

Embryologic Basis of Genitourinary tract malformations

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Addis Ababa University



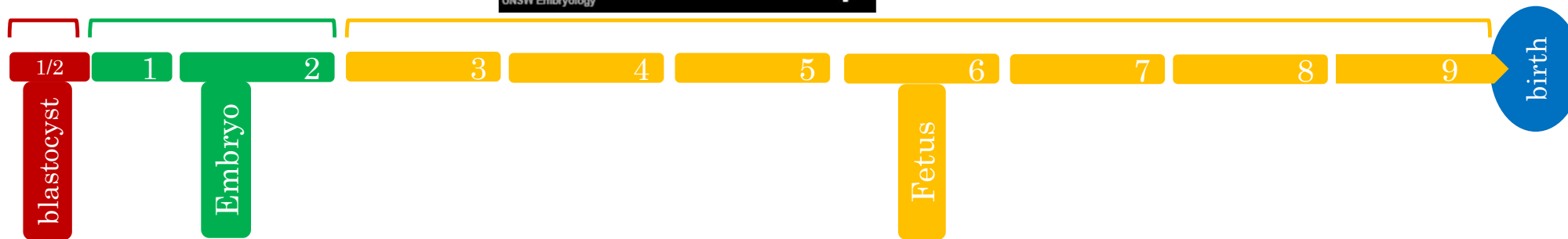
Introduction

- **U**rology accounts for a majority of pediatric surgical workload. The same has been demonstrated in our setup (*pattern of pediatric surgical conditions at Tikur Anbessa..., Prof. Milliard et al.*)

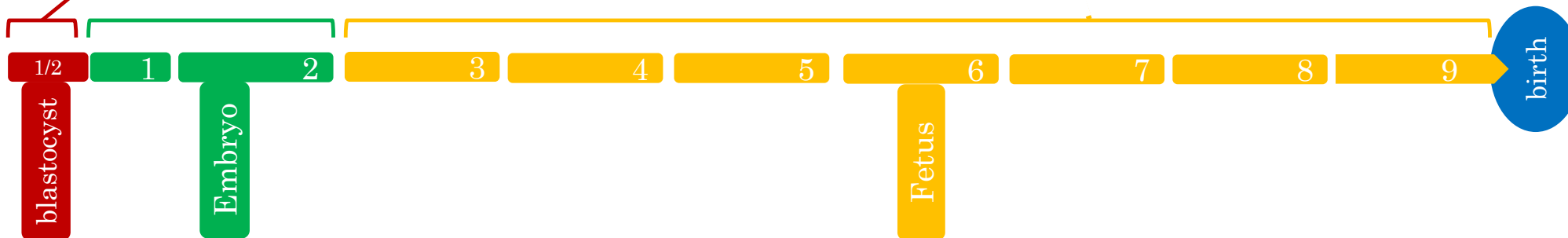
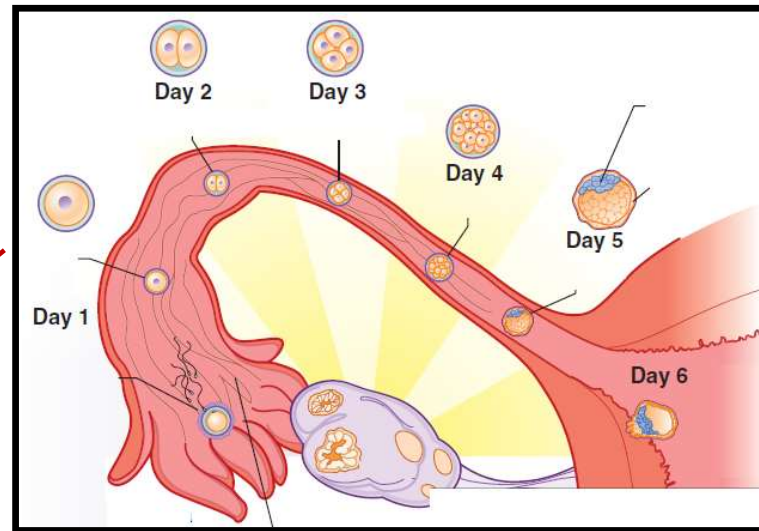
- **con.gen.i.tal** **anomaly**
existing since birth *abnormality*

- **Embryo**.logy

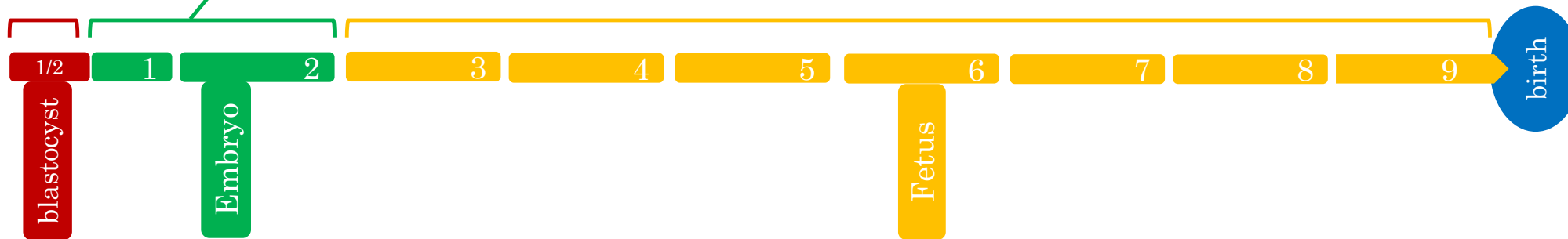
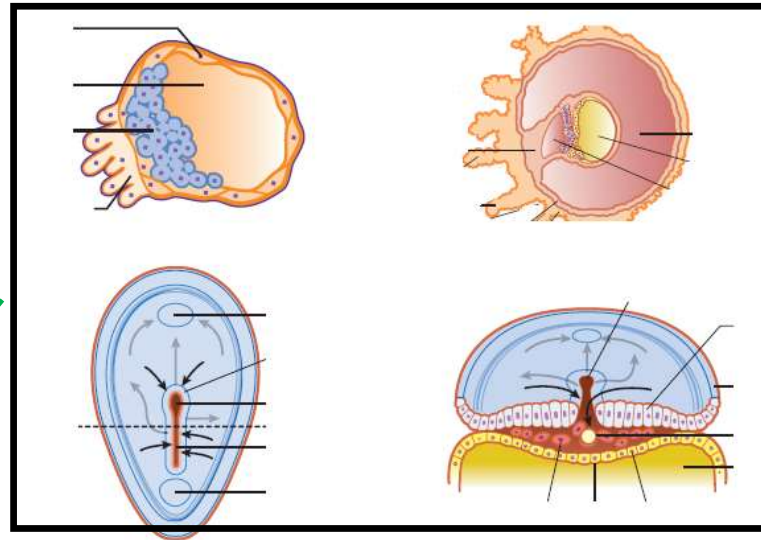
General Embryology



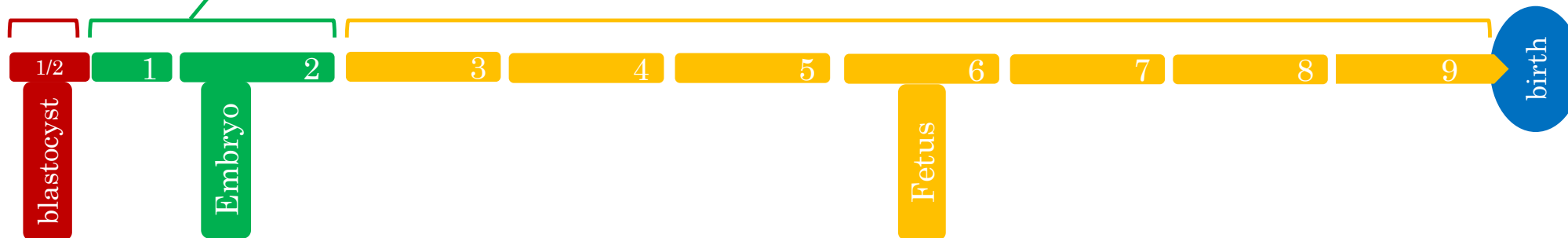
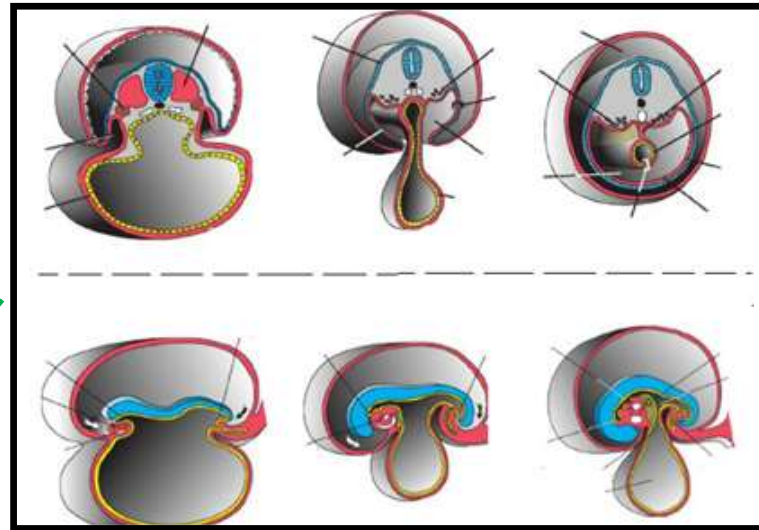
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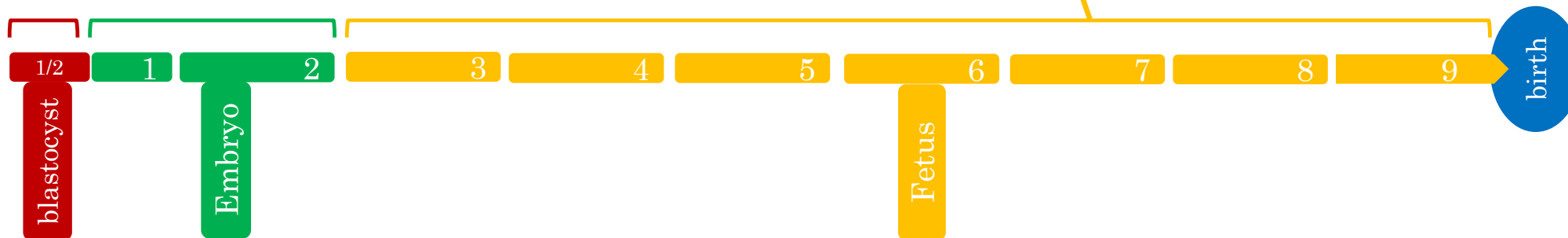
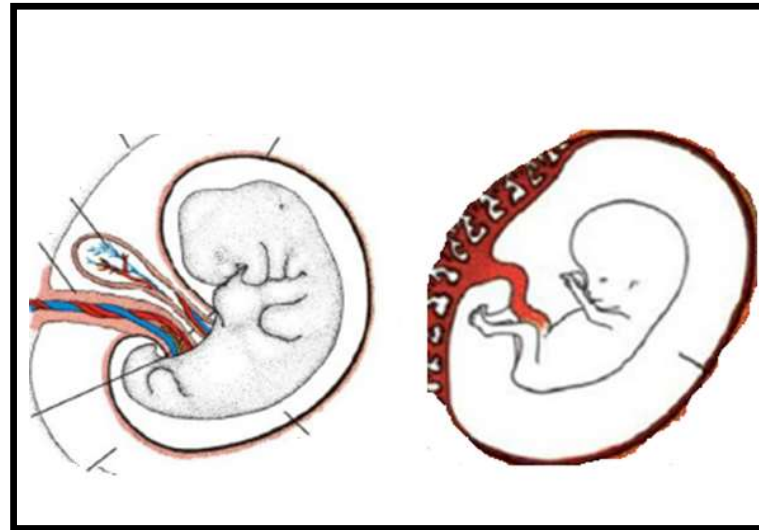
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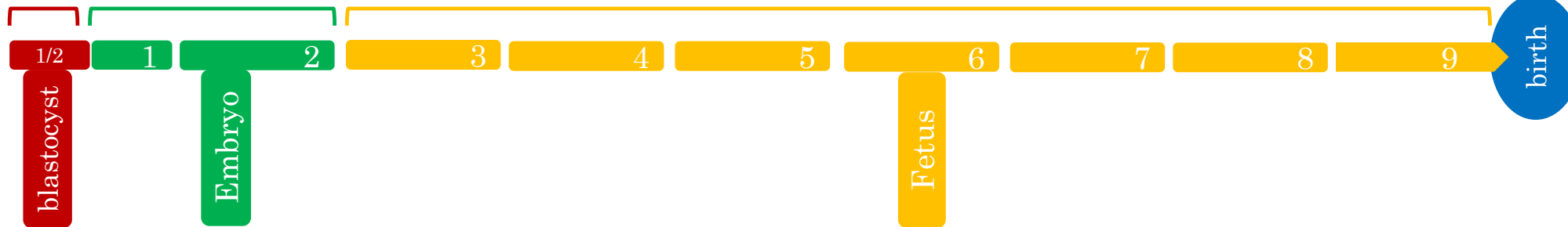
General Embryology



