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Cryptorchidism: Undescended testes

- The testes descend into the scrotum 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 3 months.

The incidence at 1 year is 1%.

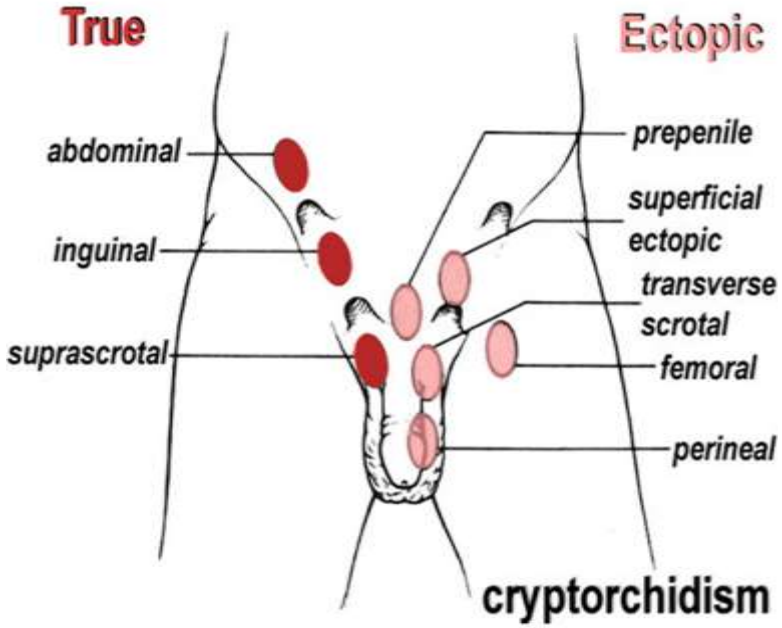
Classification

- **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)

Undescended / Incomplete descent (~95%): testis may be intra-abdominal, inguinal,or prescrotal

Atrophic/absent


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

- **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,

atrophy and abnormal spermatogenesis.

- **Long-term complications**
- Relative risk of cancer is 40-fold higher in the undescended testis. Most are seminomas; carcinoma in situ represents a small percentage (~2%).
- Reduced fertility
- Increased risk of testicular torsion
- Increased risk of direct inguinal hernias

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.
- **Surgery** consists of inguinal exploration, mobilization of spermatic cord, ligation of processus vaginalis, and securing the testis into a dartos pouch in the scrotal wall **(orchidopexy).**

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

Vesicoureteric reflux (VUR)

- **Definition**

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

- **Epidemiology**

incidence >10%;

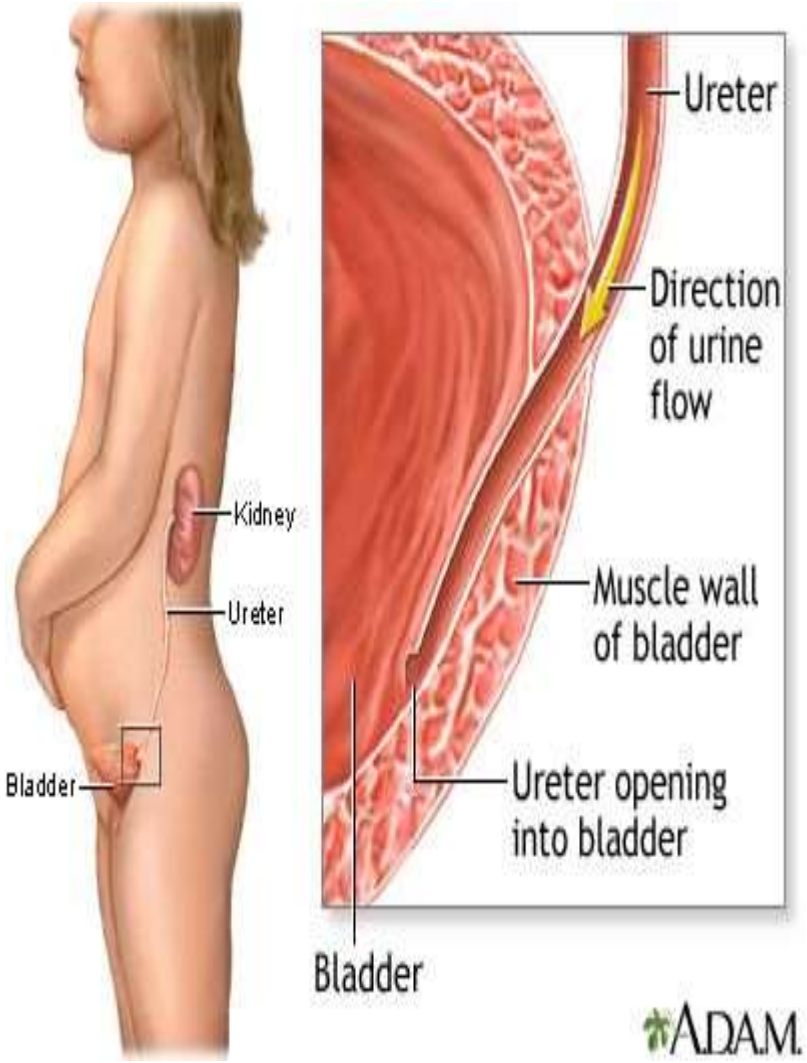
younger > older;

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. 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 of the ureterovesical junction.
- **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).

