Pediatric Urology

Dr. Saddam Al Demour
MD, MRCS, FACS, FEBU
School of Medicine
The University of Jordan

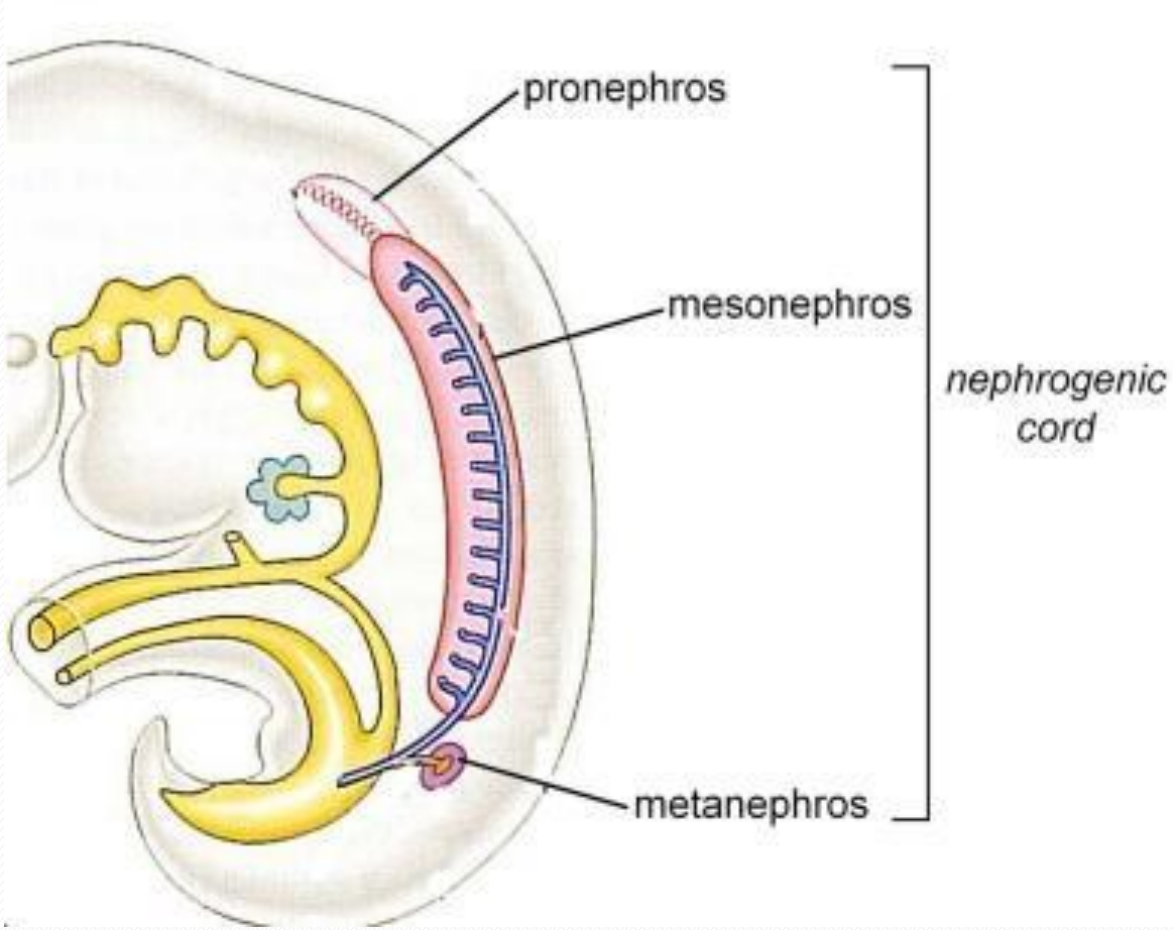
Nephric system

- The nephric system develops progressively as three distinct entities:

1. Pronephros
2. Mesonephros
3. Metanephros

Pronephros

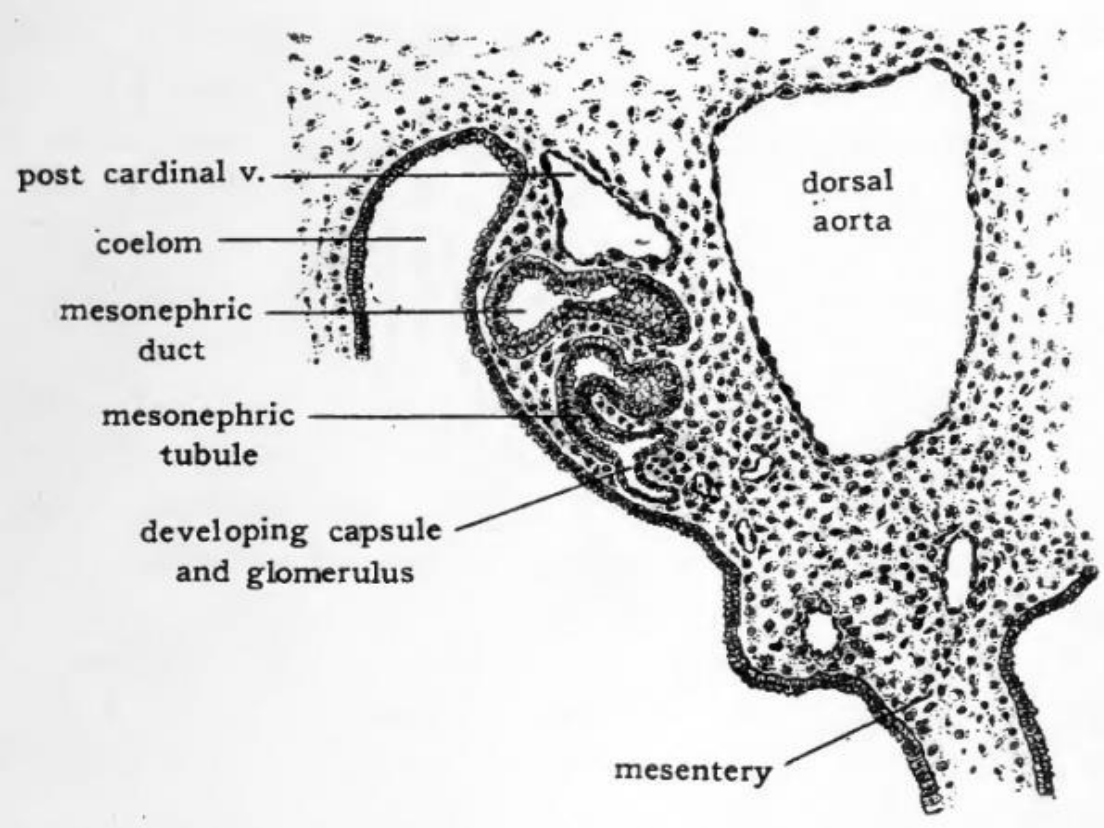
- The pronephros is the earliest nephric stage in humans
- It extends from the 4th to the 14th somites and consists of six to ten pairs of tubules
- The pronephros is a vestigial structure that disappears completely by the 4th week of embryonic life.


pronephros
mesonephros
metanephros
nephrogenic cord

Mesonephros

- The principal excretory organ during early embryonic life (4–8 weeks)
- The mesonephric tubules develop from the intermediate mesoderm caudal to the pronephros shortly before pronephric degeneration.
- The mesonephric tubules differ from those of the pronephros in that they develop a cuplike outgrowth into which a knot of capillaries is pushed.

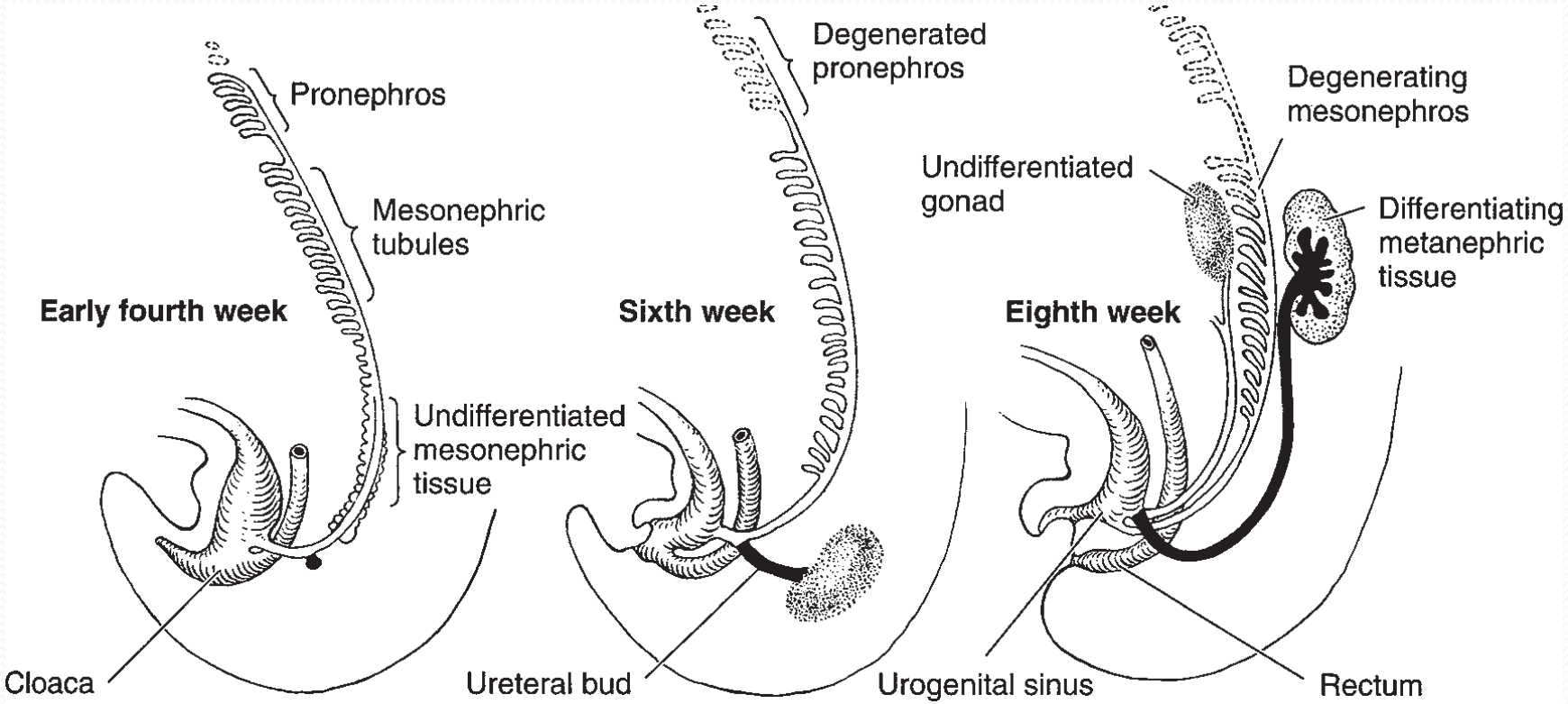
- This is called Bowman's capsule, and the tuft of capillaries is called a glomerulus.
- The mesonephros, which forms early in the 4th week, reaches its maximum size by the end of the 2nd month.