General Embryology



General Embryology



*Objectives

- To understand the embryologic basis of

- ★ Bladder Exstrophy
- ★ Posterior urethral valve
- ★ Undescended testes
- ★ Hypospadias

How many minions does it

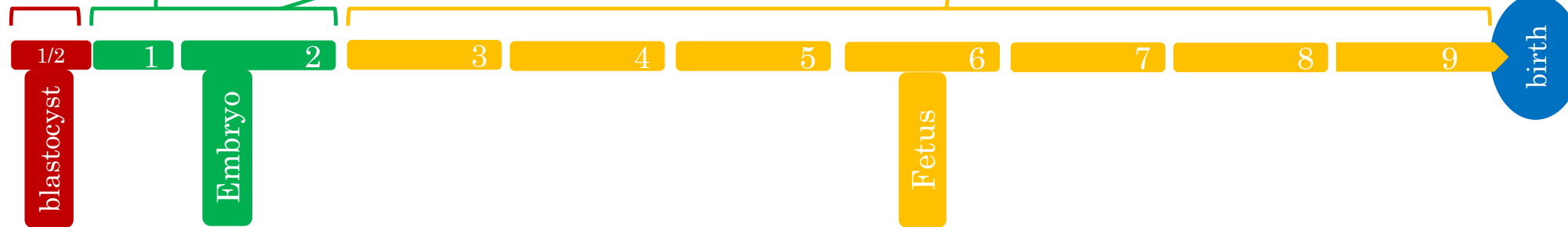
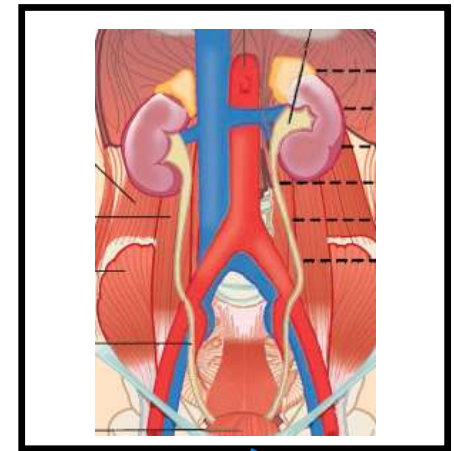
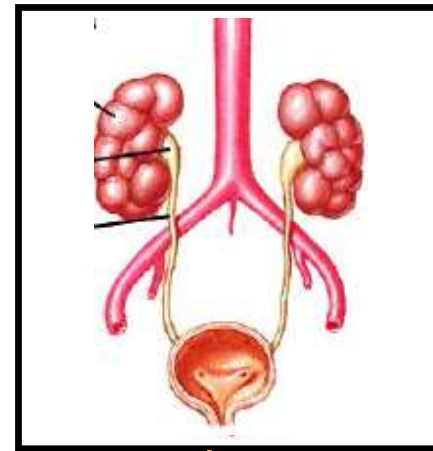
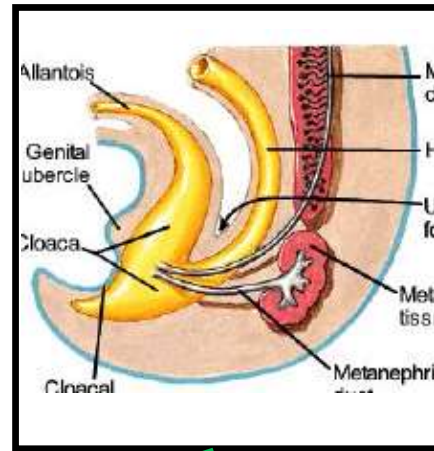
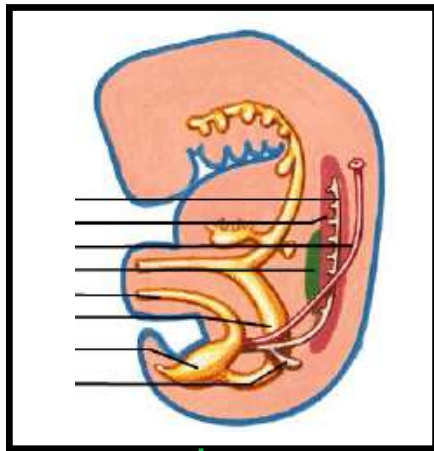


take to change a light bulb?

Part -1
Kidney & Ureter



Upper Urinary Tract: Development



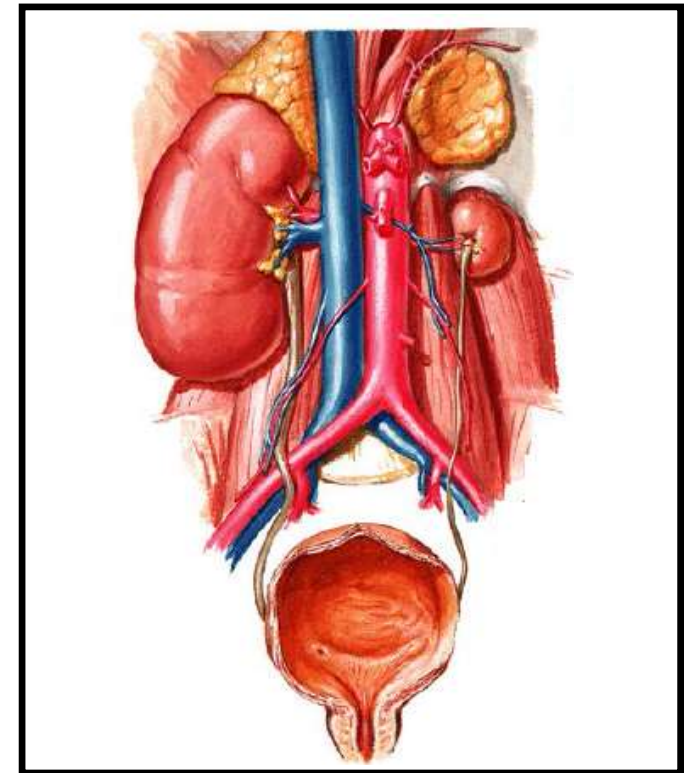
Upper urinary tract: Anomalies

- **Kidney**
 - **Renal dysgenesis and cystic disease**
 - Anomalies of number
 - Anomalies of ascent (ectopia)
 - Anomalies of rotation
 - Anomalies of form & fusion
 - Anomalies of vasculature
 - **Wilms tumor**
- **Collecting system**
 - Calyceal diverticulum
 - Megacalycosis
- **UPJ (ureteropelvic junction) obstruction**
- **Ureter**
 - Ureteral Duplication
 - Ectopic ureter
 - Ureterocele