- **Complications**

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

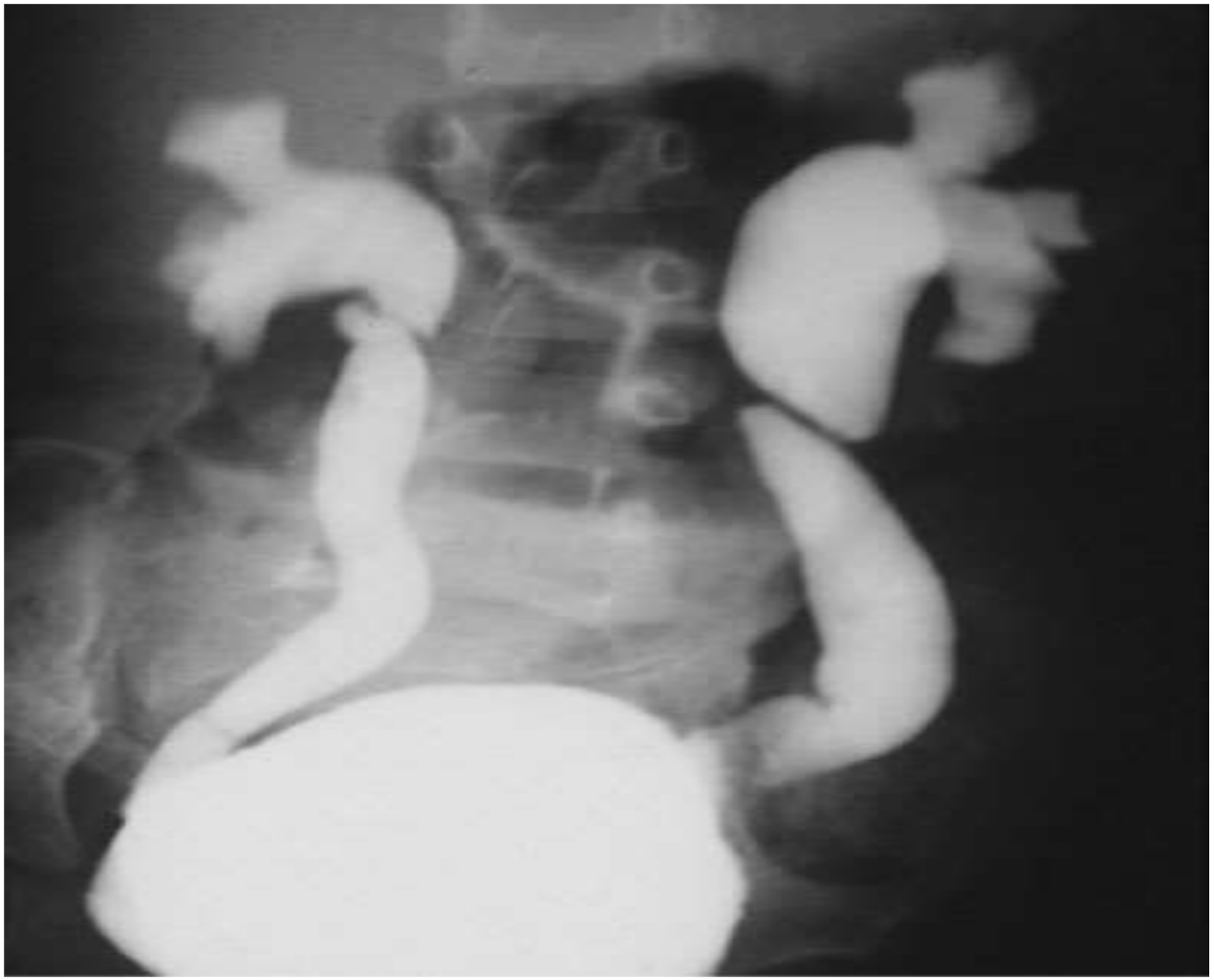
- **Presentation**

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

- Investigation
- Urinalysis and culture to diagnose UTI
- Urinary tract ultrasound scan and VCUG
- Urodynamic assessment
- DMSA 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



Massive bilateral reflux is seen on cystogram in young child.

Management

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

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

- (ureteroneocystostomy± ureteroplasty) or subureteral injection

•Indication for surgery:

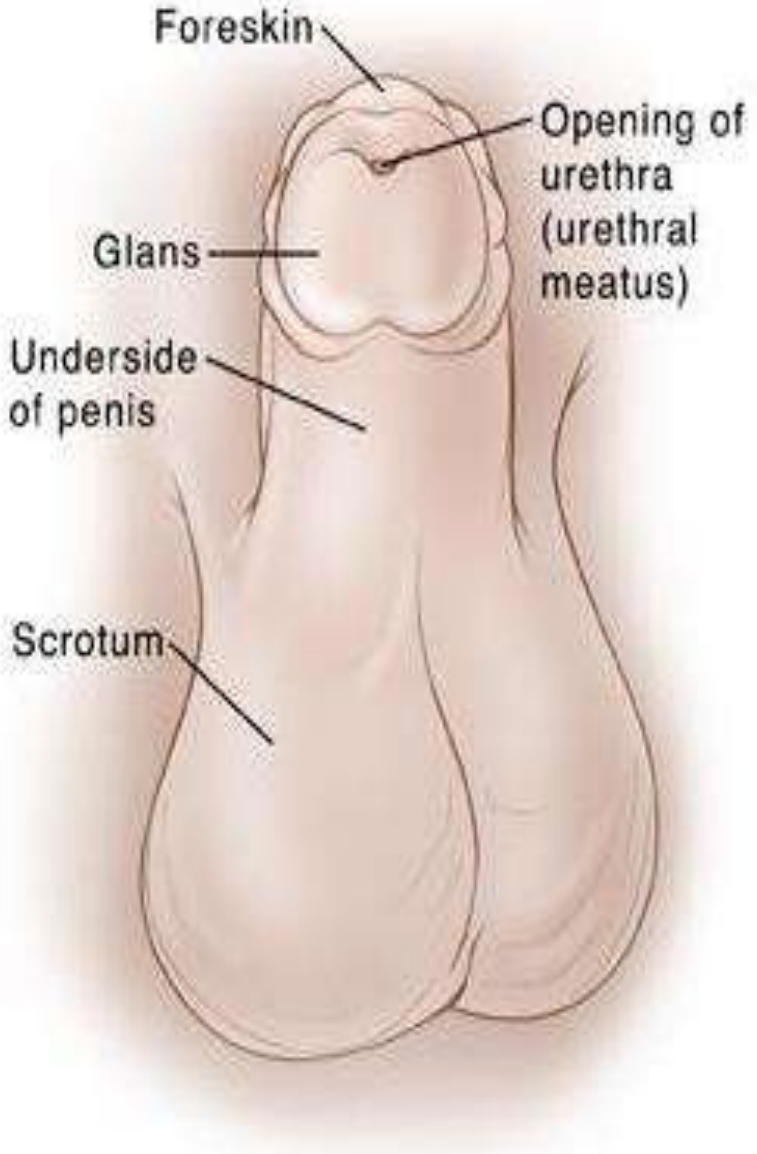
- If it is not possible to keep the urine sterile and reflux persists
- If acute pyelonephritis recurs despite a strict medical regimen and chronic suppressive antimicrobial therapy.
- If increased renal damage is demonstrated by serial excretory urograms or nuclear scan.
- High grade reflux (grade IV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

