, and twins.

- Etiology

This includes :

1)abnormal testis

2)gubernaculum

3) endocrineabnormalities (low level of androgens[hCG], (LH),calcitoningene–related peptide)
4) decreased intra abdominal pressure (prune-belly syndrome, gastroschisis).

- Pathology

There is degeneration of Sertolicells, loss of Leydigcells, and atrophy and abnormal spermatogenesis.

- Long-term complications

- Relative risk of cancer is 40-fold higher in the undescendedtestis.Mostareseminomas;carcinomain situ represents a small percentage (~2%).
- Reduced fertility (atrophy)
- Increased risk of testicular torsion
- Increased risk of direct inguinal hernias

- Increased risk for trauma

Diagnosis

- Full examination is required to elucidate if the testis is palpableand toidentify 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 processusvaginalis, 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 preservetesticular growth
- To avoid malignancies, torsion&trauma
- For psychological issues for the growing child

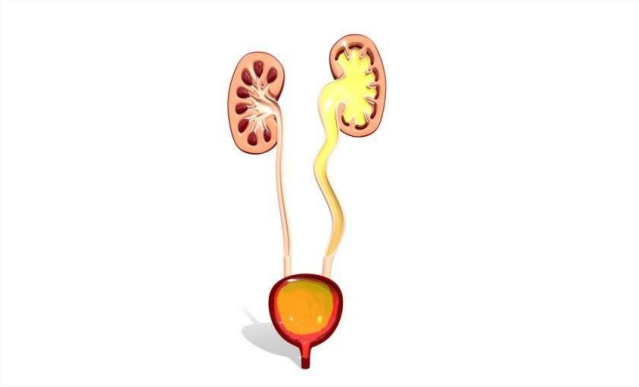
  of having asingletesticle.

- Intra-abdominal testes may require division of spermatic vessels toprovide extra length (Fowler-Stevens procedure, relying on collateral blood flow from vas), two-stage procedures, or microvascular auto transplantation

Vesicoureteric reflux (VUR)

- Definition

Results from abnormal retrograde flow of urine from the bladder into the upper urinary tract.



- Epidemiology

incidence >10%;

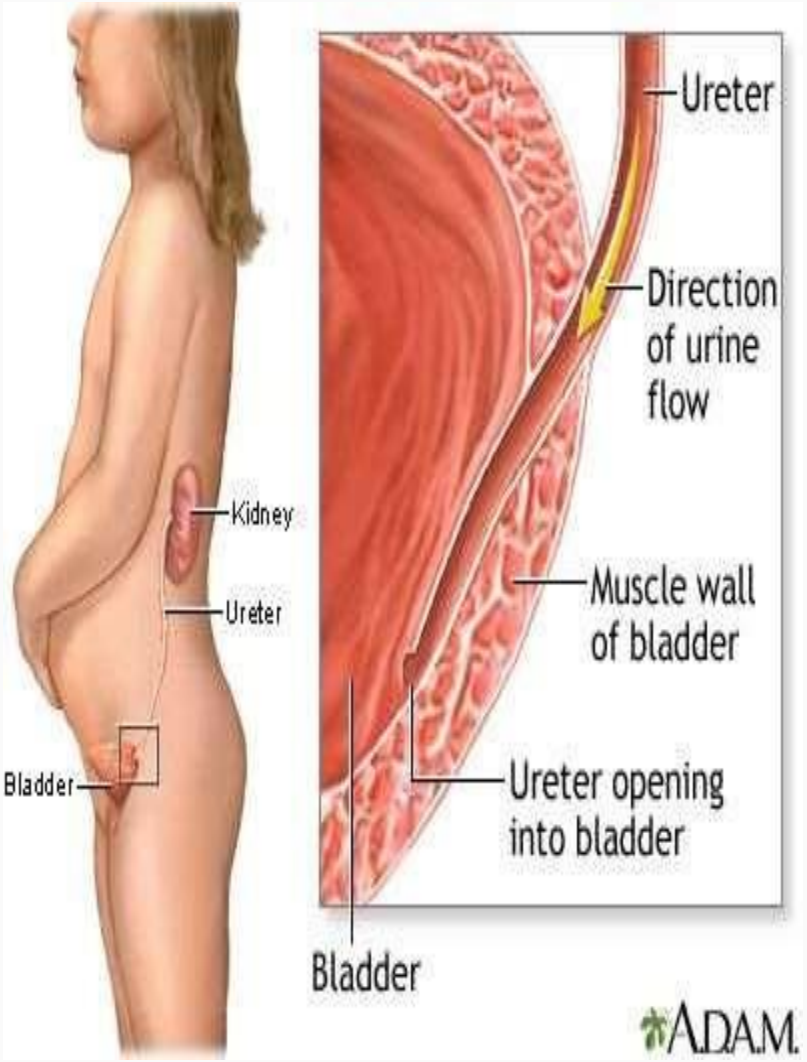
younger > older;

girls > boys (5:1)

Siblings of an affected child have a 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:

- Primary reflux (1%) results from a congenital abnormality ofthe ureterovesicaljunction.
- Secondary reflux results from urinary tract dysfunction associated with elevatedintravesical 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.

Reflexinduces Renaldamage in2 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. Waterhummereffect!Theurineflowsbackwardswithahigh flowandcauses injuryintherenalcollectingsystem.

- Presentation

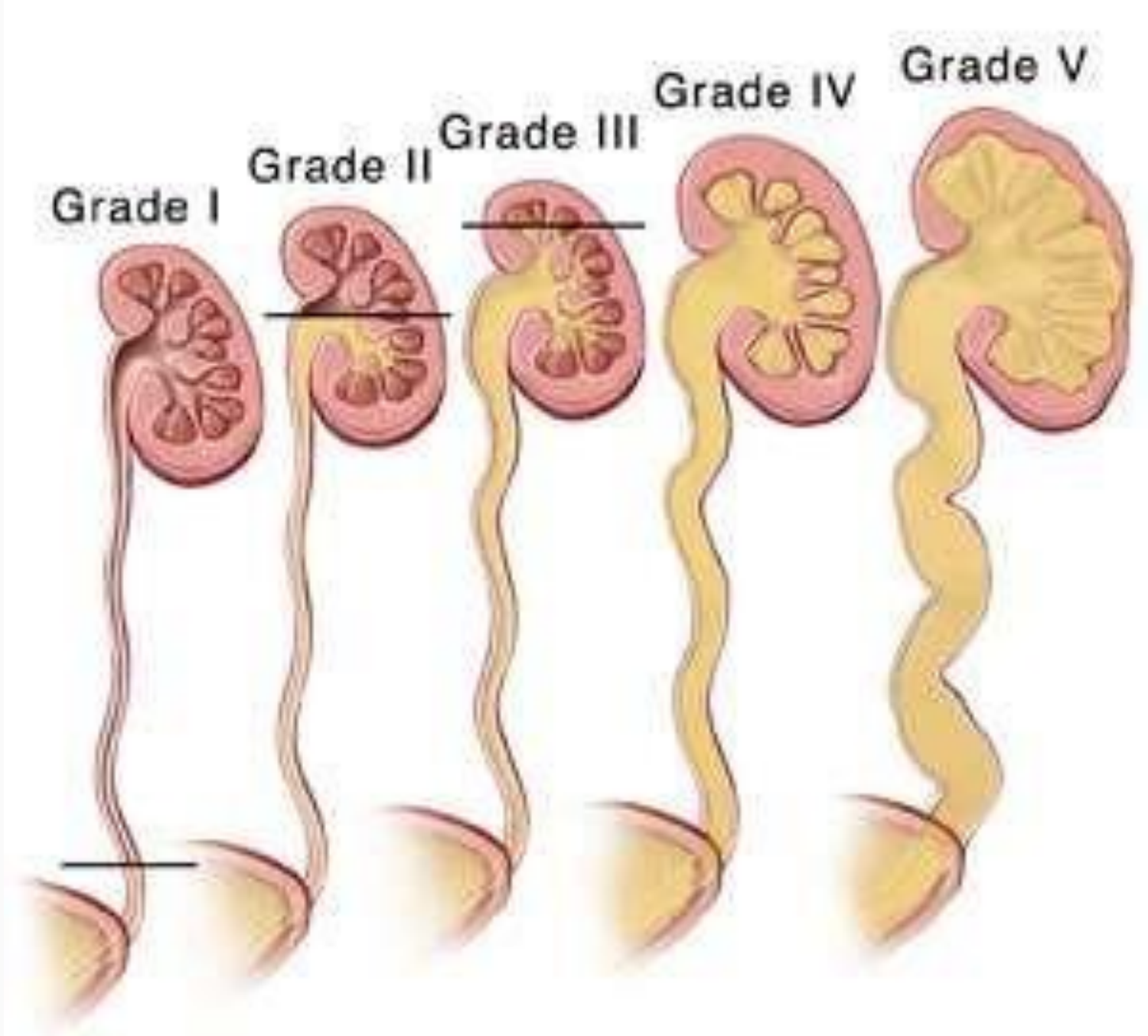
Patients have symptoms of UTI, fever, dysuria, suprapubicor abdominal pain, failure to thrive, vomiting, and diarrhea.