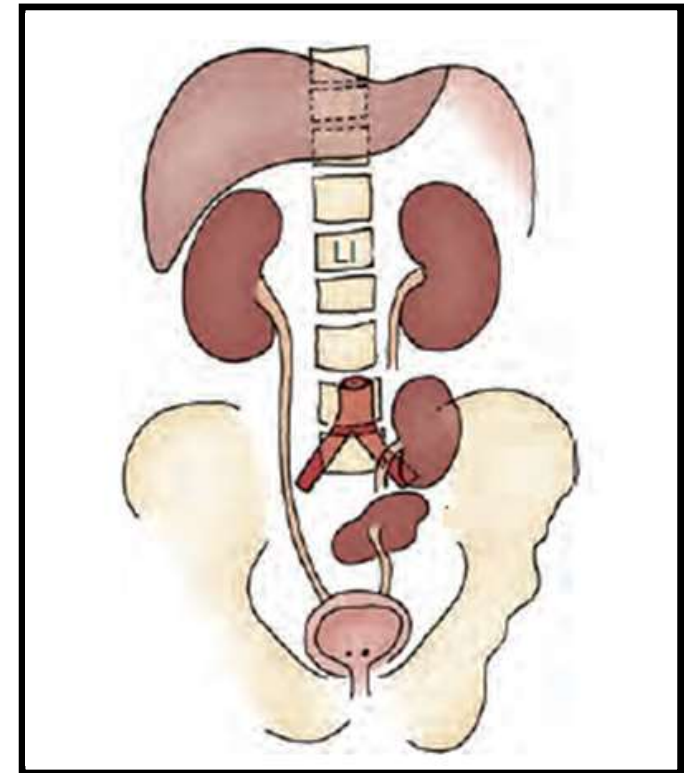
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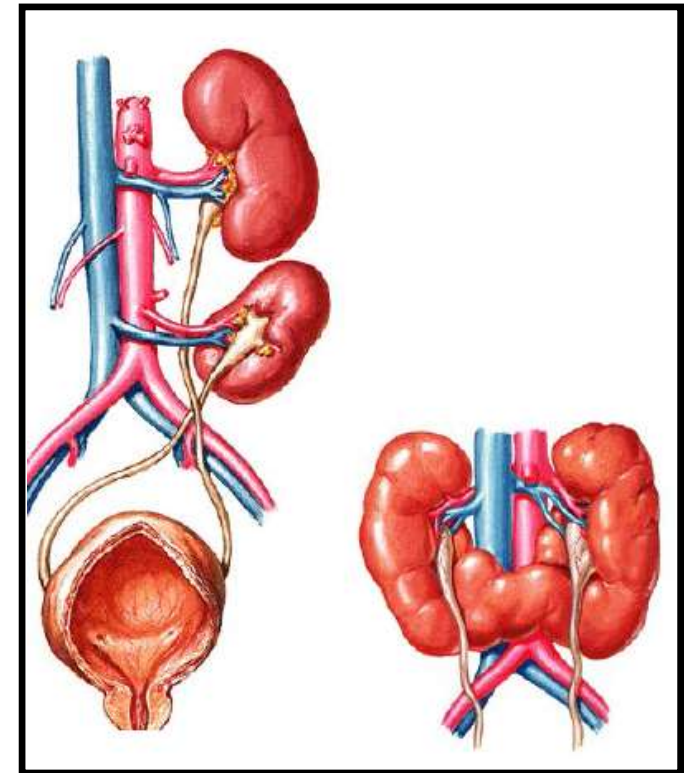
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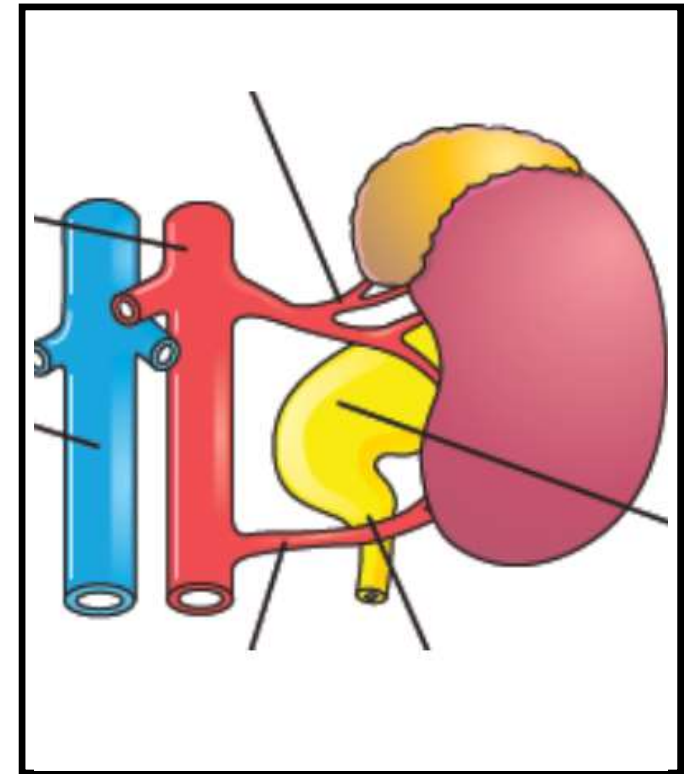
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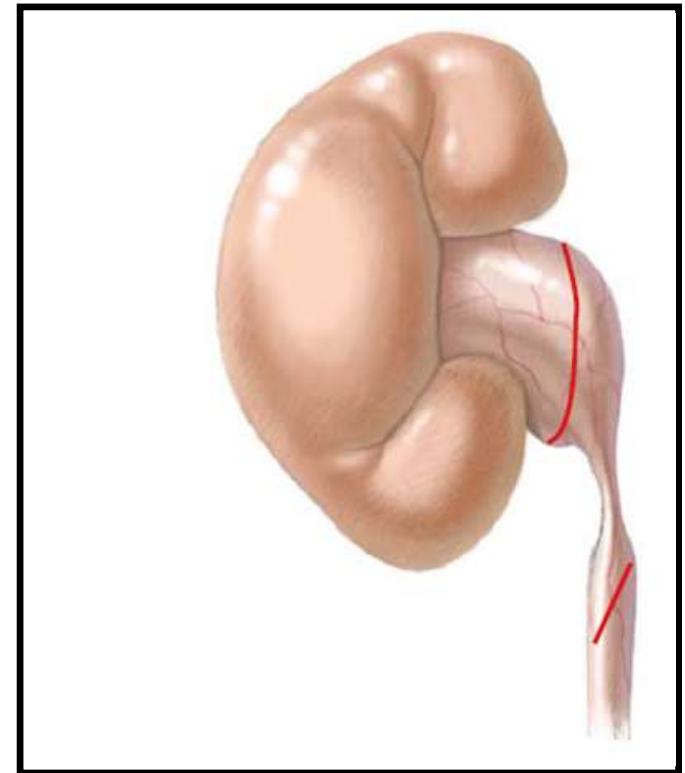
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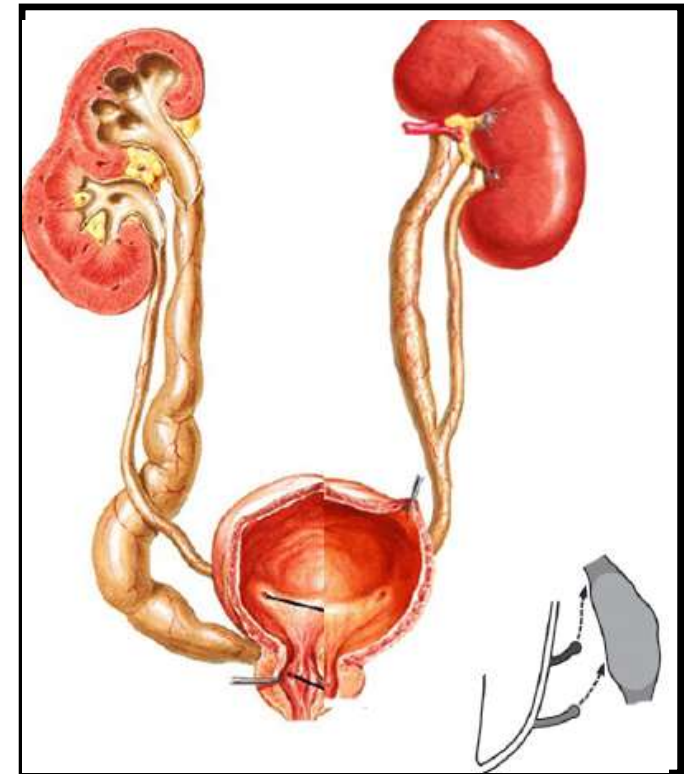
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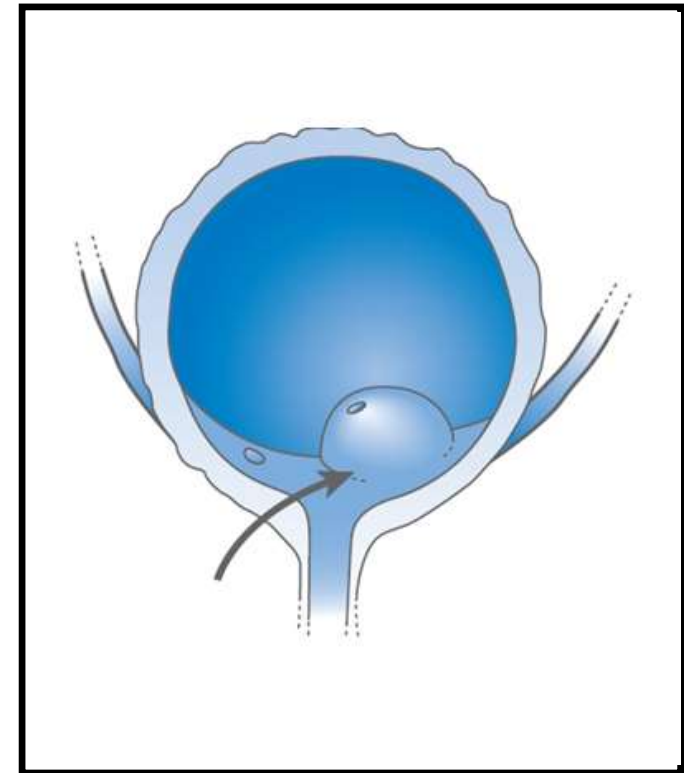
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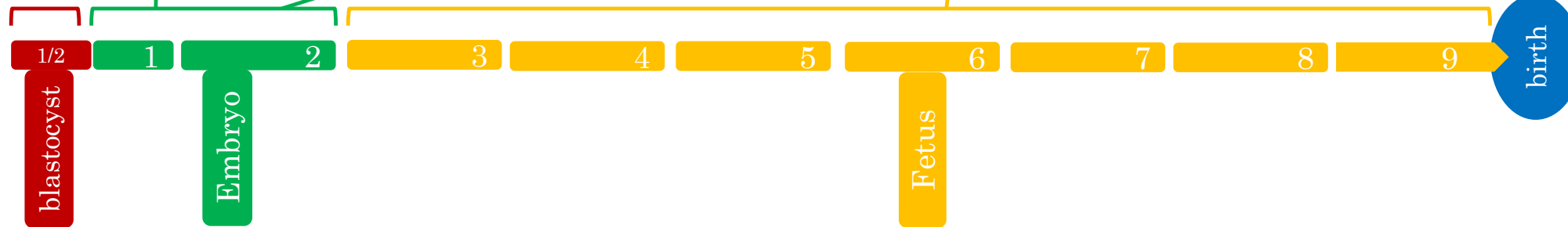
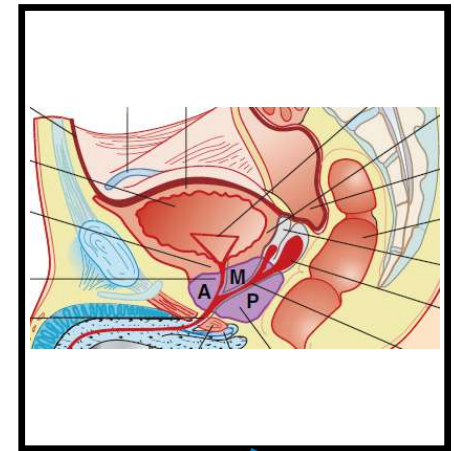
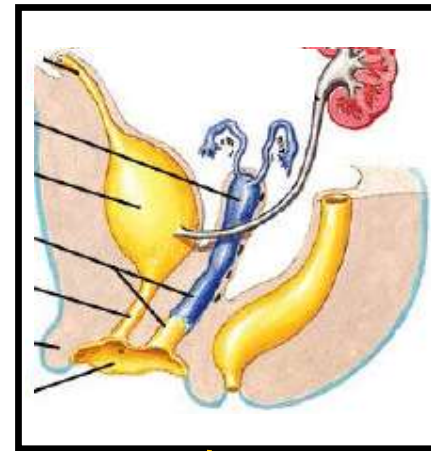
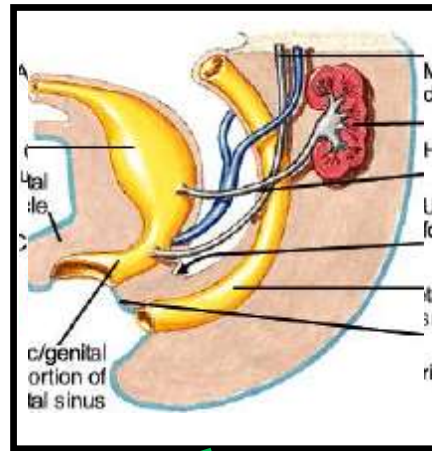
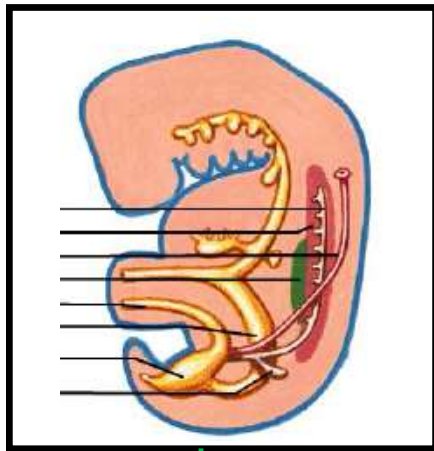
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Part -2
Bladder & Urethra