Hypospadias

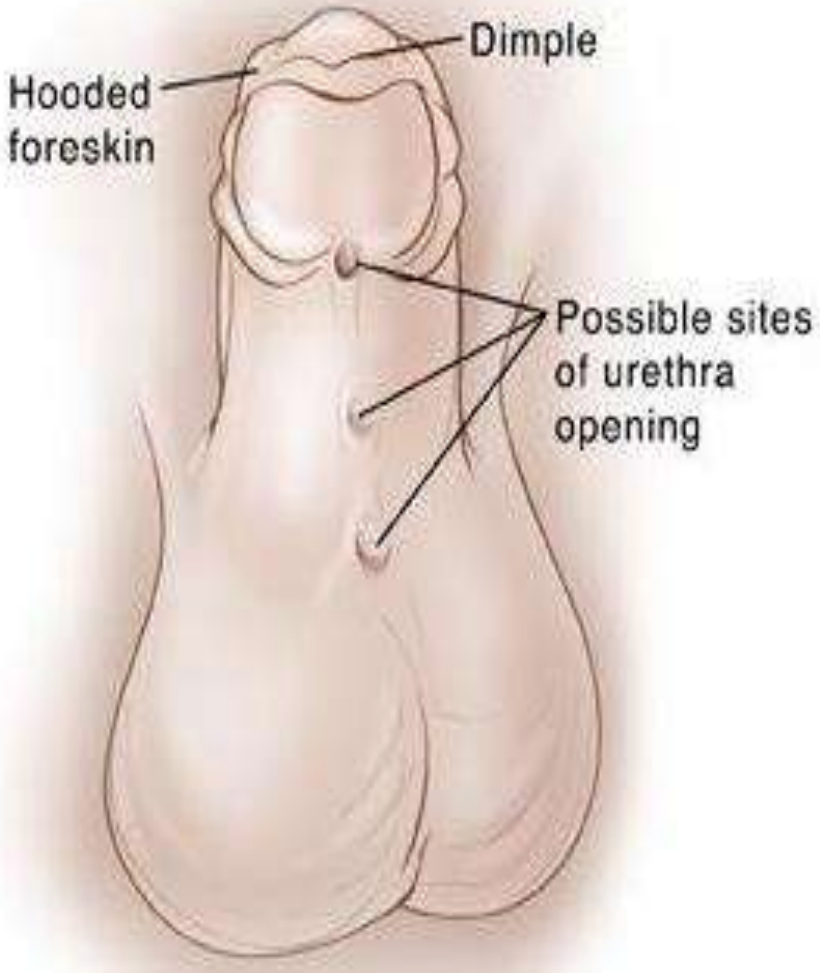
- **Definition**

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).

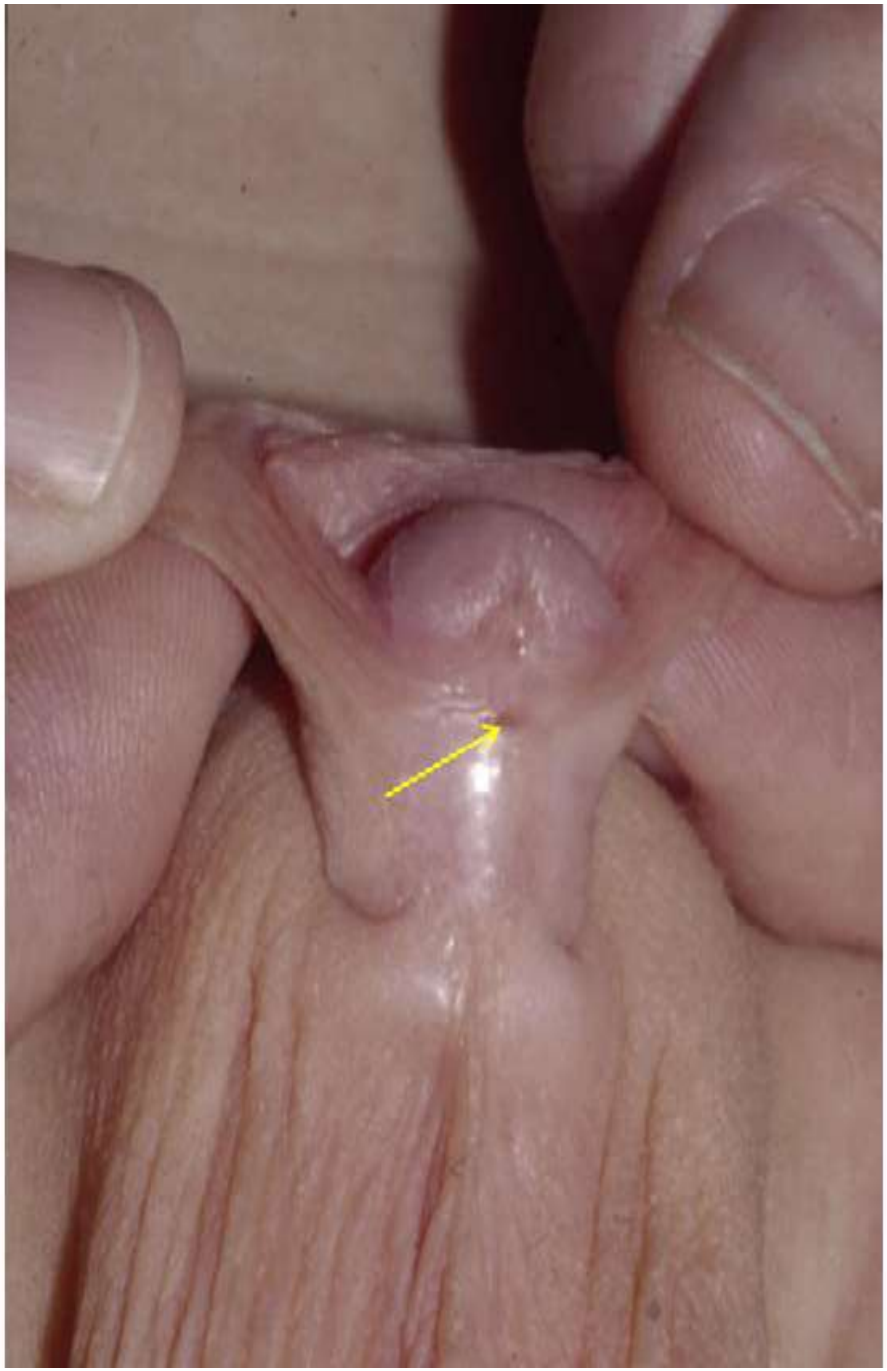
- It is the most common congenital malformation of the urethra. It occurs in 1 in 250 live male births