Metanephros

- The metanephros, the final phase of development of the nephric system, originates from both the intermediate mesoderm and the mesonephric duct.
- Development begins with a budlike outgrowth from the mesonephric duct as it bends to join the cloaca.
- This ureteral bud grows cephalad and collects mesoderm from the nephrogenic cord of the intermediate mesoderm

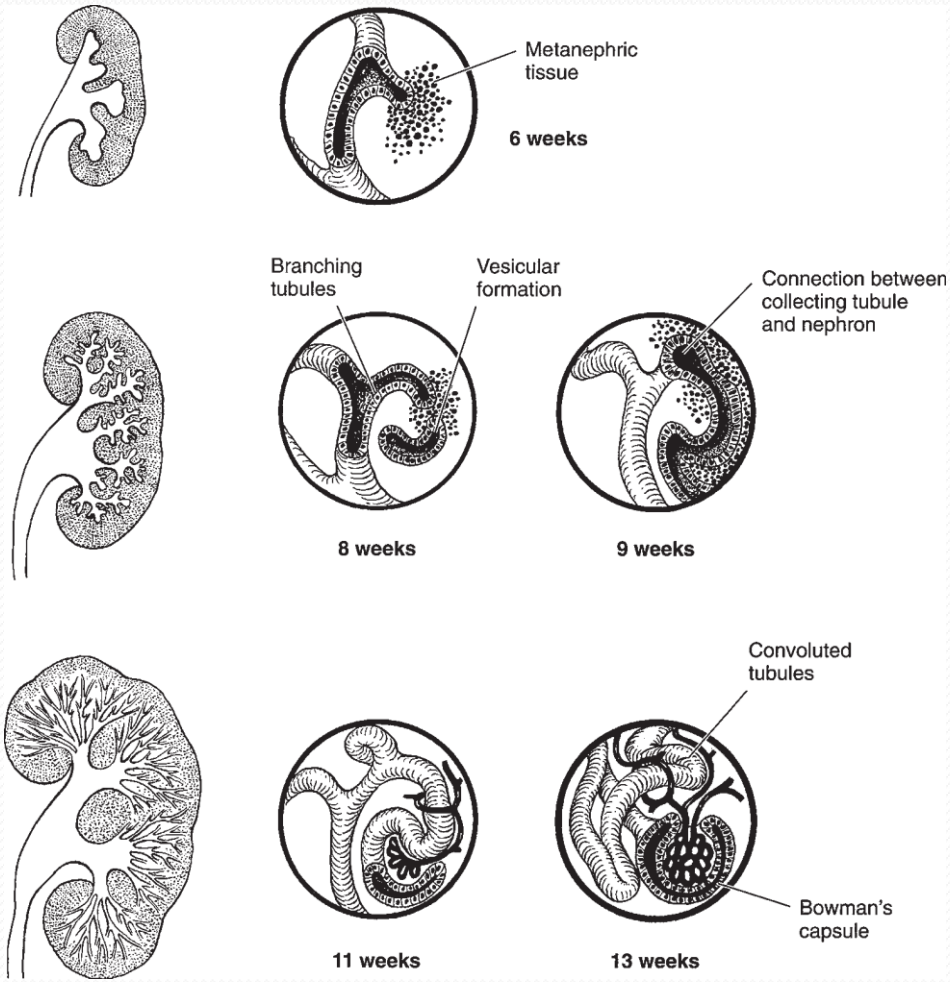
Schematic representation of the development of the nephric system. Only a few of the tubules of the pronephros are seen early in the 4th week, while the mesonephric tissue differentiates into mesonephric tubules that progressively join the mesonephric duct. The first sign of the ureteral bud from the mesonephric duct is seen. At 6 weeks, the pronephros has completely degenerated and the mesonephric tubules start to do so. The ureteral bud grows dorsocranially and has met the metanephrogenic cap. At the 8th week, there is cranial migration of the differentiating metanephros. The cranial end of the ureteric bud expands and starts to show multiple successive outgrowths



- This Mesoderm with the meta-nephric cap moves, with the growing ureteral bud, more and more cephalad
- During this cephalic migration, the metanephric cap becomes progressively larger, and rapid internal differentiation takes place
- the cephalic end of the ureteral bud expands within the growing mass of metanephrogenic tissue to form the renal pelvis
- Numerous outgrowths from the renal pelvic dilatation push radially into this growing mass and form hollow ducts that branch and rebranch as they push toward the periphery. These form the primary collecting ducts of the kidney

- Mesodermal cells become arranged in small vesicular masses that lie close to the blind end of the collecting ducts. Each of these vesicular masses will form a uriniferous tubule draining into the duct nearest to its point of origin.
- These vesicular masses develop a central cavity and become S-shaped. One end of the S coalesces with the terminal portion of the collecting tubules, resulting in a continuous canal. The proximal portion of the S develops into the distal and proximal convoluted tubules and into Henle's loop; the distal end becomes the glomerulus and Bowman's capsule.

Progressive stages in the differentiation of the nephrons and their linkage with the branching collecting tubules. A small lump of metanephric tissue is associated with each terminal collecting tubule. These are then arranged in vesicular masses that later differentiate into a uriniferous tubule draining into the duct near which it arises. At one end, Bowman's capsule and the glomerulus differentiate; the other end establishes communication with the nearby collecting tubules.

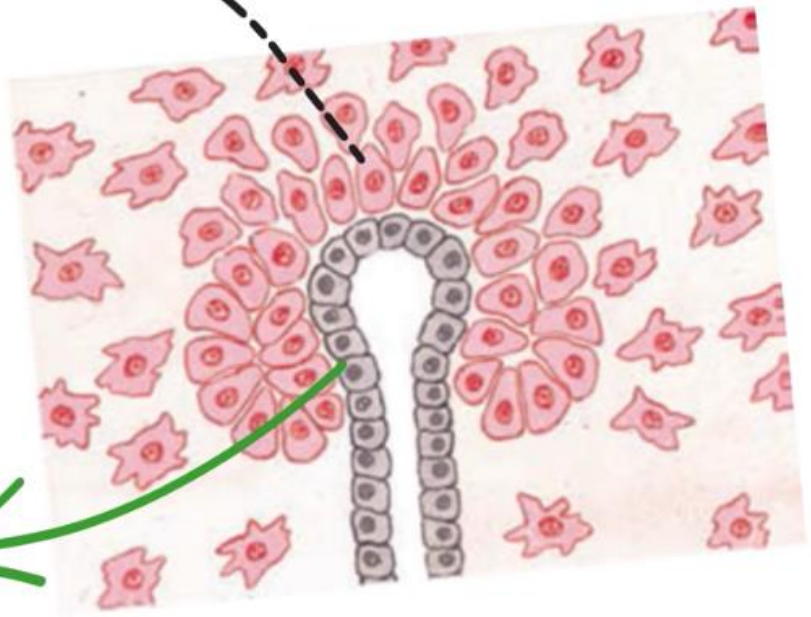


- As a Summary : Certain features of these three phases of development
- (1) The three successive units of the system develop from the intermediate mesoderm.
- (2) The tubules at all levels appear as independent primordia and only secondarily unite with the duct system.
- (3) The nephric duct is laid down as the duct of the pronephros and develops from the union of the ends of the anterior pronephric tubules.
- (4) This pronephric duct serves subsequently as the mesonephric duct and as such gives rise to the ureter.
- (5) The nephric duct reaches the cloaca by independent caudal growth.
- (6) The embryonic ureter is an outgrowth of the nephric duct, yet the kidney tubules differentiate from adjacent metanephric blastema.

Molecular Mechanisms of Renal and Ureteral Development

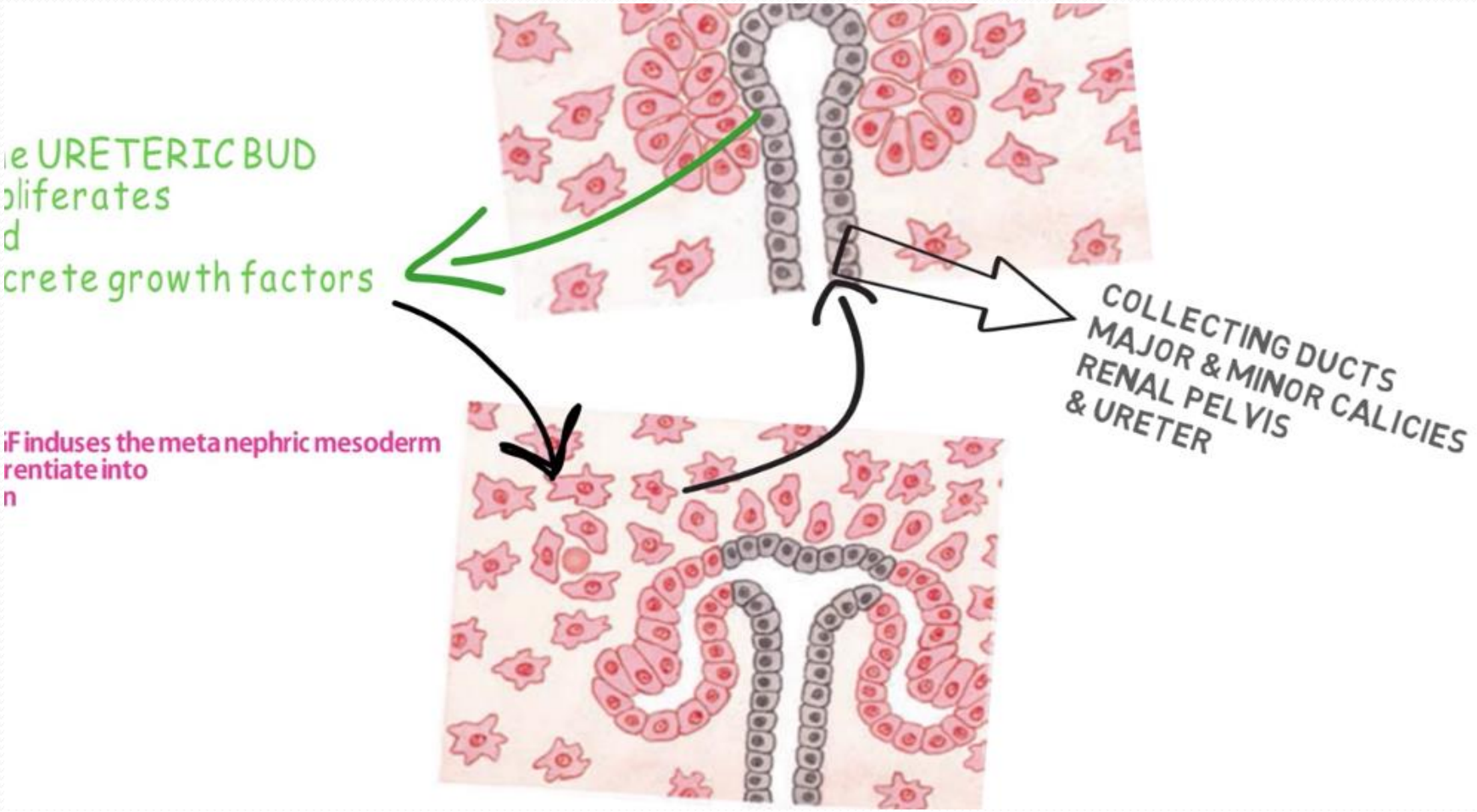
- The kidney and the collecting system originate from the interaction between the mesonephric duct(ureteric Bud) and the metanephric mesenchyme.
- Reciprocal induction between the UB and MM results in branching and elongation of the UB from the collecting system and in condensation and epithelial differentiation of MM around the branched tips of the UB.

Each newly formed collecting tubule is covered at its distal end by a metanephric tissue cap.