Lower Urinary Tract : Development



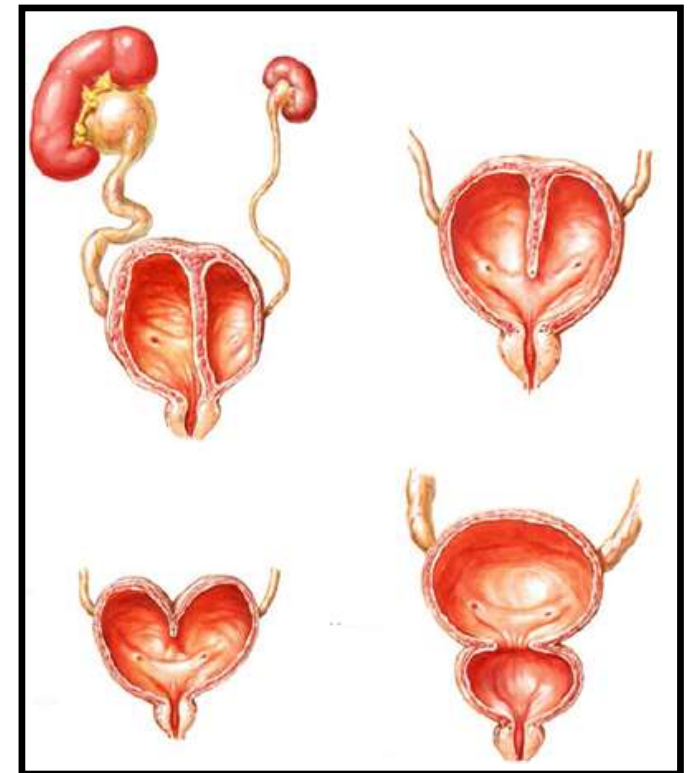
Lower urinary tract: Anomalies

- **Bladder**
 - Prenatally detected
 - Dilated (obstructive, non obstruction)
 - Non dilated or absent
 - Postnatally detected
 - **Urachal anomalies (patent, sinus, cyst)**
 - Bladder diverticulum
 - Bladder duplication
 - Other
- **EEC (Exstrophy-epispadias complex)**
- **Urethra**
 - **PUV (posterior urethral valve)**
 - Anterior urethral valve
 - Urethral Atresia
 - Megalourethra
 - Urinary fistula (with ARM)



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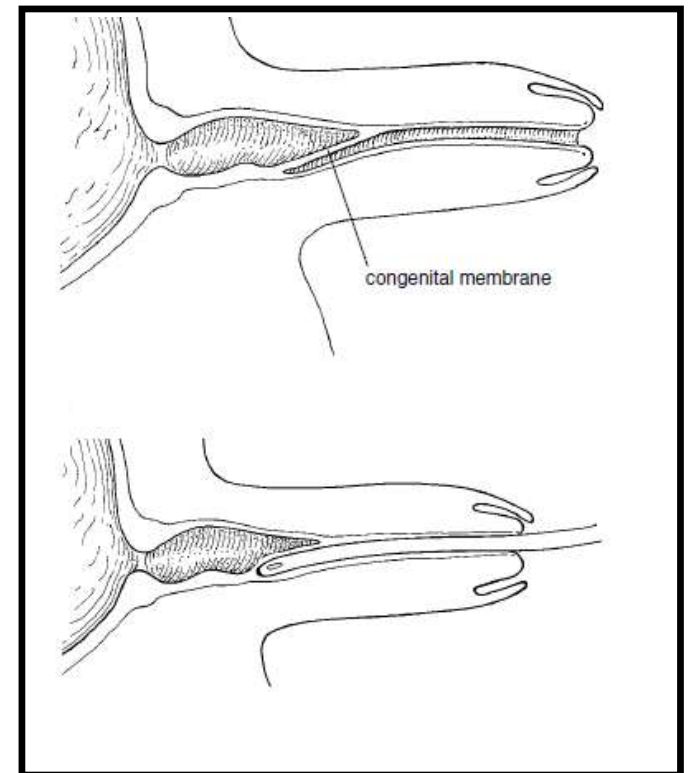
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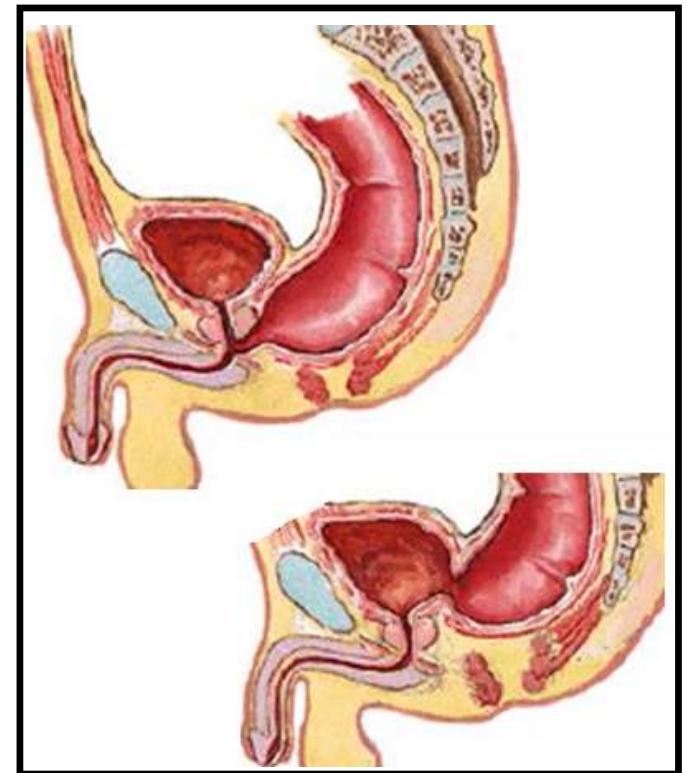
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*Bladder Exstrophy

- **Malformations in EEC**
 - Genital
 - Bladder
 - Pelvis
 - Pelvic floor
 - Anorectal
 - Other = umbilical & inguinal hernia, renal
 - **Factors leading to EEC**
 - No ingrowth of mesoderm into cloacal memb
 - Migratory failure of lateral mesodermal folds
 - Abn overdevelopment of the cloacal membrane
 - Premature rupture of the cloacal membrane
- *extent of infraumbilical defect & time of rupture
- **Effects of EEC**
 - Social (odor & hygiene)
 - UTI
 - renal failure
 - Malignancy



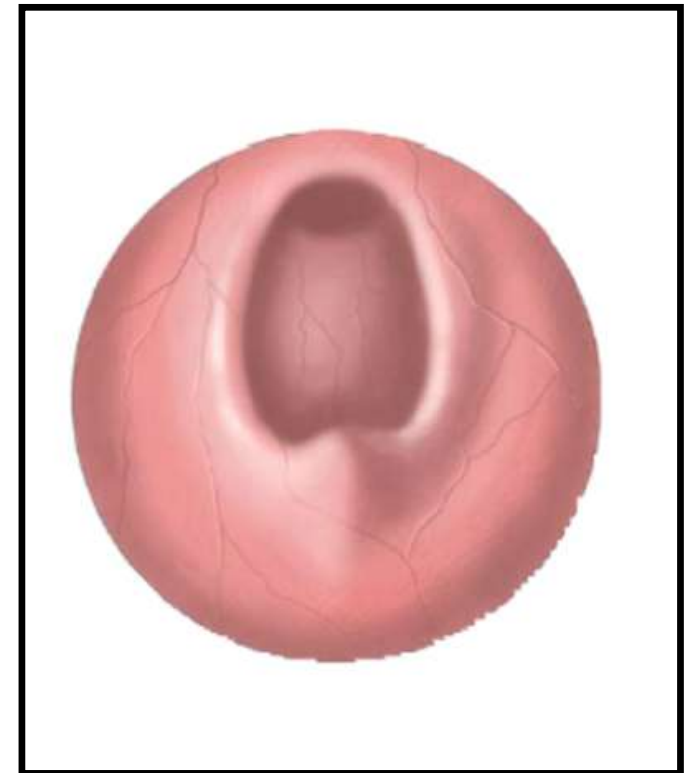
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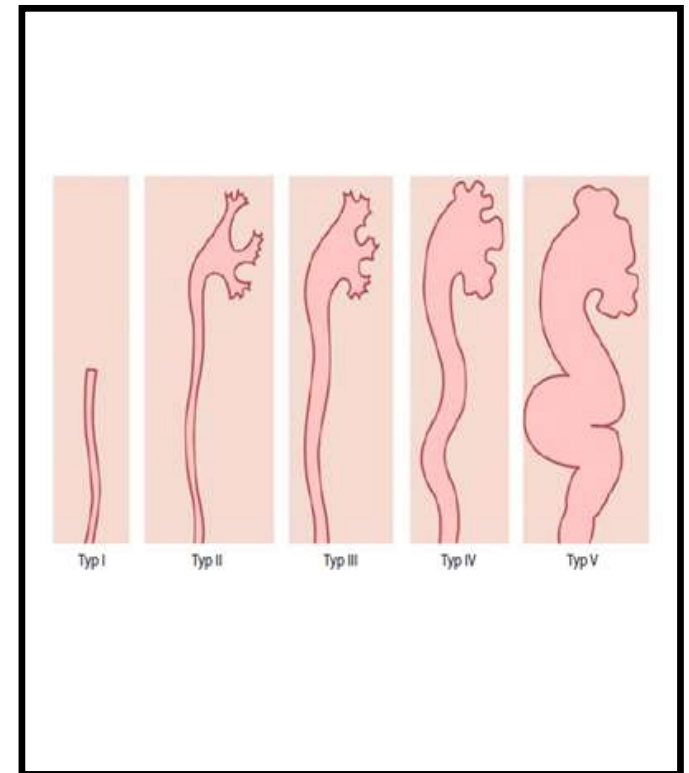
*Posterior urethral valve