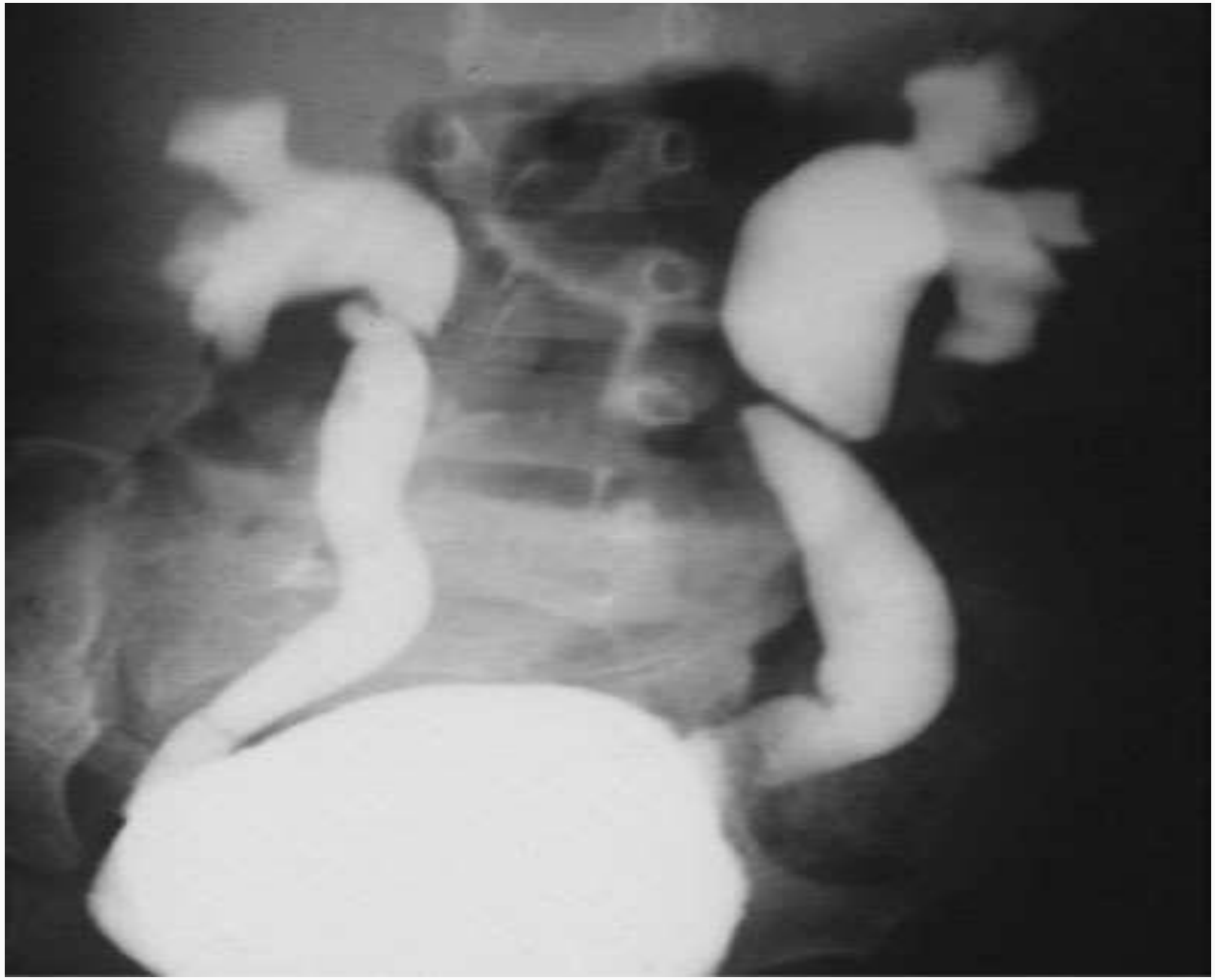
- Patients in the pediatric age group present with recurrentfebrileUTIsweshouldruleout reflex
- Pediatrics with hydronephrosiswe should rule out reflex

- 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
- Urodynamicassessment
- 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



Massive bilateral reflux is seen on cystogramin young child.

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 the risk of water hammer effect and renal injury.

Correct problems contributing to secondary reflux. MostprimaryVUR gradeI–II cases will resolve spontaneously (~85%), with 50% resolution in grade III.Observationand medical treatment areinitially recommended.

Medical treatment

Conseravitetreatment:

Low-doseantibioticprophylaxis 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 istomaintainthe5:1 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

- Definition

Hypospadiasis 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).

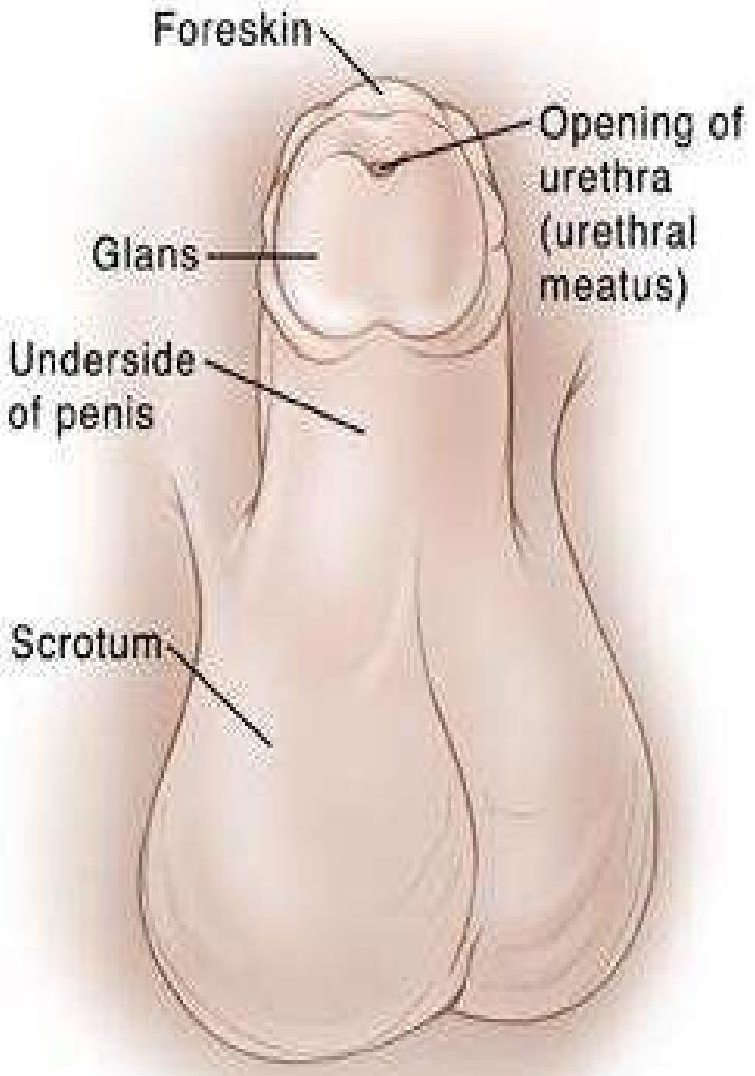
- It is the most commoncongenital malformation of the urethra. It occurs in 1 in 250 live male births

5 Associations with hypospidias:

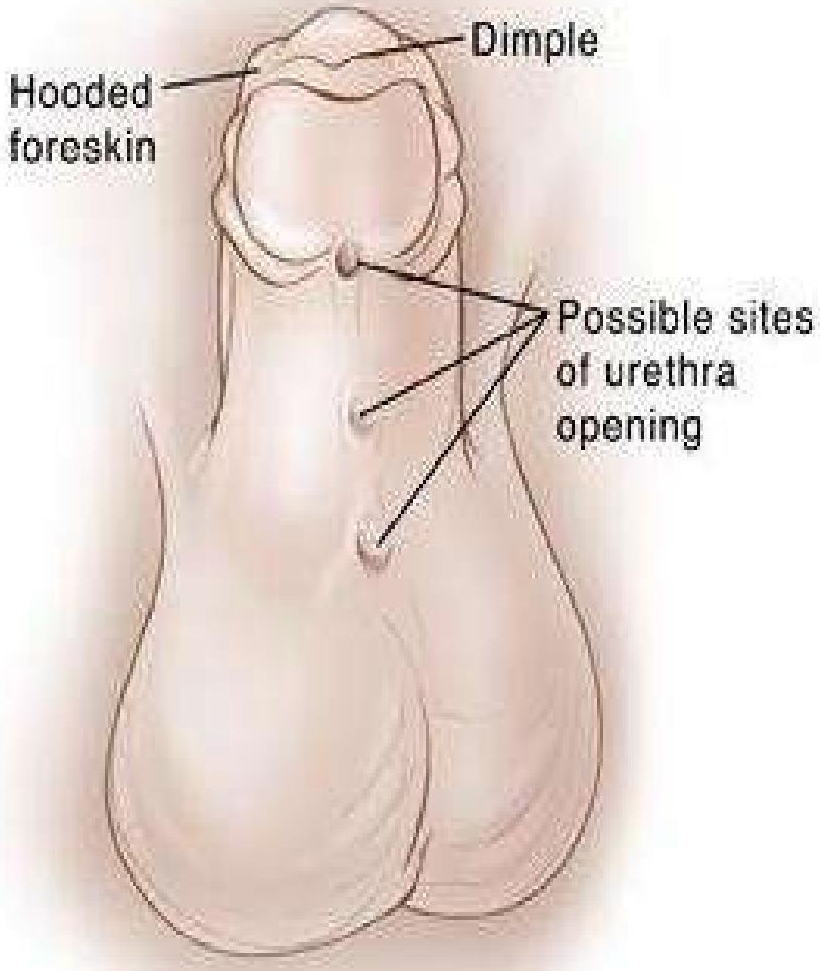
1. ventrally located meatus
- 2. absenceofventralforeskin
- 3. penile chordee(ventral curvature of the
- penile shaft).