The URETERIC BUD
proliferates
and
secrete growth factors

These GF induses the meta nephric mesoderm to differentiate into Nephron


e URETERIC BUD
liferates
d
crete growth factors
F induses the meta nephric mesoderm
rentiate into
n
COLLECTING DUCTS
MAJOR & MINOR CALICIES
RENAL PELVIS
& URETER

- This process of reciprocal induction is dependent on the expression of specific factors:
- Glial cell–derived neurotrophic factor (GDNF) is the primary inducer of ureteric budding
- GDNF interacts with several different proteins from the MM (eg, *Wt-1, Pax2, Eyal, Six1,Sall 1) and from the UB itself (Pax2, Lim1, Ret) resulting in* outgrowth of the UB.
- Proper activation of the *Ret/GDNF signaling pathway in the tip of* UB epithelium appears to be essential in the progression of branching morphogenesis

- Additional specific factors are required for (1) early branching (eg, *Wnt-4 and 11, fgf 7–10); (2) late* branching and maturation (*bmp2, activin); and (3) branching* termination and tubule maintenance (hepatocyte growth factor, transforming growth factor-alpha, epidermal growth factor receptor)

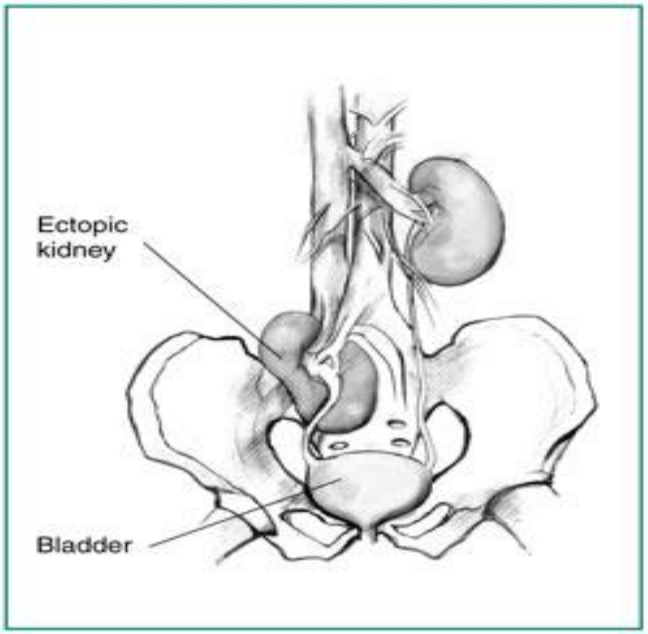
ANOMALIES OF THE NEPHRIC SYSTEM

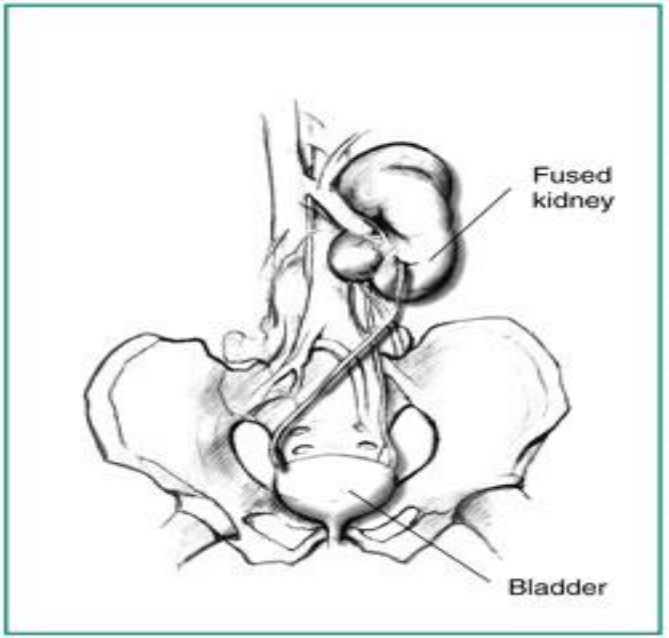
- **ectopic kidney**
- **malrotated kidney**
- **horseshoe kidney.**
- **bifid ureter**
- **duplicated ureter**
- **supernumerary kidneys**

Ectopic kidney

- Failure of the metanephros to ascend
- Two Types:

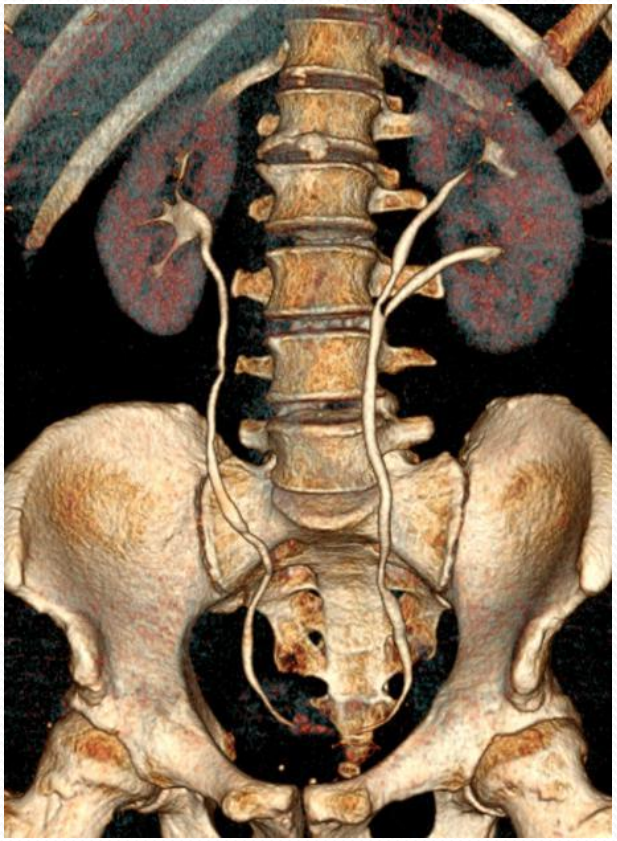
1. simple ectopy: An ectopic kidney on the proper side but low
2. crossed ectopy : on the opposite site

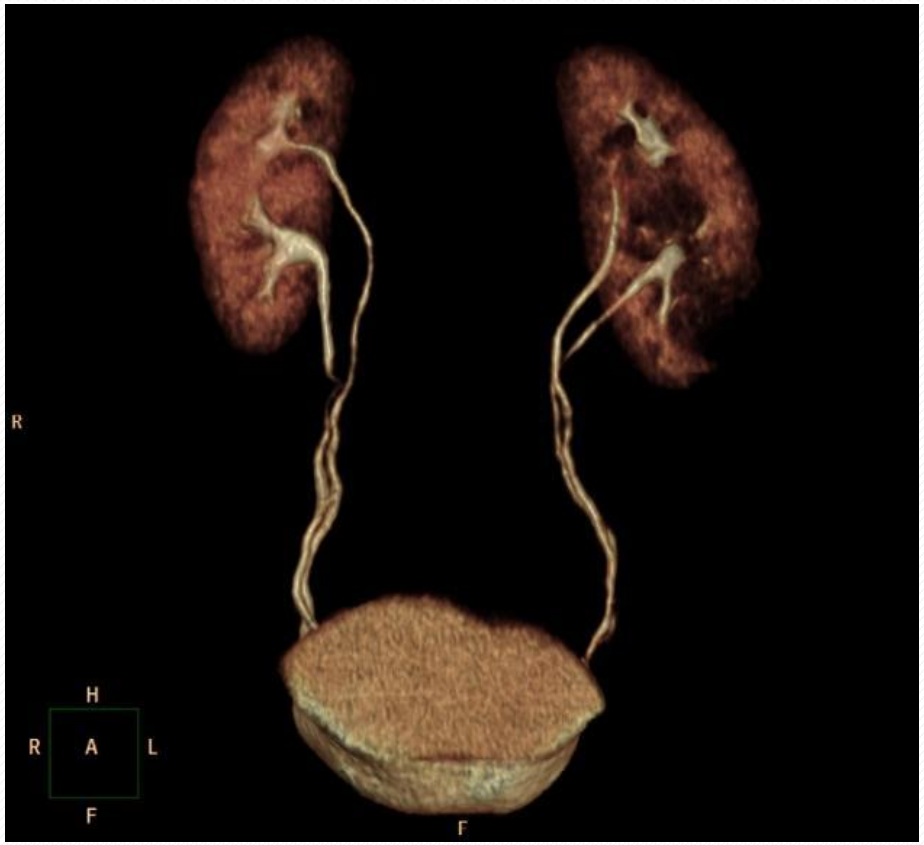




- Malrotated kidney : Failure to rotate during ascent
- **horseshoe kidney :** Fusion of the paired metanephric masses

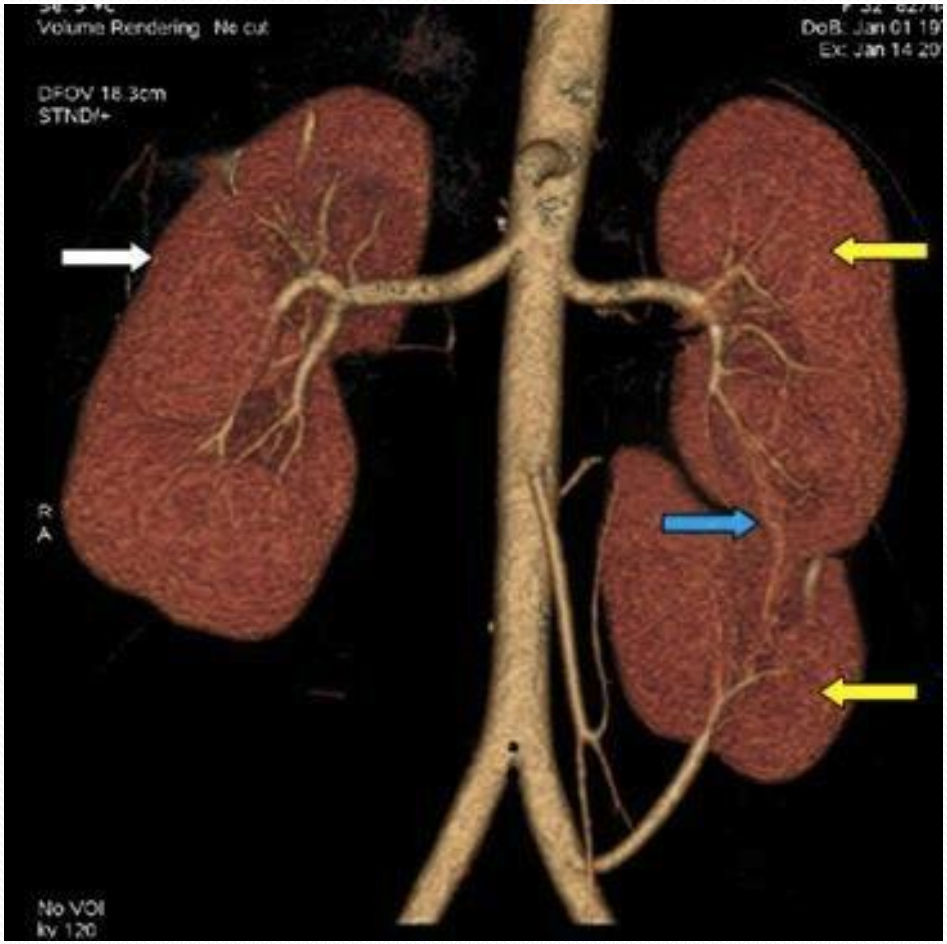
- **bifid ureter :** The ureteral bud from the mesonephric duct may bifurcate causing Bifid Ureter
- **duplicated ureter :** An accessory ureteral bud may develop from the mesonephric duct , **usually meeting the same metanephric** mass





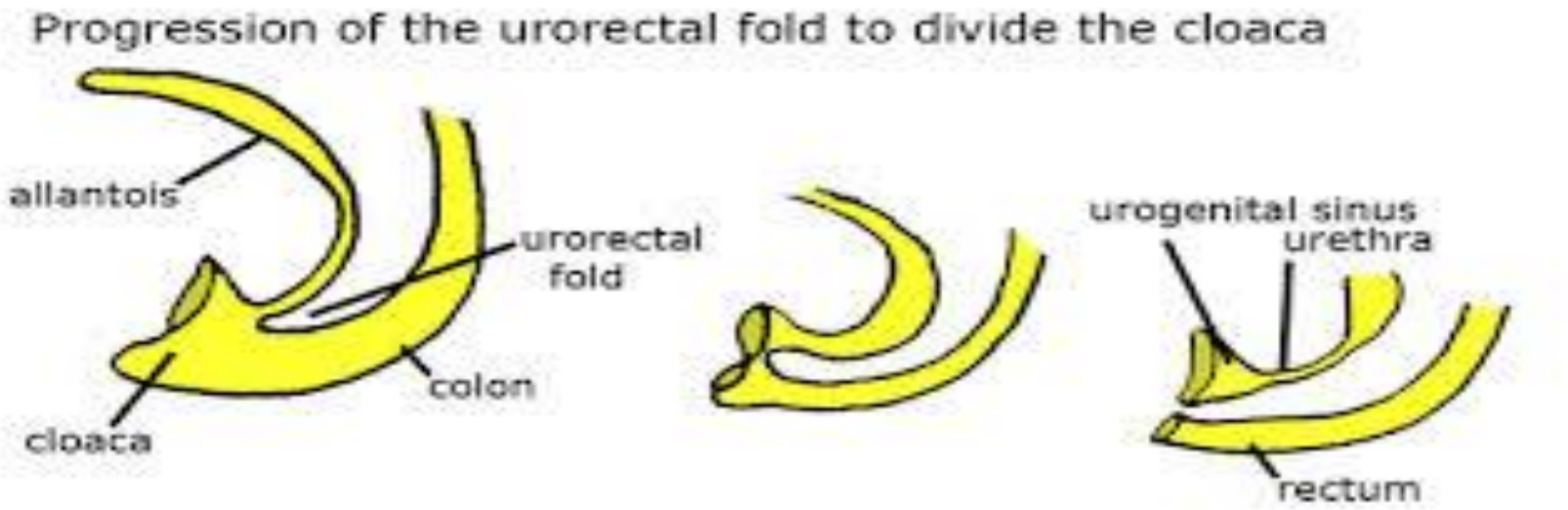
- If the double ureteral buds are close together on the mesonephric duct, they open near each other in the bladder.
- In this case, the main ureteral bud, which is the first to appear and the most caudal on the mesonephric ducts, reaches the bladder first.
- It then starts to move upward and laterally and is followed later by the second accessory bud as it reaches the urogenital sinus. The main ureteral bud (now more cranial on the urogenital sinus) drains the lower portion of the kidney.
- The two ureteral buds reverse their relationship as they move from the mesonephric duct to the urogenital sinus.
- This is why double ureters always cross (Weigert-Meyer law).

