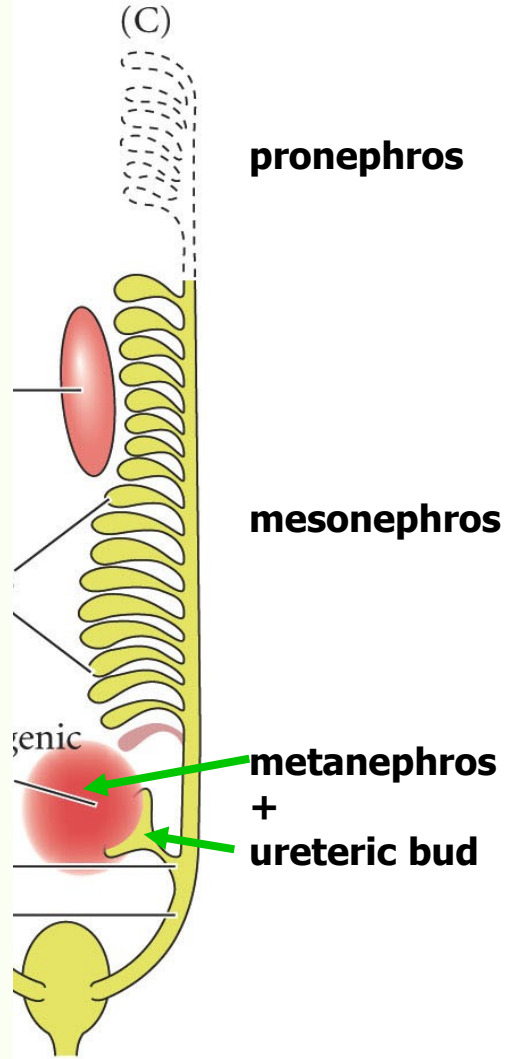
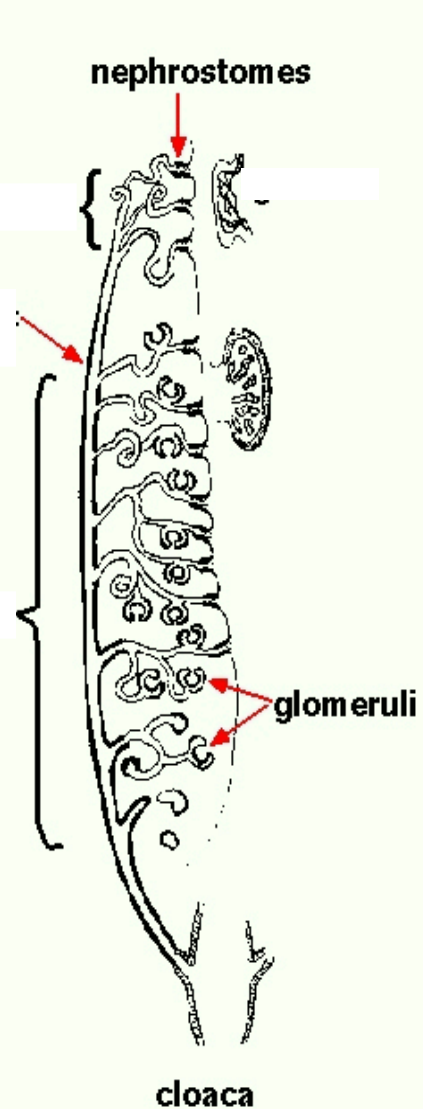
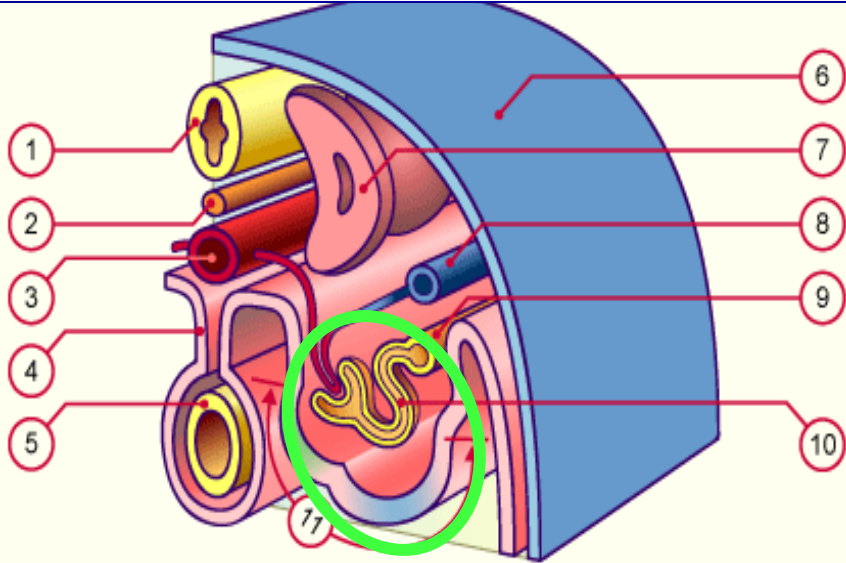
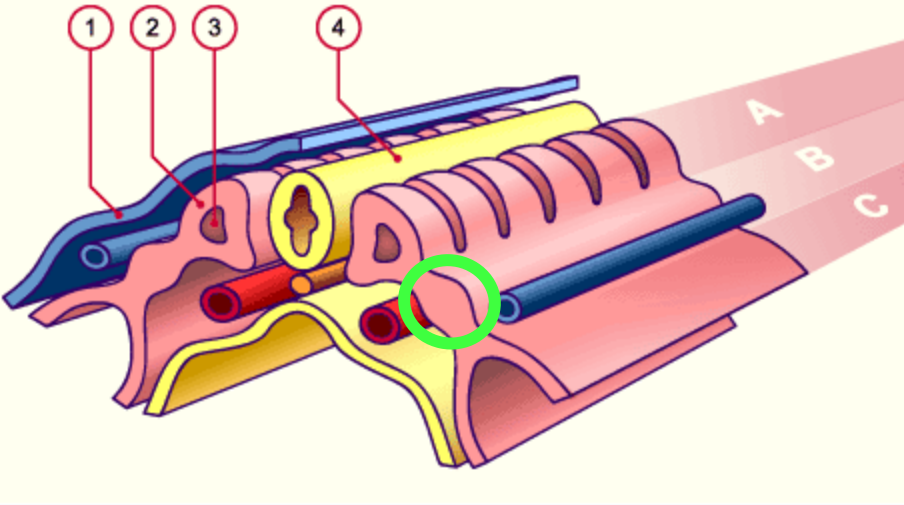