4. absence of the penile ventral skin

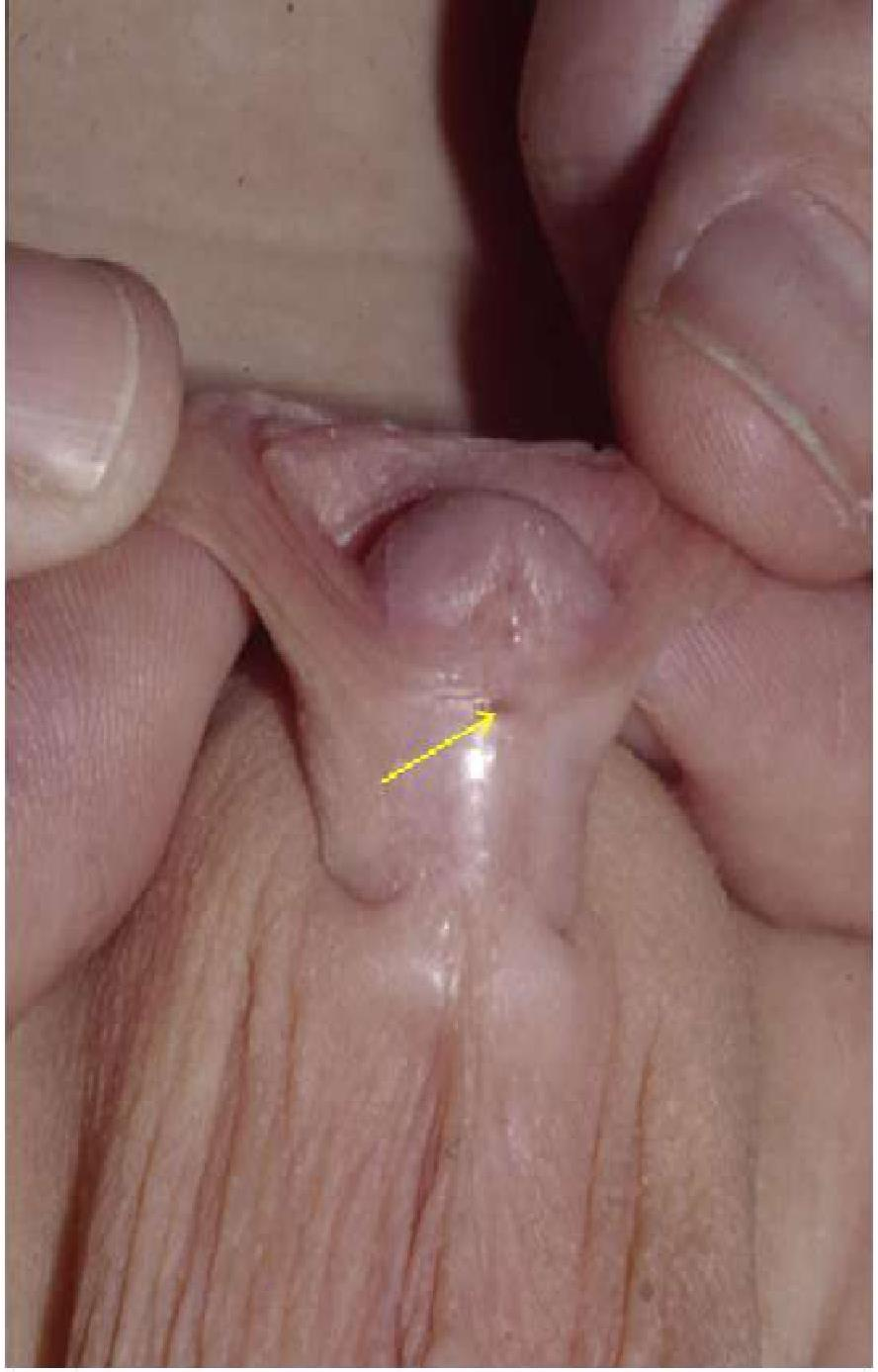
- 5. deviation of the median raphe to one side
- of the penis

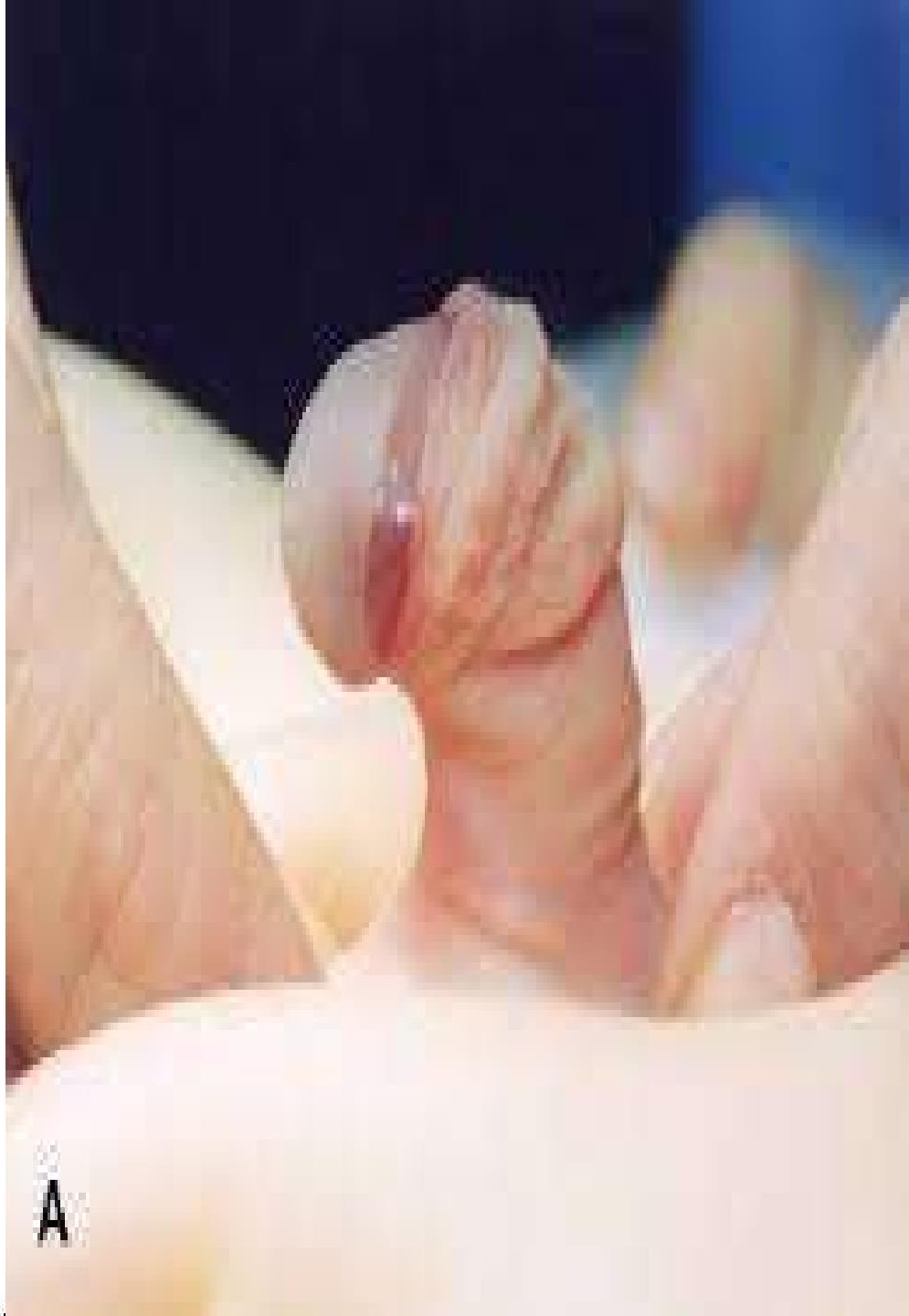


Normally, the opening of the urethral meatus is located at the tip of the penis.