- **Malformation in PUV**
 - COPUM (congenital, obstructive posterior urethral memb)
- **Factors leading to PUV**
 - Type I – Abnormal integration of the Wolffian duct
 - Type III - Persistence of the urogenital membrane.
- **Effects of PUV**
 - congenital obstructive uropathy
(bladder changes, VUR, megaureter, hydronephrosis)



*Posterior urethral valve

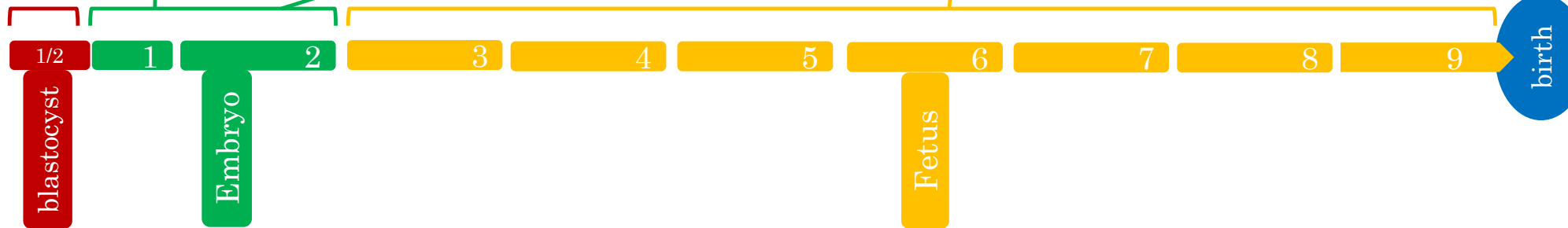
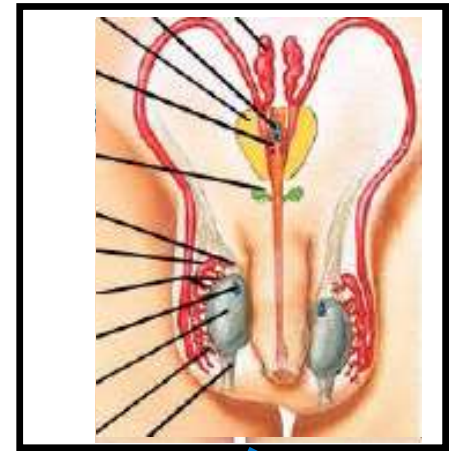
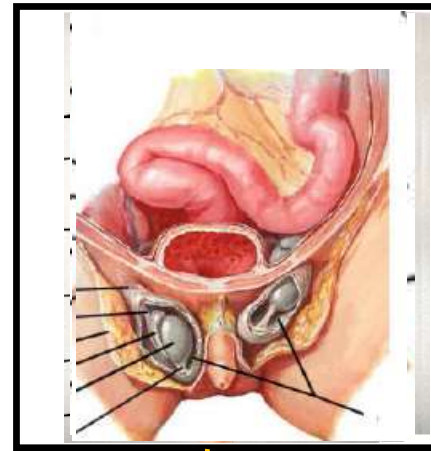
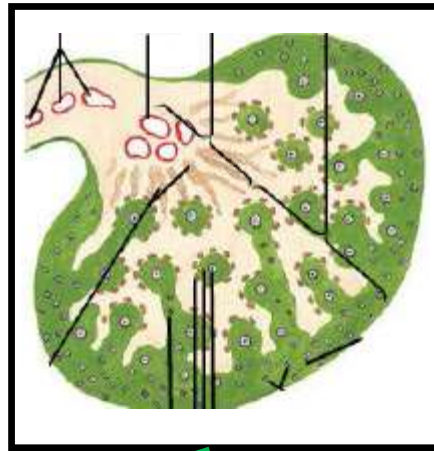
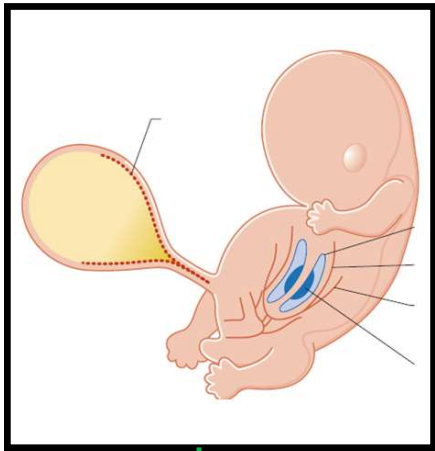
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Part - 3
Gonads

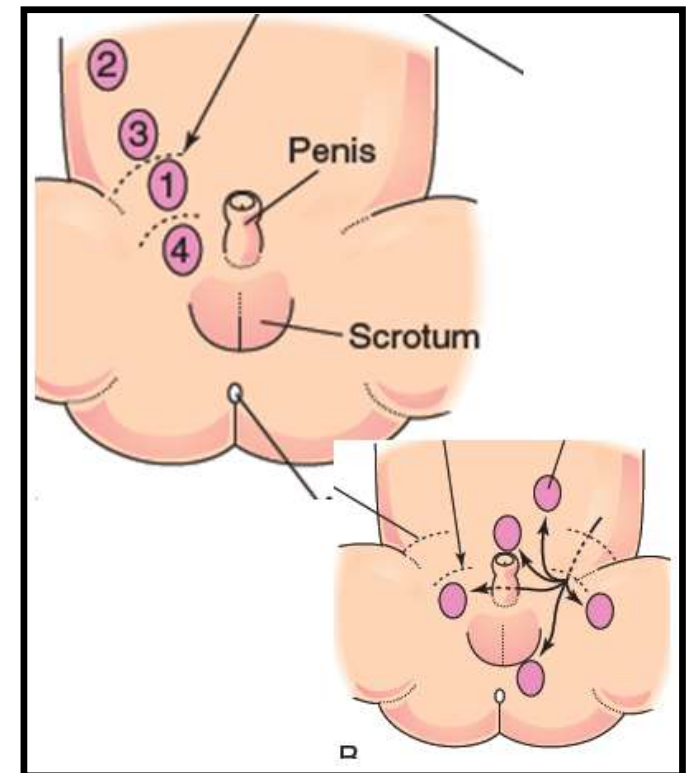


Descent of the gonads



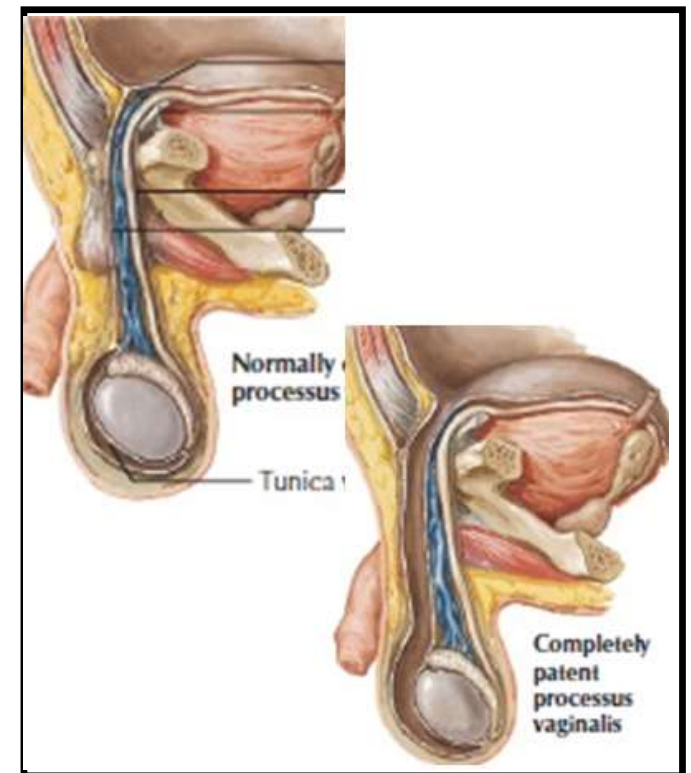
Descent of the gonads: Anomalies

- **Cryptorchidism**
 - Agenesis
 - **Ectopic**
 - **True Undescended**
 - * *Retractile*
 - * *Ascending*
- **Inguinal Hernia**
- **Hydrocele**
 - Communicating
 - Hydrocele of cord