- **supernumerary kidneys :** each bud has a separate metanephric mass. Its Rare
- Solitary Kidney : Lack of development of a ureteral bud



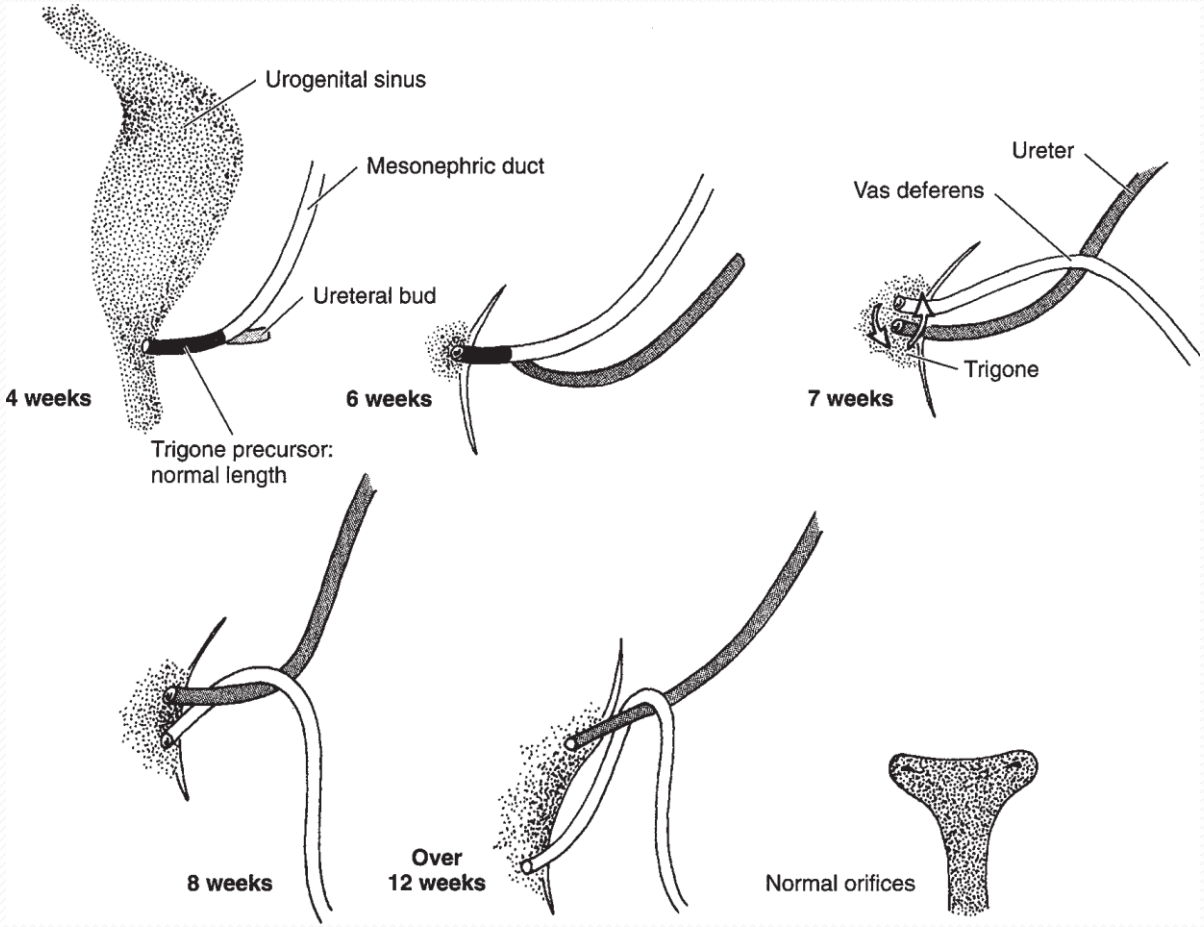
VESICOURETHRAL UNIT

- cloaca progressively divides into two compartments by the caudal growth of the urorectal fold (Mesoderm) into a ventral portion (urogenital sinus) and a dorsal portion (rectum) .

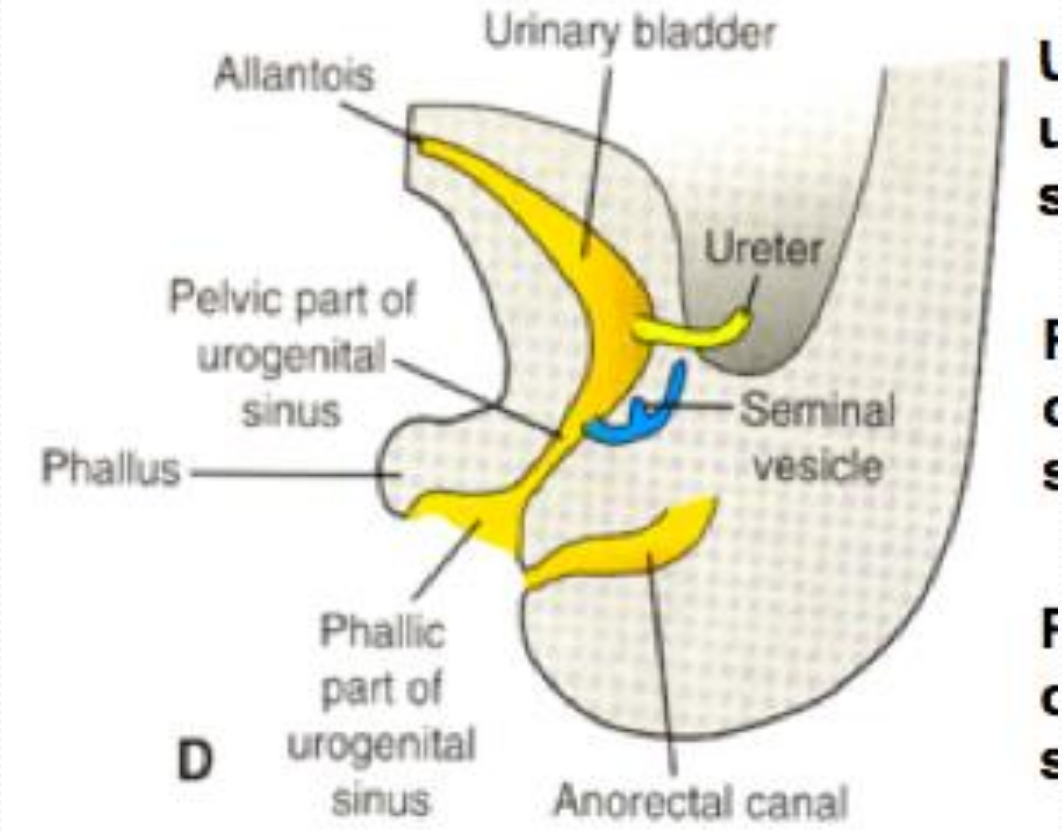


- The division completed during the 7th week
- the urogenital sinus receives the mesonephric ducts
- The caudal end of the mesonephric duct distal to the ureteral bud is progressively absorbed into the urogenital sinus
- The absorbed mesoderm of the mesonephric duct will later be differentiated as the trigonal structure, which is the only mesodermal inclusion in the endodermal vesicourethral unit.

The development of the ureteral bud from the mesonephric duct and the relationship of both to the urogenital sinus. The ureteral bud appears at the 4th week. The mesonephric duct distal to this ureteral bud is gradually absorbed into the urogenital sinus, resulting in separate endings for the ureter and the mesonephric duct. The mesonephric tissue that is incorporated into the urogenital sinus expands and forms the trigonal tissue.



- The urogenital sinus can be divided into two main segments:
- 1. The ventral and pelvic portion forms the bladder, part of the urethra in males, and the whole urethra in females. This portion receives the ureter.
- 2. The urethral, or phallic, portion receives the mesonephric and the fused Mullerian ducts. This will be part of the urethra in males and forms the lower fifth of the vagina and the vaginal vestibule in females

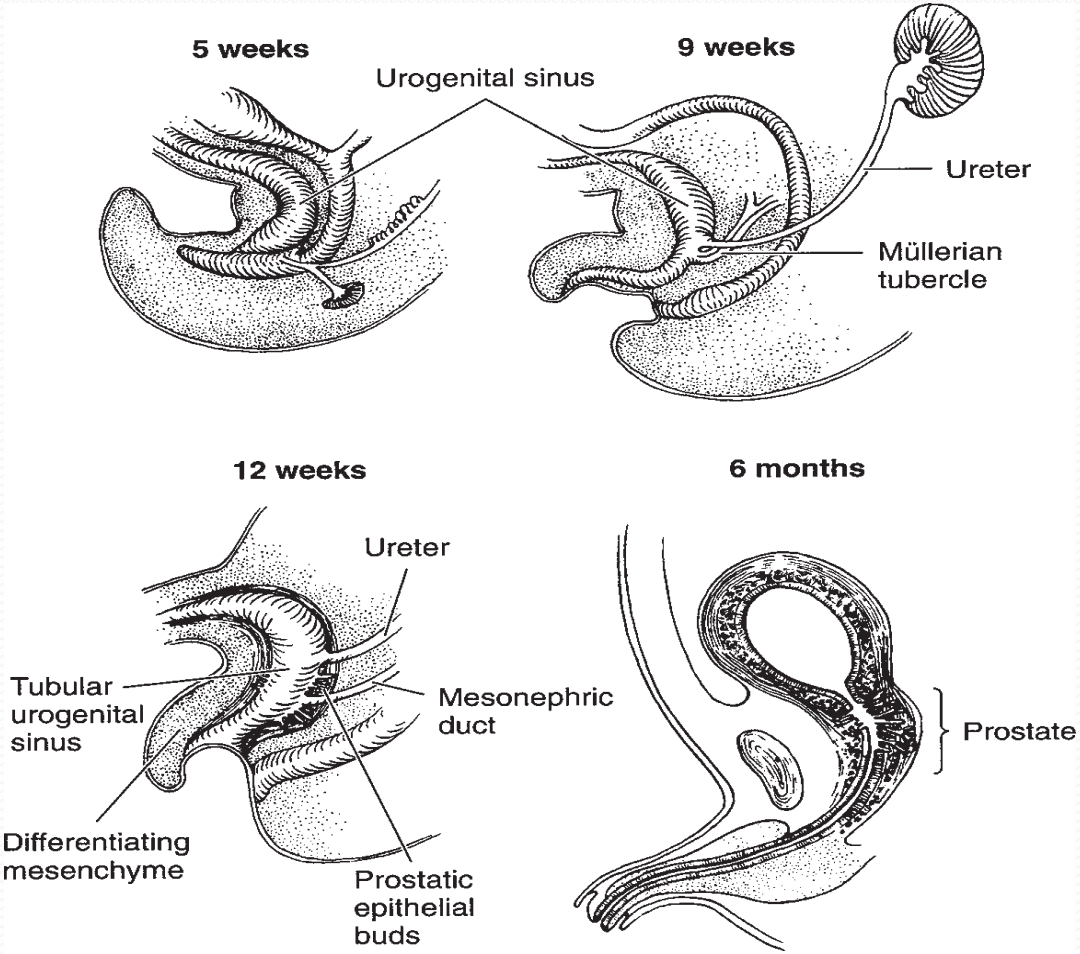

Urinary bladder
Allantois
Ureter
Pelvic part of
urogenital
sinus
Seminal
vesicle
Phallus
Phallic
part of
urogenital
sinus
Anorectal canal
D

Prostate

- The prostate develops as multiple solid outgrowths of the urethral epithelium both above and below the entrance of the mesonephric duct
- These simple tubular outgrowths begin to develop in five distinct groups at the end of the 11th week and are complete by the 16th week
- From the five groups of epithelial buds, five lobes are eventually formed: anterior, posterior, median, and two lateral Lobes
- Prostate development results from complex interaction between urogenital sinus epithelium and mesenchyme in the presence of androgens
- Several members of the fibroblast growth factor (eg, FGF7 and FGF10) play essential role in prostate development, though they do not appear to be directly regulated by androgens.