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

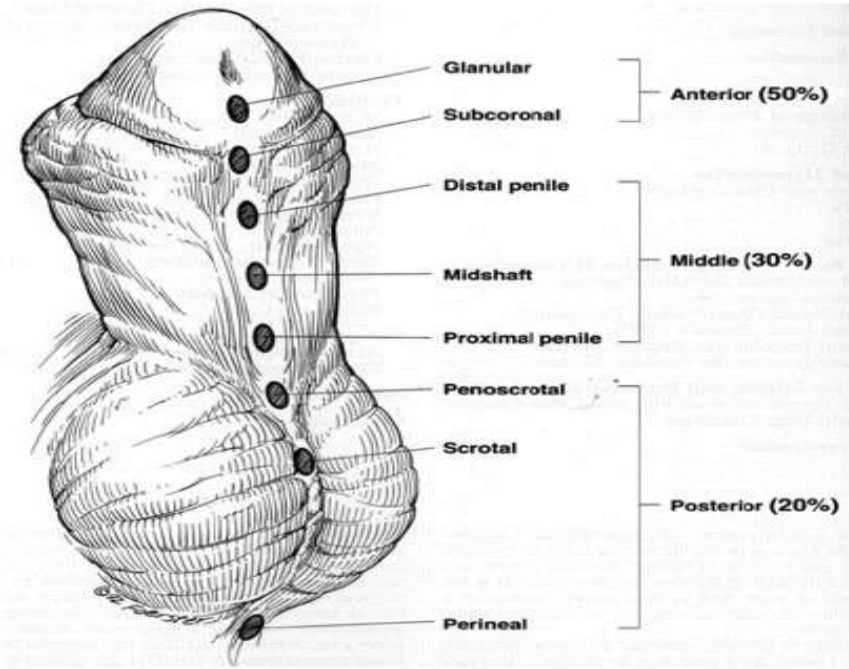
Classification

- Hypospadias can be classified according to the anatomical location of the urethral meatus

• **Anterior** (or distal)—glanular, coronal, and subcoronal (~50%)

• **Middle—**distal penile, midshaft, and proximal penile (~30%)

• **Posterior** (or proximal)—penoscrotal, scrotal, and perineal (~20%)



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, it is 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

Treatment

- 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 month 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, urethral diverticulum, and failed procedures requiring reoperation

Nocturnal enuresis

- **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**: never been 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**: frequency of episodes; daytime symptoms; new or recurrent; family history; UTIs; bowel problems; psychosocial history

- **Examination**: exclude organic causes (neurological disease)

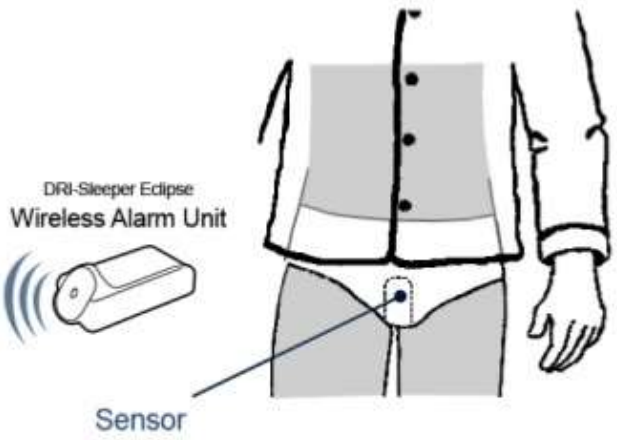
- **Investigation**: urinalysis (infection, specific gravity is reduced in nocturnal polyuria, glucose, protein); voiding diary

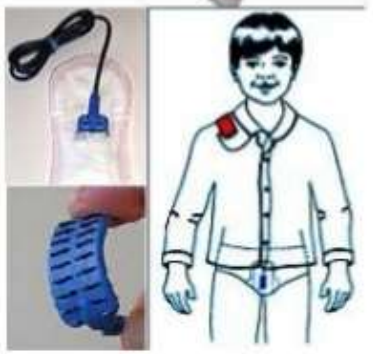
Management

- 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—a tricyclic antidepressant with anticholinergic,antispasmodic properties.

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

- Prognosis

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

Posterior urethral valves (PUV)