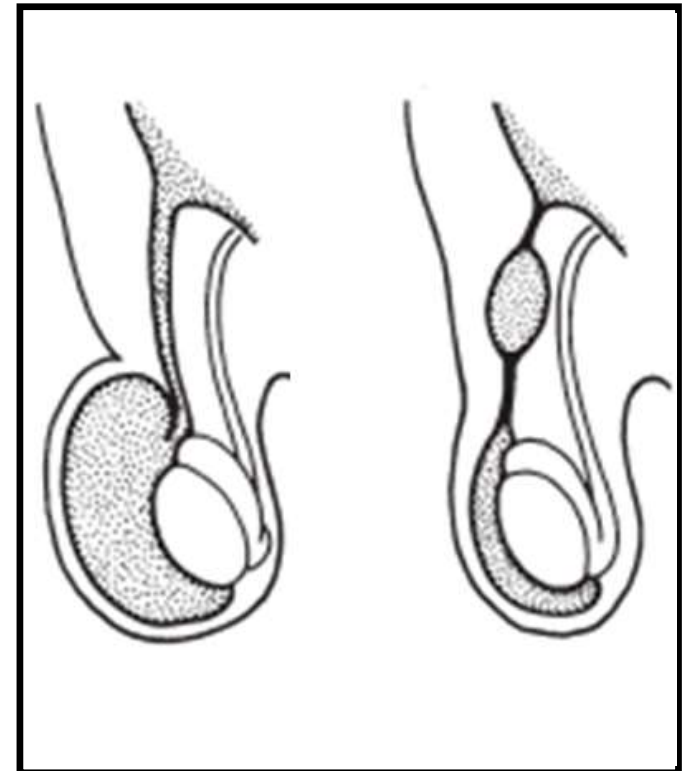
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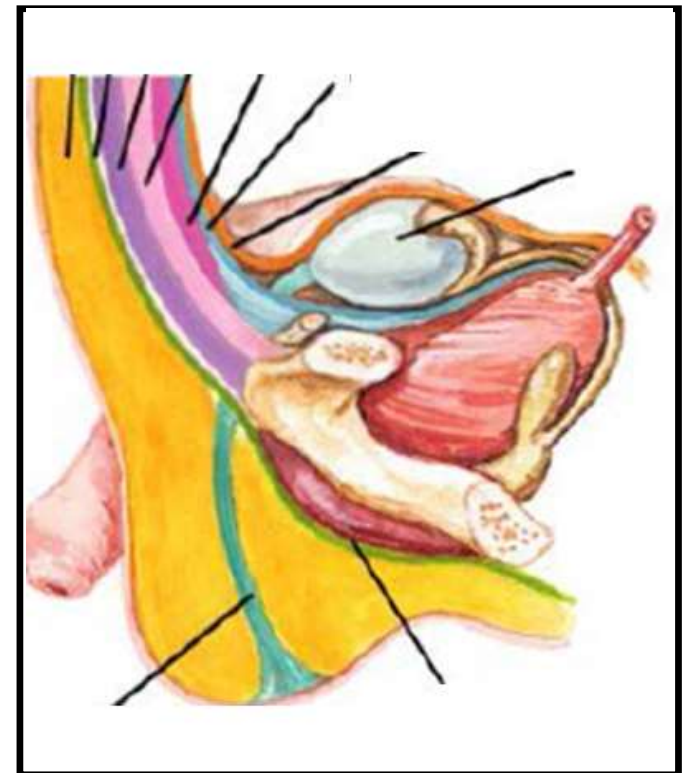
*Undescended testes

- **Malformation in UDT**
 - Testes cannot be manipulated to bottom of scrotum without tension on spermatic cord
- **Factors affecting testicular Descent**
 - *Abdominal phase (gubernacular outgrowth)*
 - Insl3 + MIS *androgen
 - *Inguinoscrotal phase (migration)*
 - androgen (>>GFN>>CGRP)
 - intraabdominal pressure + ? Cremaster
- **Effects of UDT**
 - Atrophy & Infertility
 - Trauma & torsion
 - Malignancy



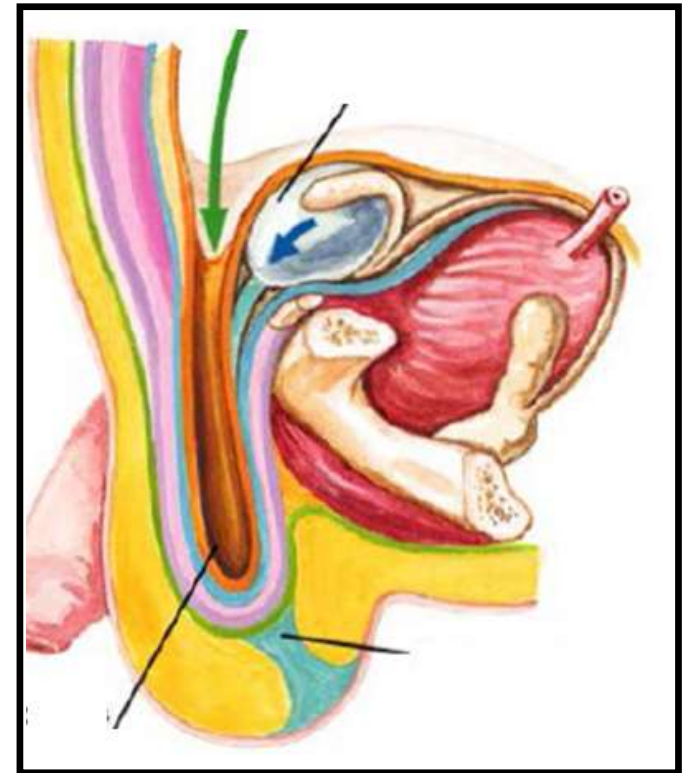
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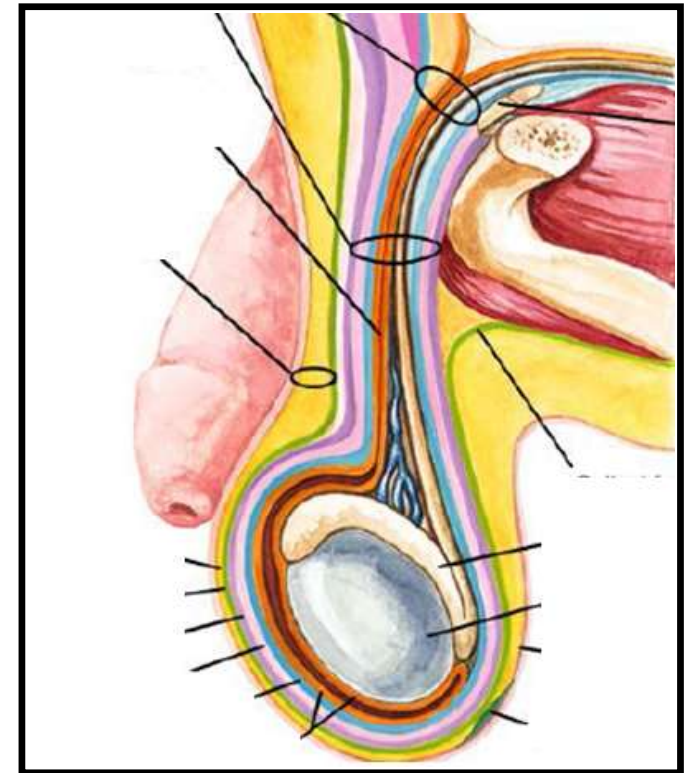
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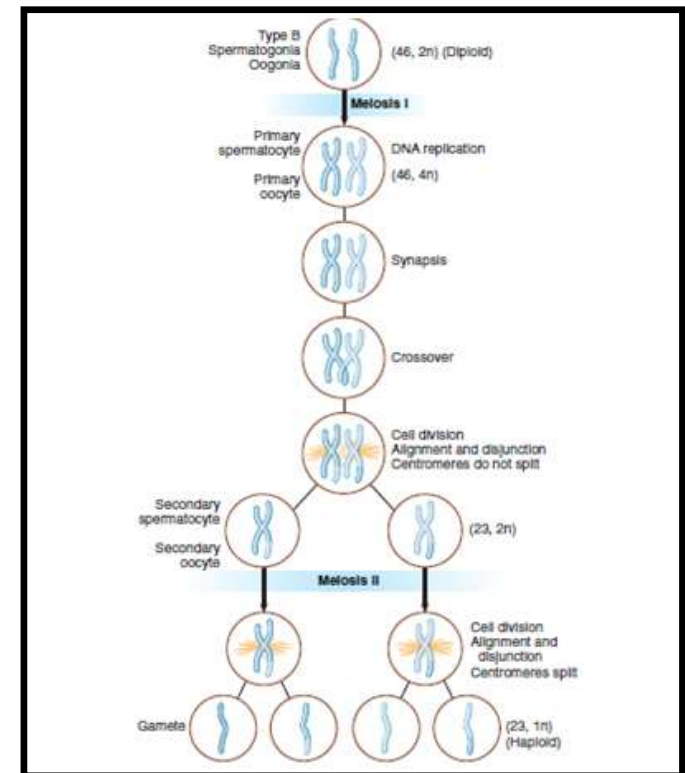


Associated anomalies

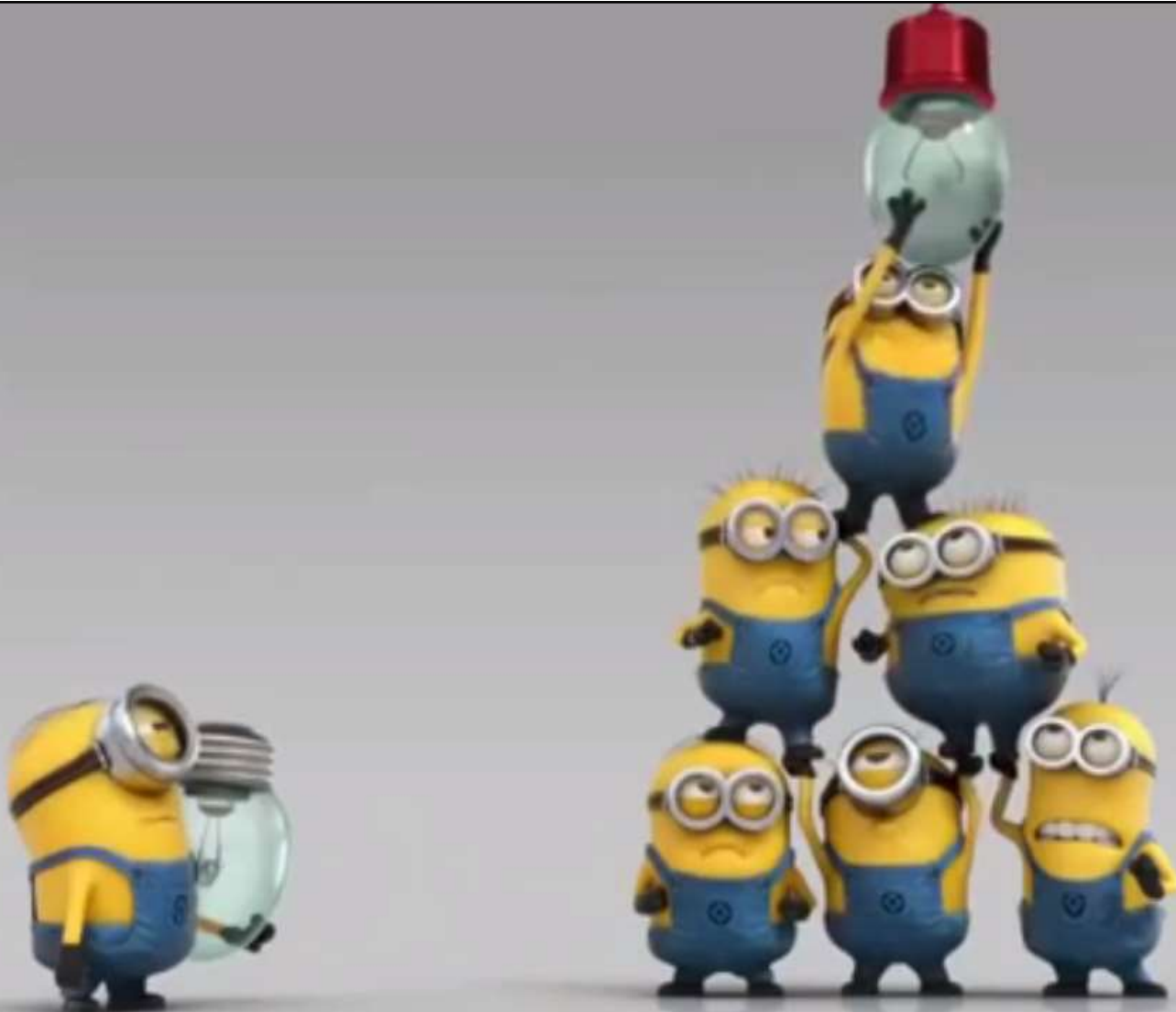
- Hypopituitary, neural tube defects
- mechanical anomalies, abdominal wall defects
- long loop vas