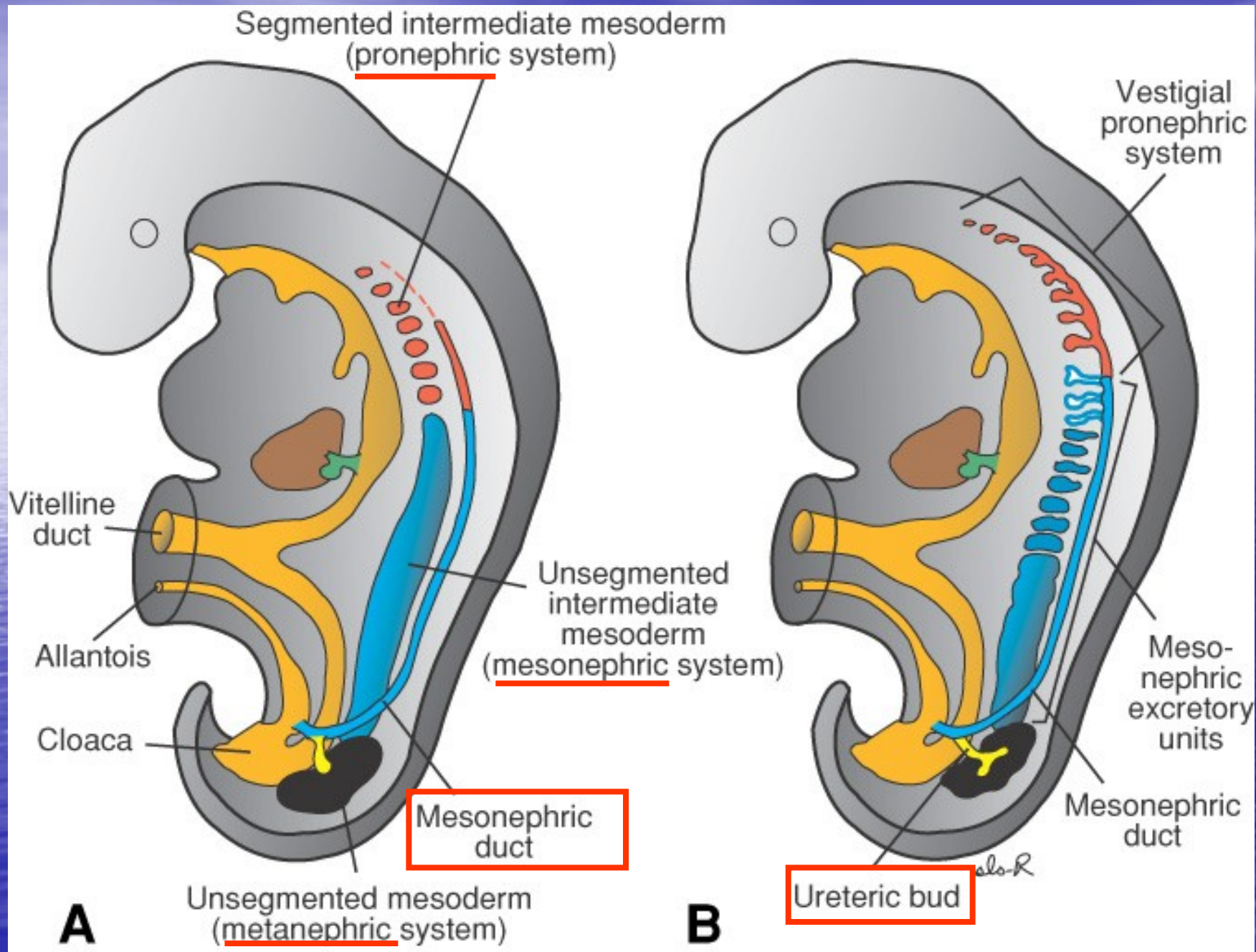
Urinary system

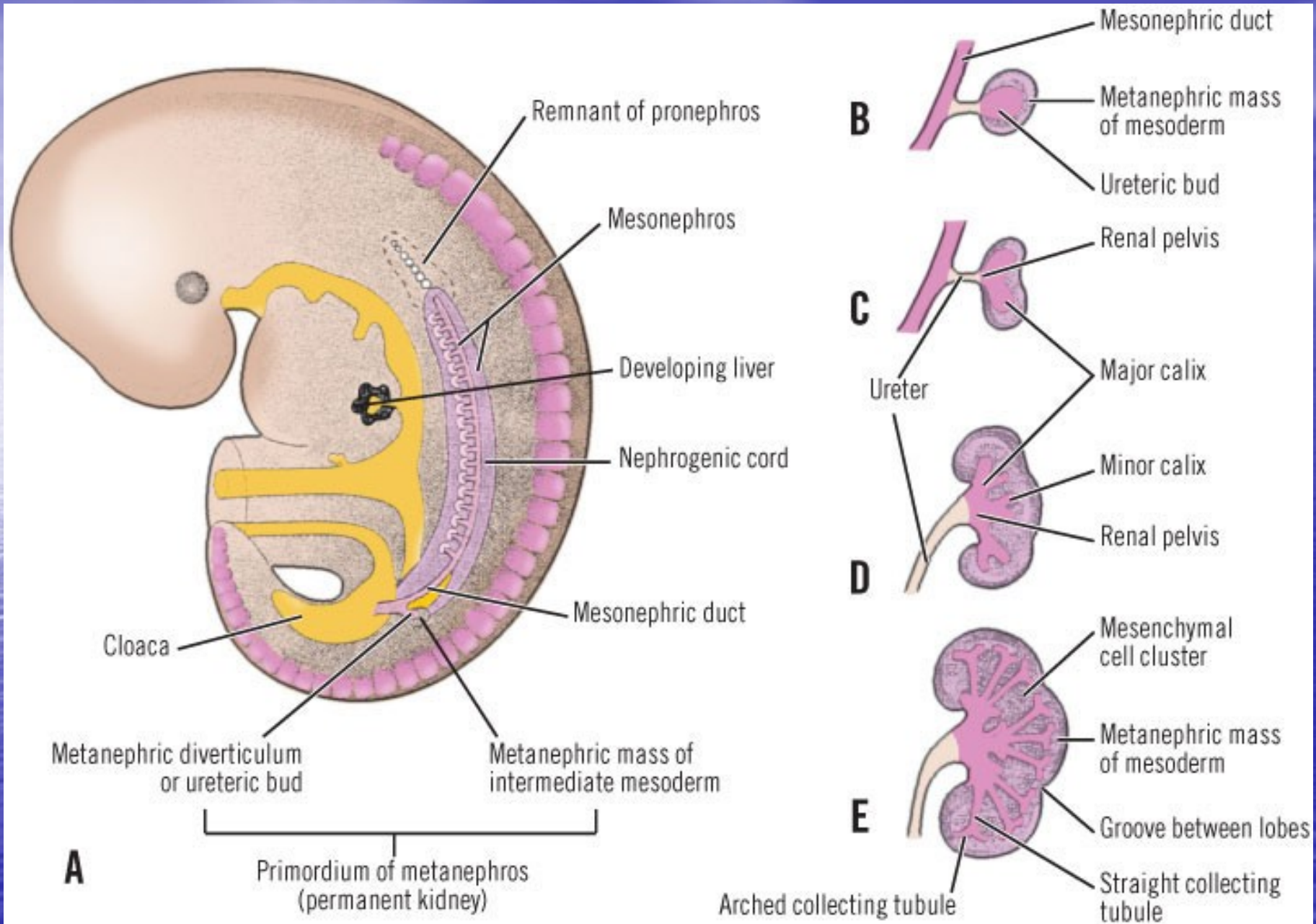


Development
Teratology

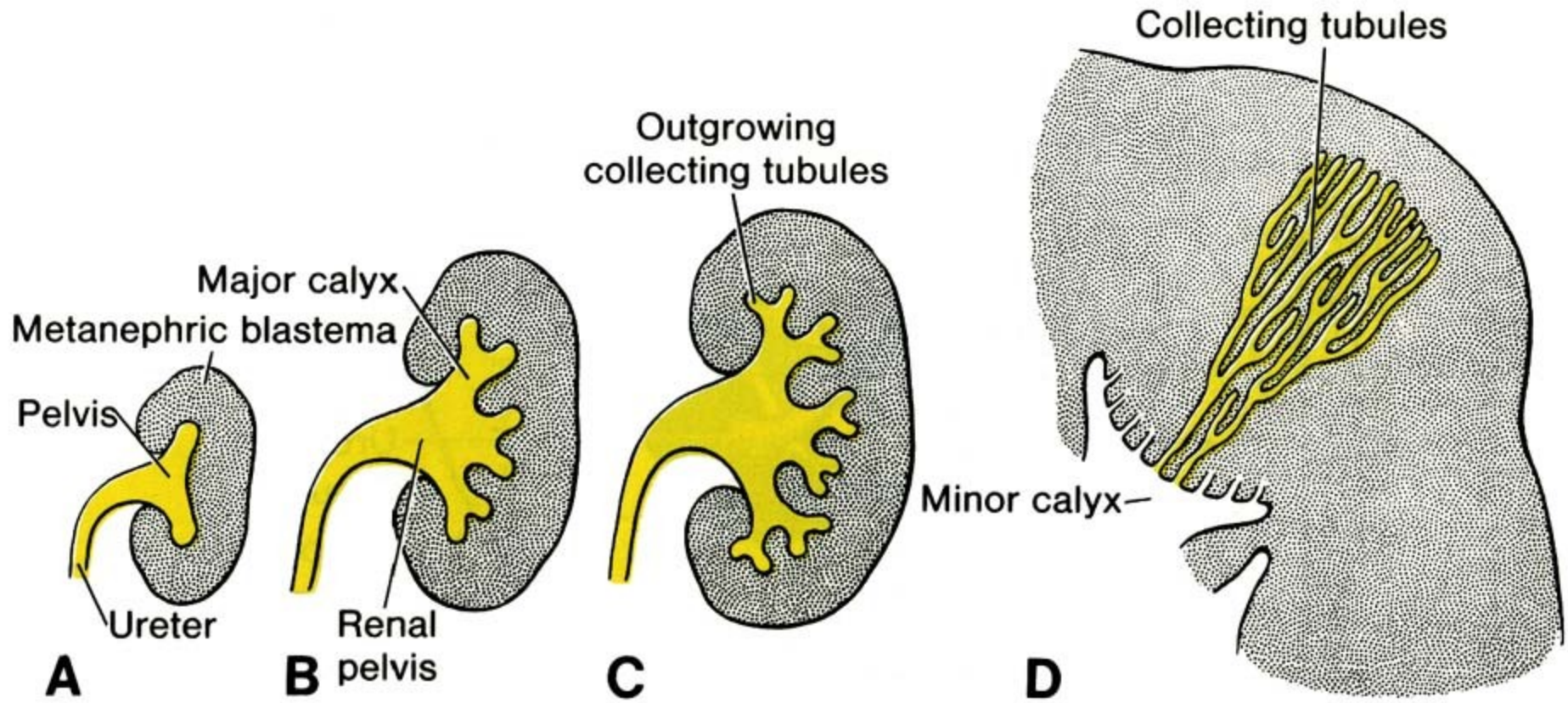
- paraaxial
 - intermediary
 - lateral
- mesoderm



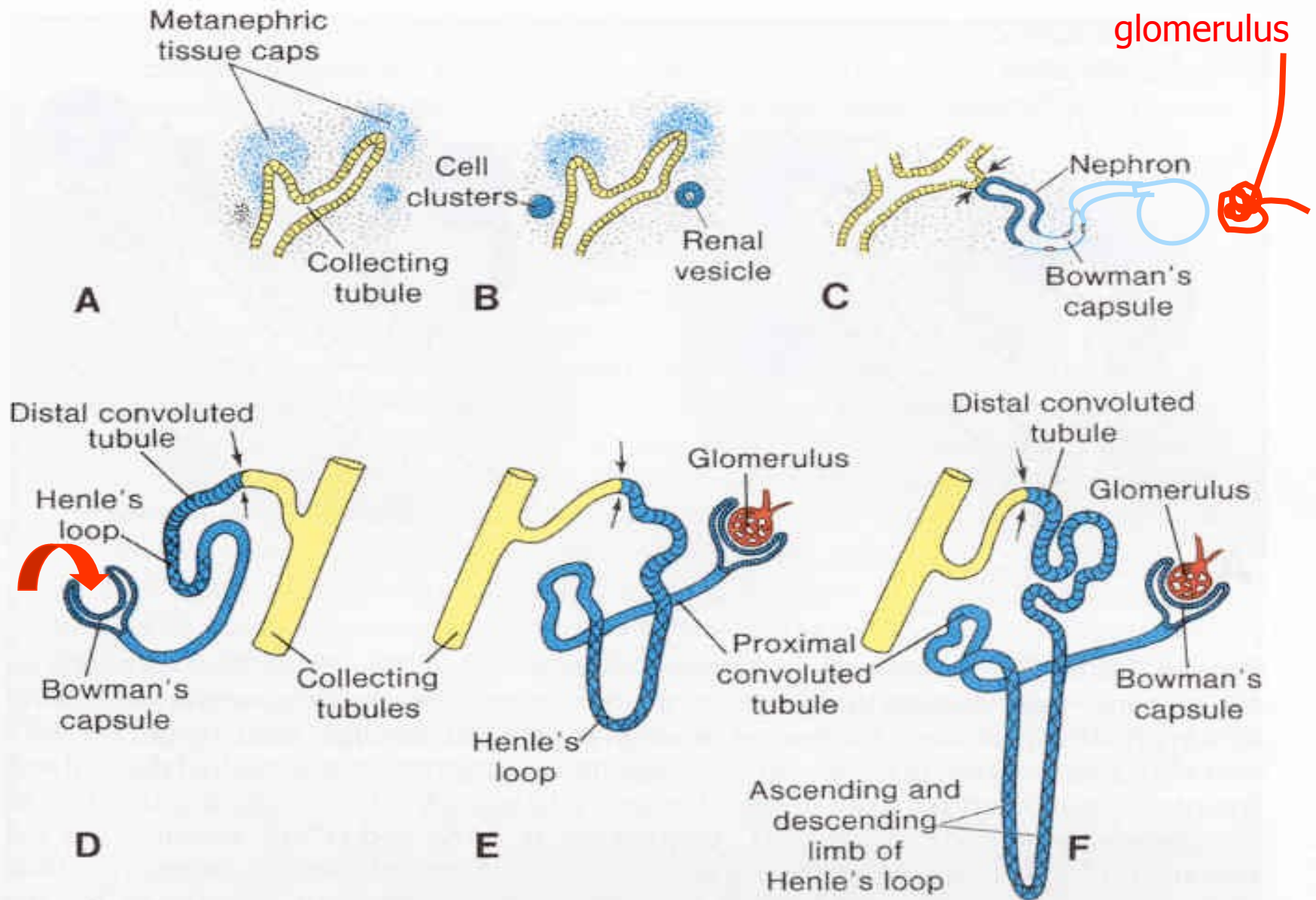




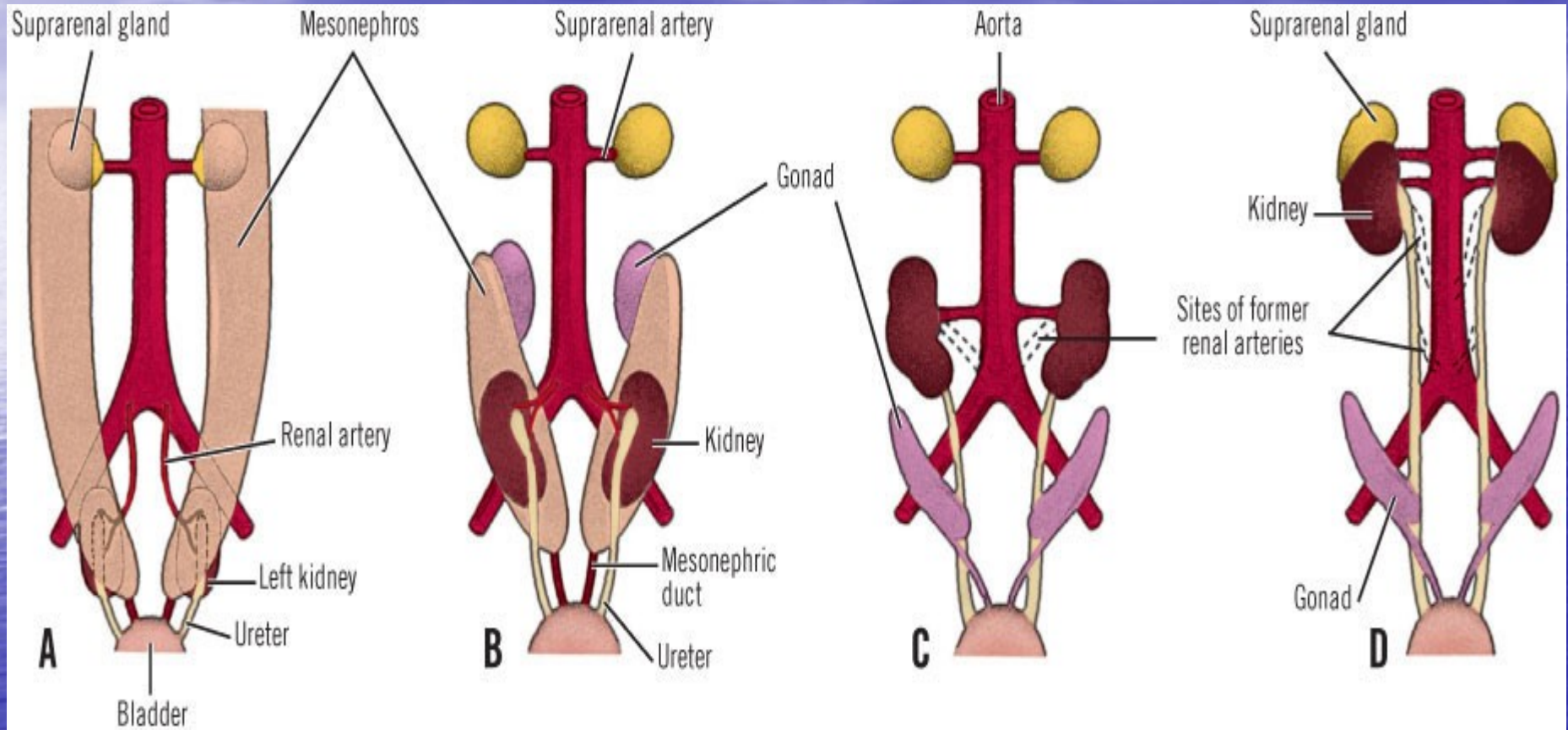
Kidney development



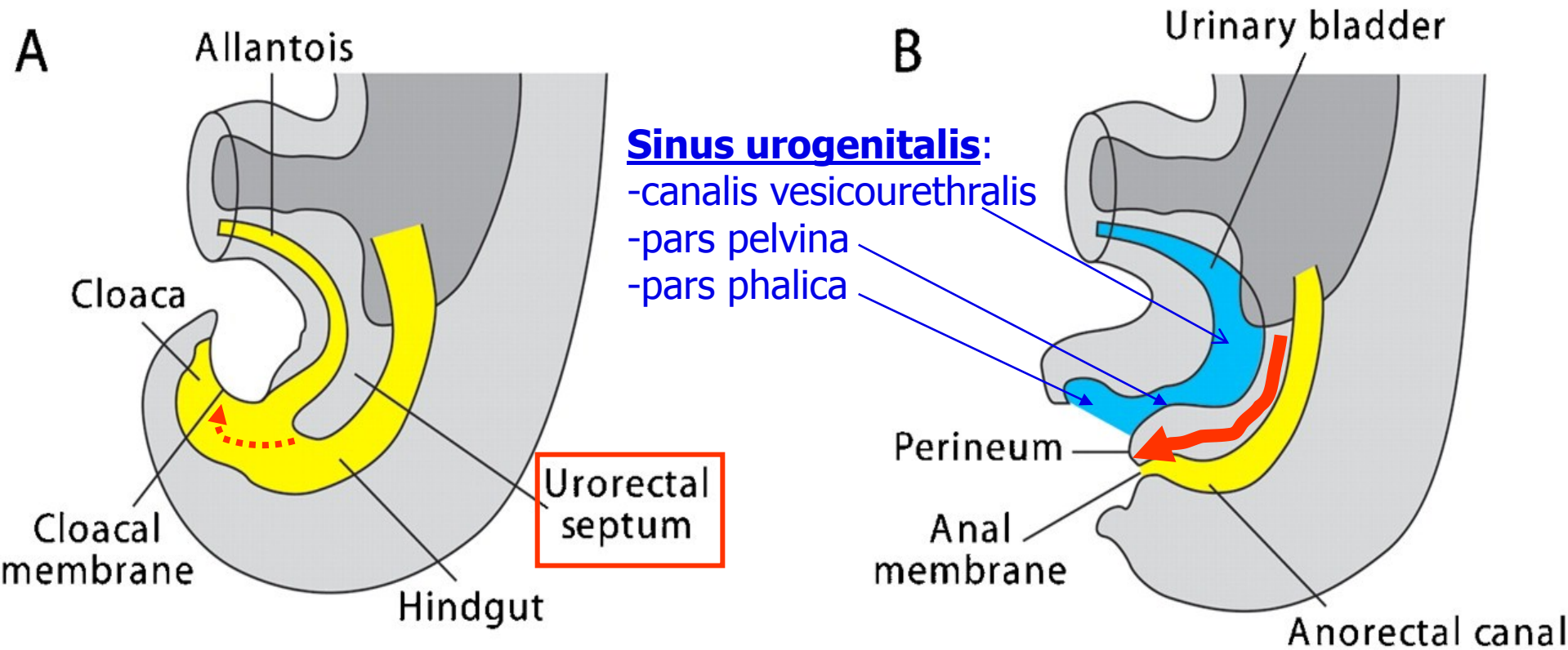
Nephron development



Ascensus renis



Cloaca development

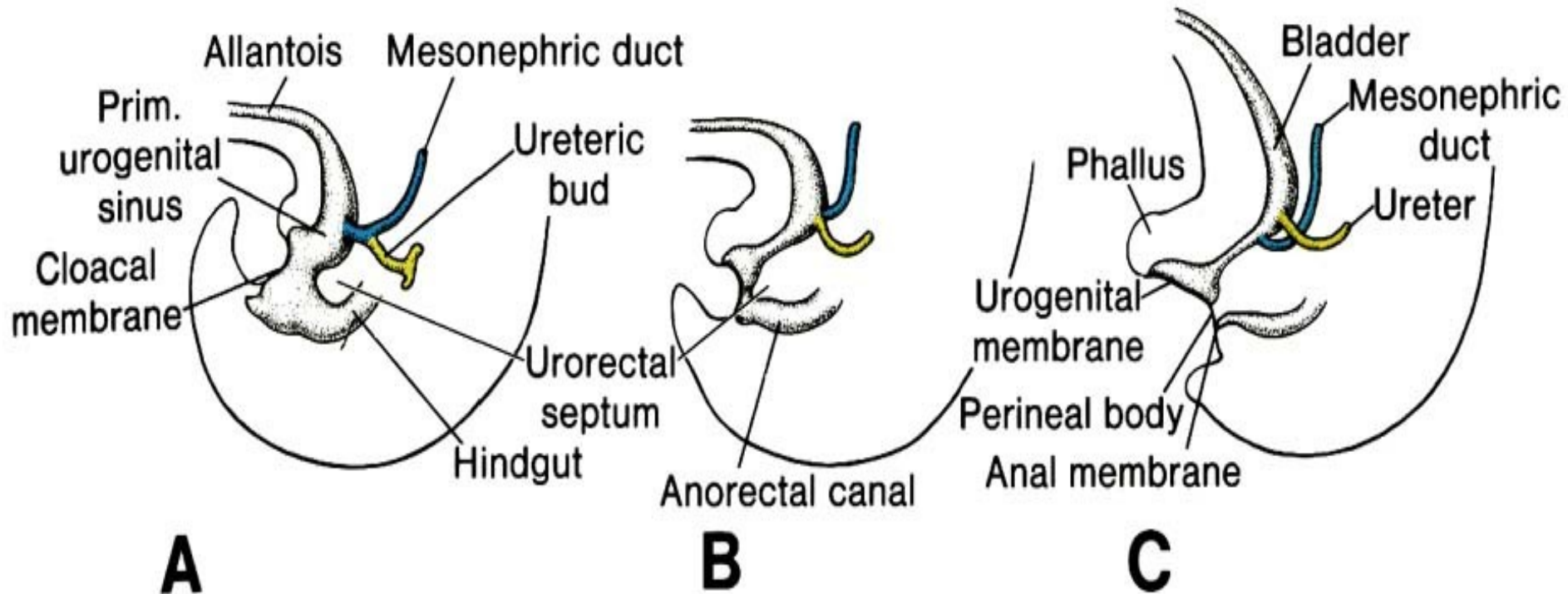


Sinus urogenitalis

- canalis vesicourethralis ⇒ urinary bladder,
- pars pelvina ⇒ f. **urethra** // m. **pars prostatica** + diaphragmatica urethrae
- pars phalica ⇒ f. vestibulum vaginae // m. pars phalica urethrae