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

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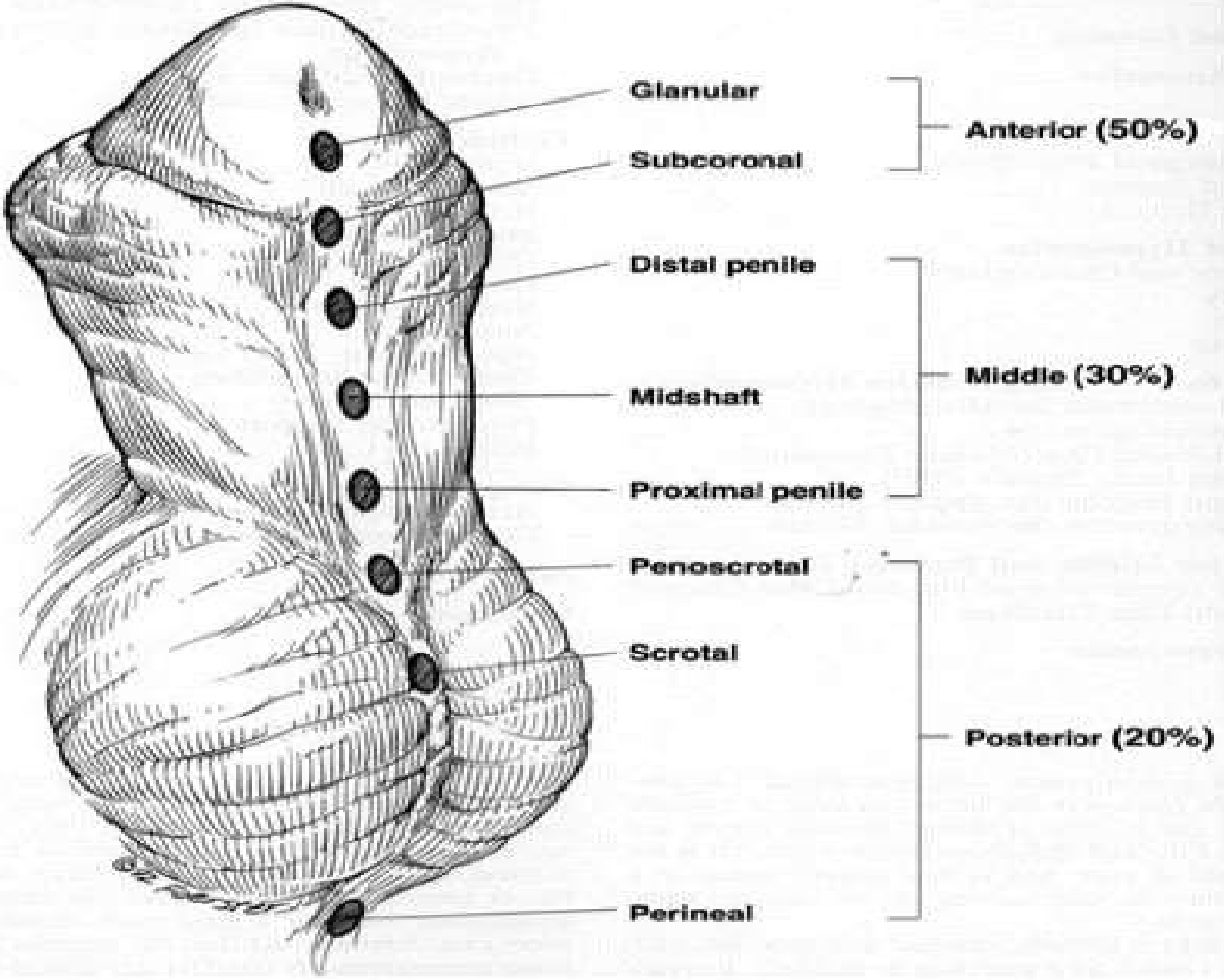
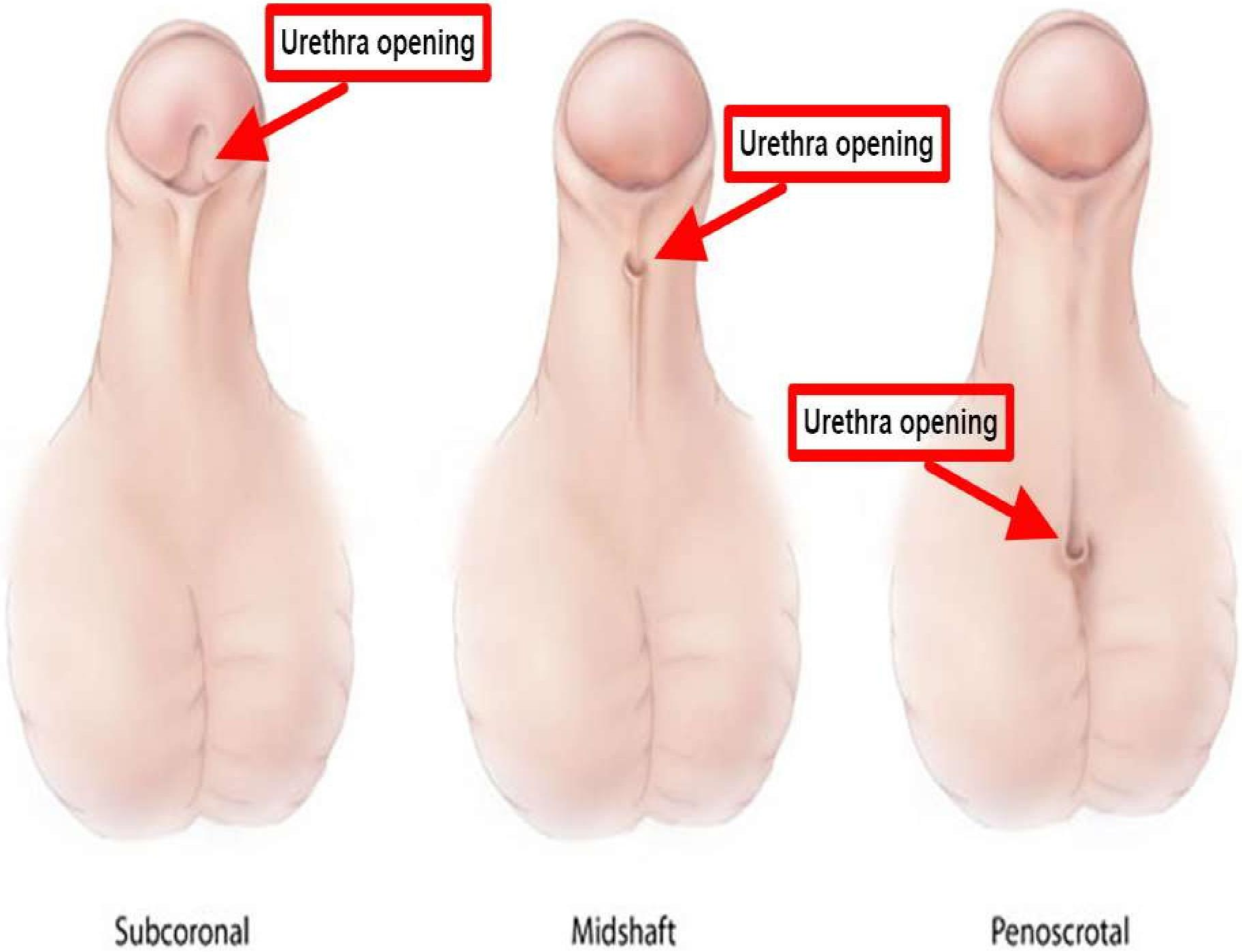
Classification

- Hypospadiascanbeclassifiedaccording to the anatomical locationof the urethralmeatus
- 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 examinationwillmakethe diagnosis.

  However,itis also important to seekout associated abnormalities that will need treatment (undescended testes, inguinal hernias, and hydroceles).
- Patients with absent testes and severe hypospadias should undergo chromosomal and endocrine investigationto excludeintersex conditions
- After diagnosis you have to inform the family that: in the case of distal or mild; circumcision should be done at the timeof the thehypospidiassurgery whichis done at 1 year of age, this condition will not increase the incideneof Utisand 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 surgeryisthe mode of treatment (cosmetically & functionally)
- Surgery is indicated where deformity is severe, interfereswithvoiding,ORispredictedtointerfere with sexual function. Surgery is now performed between 6 and 12 months of age.
- Local application of testosterone for 1 month preoperatively can help increase tissue size.
- The aim of surgery is to correct penile curvature (orthoplasty),reconstructanewurethra,andbring the new meatus to the tip of the glans using urethroplasy, glanuloplasty, and meatoplasty

Complications

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

Nocturnal enuresis

- Enuresisis normal but involuntaryvoiding

  that occurs at an inappropriate time or social setting, during the day, night, or both.