Differentiation of the urogenital sinus in males. At the 5th week, the progressively growing urorectal septum separates the urogenital sinus from the rectum. The former receives the mesonephric duct and the ureteral bud.

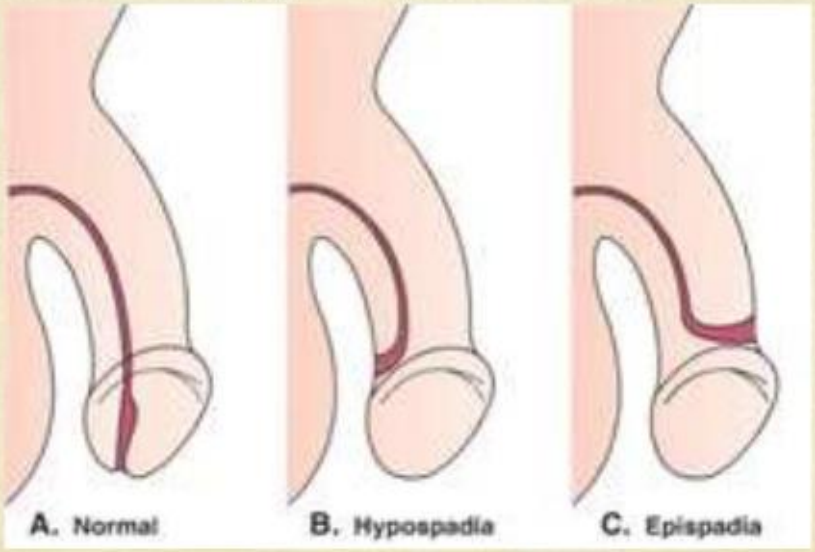
It retains its tubular structure until the 12th week, when the surrounding mesenchyme starts to differentiate into the muscle fibers around the whole structure. The prostate gland develops as multiple epithelial outgrowths just above and below the mesonephric duct. During the 3rd month, the ventral part of the urogenital sinus expands to form the bladder proper; the pelvic part remains narrow and tubular, forming part of the urethra



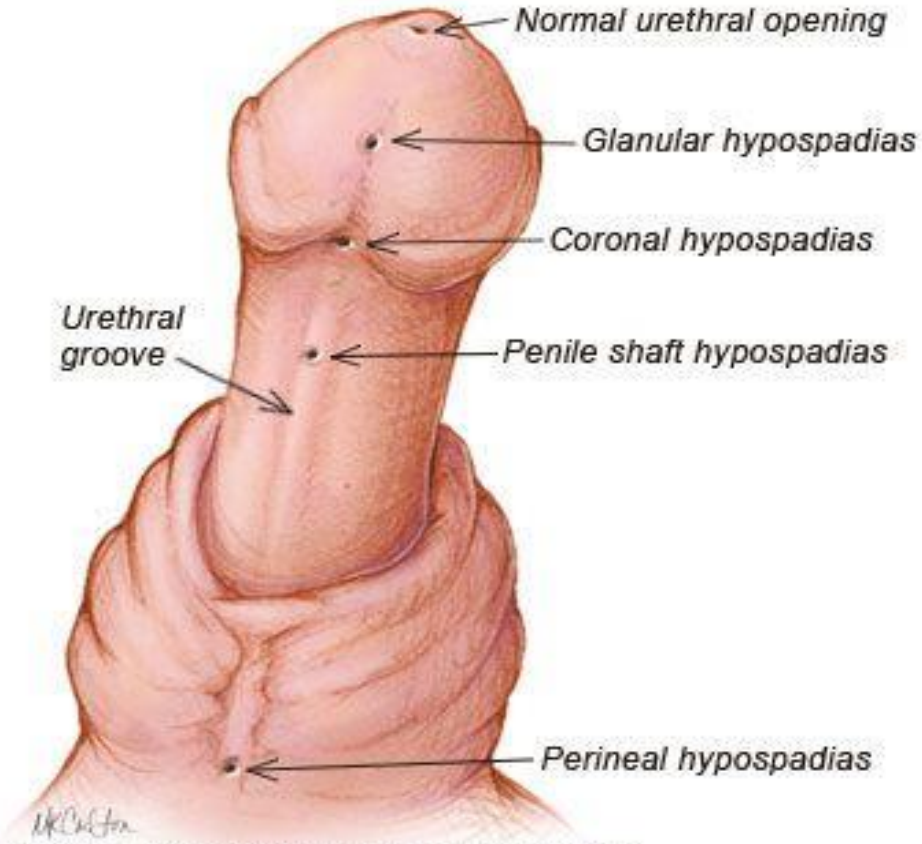
ANOMALIES OF THE VESICOURETHRAL UNIT

- Failure of the cloaca to subdivide is rare and results in a **persistent Cloaca**
- Incomplete subdivision is more frequent, ending with **rectovesical, rectourethral, or rectovestibular fistulas (usually with imperforate anus or anal atresia).**
- Failure of descent or incomplete descent of the bladder leads to a **urinary umbilical fistula (urachal fistula), urachal cyst, or urachal diverticulum depending on the stage** and degree of maldescent
- An **epispadias** is a rare type of malformation of the penis in which the urethra ends in an opening on the upper aspect (the dorsum) of the penis
- Development of the genital primordia in an area more caudal than normal can result in formation of the corpora cavernosa just caudal to the urogenital sinus outlet, with the urethral groove on its dorsal surface. This defect results in complete or incomplete **epispadias depending on its degree**

Epispadias vs Hypospadias



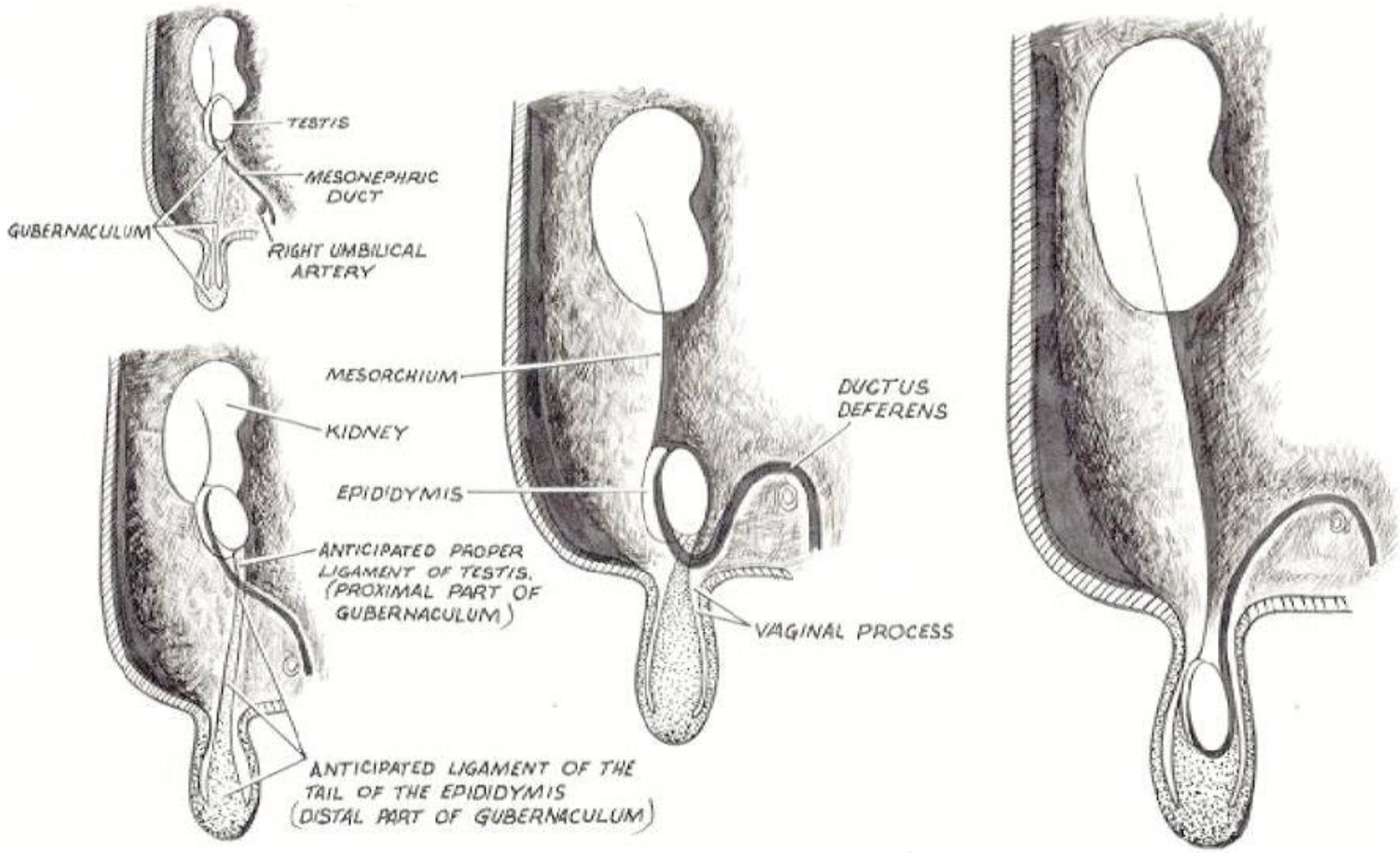
- Hypospadias is a condition in which the opening of the urethra is on the underside of the penis, instead of at the tip
- Failure of fusion of urethral folds leads to various grade
- This defect, because of its mechanism, never extends proximal to the bulbous urethra. This is in contrast to epispadias, which usually involves the entire urethra up to the internal meatus.



GONADS

- The primitive sex glands make their appearance during the 5th and 6^{th} weeks
- At the 6th week, the gonad consists of a superficial germinal epithelium and an internal blastema.
- During the 7th week, the gonad begins to assume the characteristics of a testis or ovary. Differentiation of the ovary usually occurs somewhat later than differentiation of the testis.

- By the 3rd month of fetal life, the testis is located retroperitoneally in the false pelvis
- A fibromuscular band (the gubernaculum) extends from the lower pole of the testis through the developing muscular layers of the anterior abdominal wall to terminate in the subcutaneous tissue of the scrotal swelling.
- Just below the lower pole of the testis, the peritoneum herniates as a diverticulum along the anterior aspect of the gubernaculum eventually reaching the scrotal sac through the anterior abdominal muscles (the processus vaginalis).
- The testis remains at the abdominal end of the inguinal canal until the 7th month. It then passes through the inguinal canal behind (but invaginating) the processus vaginalis and reaches the scrotal sac by the end of the 8th month.



A SCHEMATIC DIAGRAM OF THE DESCENT OF THE TESTIS.