f. – female // m. – male

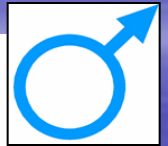
Cloaca



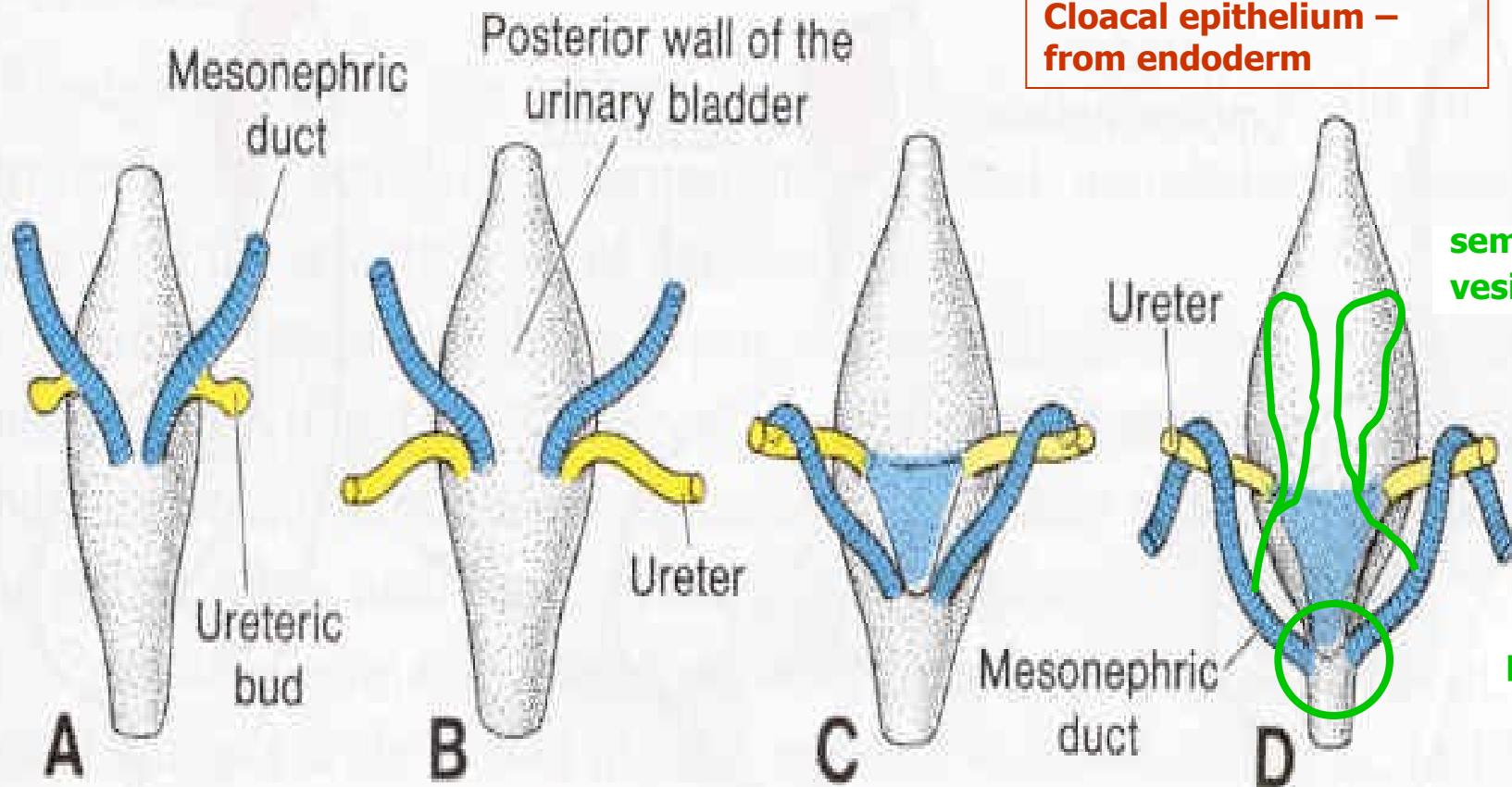
Growth of cloacal wall \Rightarrow mesonephric duct with ureteric bud (ureter) is drawn, duct and ureter and their outlets are separated

(see dorsal view of urinary bladder)

Wolffian duct (ductus mesonephricus) and ureteric bud



Cloacal epithelium – from endoderm



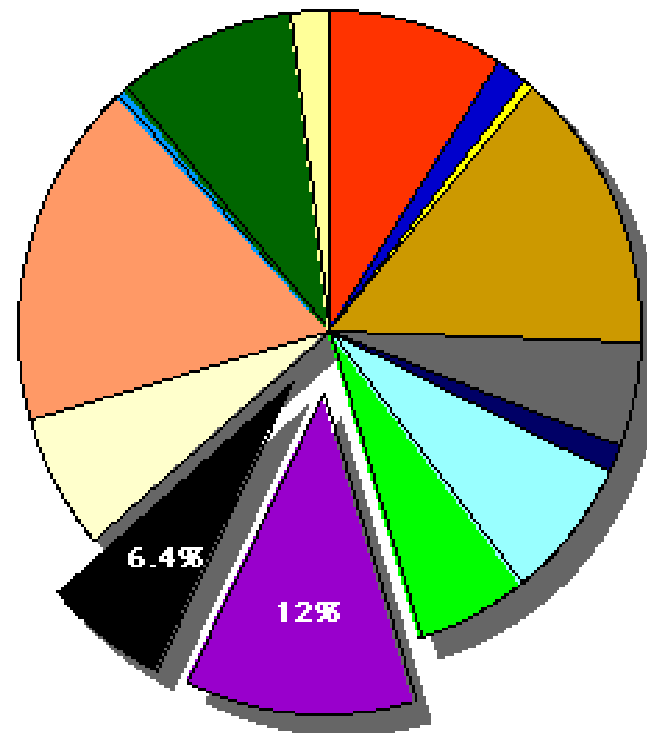
Epithelium of trigonum vesicae – from mesoderm

Congenital malformations (CM) of urinary system

Hypoplasia – small kidney

Reflux – retrograde movement (urine returns into kidney)

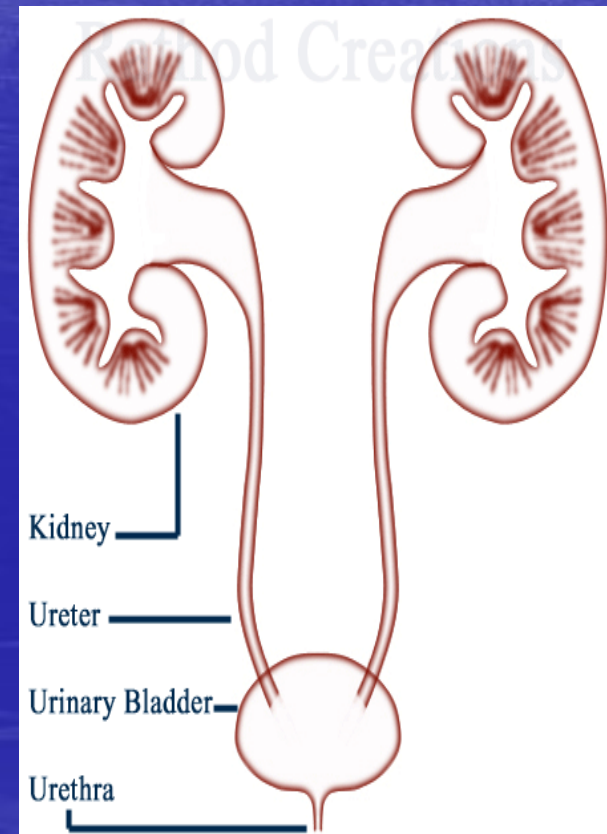
Hydronephrosis – urine stasis



Data source: Congenital Malformations Australia 1981-92

Congenital malformations (CM)

- 1. CM of kidney
- 2. CM of pelvis and ureter
- 3. CM of urinary bladder
- 4. CM of urethra




1. CM of kidney

- anomalies of number
- anomalies of shape
- anomalies of position (ectopia)
- anomalies of parenchyma (nephrodysplasia)
- anomalies of vessels

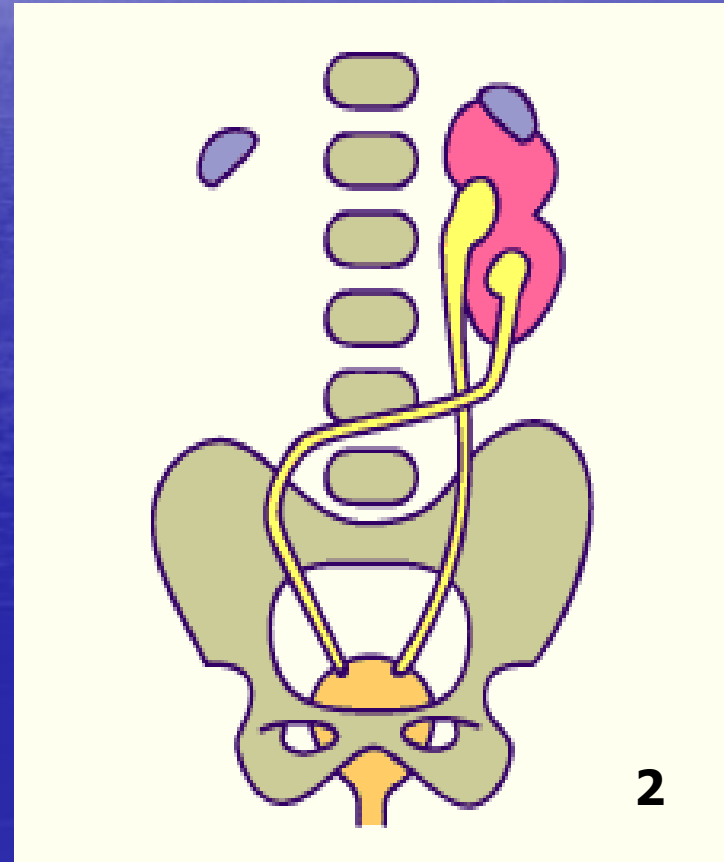
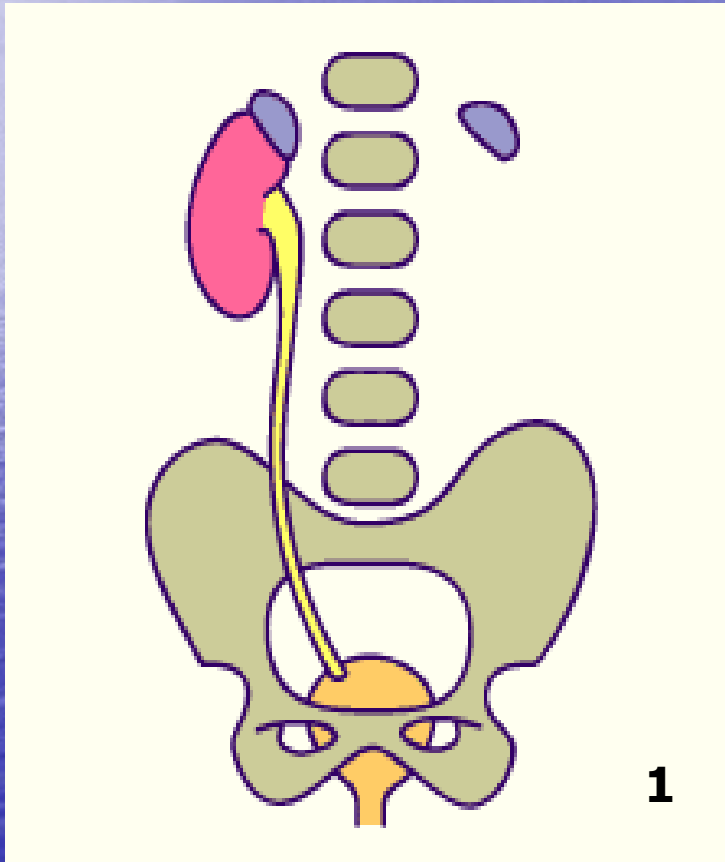
Kidney malformations arise at the beginning of development (*development of metanephros isn't induced by ureteric bud or both metanephros are closely together – before week 6*) or later (*during incomplete ascensus renis – after week 8*).