- Nocturnal enuresis :

describes any involuntary loss of urine during sleep.

Prevalence

Age (years)	Females	Males
5	10–15%	15–20%
7	7–15%	15–20%
9	5–10%	10–15%
16	1–2%	1–2%

• Classification

• Primary:neverbeen dry for more than a 6-month period

• Secondary: re-emergence of bed wetting after a period of being dry for at least 6 months

• 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

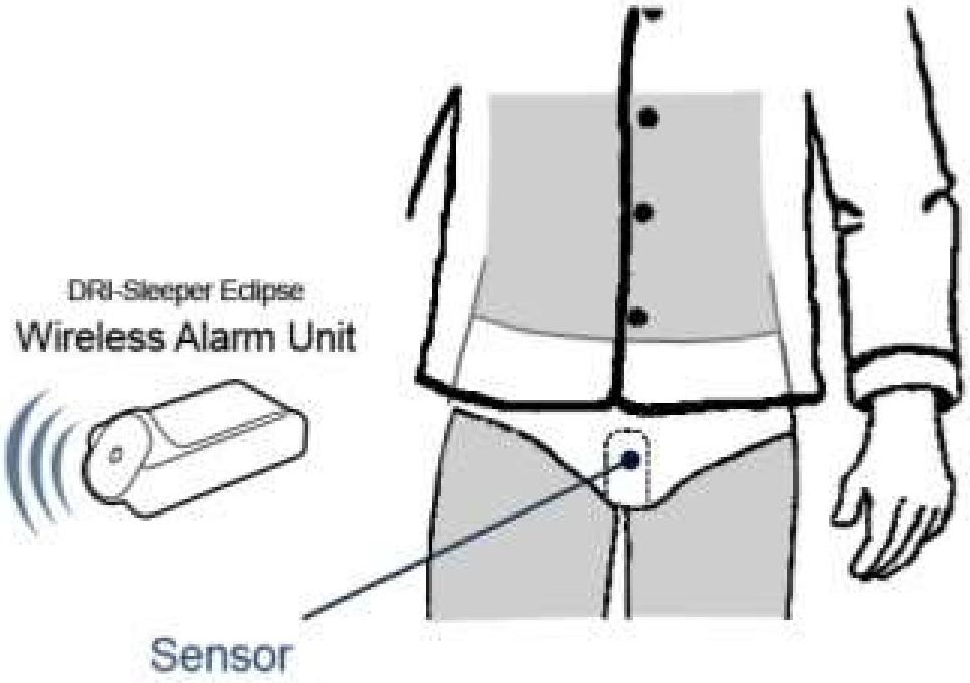
- History: frequencyof episodes; daytime symptoms; new or recurrent; family history; UTIs; bowel problems; psychosocial history
- Examination: exclude organic causes (neurological disease)
- Investigation: urinalysis (infection, specific gravity is reduced in nocturnal polyuria, glucose, protein); voiding diary

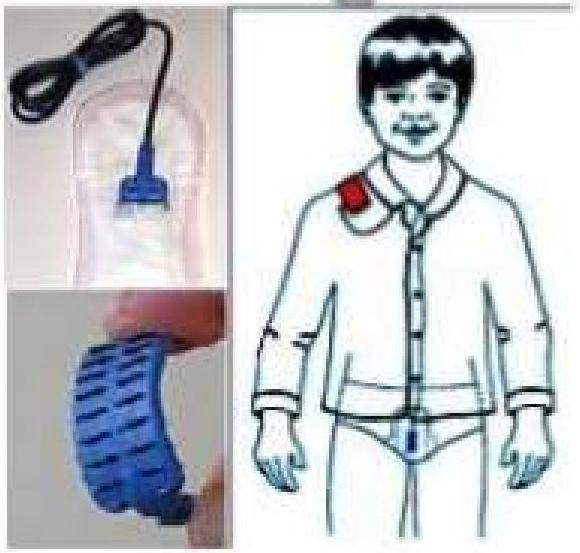
Management

- Behavioral

Provide reassurance; bladder training; motivational techniques to improve the child's self-esteem; conditioning therapy (an alarm is connected to the child's underwear, which is triggered with the first few drops of urine).

Wet Alarm Therapy





- Pharmacological

•Imipramine—atricyclic antidepressant with anticholinergic,antispasmodicproperties. Not used.

•• DDAVP or desmopressin(synthetic analogue of ADH) given intranasallyor orally

- Prognosis

□

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

Posterior urethral valves (PUV)

Definition

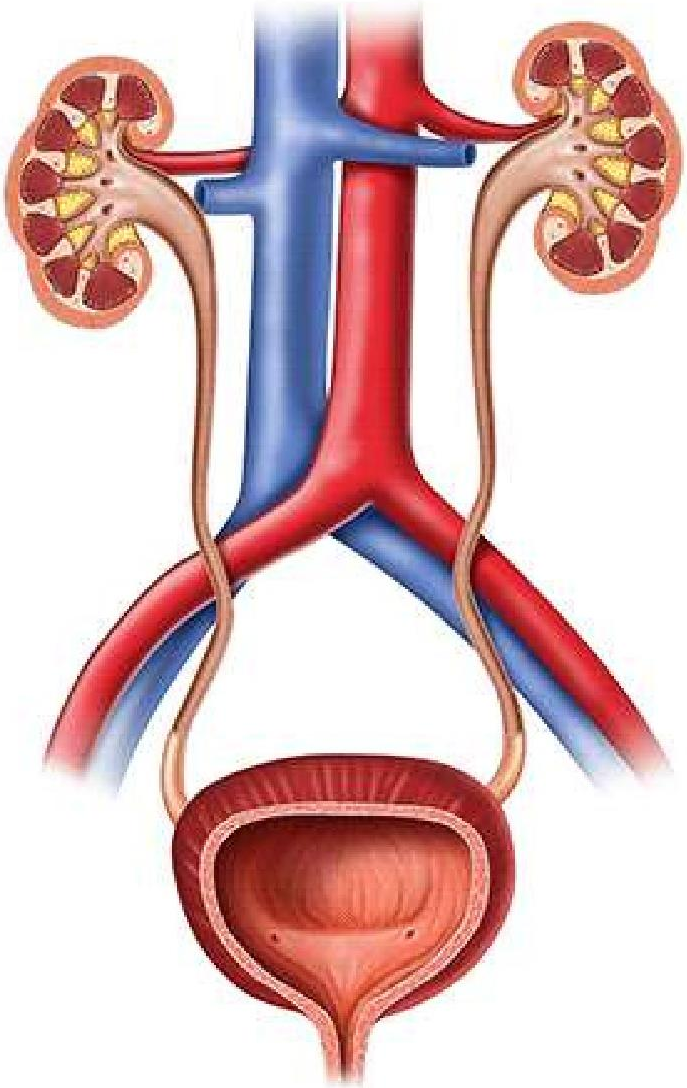
Posteriorurethral valves (PUV) are abnormal congenital mucosal fold in the prostatic (posterior) urethra causing lower urinary tract obstruction.

Incidence 1in5000 male. (Only in male patients)

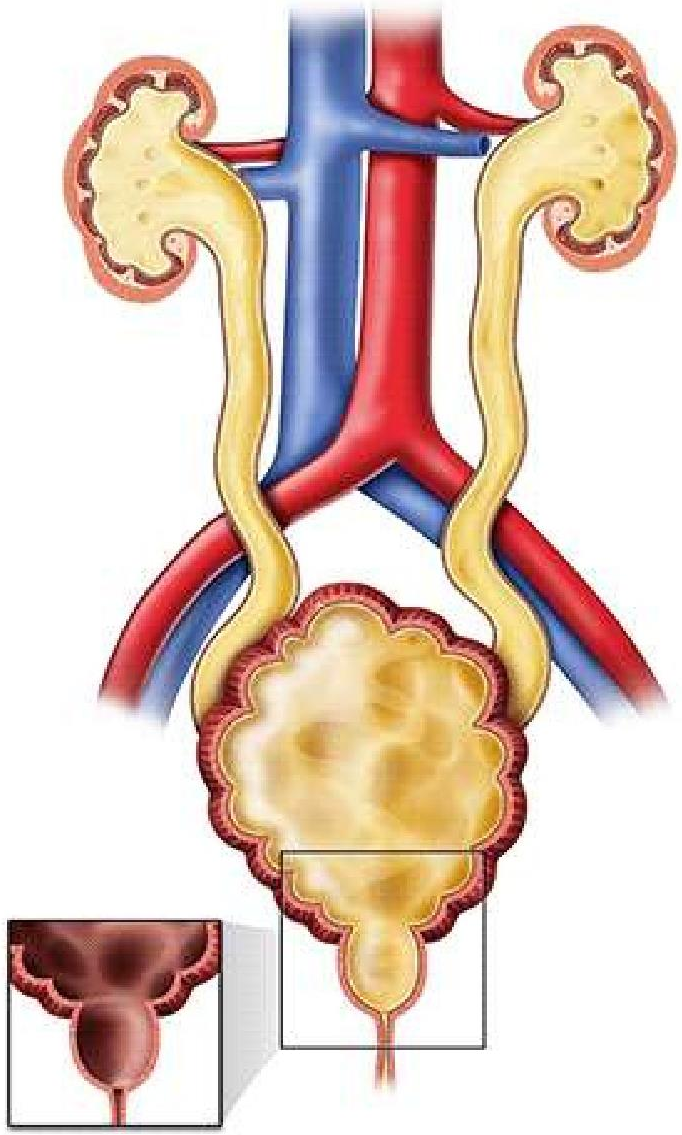
Cause:

Normal male urethra has small ,paired lateral folds(plicae colliculi) found between the lateral, distal edge of verumontanum and lateral urethral wall. PUVs probably represent a congenital overgrowth of these folds from abnormal insertion of Wolffian ducts into the posterior urethra during fetal development.