Cryptochidism

- Retention of the testis in the abdomen or arrest of its descent at any point along its natural pathway
- may be either unilateral or bilateral(about 10 percent of the time, both testicles are undescended)
- The exact cause of an undescended testicle isn't known. A combination of genetics, maternal health and other environmental factors might disrupt the hormones, physical changes and nerve activity that influence the development of the testicles

Risk factors

- Low birth weight
- Premature birth
- Family history of undescended testicle or other problems of genital development
- Conditions of the fetus that can restrict growth, such as Down syndrome or an abdominal wall defect
- Alcohol use by the mother during pregnancy
- Cigarette smoking by the mother or exposure to secondhand smoke
- Obesity in the mother
- Diabetes in the mother — type 1 diabetes, type 2 diabetes

Complications

- **Testicular cancer**
- **Fertility problems.** Low sperm counts, poor sperm quality and decreased fertility are more likely to occur among men who've had an undescended testicle
- **Testicular torsion:** Testicular torsion is the twisting of the spermatic cord, which contains blood vessels, nerves and the tube that carries semen from the testicle to the penis. Testicular torsion occurs 10 times more often in undescended testicles than in normal testicles.
- **Trauma:** If a testicle is located in the groin, it might be damaged from pressure against the pubic bone

Management

- The vast majority of the time, the undescended testicle moves into its proper position on its own, within the first few months of life.
- **Surgery(Orchiopexy) :** An undescended testicle is usually corrected with surgery. The surgeon carefully manipulates the testicle into the scrotum and stitches it into place. This procedure can be done either with a laparoscope or with open surgery. The surgeon will likely recommend doing the surgery after the child is 3 to 6 months old and before he is 12 months old.
- **Hormone treatment:** Hormone treatment involves the injection of human chorionic gonadotropin (HCG). This hormone could cause the testicle to move to the scrotum

- If the testis does not follow the main gubernacular structure but follows one of its subsidiary strands, it will end in an abnormal position, resulting in an **ectopic testis.**

Vesicoureteral Reflux

ANATOMY OF THE URETEROVESICAL JUNCTION

- **1.Mesodermal Components**
- **2.Endodermal components**

- MESODERMAL components: The mesodermal component, which arises from the Wolffian duct, is made up of two parts that are innervated by the sympathetic nervous system:
- **A. The Ureter and the Superficial Trigone**
- **B. Waldeyer's Sheath and the Deep Trigone**

The Ureter and the Superficial Trigone

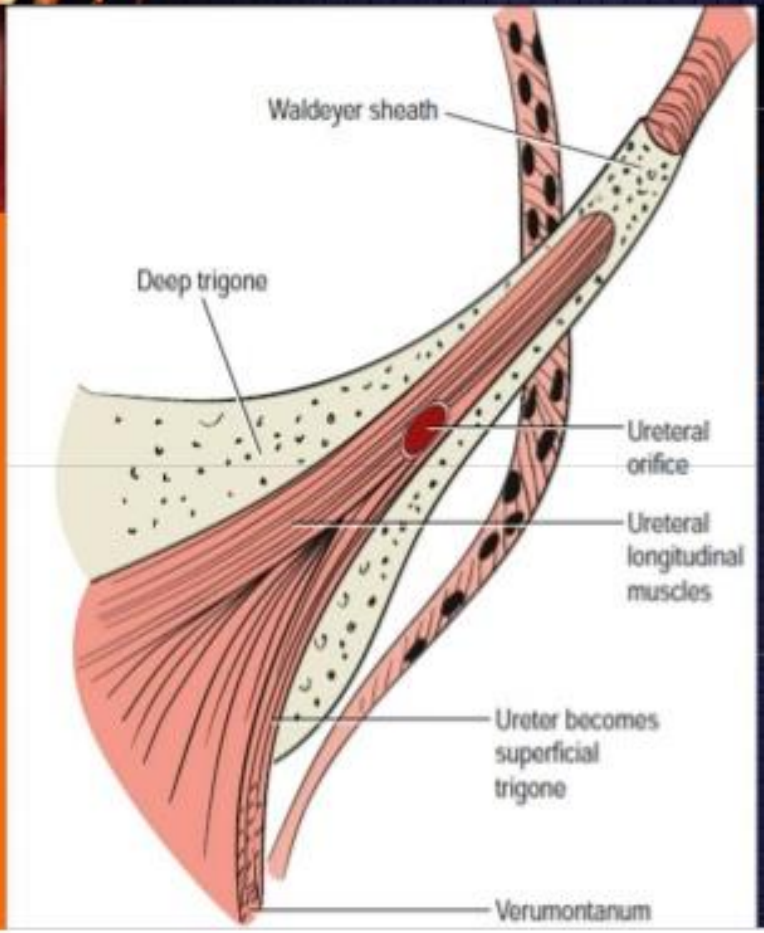
- The smooth musculature of the renal calyces, pelvis, and extravesical ureter is composed of helically oriented fibers that allow for peristaltic activity.
- As these fibers approach the vesical wall, they are reoriented into the longitudinal plane
- As these smooth-muscle fibers approach the ureteral orifice, those that form the roof of the ureter swing to either side to join those that form its floor. They then spread out and join equivalent muscle bundles from the other ureter and also continue caudally, thus forming the superficial trigone.

Waldeyer's Sheath and the Deep Trigone

- Beginning at a point about 2–3 cm above the bladder, an external layer of longitudinal smooth muscle surrounds the ureter.
- This muscular sheath passes through the vesical wall, to which it is connected by a few detrusor fibers.
- As it enters the vesical lumen, its roof fibers diverge to join its floor fibers, which then spread out, joining muscle bundles from the contralateral ureter and forming the deep trigone, which ends at the bladder neck.

Anatomy of VU Junction

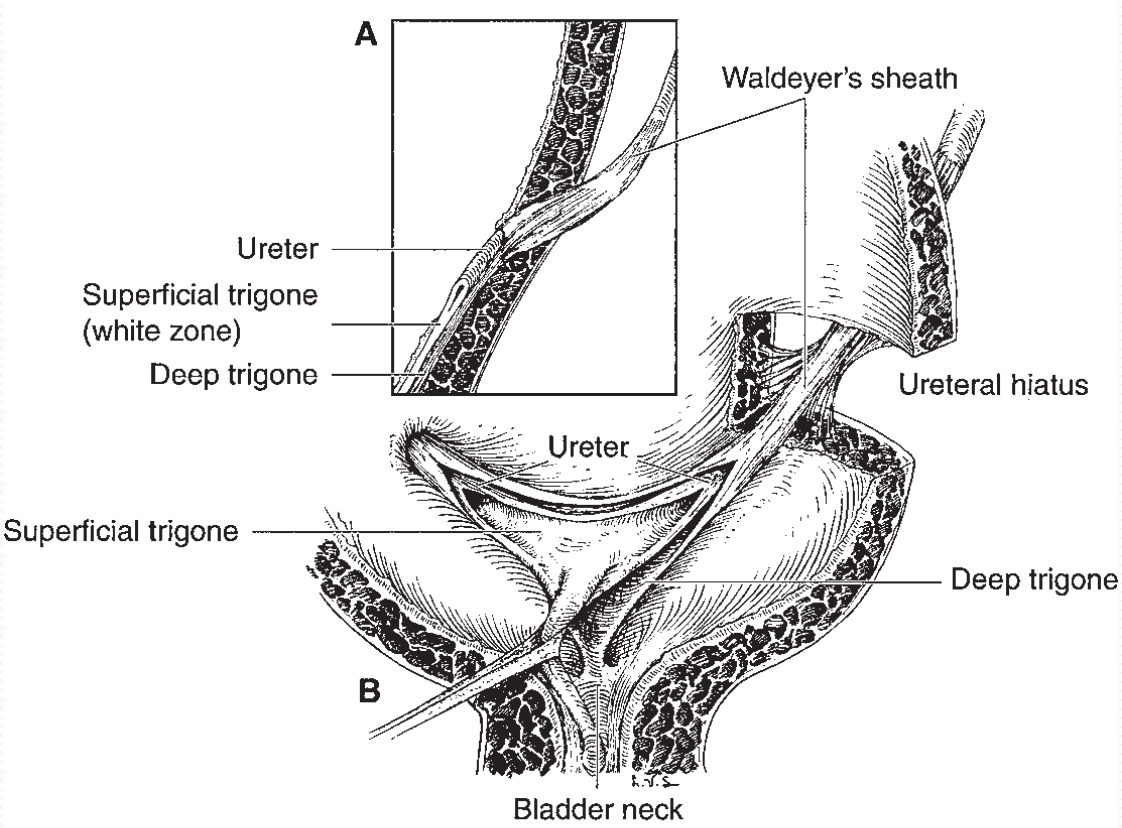
S V M C H



Ureter continues as superficial trigone

Waldeyer's fascia continues as deep trigone

Normal ureterotrigonal complex. ***A: Side view of ureterovesical junction. Waldeyer's muscular sheath*** invests the juxtavesical ureter and continues downward as the deep trigone, which extends to the bladder neck. The ureteral musculature becomes the superficial trigone, which extends to the verumontanum in the male and stops just short of the external meatus in the female. ***B: Waldeyer's sheath is connected by a few fibers to the detrusor muscle in*** the ureteral hiatus. This muscular sheath, inferior to the ureteral orifices, becomes the deep trigone. The musculature of the ureters continues downward as the superficial trigone.



The ureteral muscle extends downward and becomes the superficial trigone.

Waldeyer's sheath extends downward and becomes the deep trigone.

Endodermal Component

- The vesical detrusor muscle bundles are intertwined and run in various directions. As they converge on the internal orifice of the bladder, however, they tend to become oriented into three layers.
- **A. Internal Longitudinal Layer:** continues into the urethra submucosally and ends just inside the external meatus in the female and at the caudal end of the prostate in the male.
- **B.Middle Circular Layer:** thickest anteriorly and stops at the vesical neck.
- **C.Outer Longitudinal Layer,** which constitute the true vesicourethral sphincter

PHYSIOLOGY OF THE URETEROVESICAL JUNCTION

- Using nonrefluxing dogs, they demonstrated the following:
- 1. Interruption of the continuity of the trigone resulted in reflux. (Incision in the trigone 3 mm below the ureteral orifice, resulting in an upward and lateral migration of the ureteral orifice with shortening of the intravesical ureter.)
- 2. Unilateral lumbar sympathectomy resulted in paralysis of the ipsilateral trigone. This led to lateral and superior migration of the ureteral orifice and reflux.
- 3. Electrical stimulation of the trigone caused the ureteral orifice to move caudally, thus lengthening the intravesical ureter. This maneuver caused a marked rise in resistance to flow through the ureterovesical junction.

- 4.During gradual filling of the bladder, intravesical pressure increased only slightly, whereas pressure within the intravesical ureter rose progressively—owing, apparently, to increasing trigonal stretch. A few seconds before the expected sharp rise in intravesical pressure generated for voiding, the closure pressure in the intravesical ureter rose sharply and was maintained for 20 seconds after detrusor contraction had ceased. This experiment demonstrated that ureterovesical competence is independent of detrusor action and is governed by the tone of the trigone.

- ***Conclusion: <u>normal ureterotrigonal tone</u> prevents VUR. Electrical or pharmacologic stimulation of the trigone caused increased occlusive pressure in the intravesical ureter and increased resistance to flow down the ureter, whereas incision or paralysis of the trigone led to reflux***.