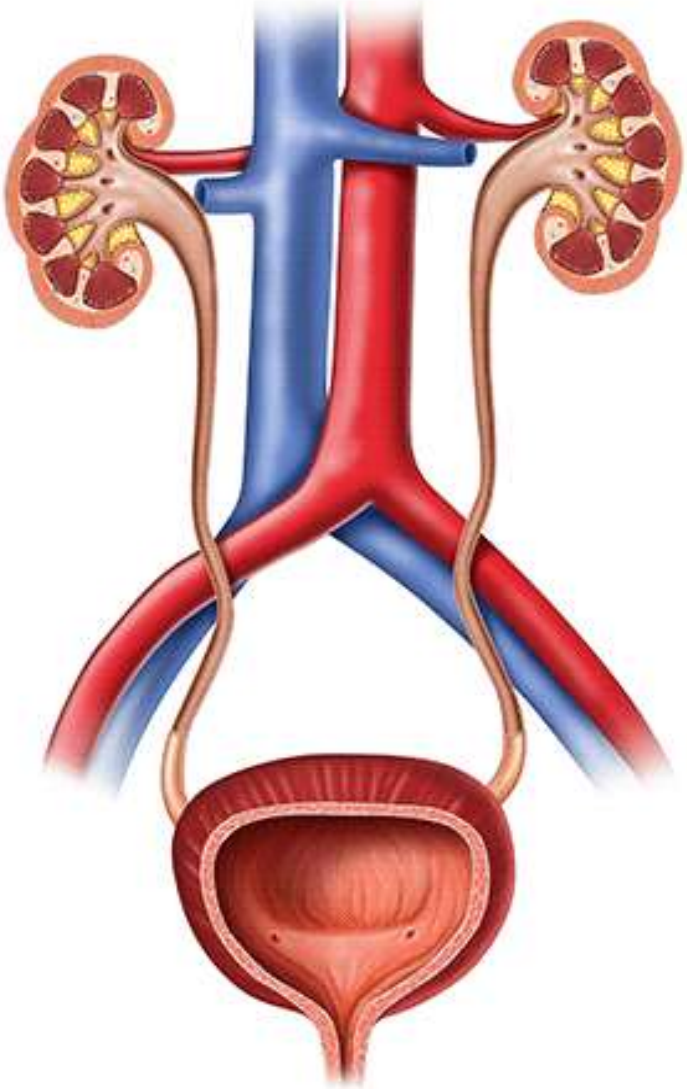
Definition

Posterior urethral valves (PUV) are abnormal congenital mucosal fold in the prostatic (posterior) urethra causing lower urinary tract obstruction. Incidence 1in5000 male.

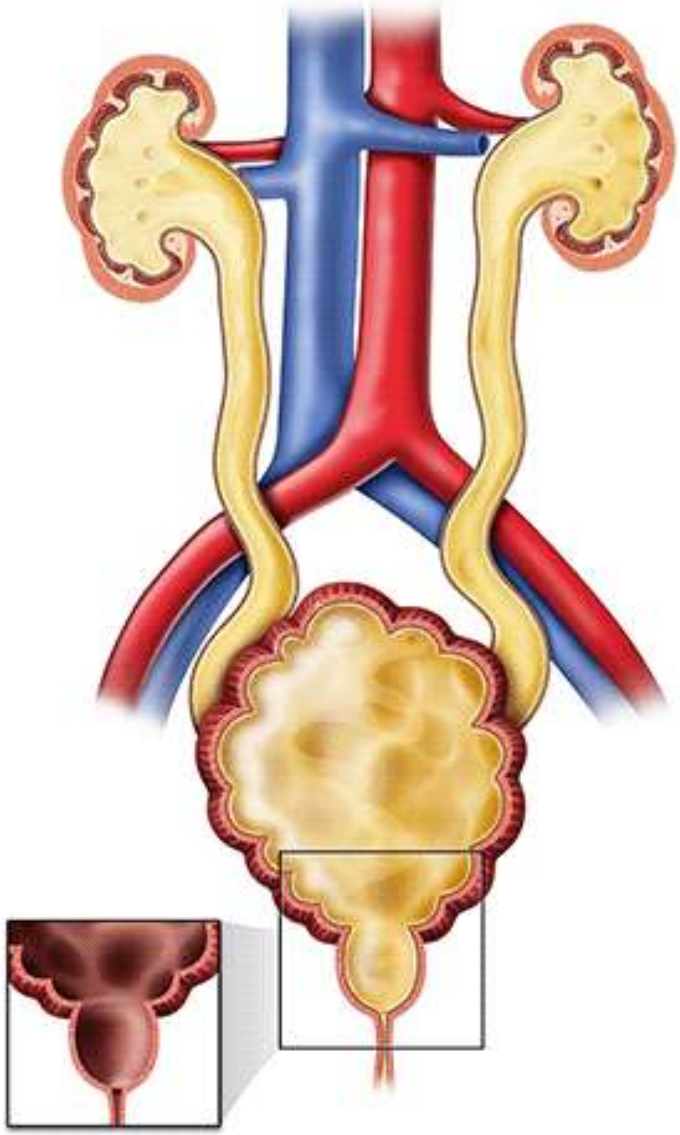
Cause:

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 prostatic urethra, important in the systemic division of posterior valve disorders:

1. Type I - Most common type; due to anterior fusing of the *plicae colliculi*, mucosal fins extending from the bottom of the verumontanum distally along the prostatic and membranous urethra
2. Type II - Least common variant; vertical or longitudinal folds between the verumontanum and 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

Investigation

- Ultrasound scan of kidneys and bladder.
- VCUG shows distended and elongated posterior urethra, partially filled anterior urethra, bladder neck hypertrophy; GOLD STANDARD FOR DIAGNOSING PUV.
- Isotope renal scan (MAG-3, DMSA) …… renal function.
- Video urodynamics allows diagnosis of associated voiding dysfunction, urethra, bladder neck