Normal System



Posterior Urethral Valves (PUV)



- **The verumontanum, or mountain ridge, is a distinctive landmark in the prostaticurethra, important in the systemic division of posterior valve disorders:**

1. **Type I** -Most common type; due to anterior fusing of the plicaecolliculi, mucosal fins extending from the bottom of the verumontanumdistally along the prostatic and membranous urethra

2. **Type II** -Least common variant; vertical or longitudinal folds between the verumontanumand proximal prostatic urethra and bladder neck

3. **Type III -**Less common variant; a disc of tissue distal to verumontanum, also theorized to be a developmental anomaly of congenital urogenital remnants in the bulbar urethra

Presentation

Prenatal US features

These include bilateral hydroureteronephrosis, dilated bladder with elongated ectatic posterior urethra, thick-walled bladder, oligohydramnios (reduced amniotic fluid), and renal dysplasia.

Early features are associated with poor prognosis.

Newborn and infants

These children have respiratory distress (secondary to pulmonary hypoplasia), palpable abdominal mass (hydronephrotic kidney or distended bladder), ascites, UTI, electrolyte abnormalities, and failure to thrive.

Older children

Milder cases may present later with recurrent UTI, poor urinary stream, incomplete bladder emptying, poor growth, and incontinence. There is a risk of renal failure, vesicoureteric reflux, and voiding dysfunction (overactive or underactive bladder), also described as valve bladder syndrome.

Investigation

- **Ultrasound scan** of kidneys and bladder.
- **VCUG** while voiding shows :

distended and elongated posterior urethra, partialy filled anterior urethra, bladder neck hypertrophy; GOLD STANDARD FOR DIAGNOSING PUV.

- **Isotope renalscan (MAG-3, DMSA) assesses renal** function.

- **Video urodynamic** sallows diagnosis of associated voiding dysfunction, urethra, bladder neck

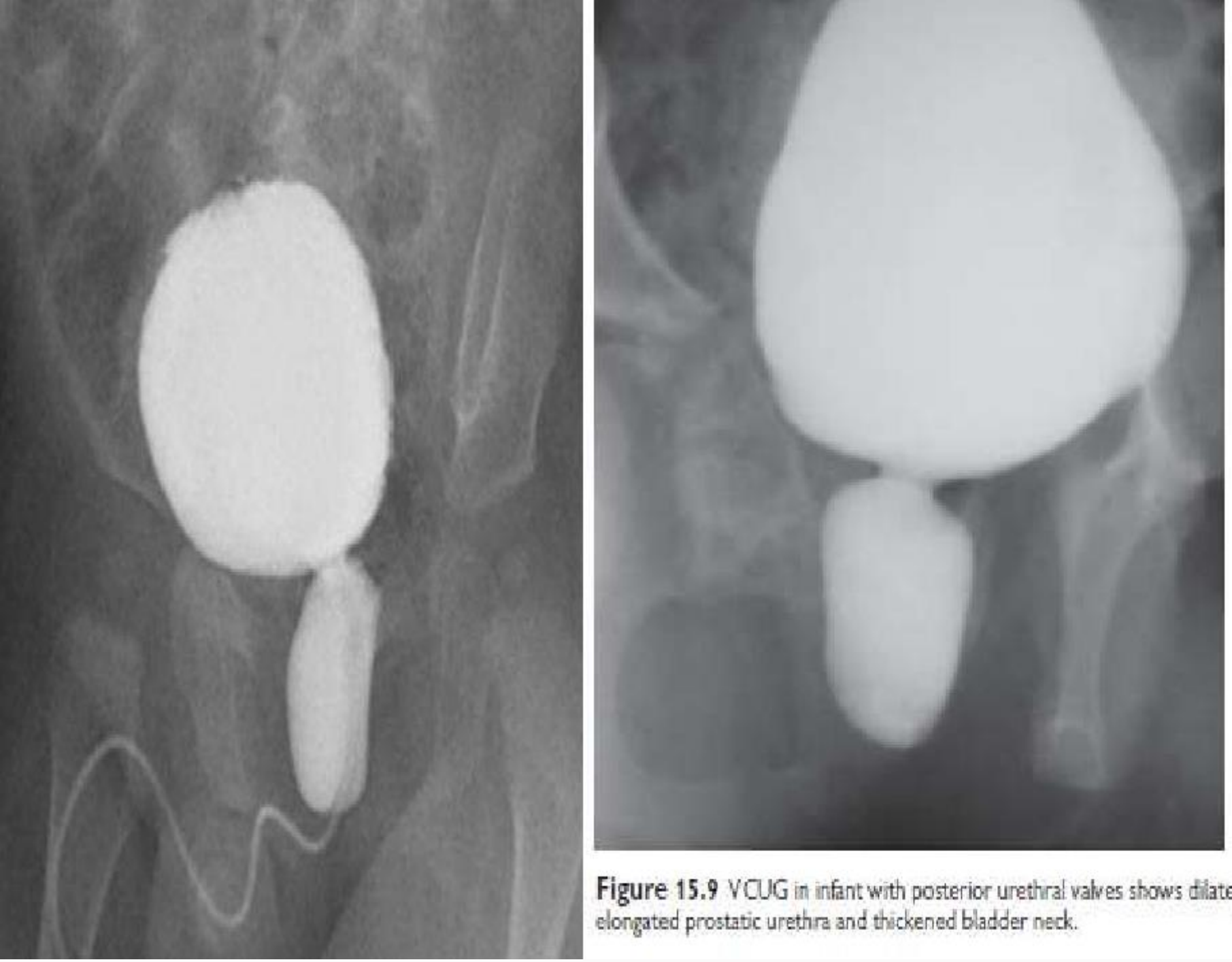


Figure 15.9 VCUG in infant with posterior urethral valves shows dilated elongated prostatic urethra and thickened bladder neck.

Treatment:

Commence prophylactic antibiotics immediately, check serum .electrolytes drain the bladder with a pediatric feeding tube. If there is improvement,

cystoscopy and transurethral ablation of valve (cuts at 5 and 7 o'clock with electrocautery) is recommended (complications include urethral strictures).

- If upper tracts remain dilated with raised creatinine after bladder drainage, a temporary cutaneous vesicostomyis indicated (communicating stoma between the bladder dome and suprapubic abdominal wall, allowing free drainage of urine).