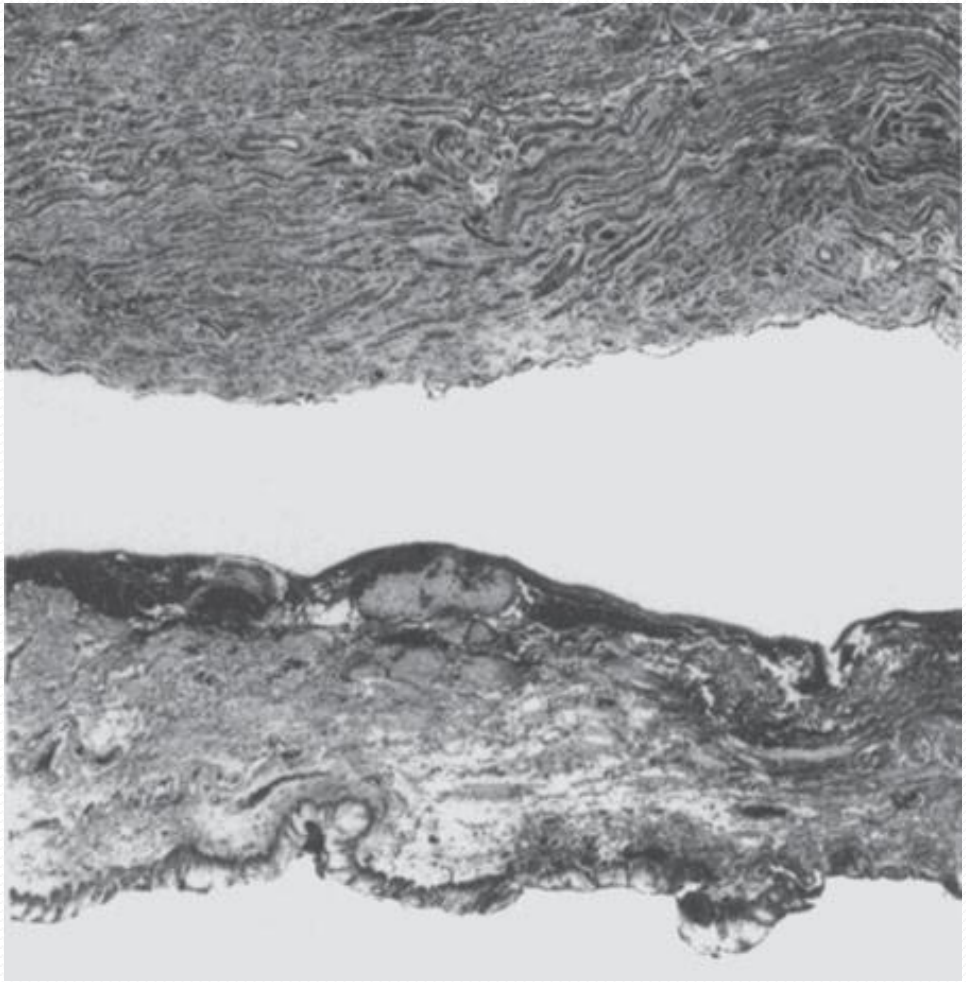
Causes

- Congenital
- **A. Trigonal Weakness (Primary Reflux):**
- Trigonal weakness is by far the most common cause of ureteral Reflux
- Most often seen in young children, more in girls
- In the normal state, the intravesical ureterotrigonal muscle tone exerts a downward pull, whereas the extravesical ureter tends to pull cephalad
- If trigonal development is deficient, not only its occlusive power diminished but the ureteral orifice tends to migrate upward toward the ureteral hiatus.

Histology of the trigone in primary reflux.

Top: Normal trigone demonstrating wealth of closely packed smooth-muscle fibers.

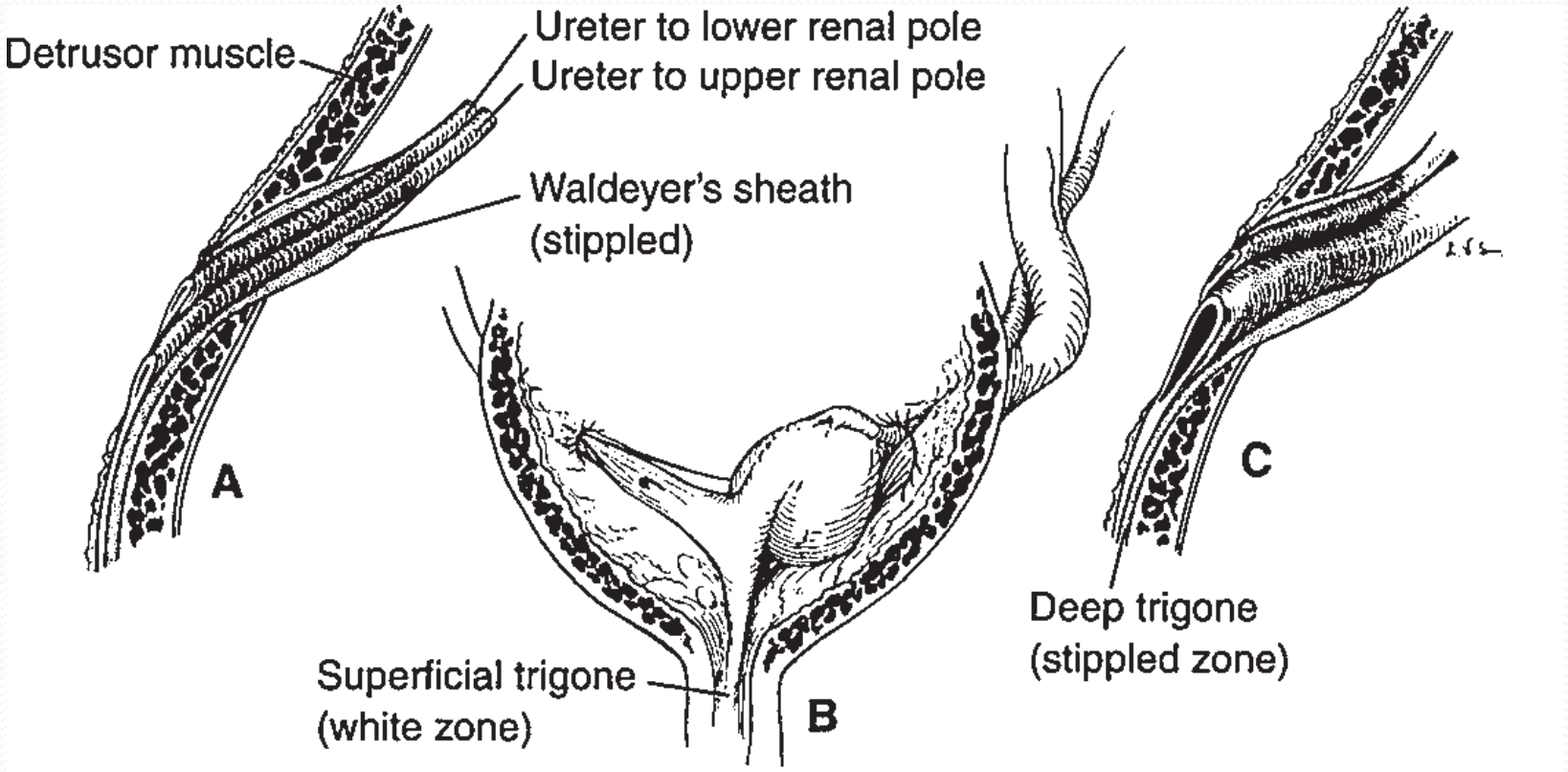
Bottom: The congenitally attenuated trigonal muscle that accompanies vesicoureteral reflux



- **B. Familial Reflux**
- There appears to be genetic predisposition for reflux. The reported prevalence of VUR among siblings of index patients with reflux has ranged from 4.7% to 51%, which is significantly higher than the incidence of reflux in the general population (1%)
- Several genes have been associated with VUR, including uroplakin-3, SLIT2/ROBO2, and TGF-β.

- **C. Ureteral Abnormalities**
- **1. Complete ureteral duplication**
- The intravesical portion of the ureter to the upper renal segment is usually of normal length, whereas that of the ureter to the lower pole is abnormally short; this orifice is commonly incompetent.

Ureteral duplication and ureterocele as causes of vesicoureteral reflux. ***A: Ureteral duplication showing*** juxtavesical and intravesical ureters encased in common sheath (Waldeyer's). The superior ureter, which always drains the lower renal pole, has a shorter intravesical segment; in addition, it is somewhat devoid of muscle. It therefore tends to allow reflux. ***B: Duplication with ureterocele that always involves caudal ureter, which drains upper renal pole. Pinpoint*** orifice is obstructive, causing hydroureteronephrosis. Resulting wide dilatation of ureter and ureteral hiatus shortens the intravesical segment of the other ureter, often causing it to reflux. ***C: Resection of ureterocele allows reflux into*** that ureter.



Ureteral and superficial trigonal muscles are one and the same.

Waldeyer's and deep trigone are stippled because they are one and the same.

- **2. Ectopic ureteral orifice—Single ureter or one of a pair** may open well down on the trigone, at the vesical neck, or in the urethra. In this instance, VUR is the rule. This observation makes it clear that the length of the intravesical ureter is not the sole factor in reflux. Such intravesical ureteral segments are usually devoid of smooth muscle. Thus, they have no occlusive force.

- **3.Ureterocele:**
- Usually involves the ureter that drains the upper pole of a duplicated kidney.
- Because the ureteral orifice is obstructed, the intramural ureter becomes dilated. This increases the diameter of the ureteral hiatus, further shortening the intravesical segment of the other ureter

- **Vesical Trabeculation:** Occasionally, a heavily trabeculated bladder may be associated with reflux. The causes include spastic neurogenic bladder and severe obstruction distal to the bladder

- **Edema of the Vesical Wall Secondary to Cystitis:**
- Valves vary in their degrees of incompetence.
- A “borderline” junction may not allow reflux when the urine is sterile, but valvular function may be impaired when cystitis causes associated edema involving the trigone and intravesical ureter.

- **Eagle–Barrett (Prune Belly) Syndrome**
- The Eagle–Barrett syndrome is a relatively rare condition in which there is failure of normal development of the abdominal muscles and the smooth muscle of the ureters and bladder.
- Because the smooth muscle of the ureterotrigonal complex is deficient, reflux is to be expected

Iatrogenic

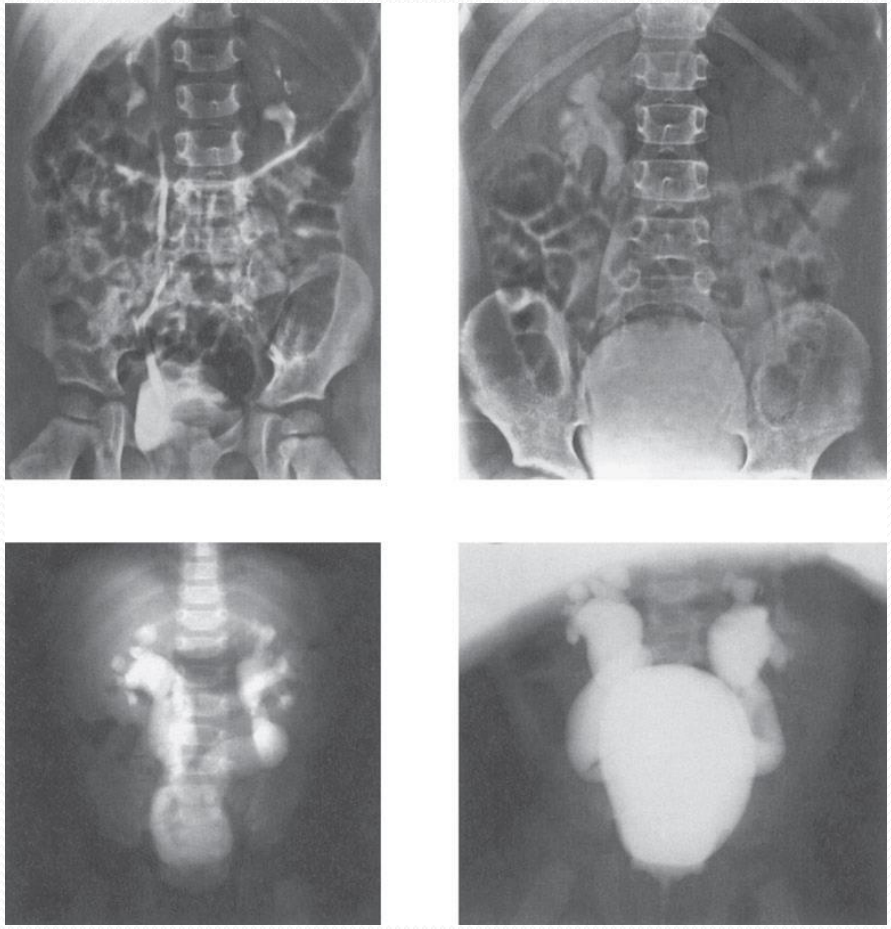
- **A. Prostatectomy**
- With any type of prostatectomy, the continuity of the superficial trigone is interrupted at the vesical neck. If the proximal trigone moves upward, temporary reflux may occur.
- Fortunately, in 2–3 weeks, the trigone again becomes anchored and reflux ceases.
- Other Iatrogenic causes: **Wedge Resection of the Posterior Vesical Neck, Ureteral Meatotomy, Resection of Ureterocele**
- **Contracted bladder** (secondary to interstitial cystitis,
- tuberculosis, radiotherapy, carcinoma, or schistosomiasis)