Aggenesis renis

- **bilateral** (1 : 3000; prenatal dg. – **oligohydramnion, hypotrophic fetus,** *skeleton deformities and lung hypoplasia due to fetus oppresion*) - (*death by uremia and respiratory distress*)
- **unilateral** (1 : 1500) + aggenesis of ipsilateral ureter and renal vessels;
- **etiology**: 
absence of metanephros, ureteric bud did not develop or did not reach metanephros (regression)
– metanephros development was not induced
- – *genetic disposition*
-

1 – kidney agenesis

2 – kidney agenesis + cross ectopia of ureter

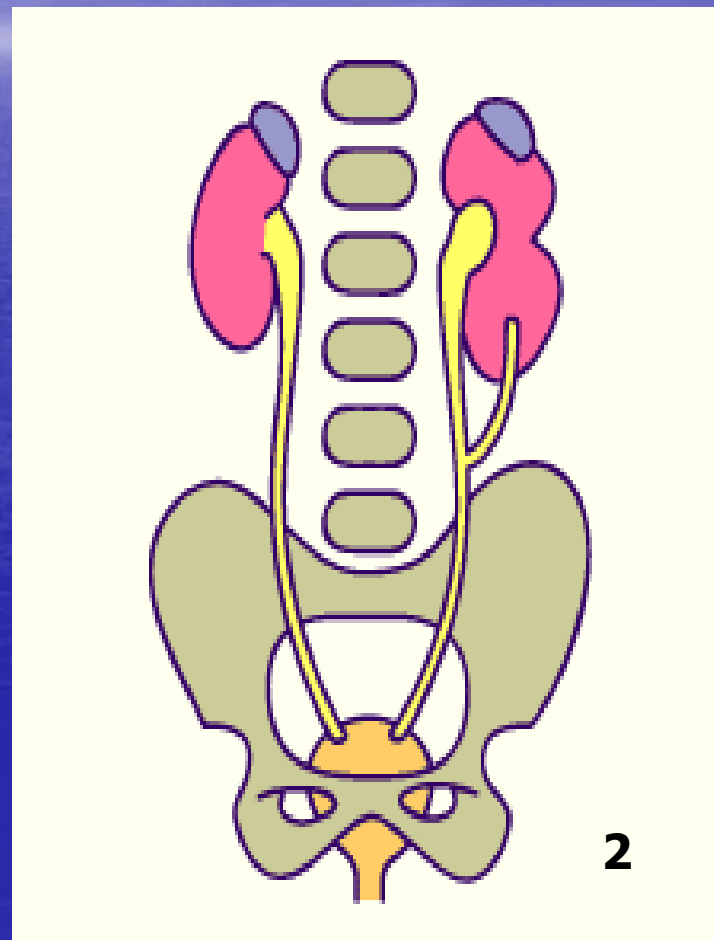
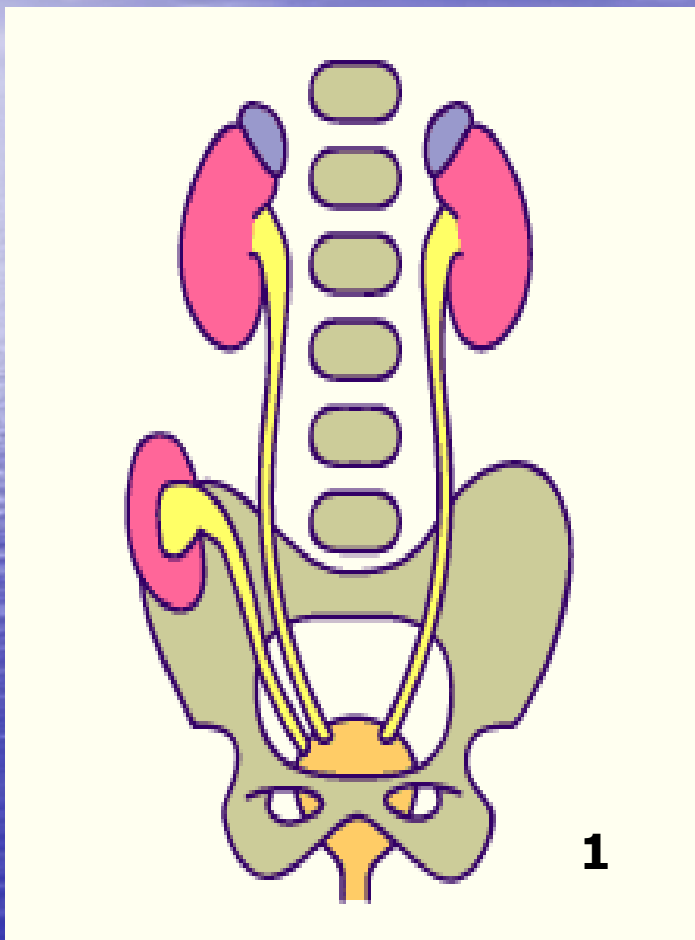


Supernumerary kidney (2-3 % newborns)

Ren duplex

- unilateral or bilateral
- + **pelvis duplex** and partially or completely **ureter fissus** or **ureter duplex**
- etiology: 2 ureteric buds from one mesonephric duct or branched ureter

1 – ren duplex et ureter duplex, 2 – ureter fissus

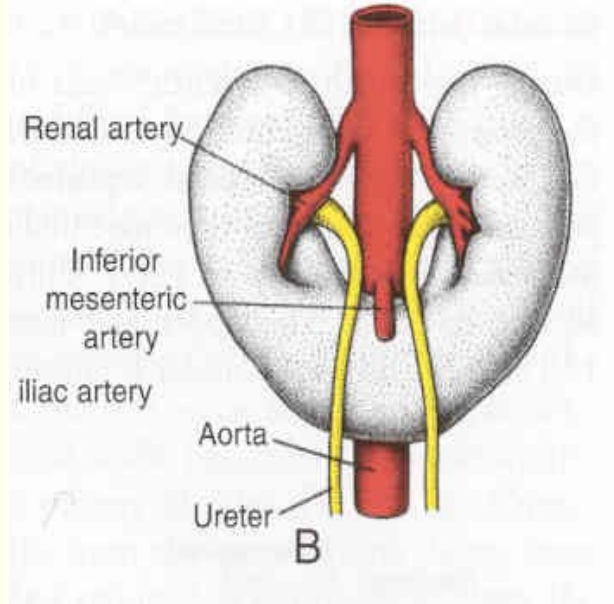
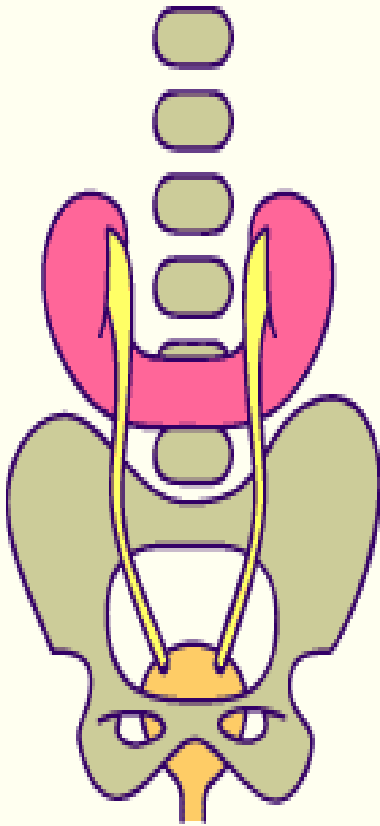


Shape malformations of kidney:

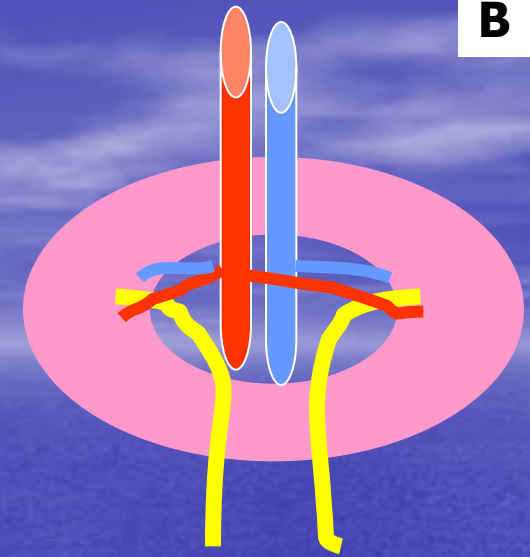
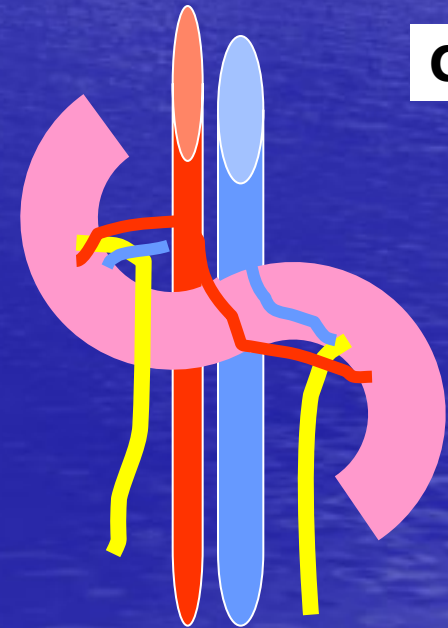
Horse-shaped kidney (*ren arcuatus*)

1:500

- **etiology**: fusion of lower pole of both metanephros in front of large vessels (aorta + v.cava inf.)
- fused parenchyma = isthmus „brakes“ ascensus renis bellow detachment of a. mesenterica inf. (+**position anomaly - ectopia**) and rotation (+**malrotation**; hilus – ventrally), ureters run in front of isthmus – **+ renal vessels duplication**

A

A – ren arcuatus
B – ren fungiformis
C – ren sigmoideus

B**C**

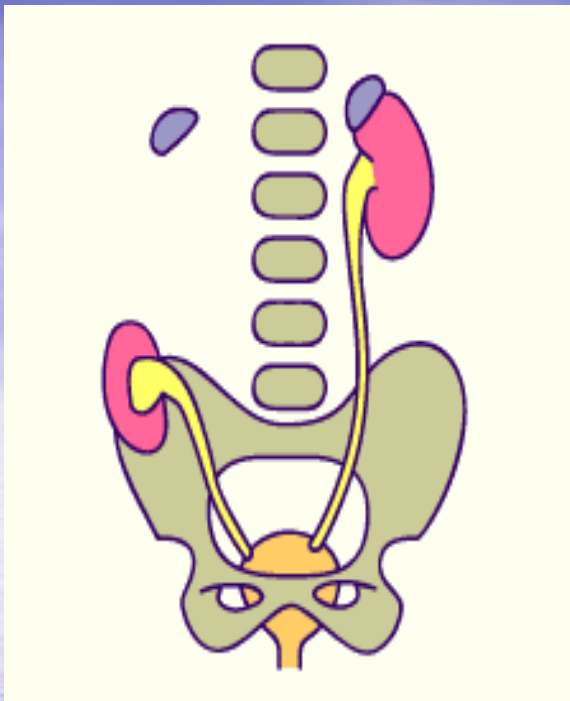
Anomaly of the shape + ektopia:

+ [urine stasis – hydronefrosis
vesicaureteric reflux
secondary infections]

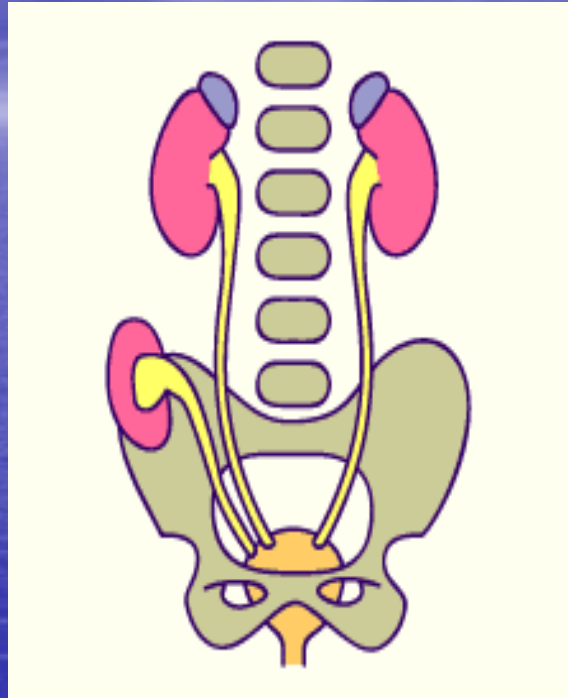
Position anomalies:

Ectopia of kidney uni-, bilat.

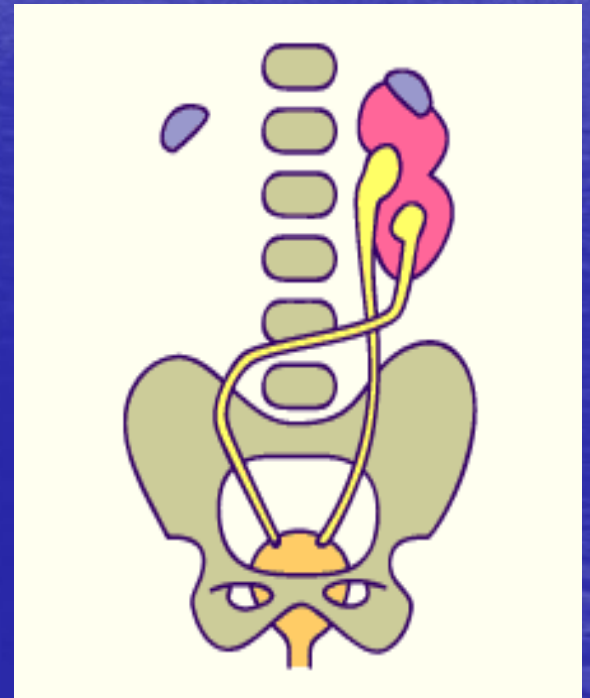
- - **ren pelvicus (ren sacralis, ren lumbalis)**: retention of kidney during ascensus renis
- - **cross ectopia**: both ureters grow into metanephros on one side or during ascensus renis one kidney transfers on the opposite side and fuse with the other kidney



Ren pelvici

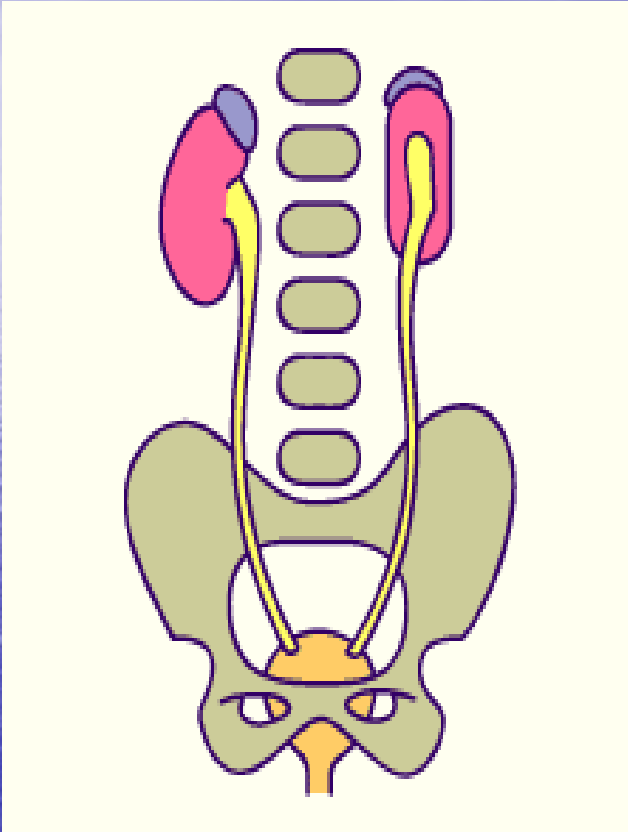


+ ren + ureter duplex



Cross ectopia

Malrotation (or hyperrotation) of kidney



- is connected with ectopia or anomaly of kidney shape
- hilus – ventrally (embryonic position) or dorsally
- *Notice:(normal adult position of hilus is medial)*