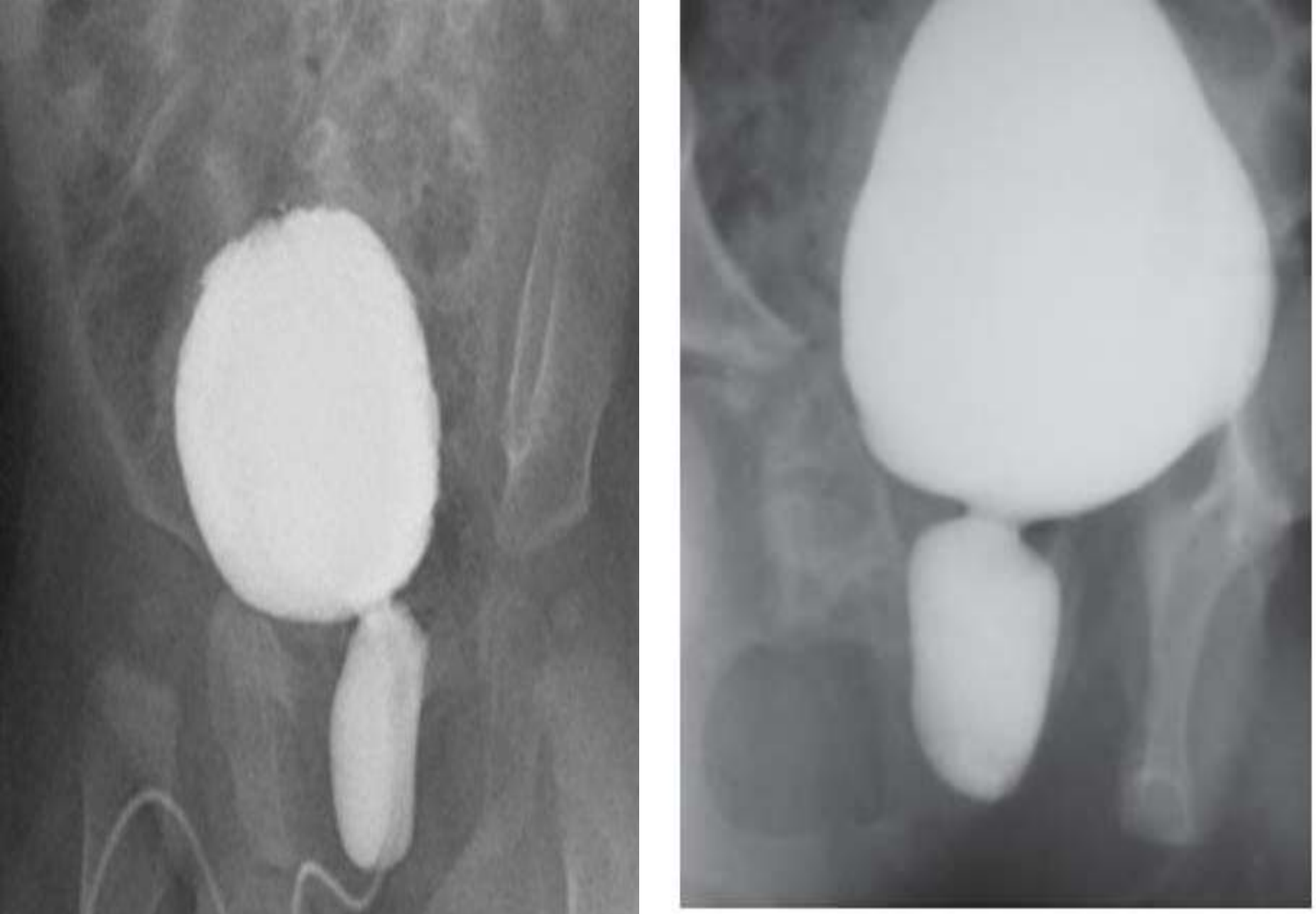


Figure 15.9 VCUG in infant with posterior urethral valves shows dilated elongated prostatic urethra and thickened bladder neck.

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 vesicostomy is indicated (communicating stoma between the bladder dome and suprapubic abdominal wall, allowing free drainage of urine). An alternative is ureterostomy drainage. Valve ablation is performed at a later stage.

Extrophy of the Bladder

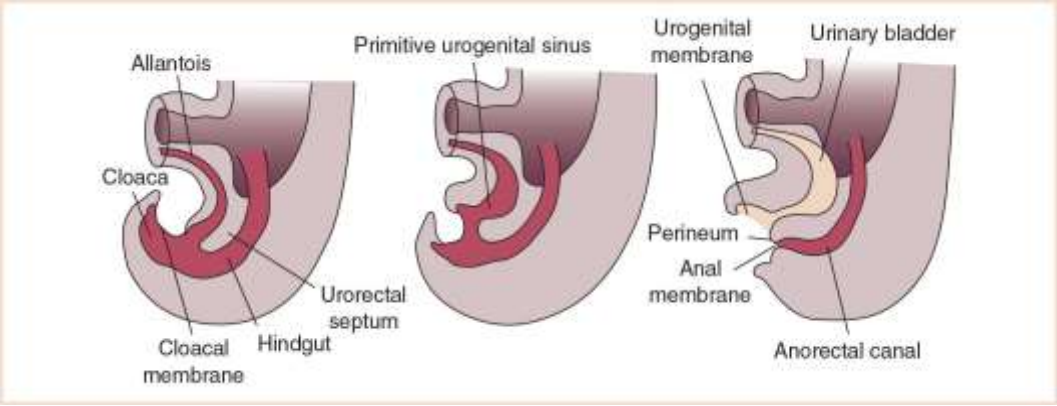
Bladder extrophy results from defective development of the anterior bladder and lower abdominal walls, leaving the posterior bladder wall lying exposed on the abdomen.

M:F >2:1

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

Embryology

- An embryological malformation results in the abnormal overdevelopment of the cloacal membrane, which prevents in-growth of lower abdominal tissues.
- The cloacal membrane 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 (bladder extrophy, cloacal extrophy, OR epispadias).



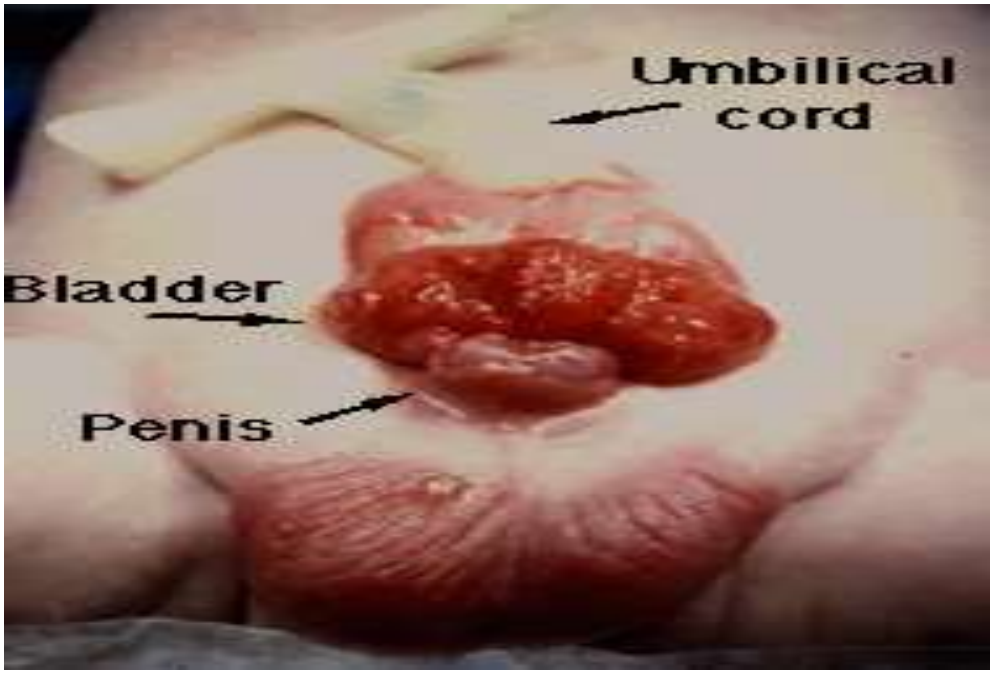
Associated anomaly:

Urinary tract defect:VUR.