COMPLICATIONS

- **Pyelonephritis**
- **Hydroureteronephrosis:** Dilation of the ureter, renal pelvis, and calyces is usually observed in association with reflux.
- There are three reasons for the dilatation:

1. **Increased workload on ureter**
2. **High hydrostatic pressure**
3. **Weak ureteral musculature**

Excretory urogram with changes that imply right vesicoureteral reflux. ***Upper left: Excretory urogram*** showing normal right urogram and a ureter that is mildly dilated and remains full through its entire length. The ureteral change implies reflux. ***Upper right: Cystogram demonstrates the reflux. Note, now, the degree of dilatation of the*** ureter, pelvis, and calyces. ***Lower left: Excretory urogram shows bilateral hydroureteronephrosis with pyelonephritic*** scarring. These findings imply the presence of reflux. ***Lower right: Voiding cystourethrogram.Free reflux bilaterally.***



Incidence

- VUR occurs in 25–40% of children with urinary tract infection but in only 8% of adults with bacteriuria.
- Infection associated with reflux occurs during the first few weeks of life >> Most are boys , but After 1 year of age, the female:male ratio of children with infection and reflux is approximately 3:1–4:1

Symptoms

- **Symptoms Related to Reflux**
- **A. Symptomatic Pyelonephritis :** (*In Adults*: chills ,high fever, renal pain, nausea and vomiting, and symptoms of cystitis. *In children*: only fever, vague abdominal pains, and sometimes diarrhea)
- **B. Asymptomatic Pyelonephritis**(The incidental findings of pyuria and bacteriuria may be the only clues)

- **C. Symptoms of Cystitis Only (LUTS)**
- **D. Renal Pain on Voiding** (a rare complaint in patients with VUR.)
- **E. Uremia**
- The last stage of bilateral reflux is uremia due to destruction of the renal parenchyma by hydronephrosis or pyelonephritis (or both).
- **F. Hypertension**
- In the later stages of atrophic pyelonephritis, a significant incidence of hypertension is observed.

- **Symptoms Related to the Underlying Disease**
- **A. Urinary Tract Obstruction** (hesitancy, impaired / intermittent stream)
- **B. Spinal Cord Disease**
- The patient may have a serious neurogenic disease such as paraplegia, quadriplegia, multiple sclerosis, or meningomyelocele.
- Symptoms may be limited to those of neurogenic bladder: incontinence of urine, urinary retention or large residual volume, and vesical urgency

Physical Findings

- renal tenderness (If acute pyelonephritis)
- distended bladder on Palpation and percussion of the suprapubic area secondary to obstruction or neurogenic disease
- Examination may reveal a neurologic deficit compatible with a paretic bladder

Laboratory Findings

- The most common complication of reflux, particularly in females, is infection. Bacteriuria without pyuria is not Uncommon
- The serum creatinine may be elevated in the advanced stage of renal damage, but it may be normal even when the degree of reflux and hydronephrosis is marked

X-Ray Findings

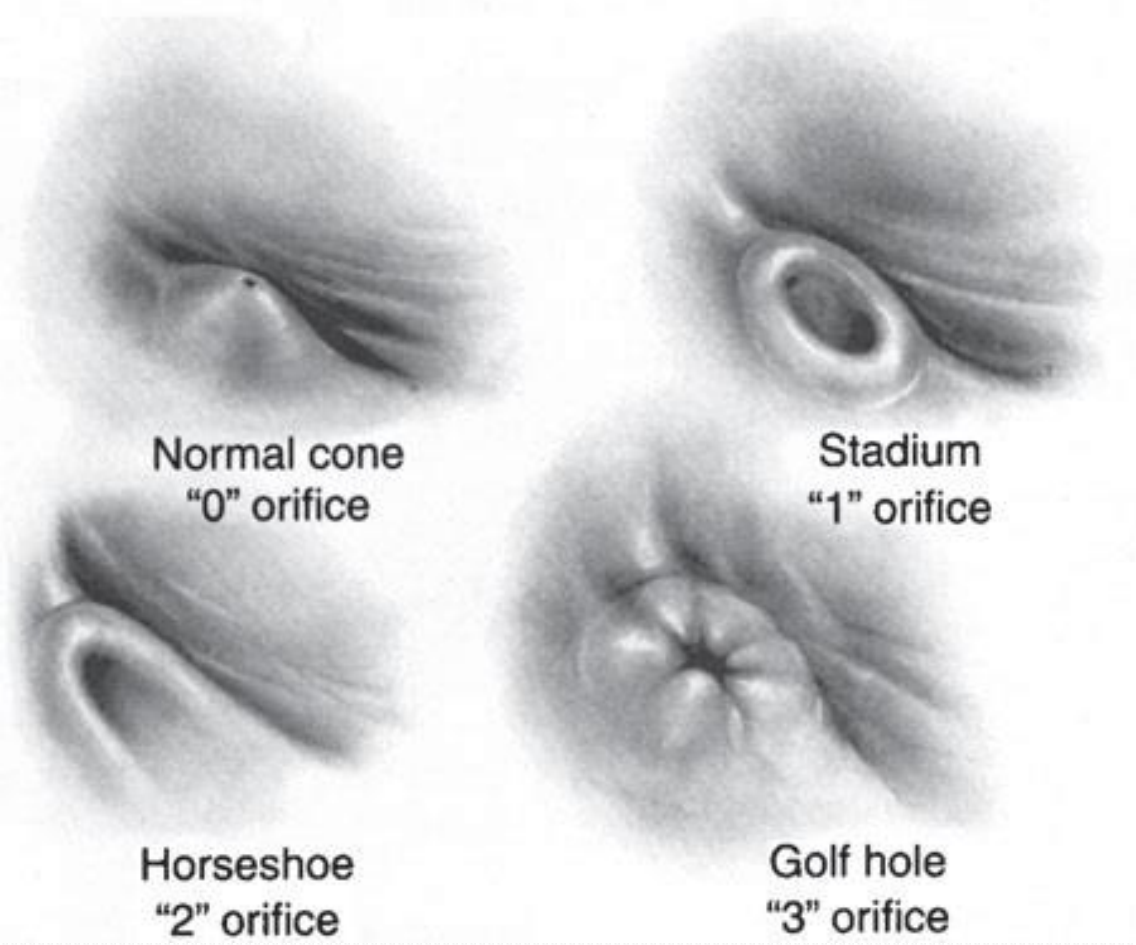
- The plain film may reveal evidence of spina bifida, meningomyelocele, or the absence of the sacrum and thus point to a neurologic deficit
- in VUR, excretory urograms may be normal, but usually, one or more of the following clues to the presence of reflux is noted: (1) a persistently dilated lower ureter (2) areas of dilatation in the ureter, (3) ureter visualized throughout its entire length, (4) the presence of hydroureteronephrosis with a narrow juxtavesical ureteral segment, or (5) changes of healed pyelonephritis (caliceal clubbing with narrowed infundibula or cortical thinning).
- ***A normal intravenous urogram does not rule out reflux.***

- Reflux is diagnosed by demonstration of its existence with one of the following techniques: simple or delayed cystography, voiding cystourethrography, or voiding cinefluoroscopy. Radionuclide scanning can be used (Tc is instilled into the bladder along with sterile saline solution, and the gamma camera will reveal ureteral reflux)
- Reflux can be demonstrated by a technique using indigotindisulfonate sodium (indigo carmine), a blue dye. The bladder is filled with sterile water containing 5 mL of indigo carmine per 100 mL, after which the patient voids and the bladder is thoroughly flushed out with sterile water. The ureteral orifices are then viewed cystoscopically for blue-tinged efflux. This technique has the advantage that no ionizing radiation is used, and its efficiency is equal to that of voiding cystourethrography

Instrumental Examination

- ***Cystoscope:*** The major contribution of cystoscopy is to allow study of the morphologic characteristics of the ureteral orifice and its position in relation to the vesical neck
- **1. Morphology**—The orifice of a normal ureter has the appearance of a ***volcanic cone***. That of a slightly weaker valve looks like a ***football stadium***; an even weaker one has the appearance of a ***horseshoe*** with its open end pointing toward the vesical neck. The completely incompetent junction has ***a golf-hole*** orifice that lies over the ureteral hiatus.

Cystoscopic appearance of the normal ureteral orifice and 3° of incompetence of the ureterovesical junction



DIFFERENTIAL DIAGNOSIS

- Functional (nonocclusive) vesicoureteral obstruction may cause changes similar to those suggesting the presence of reflux on excretory urography. this congenital obstruction is due to an *abundance of circularly oriented smooth-muscle fibers in the ureteral musculature* at this point. Its action is sphincteric.
- Significant obstruction distal to the vesical neck leads to *hypertrophy of both the detrusor and trigonal muscles*. The latter exert an exaggerated pull on the intravesical ureter and thus cause functional obstruction. Hydroureteronephrosis is therefore to be expected.
- Other lesions that may cause hydroureteronephrosis without reflux include low ureteral stone, occlusion of the ureter by cervical or prostatic cancer, urinary tract tuberculosis, and schistosomiasis

TREATMENT

- ***Medical Treatment***
- **Indications:**
- majority of the cases, children with primary reflux are initially treated medically, since there is a chance of spontaneous resolution. Positive predictors for reflux resolution include unilateral reflux, the lower grades of reflux, the earlier age of presentation, and male gender
- In a woman who occasionally develops acute pyelonephritis following intercourse but whose urine quickly clears on antimicrobial therapy, reflux will probably be controlled if she takes steps to prevent vesical infections

- Methods of treatment:
- Urinary infection should be definitively treated with *antimicrobial drugs*
- "void by the clock" Method - every 3–4 hours: Children with reflux often have voiding dysfunction due to thin-walled bladders and do not perceive the normal urge to void when the bladder is full. Further detrusor tone is lost with overfilling, increasing the likelihood of residual urine. Such children should "void by the clock" every 3–4 hours whether they have the urge or not. Vesical residual urine may then be minimized.

- **Evaluation of Success of Medical Treatment:**
- Cystograms should be repeated every 12–18 months

Surgical Management

- ***Absolute indications*** for surgery include the following conditions: (1) if it is not possible to keep the urine sterile and reflux persists, (2) if acute pyelonephritis recurs despite a strict medical regimen and chronic suppressive antimicrobial therapy, (3) if increased renal damage is demonstrated by serial excretory urograms or nuclear scan, or (4) if noncompliance with medical treatment.
- ***Relative indications*** for surgery include failure to resolve after prolonged observation period (ie, >3 years), parental decision (avoidance of chronic antibiotic usage or radiologic evaluation), or the presence of a diverticulum.

- **B. Types of Surgical Treatment**
- **1. Temporary urinary diversion—If refluxed urine drains** freely into the bladder, cystostomy (or an indwelling urethral catheter in girls) may prove helpful. If the ureters are dilated and kinked, *a low redundant loop can be brought to the skin*. The ureter is opened at this point and urine collected into an ileostomy bag. Later, the loop and the section of ureter distal to it can be resected and the ureter proximal to the loop reimplanted into the bladder.
- **2. Permanent urinary diversion—If it is felt that successful** ureterovesicoplasty cannot be accomplished, a Bricker type of diversion is indicated. If renal function is poor and the ureters are widely dilated and atonic, ureterocutaneous diversion may be the procedure of choice.
- **3. Other surgical procedures**
- a. If reflux is unilateral, with the affected kidney badly damaged and the other kidney normal, nephrectomy is indicated.
- b. If one renal pole of a duplicated system is essentially functionless, heminephrectomy with removal of its entire ureter should be done.
- c. In unilateral reflux, anastomosis of the lower end of the refluxing ureter into the side of its normal mate (transureteroureterostomy) has a few proponents.