*Undescended testes

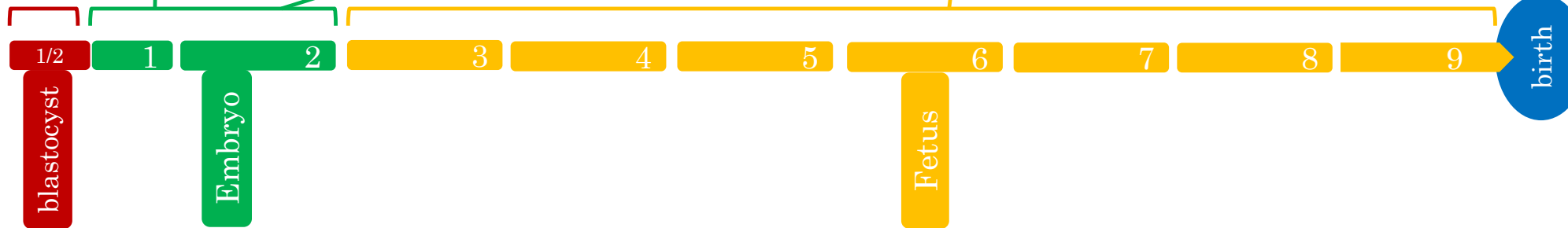
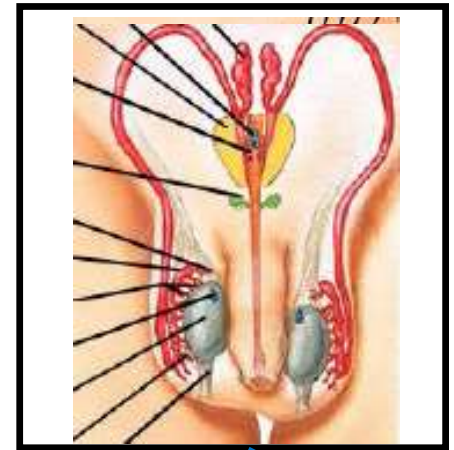
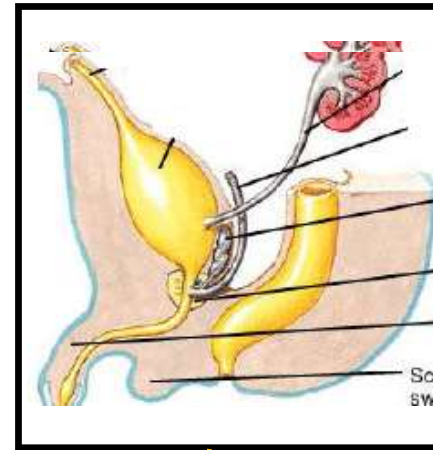
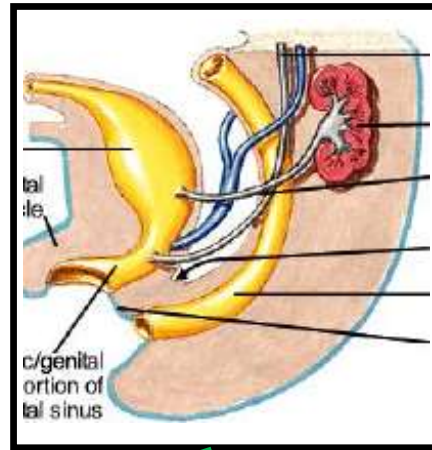
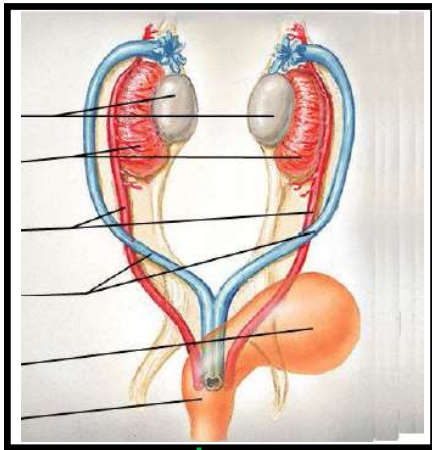
- **Malformation in UDT**
 - Testes cannot be manipulated to bottom of scrotum without tension on spermatic cord
- **Factors affecting testicular Descent**
 - *Abdominal phase (gubernacular outgrowth)*
 - Insl3 + MIS *androgen
 - *Inguinoscrotal phase (migration)*
 - androgen (>>GFN>>CGRP)
 - intraabdominal pressure + ? Cremaster
- **Effects of UDT**
 - Atrophy & Infertility
 - Trauma & torsion
 - Malignancy