Defekts of parenchyma:

Polycystic kidneys

nephrodysplasia polycystica



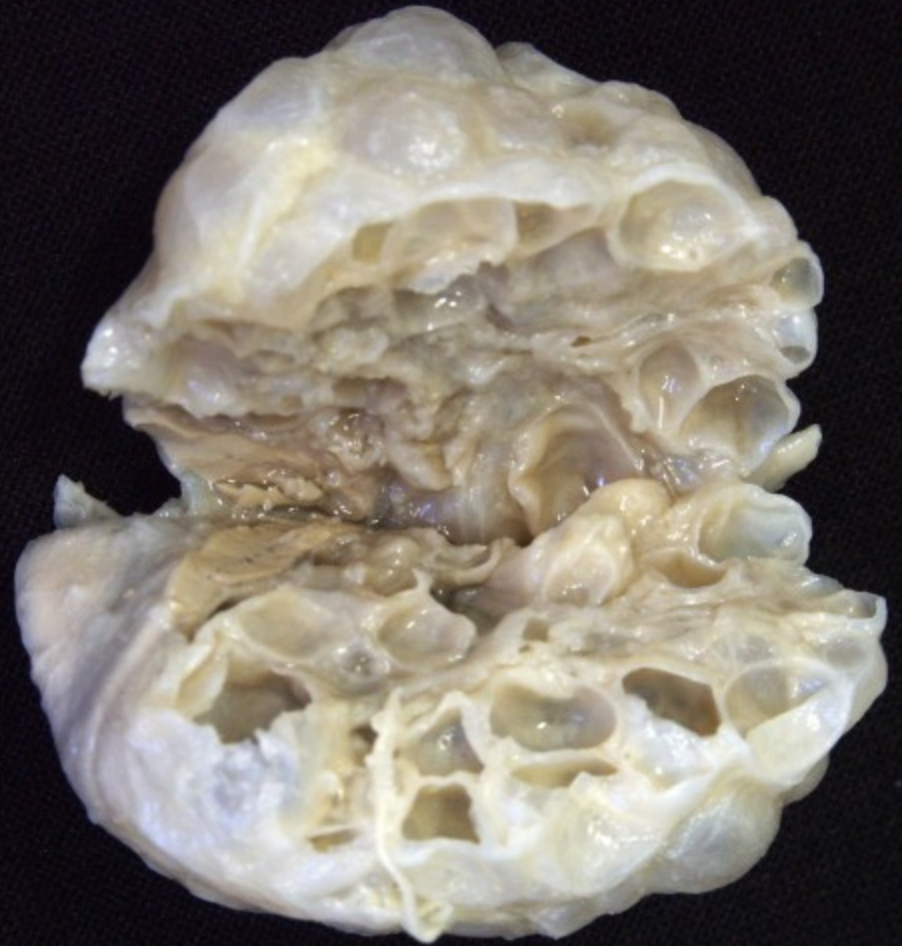
- diffuse cystic malformation (always bilat.)
 - cystic degeneration of kidney
- 2 forms of polycystic disease:
 - **autosomally dominant** type
 - 'adult' macrocytic form ■
 - **autosomally recessive** type
 - 'infantil' microcytic form ■

autosomally dominant type

■ APCD – Adult Polycystic Disease ■

- Disease manifests in adulthood (after 30th); 1:400 - 1000, probability of transmission to offspring is 50 %;
- etiol.: patol. genes on 4th and 16th chromosomes – **insufficient polycystin production** (membrane protein necessary for differentiation of cells in renal tubules).
- **Klinic manifestation:** bilat. enlarged kidney, macroscopic cysts, abdominal and/or lumbal pain, hematuria, hypertension, infections, renal insufficiency and failure.
- **Dg.:** (FA), abdomen palpation, sono event. CT
- **Th.:** symptomatic, decelerate progression of disease, renal failure – renal functions have to be compensated (hemodialysis, peritoneal dialysis, transplantation)

Polycystic kidney – macroscopic cysts are seen also on the kidney surface

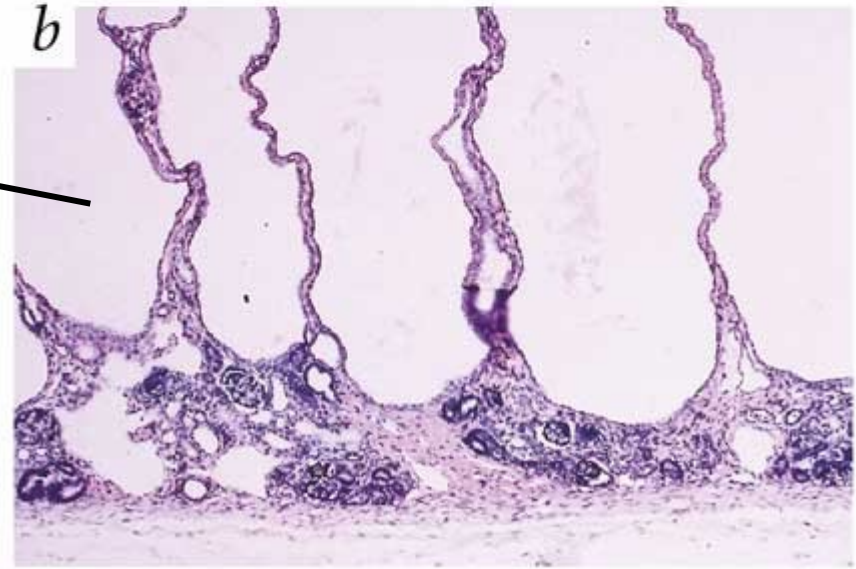
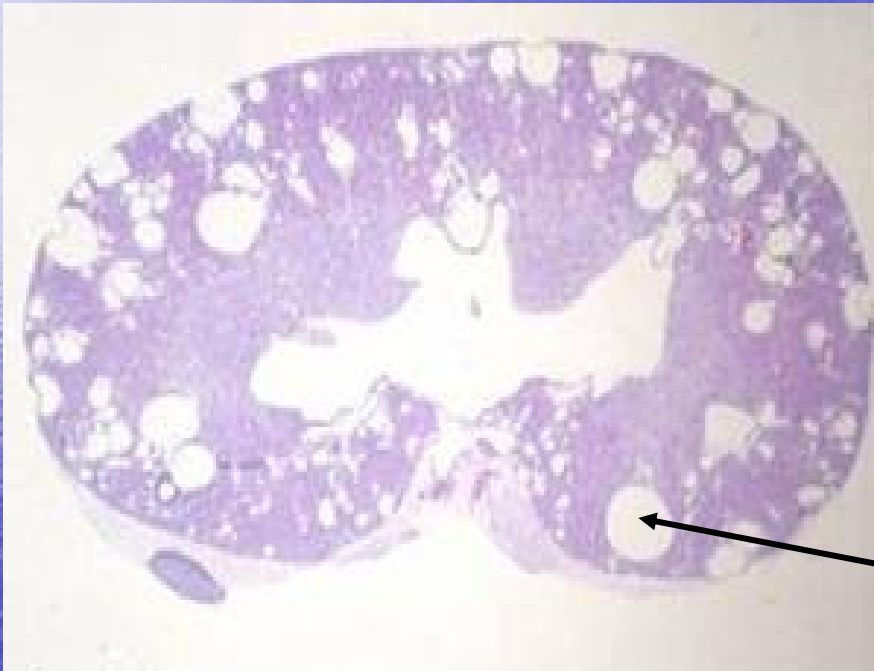


autosomally recessive type

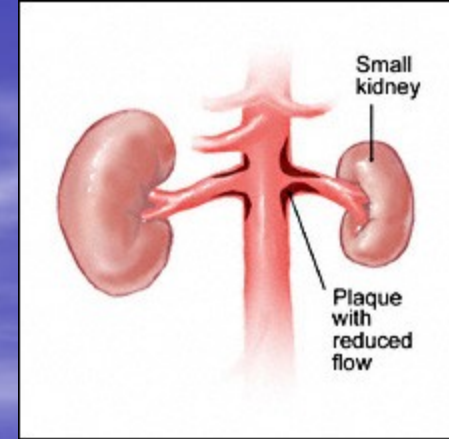
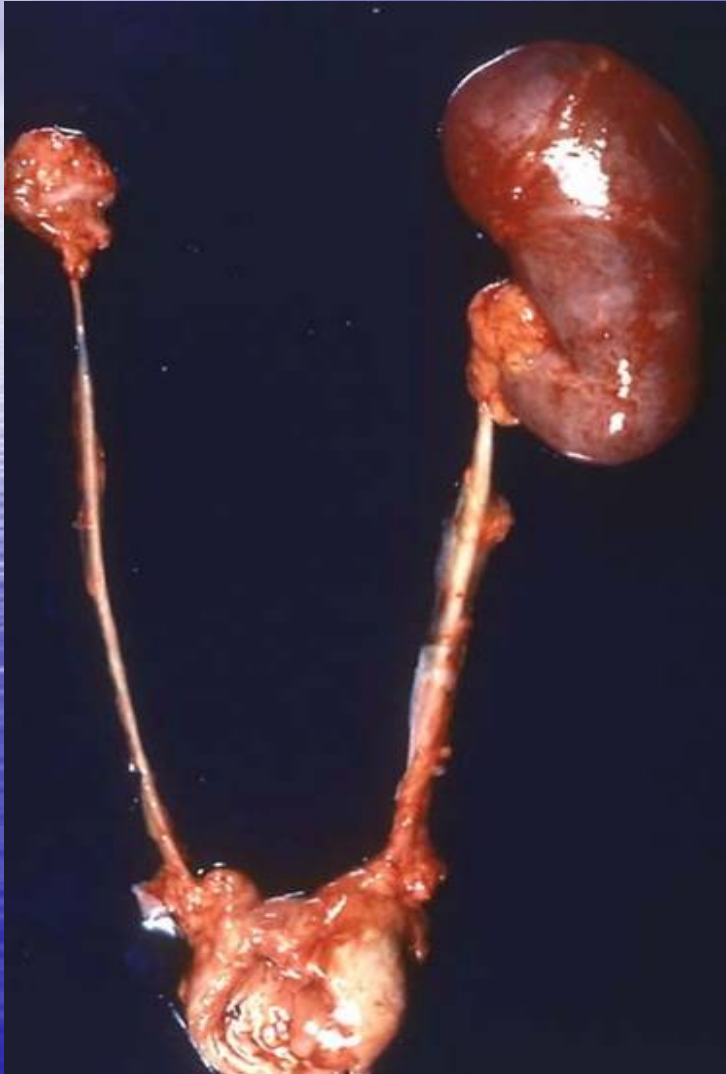
■ *PCD* - "infantil" form ■

- 1 : 40.000, probability of transmission to offspring - 25 % children of healthy parents „disease carriers“ ;
- + anomalies also in liver, spleen, lungs,
- etiol.: unclear - defect of ureter development (nephrons are not connected with collecting ducts)
- Klinic manifestation: bilat. enlarged kidney, hypertension, decreased glomerular filtration, renal failure. To a lesser extent of damage 50-80 % children can live about 15 years. Some children die shortly after birth by lung failure.
- Prenatal dg. in week 9 of i.u.dev. – FA, DNA markers.
- Th.: same as in *PCHLAD*

Polycystic kidney – cysts are not seen on the surface of kidney




Hypoplasia renis



- Insufficiently developed kidney – small amount of histologically normal and functional nephrons
- usually unilateral
- compensational hypertrophy of the other kidney

Wilms' tumor (nephroblastom)

- The most frequent type of tumors in children under 5 years, rare in adulthood
- 90% treatment success, also in case of greater distribution (metastasis)
- familial occurrence – tumor contains cells of mesonephros
- etiology:  - hereditary basis

Thesaurismosis („storage disease“)

A metabolic disorder in which a substance is stored in certain cells of some organs, usually in large amounts, due to defect production of enzymes splitting this substance. It causes functional failure of storing organs

Etiol.: *defected gen in auto- or heterosomes, usually recessive inheritance*

- **Anderson-Fabry disease** (storage of **cerebrosides** = *neutral sphingolipids*),
- **von Gierke disease** (storage of **glycogen**),
- **Gaucher disease** (storage of **glukocerebrosides**),
- **Fanconi sy.** (storage of **cystine**; **cystinóza, cystinurie**)
- **Primary hyperoxaluria** – cong. defect of glykooxalates production (storage of **oxalates**; **urolithiasis**).
- **Cong. defects of metabolism of purines** – familiar gouty juvenile nephropathy + arthritis already in the 2nd decade of life.