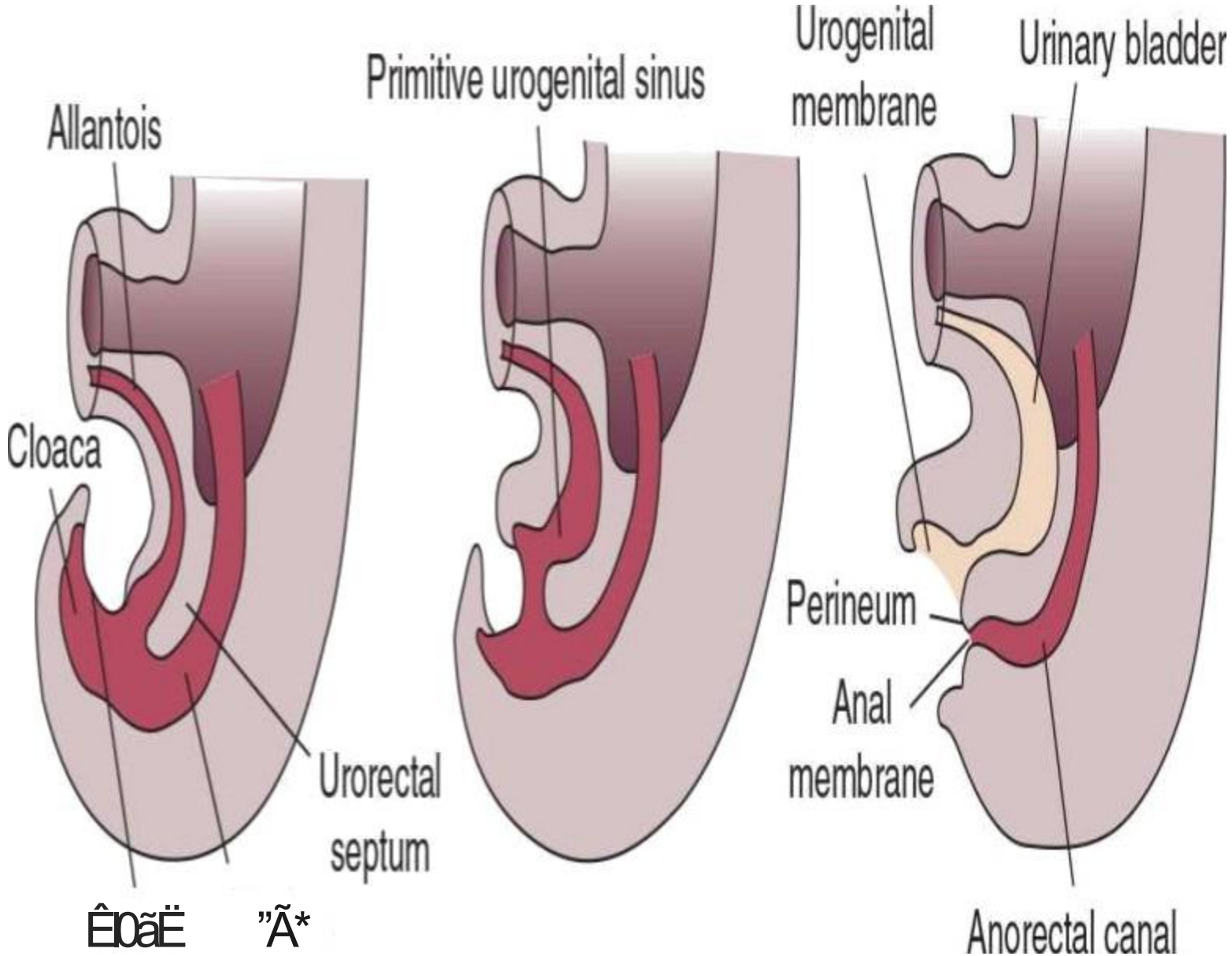
An alternative is ureterostomydrainage. Valve ablation is performed at a later stage.

Extrophy of the Bladder

- **Bladder extrophy results from defective development of the anterior bladder and lower abdominal walls, leaving the posterior bladder wall lying exposed on the abdomen.**

- **M:F >2:1**

- **Risk increased** in family ,younger maternal age and increased parity.



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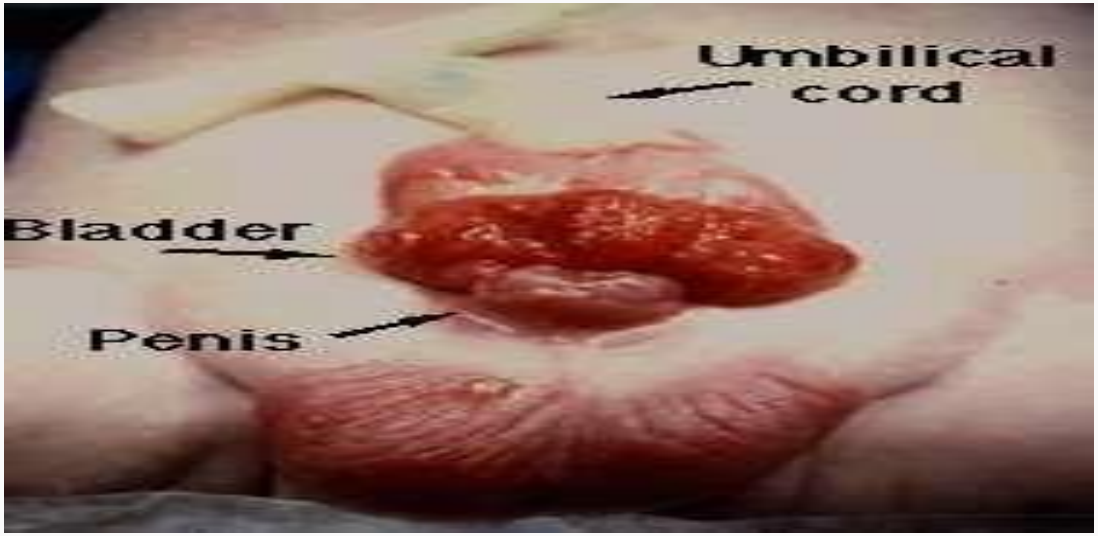
Embryology

- Anembryological malformation results in the **abnormal** over development of the cloacal membrane, which prevents in-growth of lower abdominal tissues.

The cloacalmembrane normally perforates to form the urogenital and anal openings, but in **extrophy there is** premature rupture, resulting in a triangular defect below the umbilicus.
The timing of this rupture determines the type of defect (bladderextrophy, cloacal extrophy, OR epispadias).

Associated anomaly:

- Urinary tract defect: VUR.
- Bone defect: Widening of thepubic symphysis
- Genital defect: Epispadias.
- Musculofascial defect: Inguinal , femoral hernia.



Investigation

- Typical features seen on prenatal ultrasound scan include:

-lower abdominal wall mass

- absent bladder filling

-low-set umbilicus

- small genitalia

-abnormal iliac crest widening.

Management

- At birth, cover the bladder with plastic film and irrigate regularly with sterile saline. Then refer to a tertiary center.

- Trauma to the bladder mucosa can eventually result in squamousmetaplasia, cystitis cystica, or adenocarcinoma and squamouscell carcinoma after chronic exposure.

- Selected cases are suitable for one-stage repair, but most require a three-stage procedure:

- ***Newborn***: pelvic osteotomy (cutting bone to correct deformity) with external fixation with closure of bladder, abdominal wall, and posterior urethra

- ***6–12 months:*** epispadias repair

- ***4–5 years:*** Bladder neck reconstruction (Young–Dees–Leadbetter procedure) and anti-reflux surgery (ureteric reimplantation) are performed when there is adequate bladder capacity and children can participate in voiding protocols.

- Where bladder capacity is too small, bladder augmentation or urinary diversion is required.

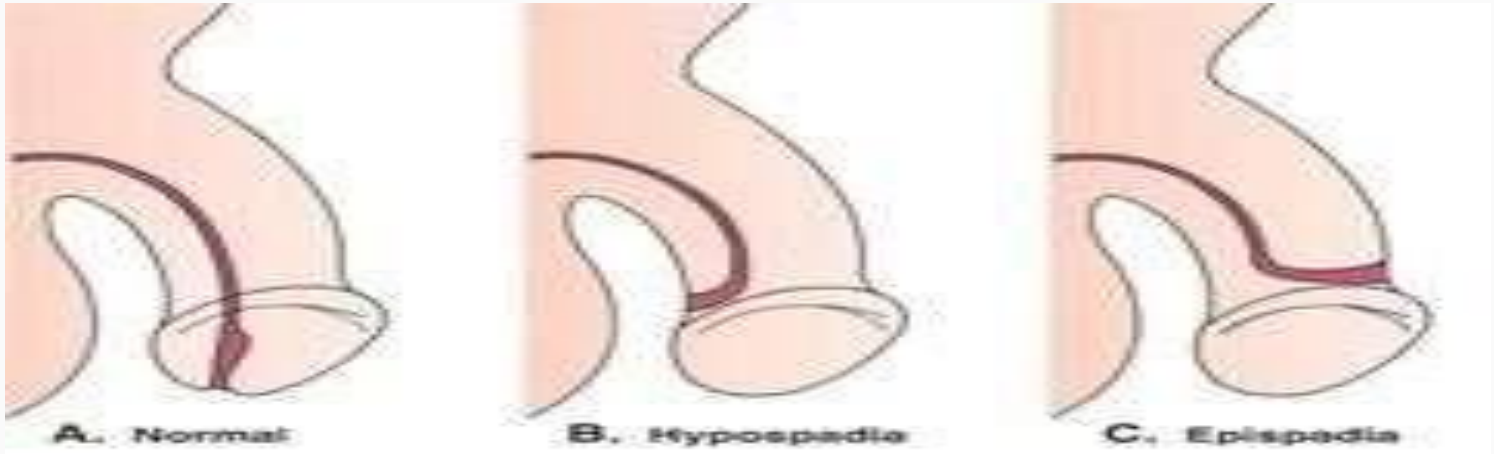
Prognosis

- Even with successful surgery, patients may have long-termproblemswith
  - Incontinence Urinary
  - reflux Repeated UTI
  - Bladder
  - adenocarcinoma Colonic
  - adenocarcinoma
  - Uterine prolapse

- Sexual function and libidoare normal in extrophy patients.