Bone defect: Widening of the pubic 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.

- Trauma to the bladder mucosa can eventually result in squamous metaplasia, cystitis cystica, or adenocarcinoma and squamous cell 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.

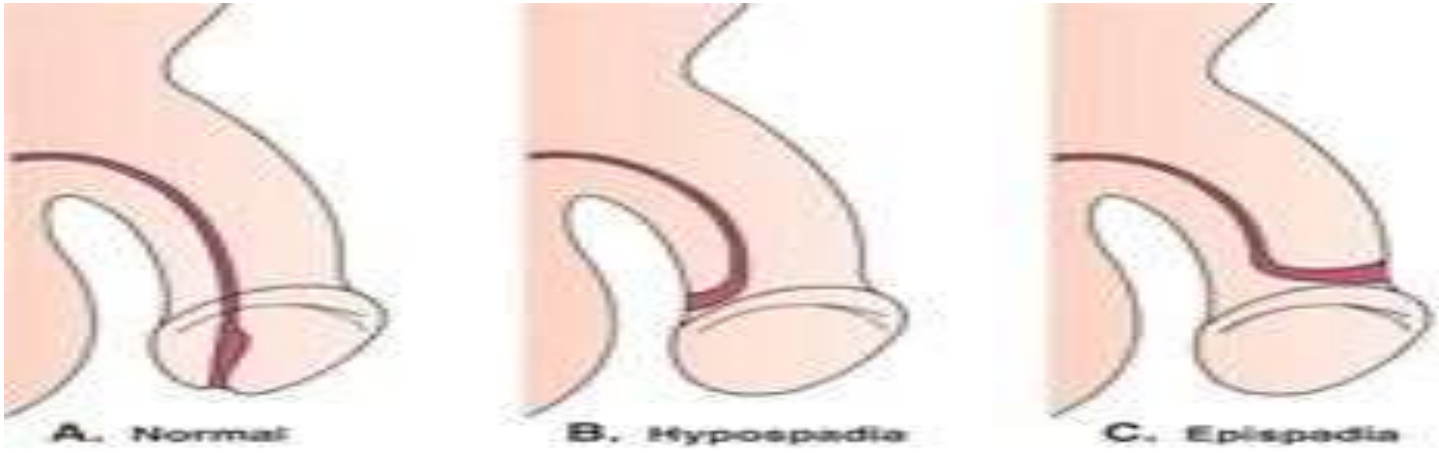
Prognosis

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

- Sexual function and libido are normal in extrophy patients.

Epispadias

- The urethra open in the dorsal surface of the penis any where from the glans, penile shaft or most commonly the penopubic region .
  - 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

- **at 6–12 months >>** This involves ***urethroplasty*** with functional and cosmetic reconstruction of the external genitalia (penile lengthening and correction of chordee) .

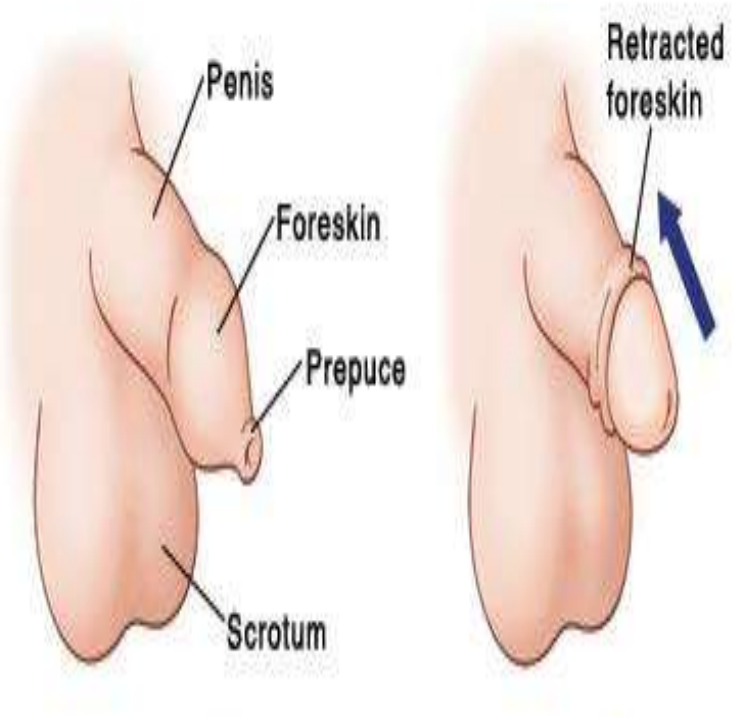
- The modified Cantwell–Ransley technique is commonly used in males. It describes mobilizing the urethra to the ventral aspect of the penis, with advancement of the urethral meatus onto the glans with a reverse MAGPI (meatal advancement-glanuloplasty) .

- **From age 4–5 years** >> when children can be toilet trained, ***bladder neck reconstruction*** can be performed (Young–Dees–Leadbetter procedure). This achieves continence, and any bladder residuals may then be emptied by urethral catheterization.