Anomalies of renal vascularization

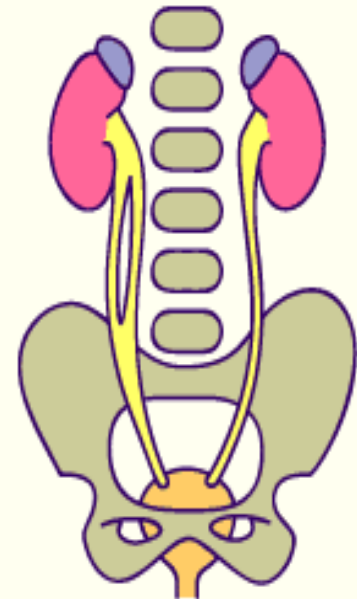
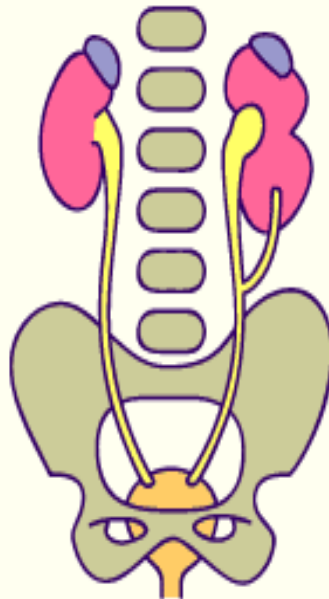
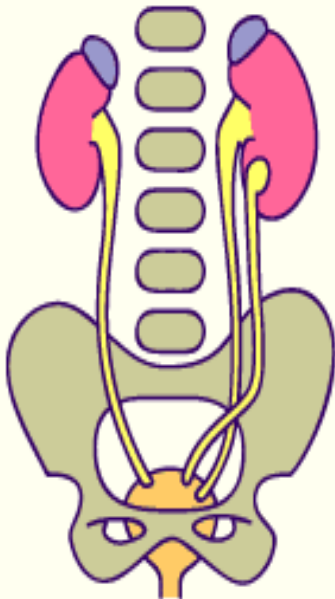
- Arise during ascensus renis – accessory arteries from a. iliaca and aorta (there are NOT collaterals between arteries! – obstruction causes infarction of renal parenchyma)
- supernumerary veins (with collaterals)
- accessory arteries – 25 %, veins - 12,5 %

Renal renculi



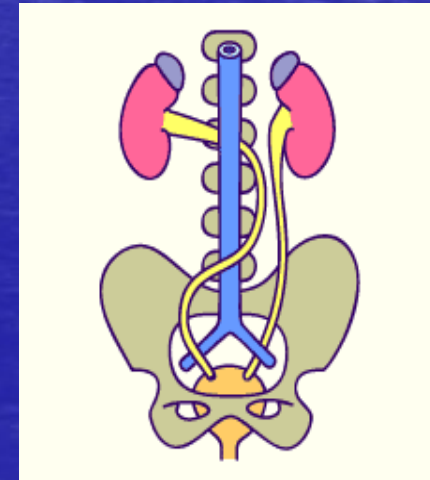
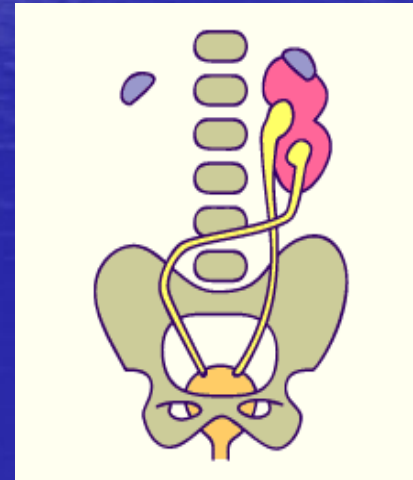
2. CM of pelvis and ureter

- **Ureter duplex, ureter fissus (+ pelvis duplex, ren duplex)**
- unilat. or bilat., partial or completel
- etiology: branching or accesory ureteric bud, splitting



Ectopic defects of ureter

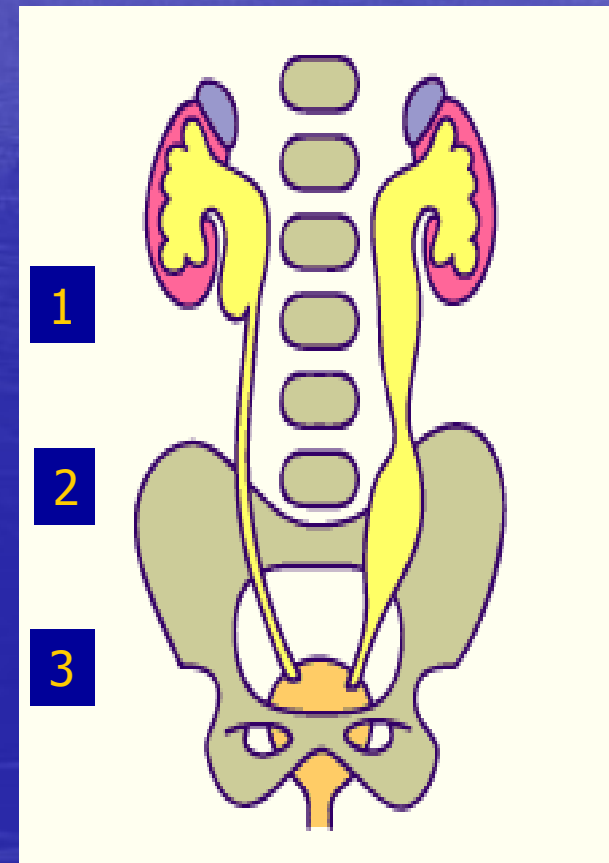
- ectopia of orificium ureteris
–ureter opens into urethra,
uterus or vagina (*rarely
into ductus deferens*)
- cross ectopia of ureter,
„retrocaval” ureter –



Congenital stenosis, obstructions, atresis

Physiol. ureter narrowings:

1. pelvi-uretral junction,
2. crossing with vasa iliaca,
3. pars intramuralis – ureter-vesical junction.




3. CM of urinary bladder

- **Extrophia** 1 : 40.000
(2-3 ♂ : 1♀)
- Ventral abdominal wall and ventral wall of urinary bladder are not formed; urinary bladder is opened and inner surface of its dorsal wall is visible (+ epispadia and cleft of symphysis (diastasis))

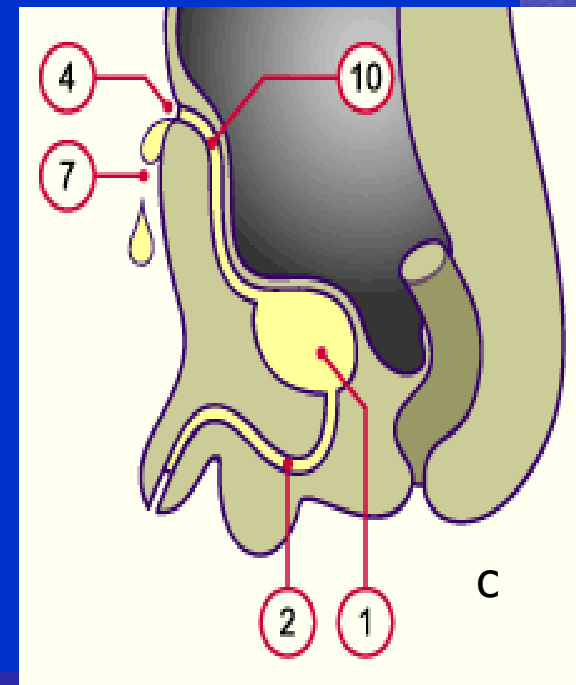
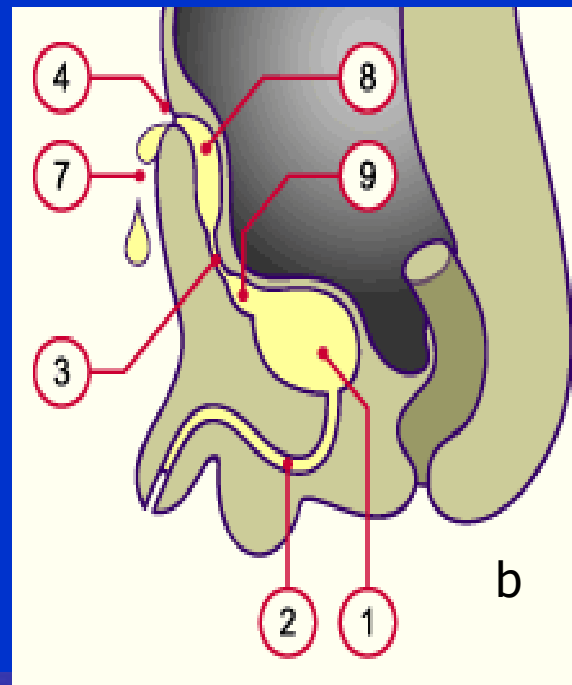
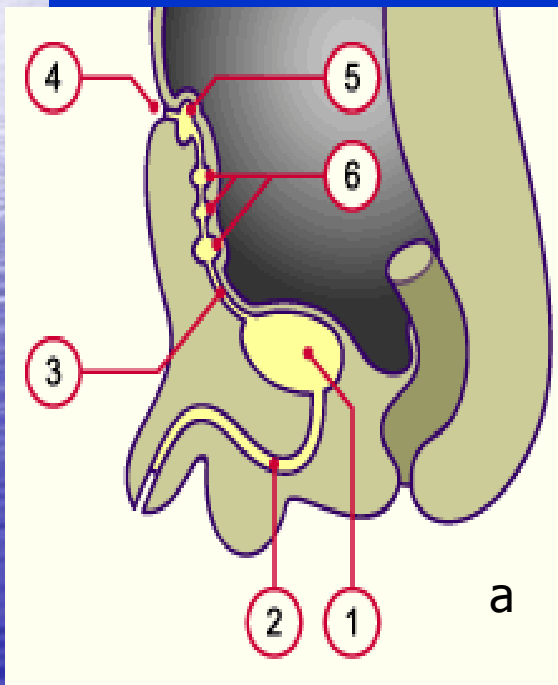


Extrophia

- etiol.:  defect of mesenchyme migration between ectoderm of abdominal wall and cloaca in week 4
- Reconstruction of the wall (24 - 48 h after birth), epispadia (about 2nd year).

defect obliteration of allantois

- urachal cysts and fistulae (a)
- Urachal sinus (b)
- urachus persistens (c)



4. CM or urethra



- Clefts of urethra:

Hypospadia

insufficient fusion of plicae genitales

Epispadia

see extrophia

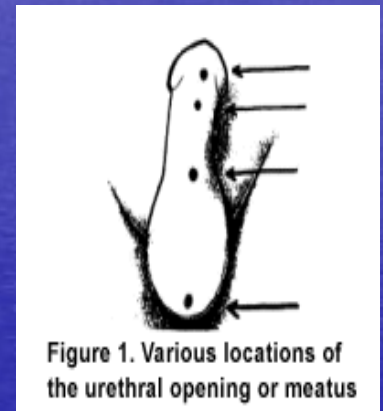


Figure 1. Various locations of the urethral opening or meatus



Sources of pictures:

- <http://www.embryology.ch/genericpages/moduleorganoen.html>
- embryology.med.unsw.edu.au/.../BGDlabXYXX_5.htm
- www.embryology.ch/.../genitinterne06.html
- www.emedicine.com/ped/topic704.htm
- embryology.med.unsw.edu.au/Defect/page4.htm
- www.childrenskidneydisease.org/Stories.asp

Vývoj zevního genitálu a uzavření uretry ♂

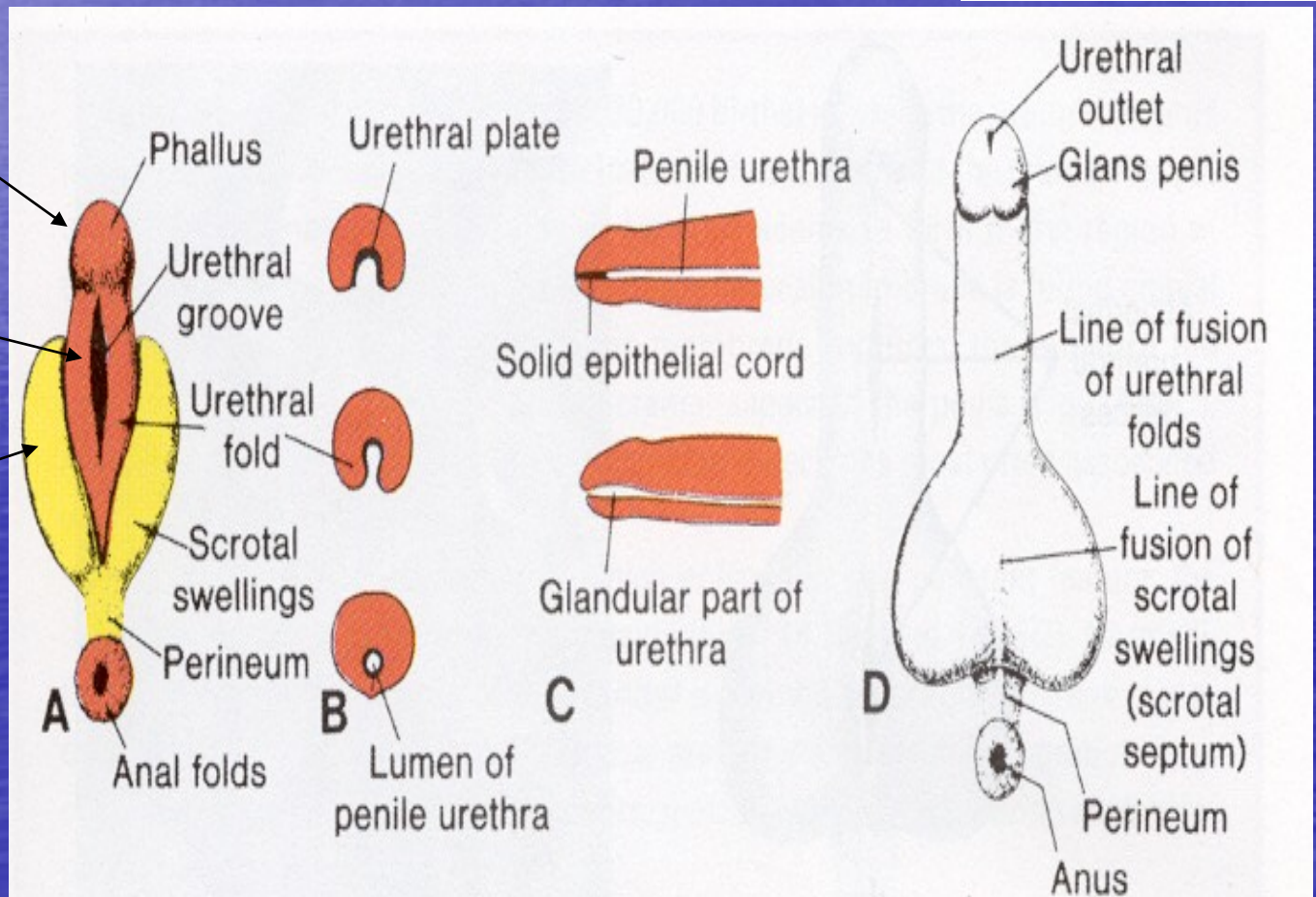


Zdroje pro vývoj
zevního genitálu:

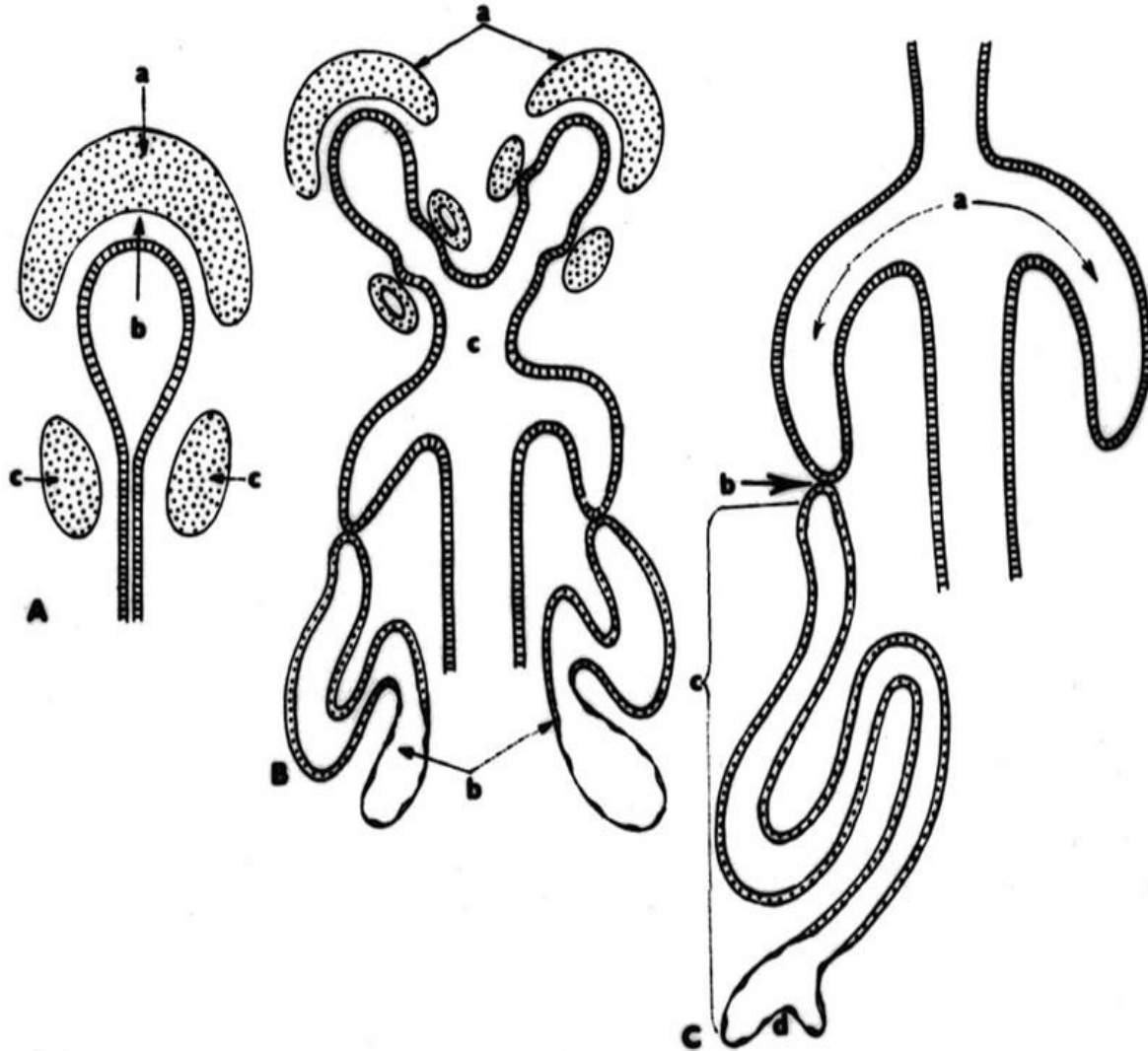
Tuberculum genitale

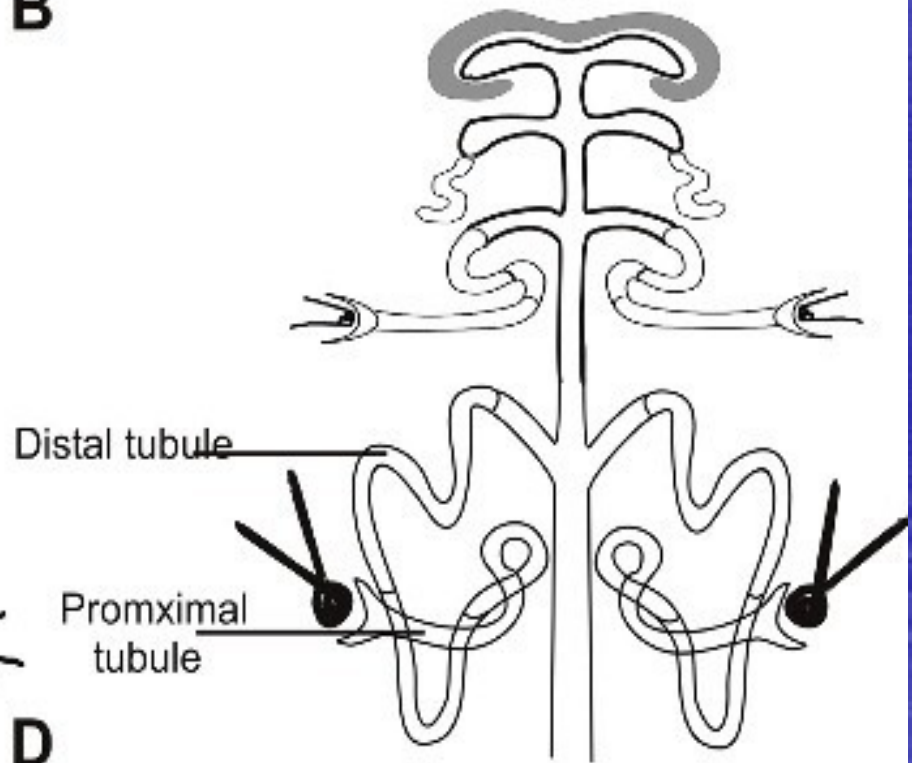
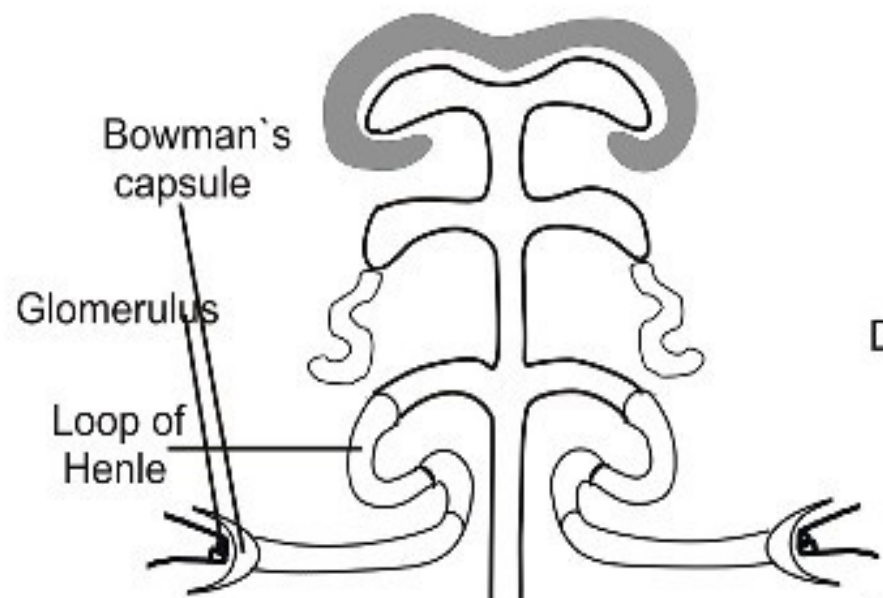
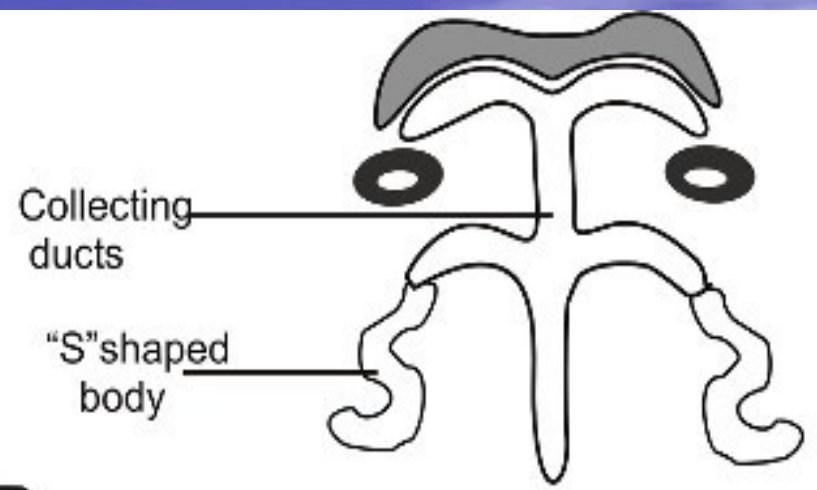
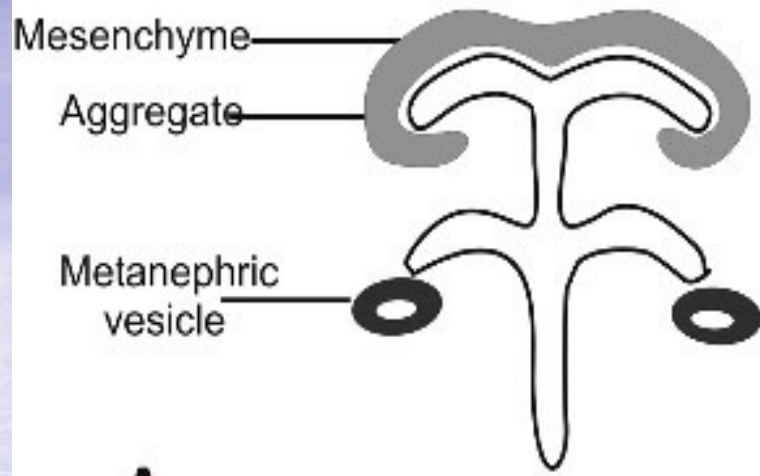
Plicae genitales
(urethrales)

Tori genitales
(plicae labioscrotales)