Epispadias

- The urethraopen inthedorsalsurface of the penis any where from the glans, penile shaft or most commonly the 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

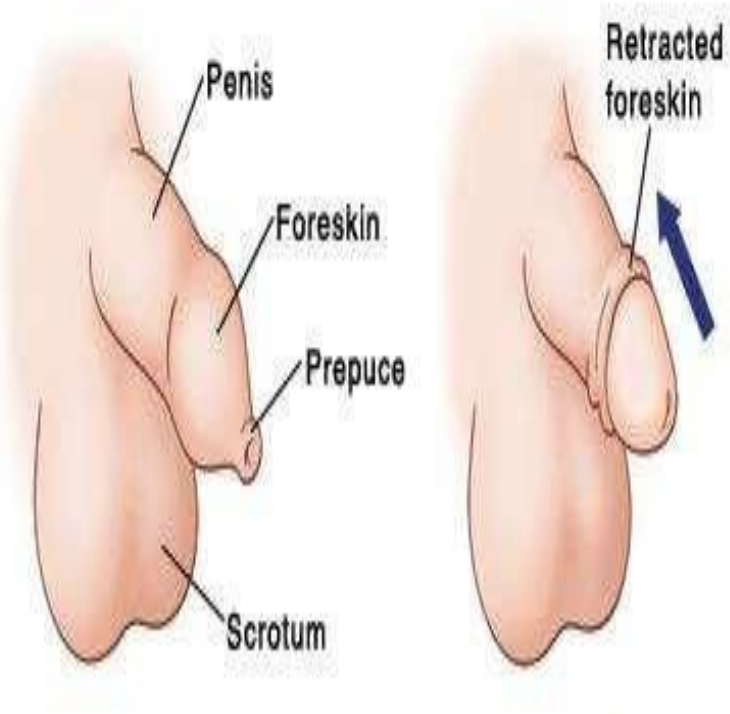
- at6–12 months >>This involves urethroplastywith functional and cosmetic reconstruction of the external genitalia (penile lengthening and correction of chordee) .
- The modified Cantwell–Ransleytechnique is commonly used in males. It describes mobilizing the urethra to the ventral aspect of the penis, 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.

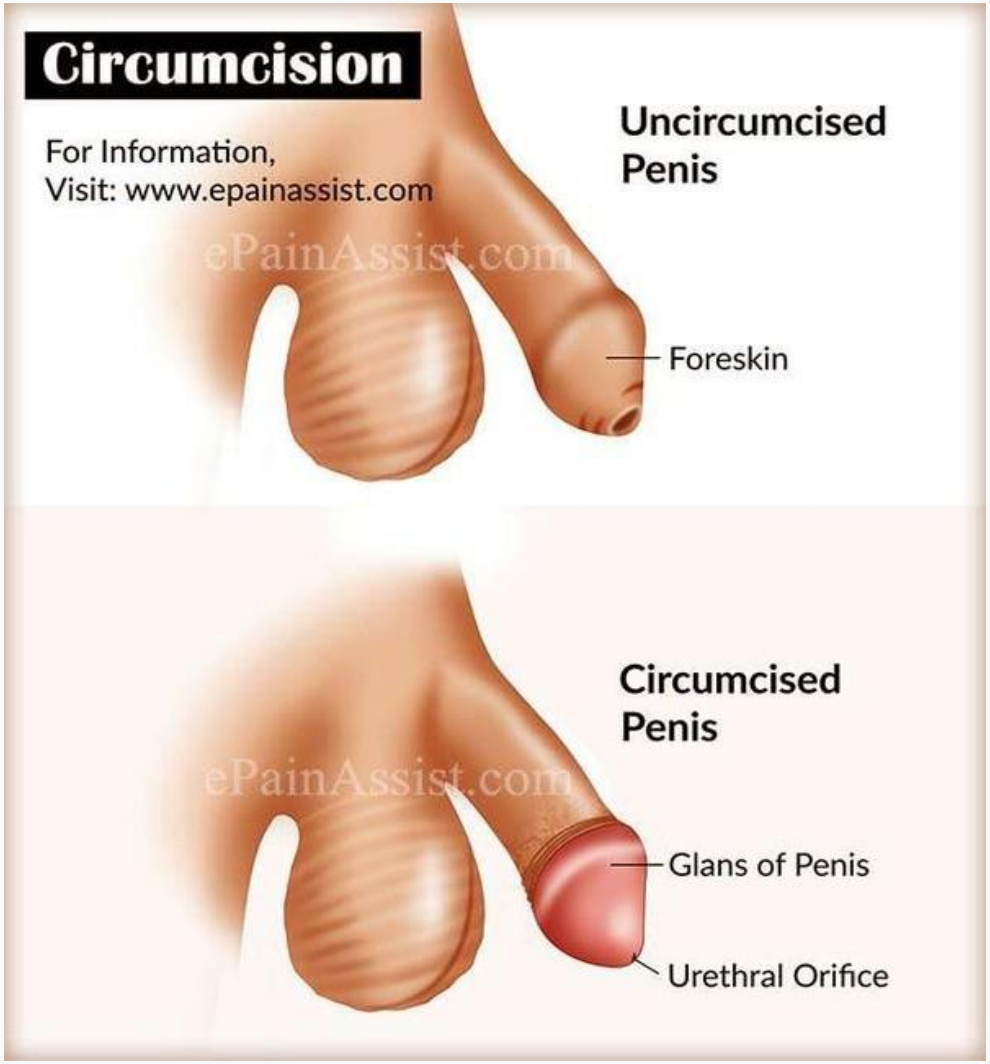
- A physiological phimosisis present at birth due to adhesions between the foreskin and glans.
- As the penis develops, epithelial debris (smegma) accumulates under the foreskin, causing gradual separation.
- 90% of foreskins are retractile at age 3 years , (<1% of phimosis at age 17)
- Resolves with 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

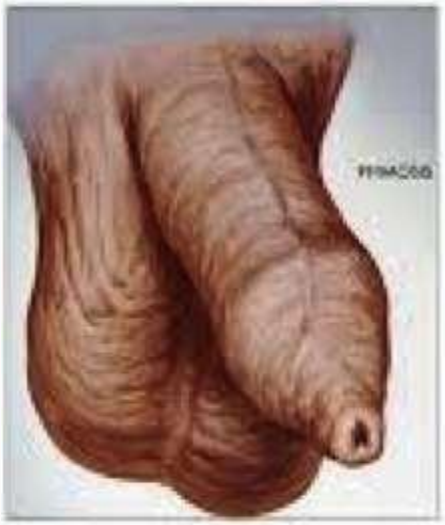


Treatment

- Olderchildrenwith 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

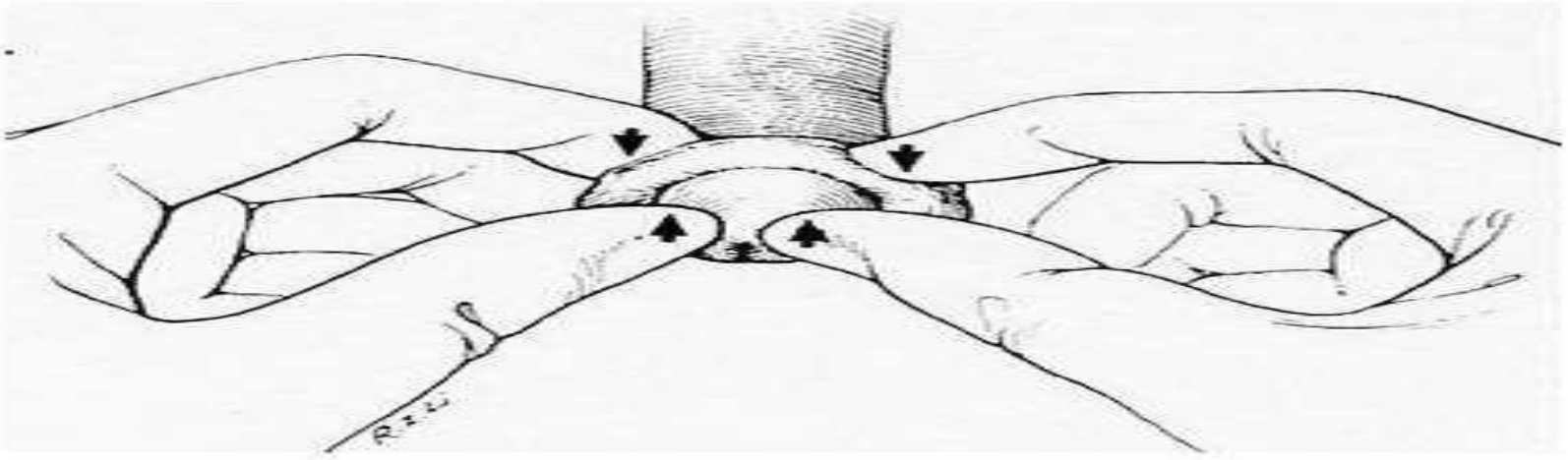
paraphimosis

- 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 dysgenesis(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