Phimosis

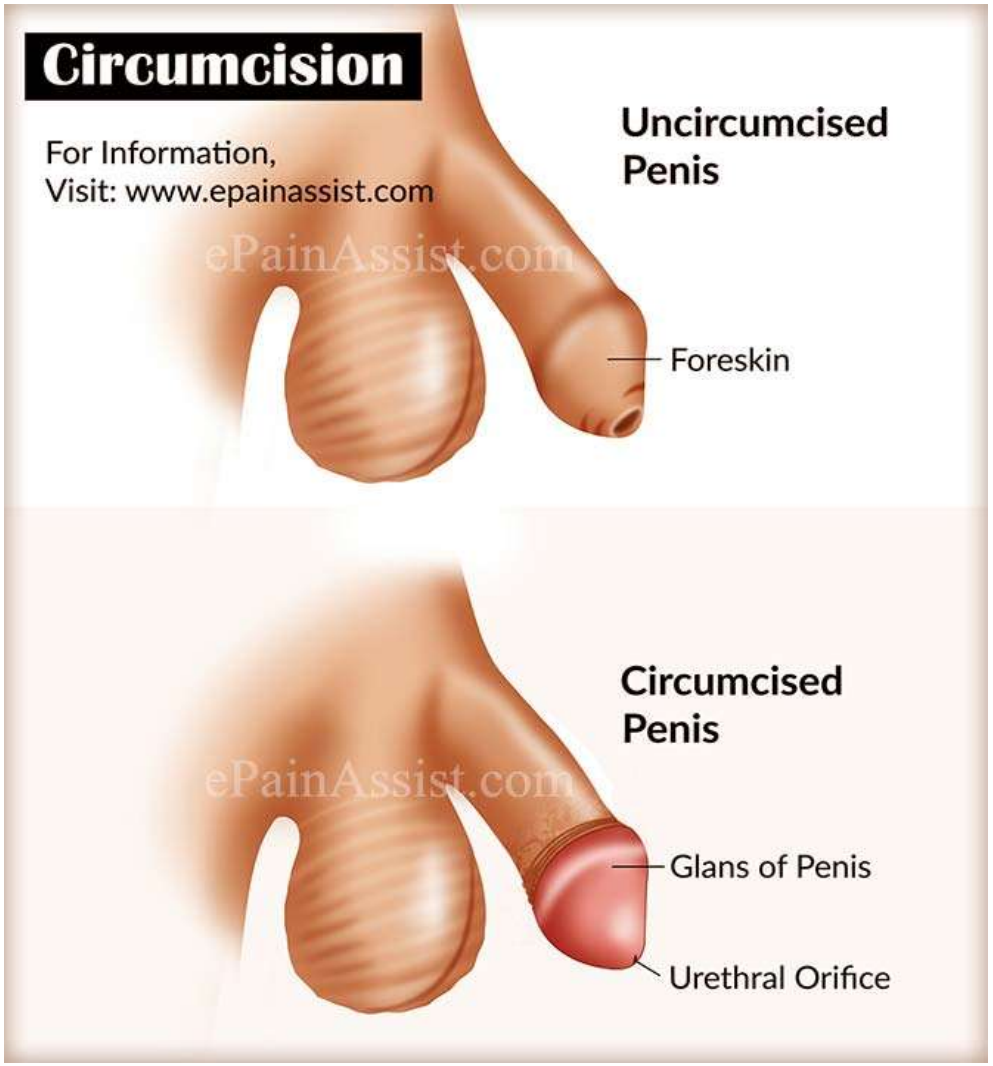
Is when the foreskin cannot be retracted behind the glans.

- A physiological phimosis is 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)



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

- **Recurrent balanitis in uncircumcised males can cause new phimosis**

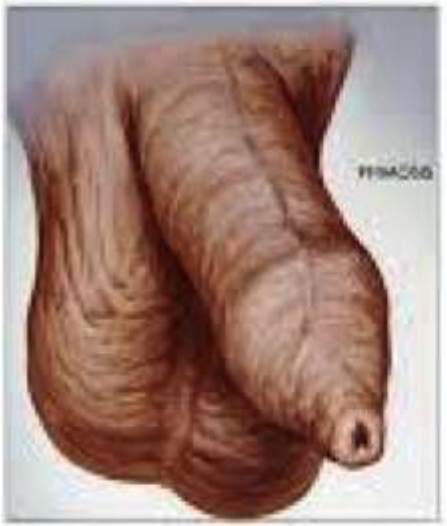


Treatment

- **Older children** with phimosis, suffering recurrent infection (balanitis), can be treated with a 6-week course of topical 0.1% dexamethasone cream, which acts to soften the phimosis and allow foreskin retraction (avoid circumcision where possible).
- **Adults** may require a dorsal slit or circumcision for recurrent balanitis, voiding obstruction, or difficulties with sexual intercourse.

Paraphimosis

- Paraphimosis is 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.



phimosis

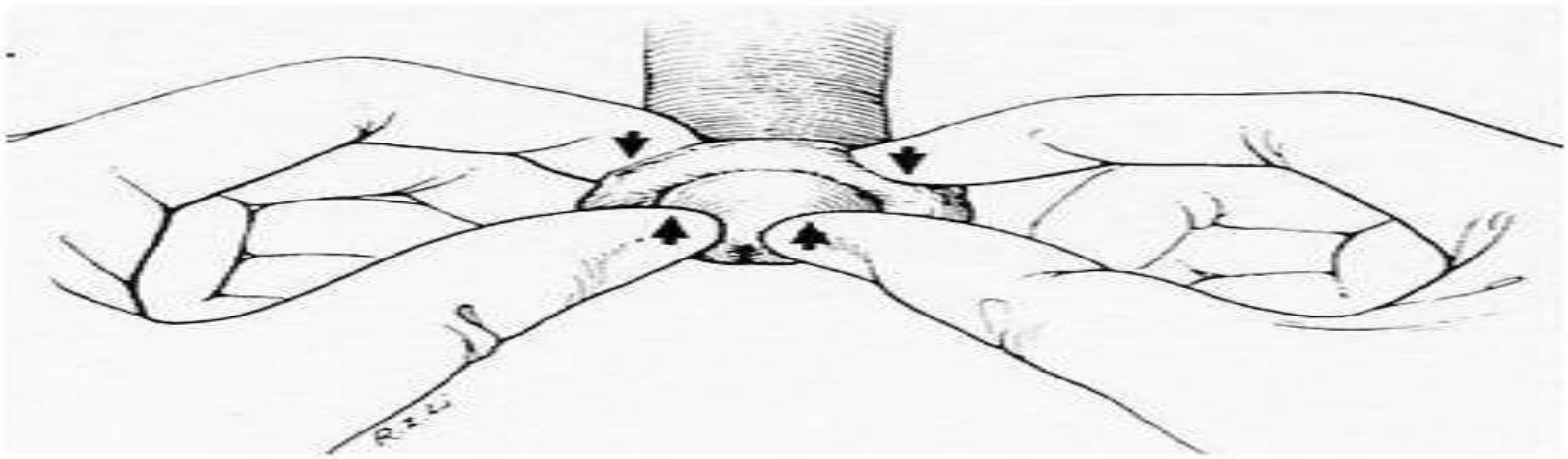
paraphimosis

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

Treatment :

- Manual reduction is preferred using ice packs, elastic compression, and topical anesthetic such as 2% lidocaine gel.
- 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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