Part - 4
Genital Ducts

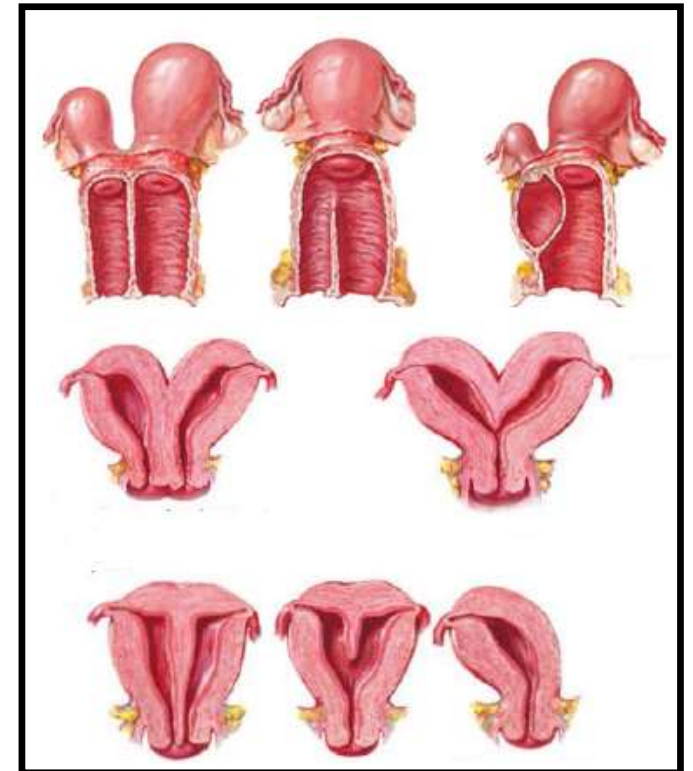


Genital ducts: Development



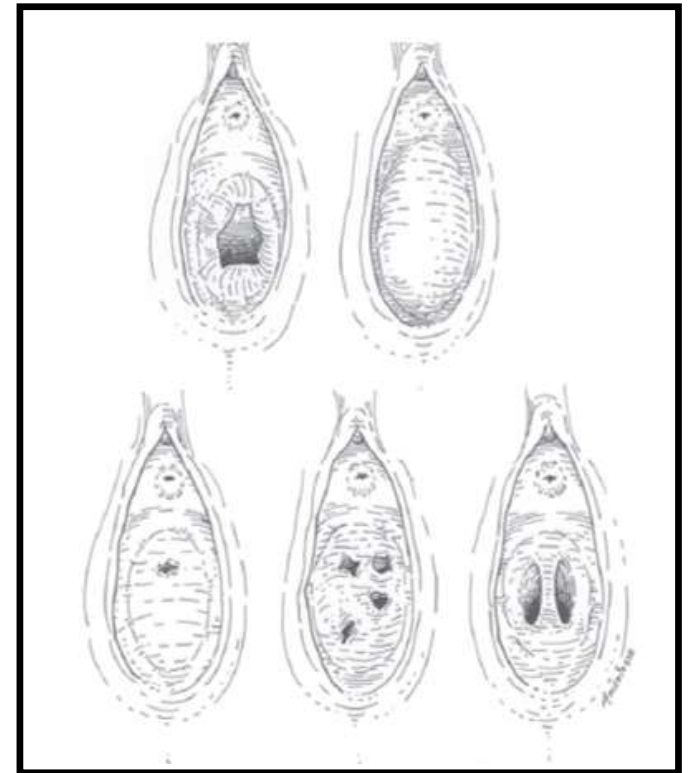
Genital ducts: Anomalies

- **Abnormalities of fusion**
 - Uterine duplication, rudimentary horns
 - Vaginal duplication, OHVIRA
- **Congenital vaginal obstruction**
 - Imperforate hymen
 - Transverse vaginal septum
 - Vaginal atresia
 - Vaginal agenesis (ALV)
- * Labial adhesion
- **Introital mass**
 - Urethral prolapse, polyp & cyst
 - Prolapsed ureterocele
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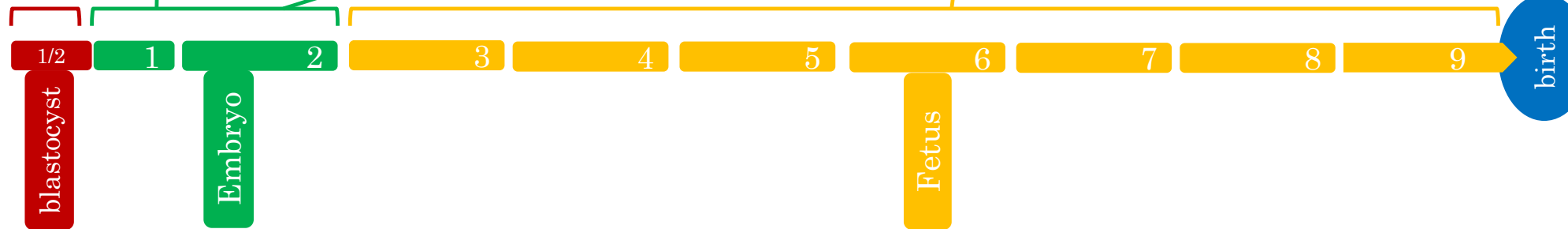
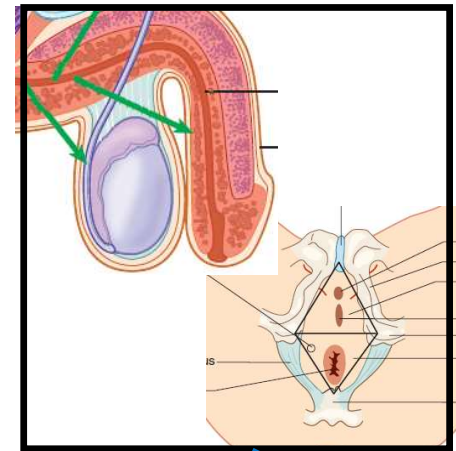
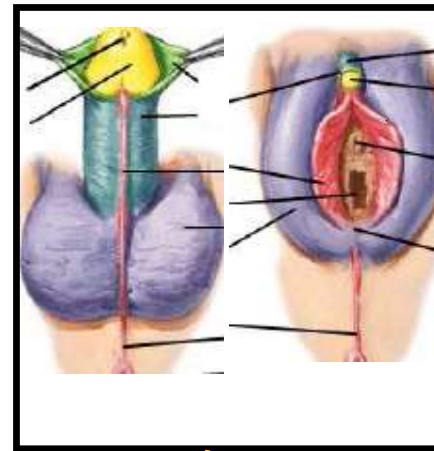
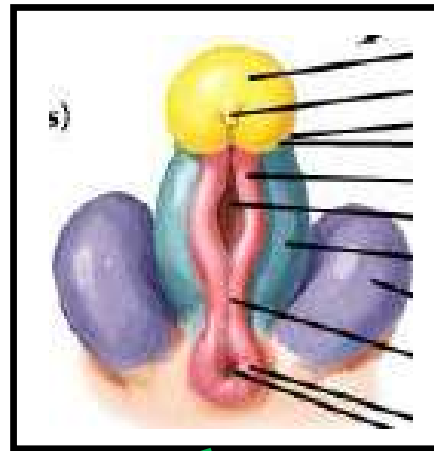
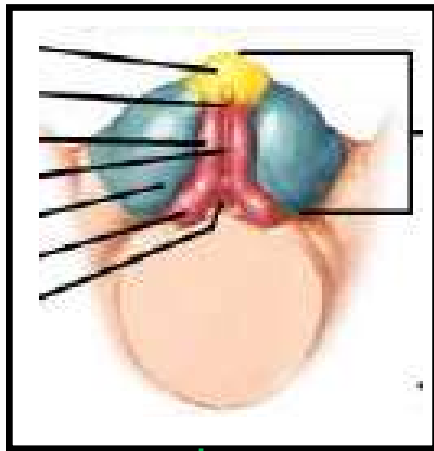
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Part - 5
External Genitalia

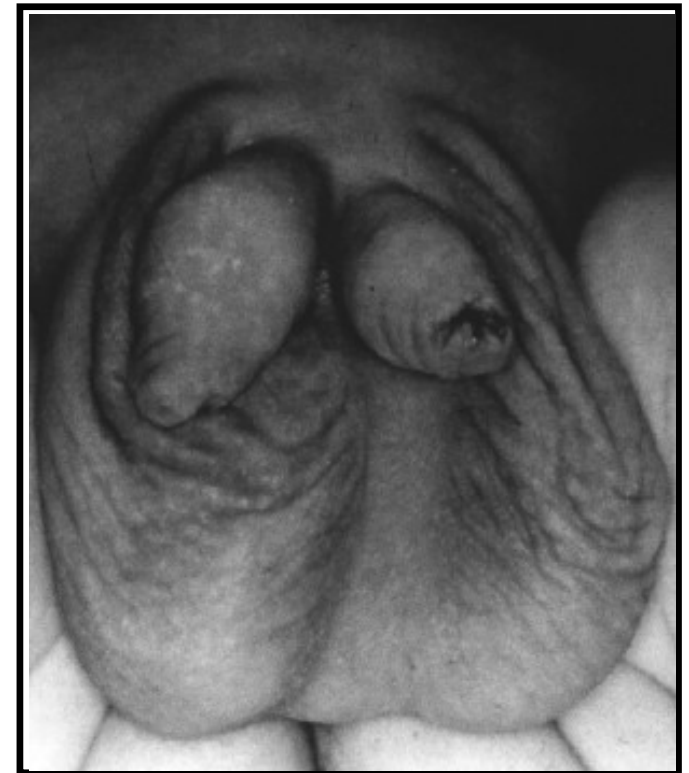


External Genitalia: Development



External genitalia: Anomalies

- **Male**
 - Phimosis (persistent primary)
 - **Abnormal number** (Aphallia, Diphallia)
 - Abnormal urethral opening (**Hypospadias**)
 - Accessory opening (congenital UCF, duplication)
 - Abnormal orientation (chordee, torsion)
 - Micropenis, Inconspicuous penis (buried, webbed)
 - Scrotal anomalies (bifid, ectopic, agenesis, scrotoschisis)
 - Penile mass (cysts)
- **DSD (Disorders of sex development)**
 - 46 XX DSD
 - Virilized female (*female pseudohermaphrodite*)
 - 46 XY DSD
 - Undervirilized male (*male pseudohermaphrodite*)
 - Sex chromosome DSD
 - Turner syndrome (45 X), Klinefelter (47 XY)
 - Mixed gonadal dysgenesis
 - Ovotesticular DSD (*true hermaphrodite*)



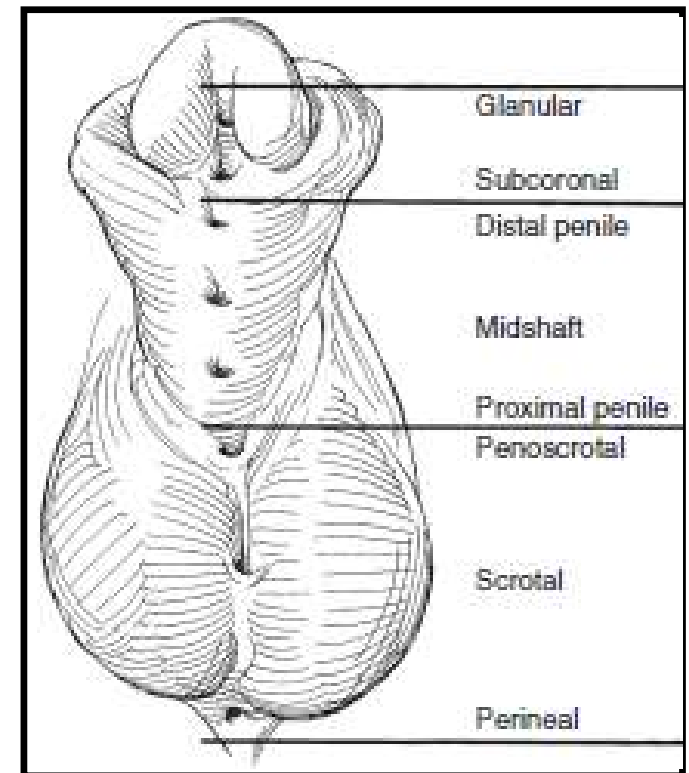
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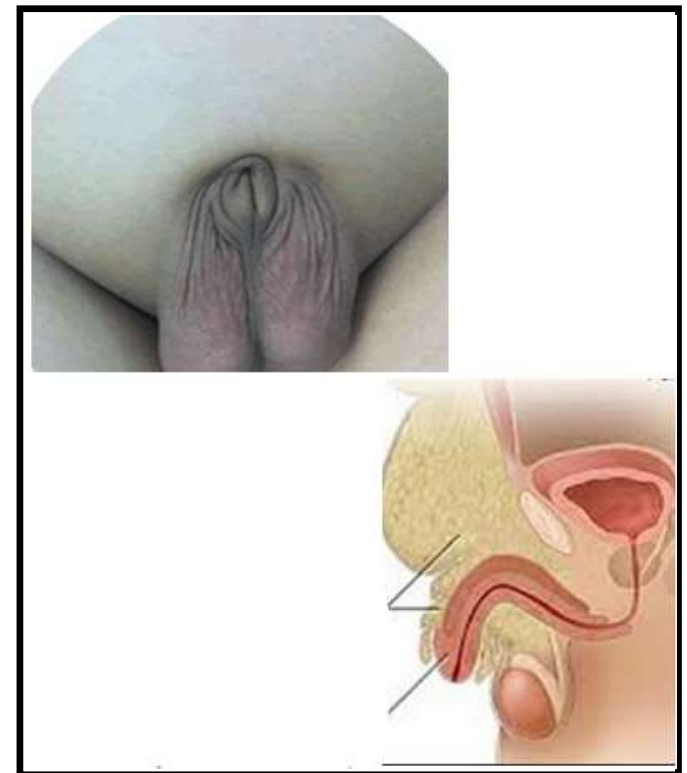
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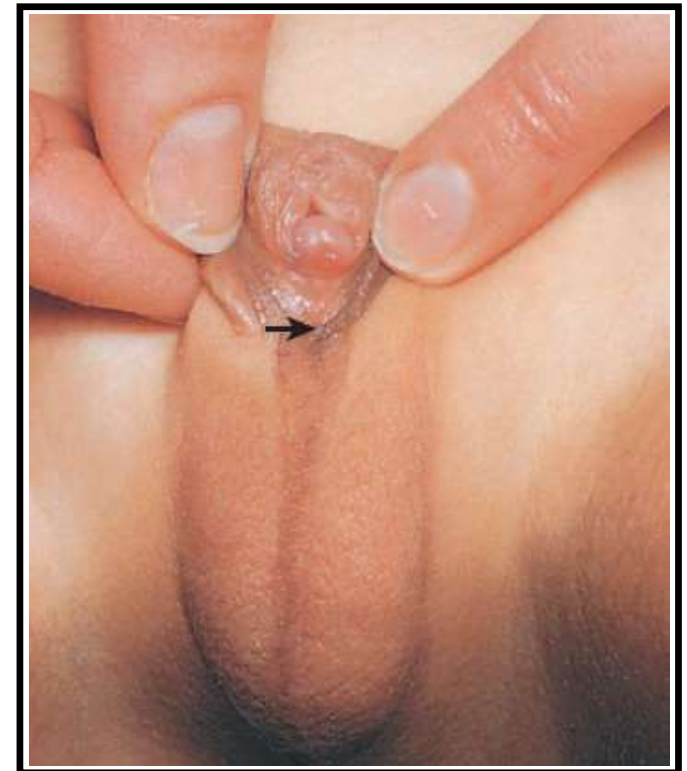
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*Hypospadias

- **Malformations in hypospadias**
 - Abnormal urethral opening (ventral)
 - Hooded prepuce, Chordee, bifid scrotum
 - Ass. anomalies - Microphalus, UDT, DSD
- **Factors leading to hypospadias**
 - Incomplete fusion of urethral folds
 - ?Hormonal/receptor, ?environmental
 - ?Genetic, ?enzymes
- **Effects of hypospadias**
 - Cosmetic
 - Infertility
 - Micturation
 - Erection
 - UTI
 - Meatal stenosis



THE

END



References

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