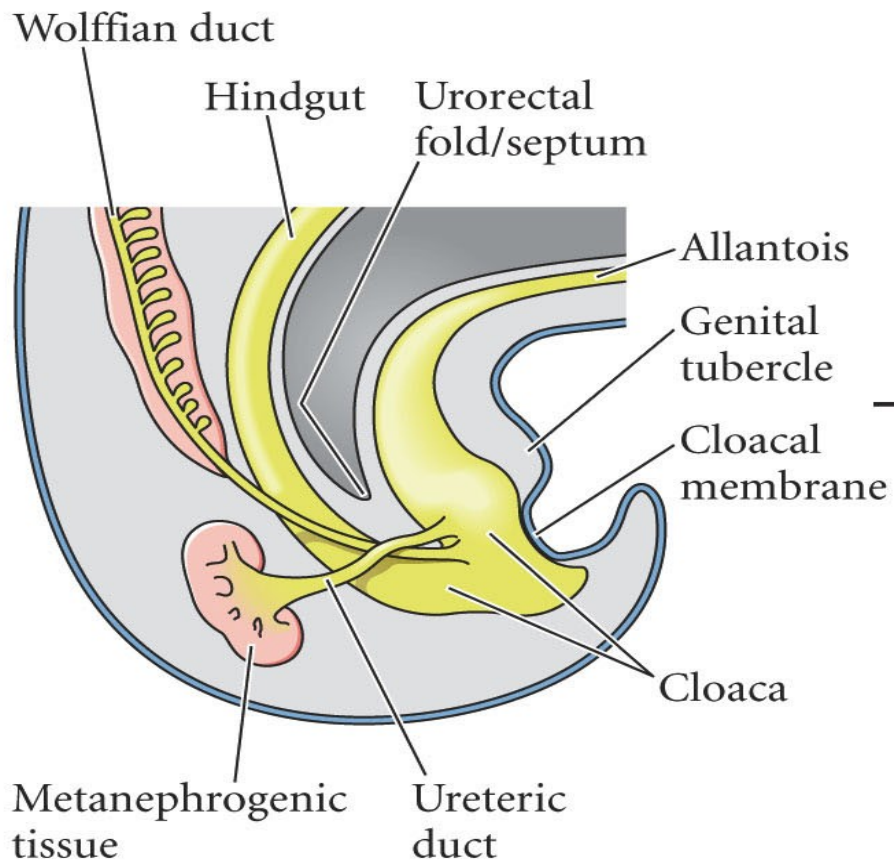
Vývoj nefronu



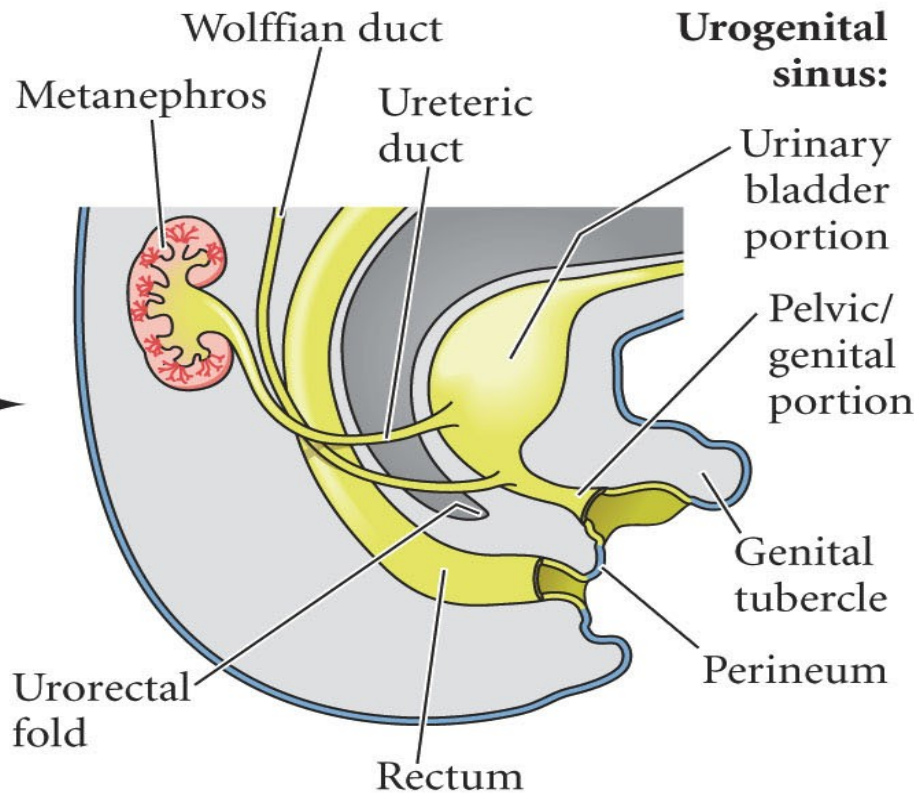


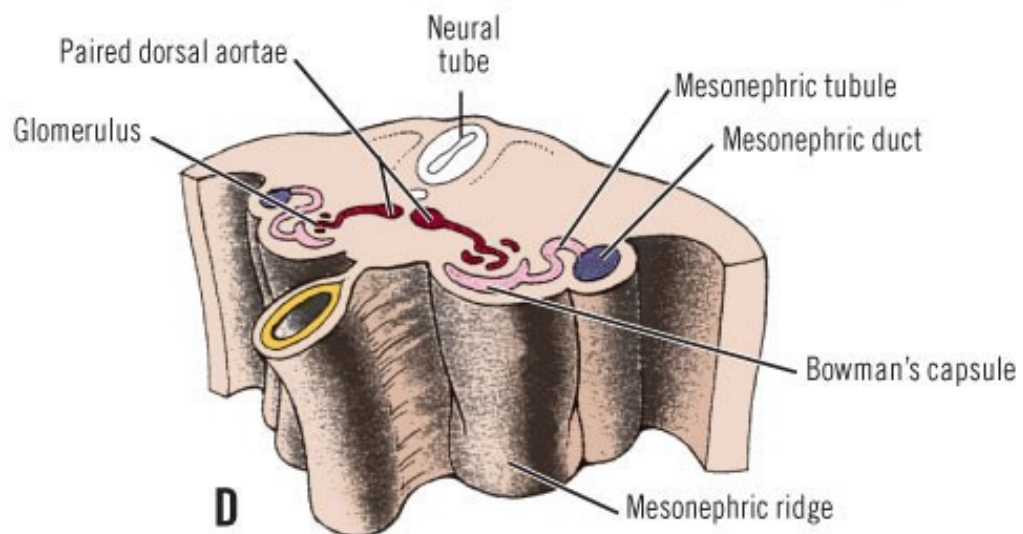
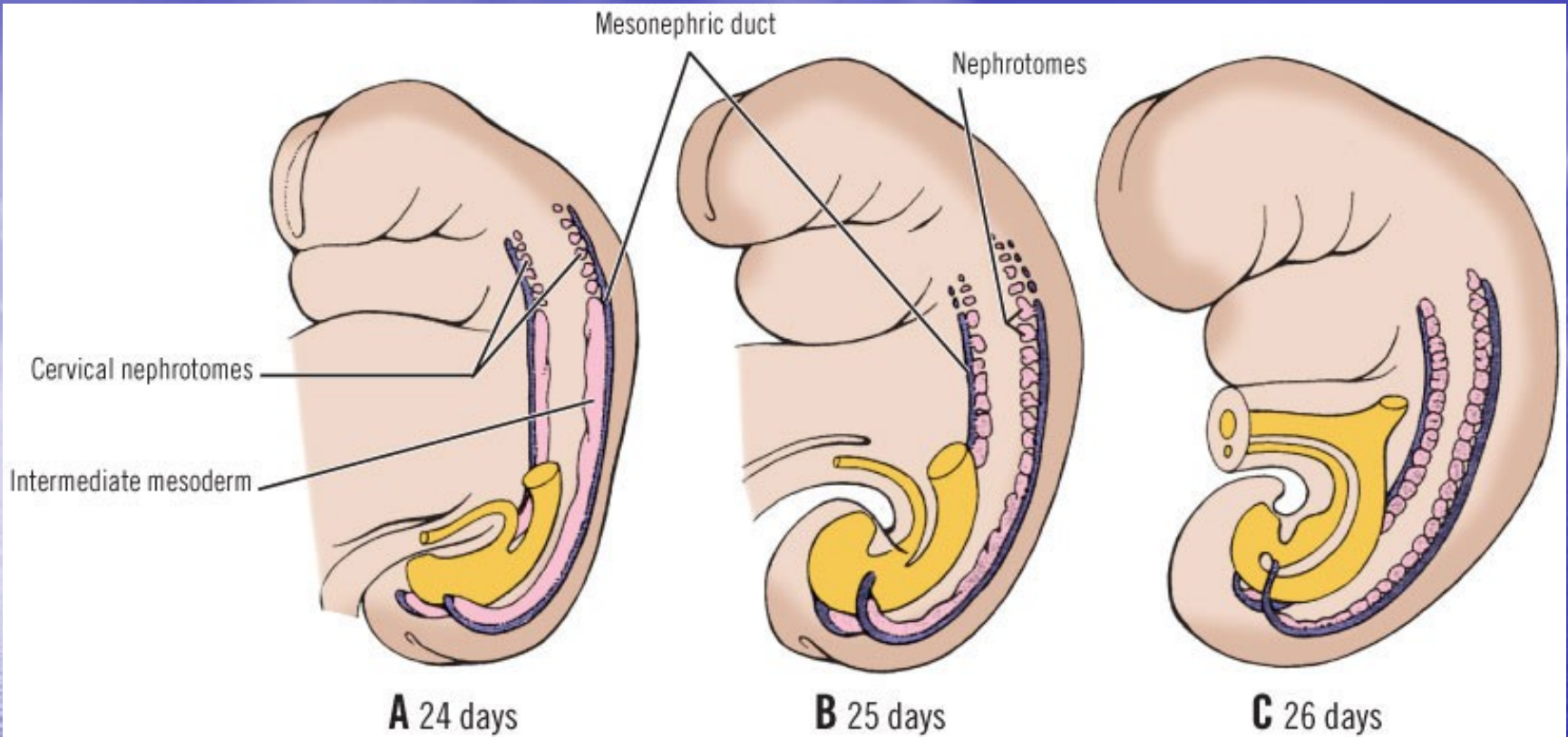
Močový měchýř, ureter

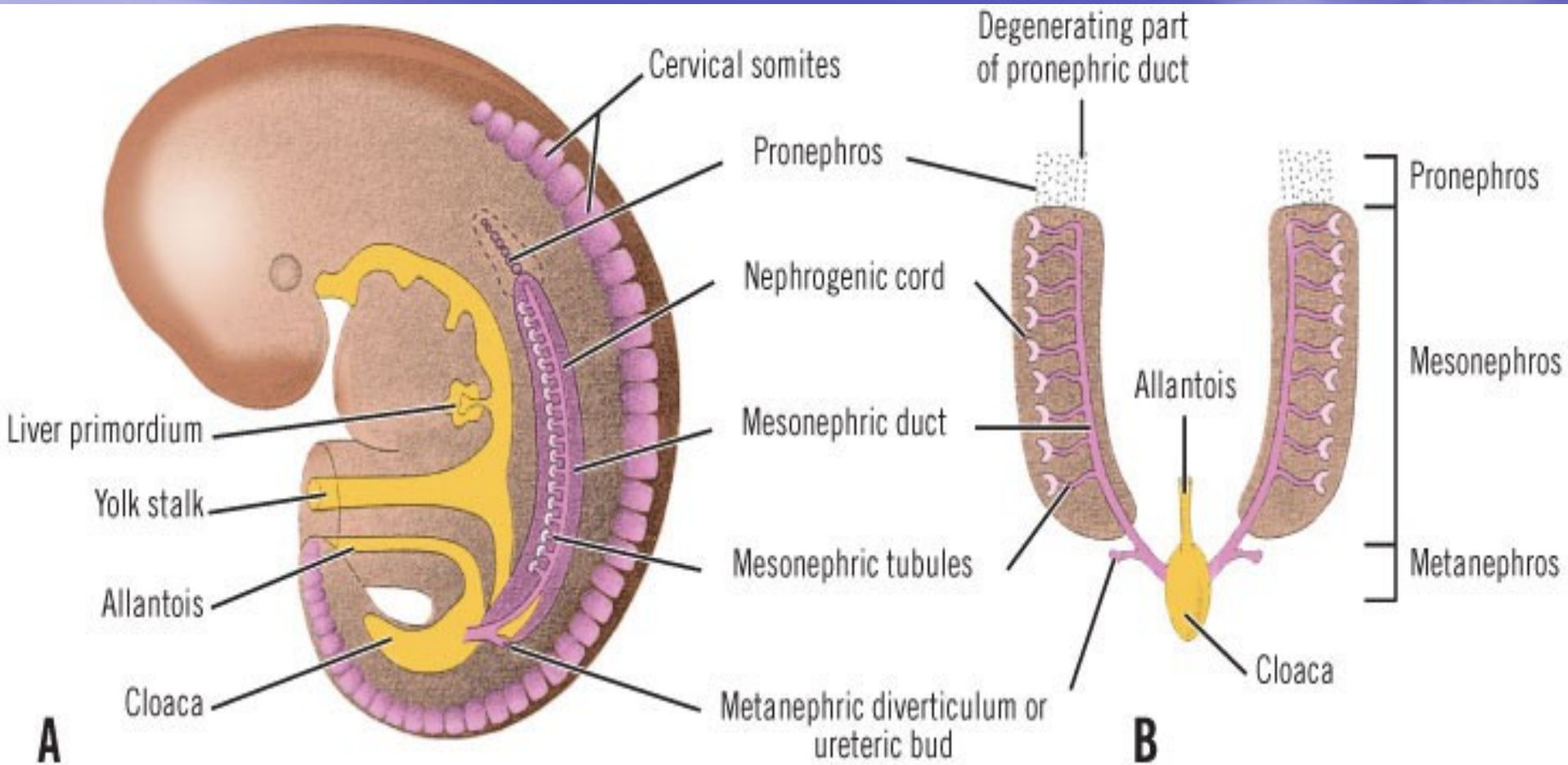
(A)

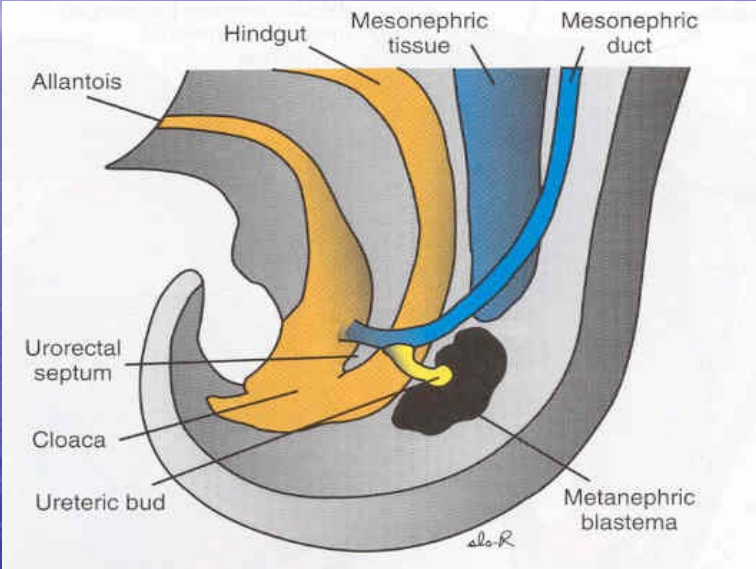


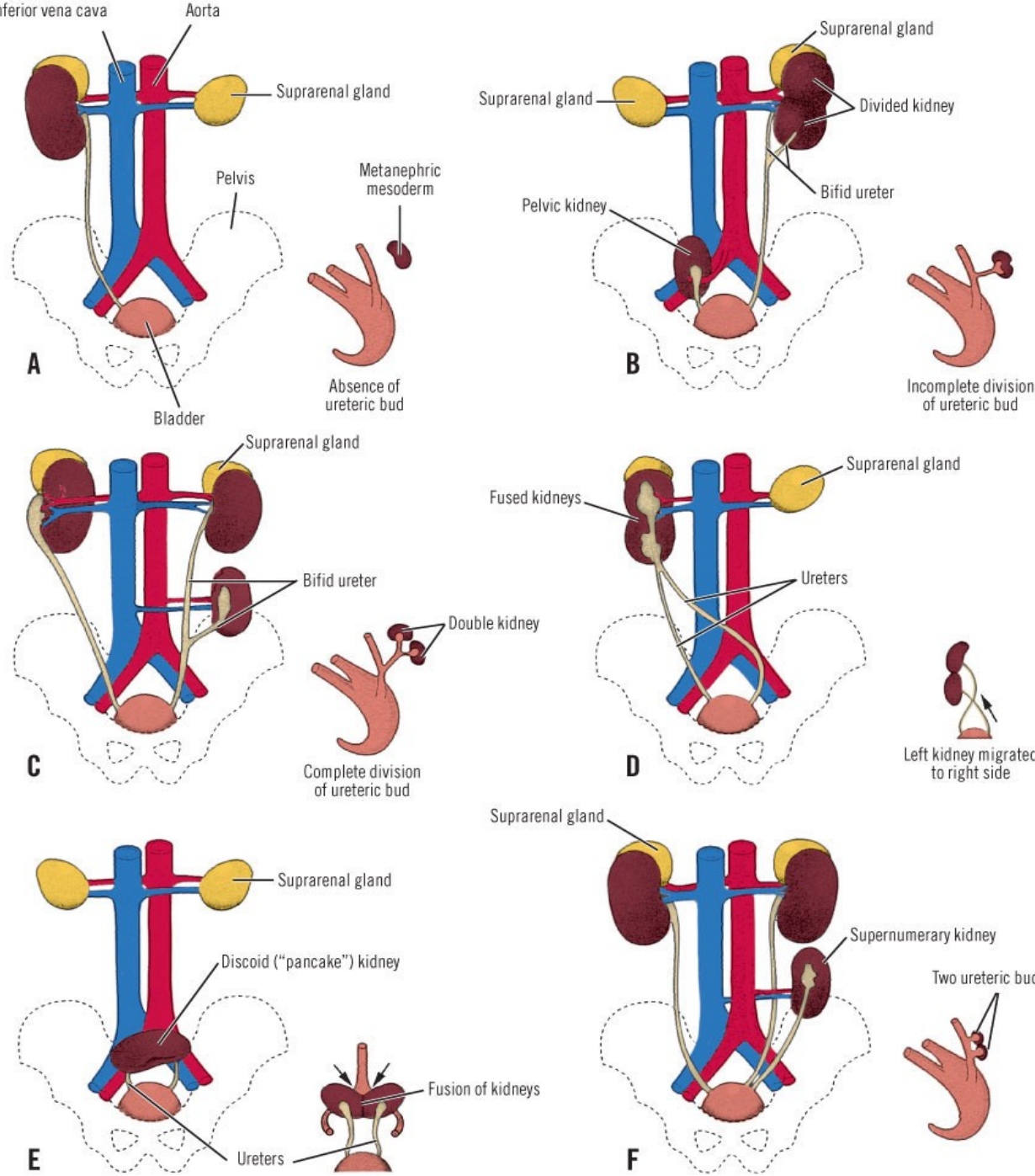
(B)












- A – unilateral renal agenesis
- B – pelvic ren + ureter bifidus
- C – kidney malrotation + ren duplex
- D – crossed ectopia
- E – „pancake“ kidney
- F – supernumerary kidney

Sy. prune – belly



- 1 : 40 000
- Trias of syndromes:
 1. agenesia of muscles in abdominal wall
 2. obstruction of urinary bladder in region of internal urethral ostium (dilatation of bladder, megaureters and hydronephrosis – above obstruction)
 3. bilateral **kryptorchism**
- Etiol.:  unclear
hormonal treatment (estrogens) of mother during the 1st trimester was frequently found in anamnesis
- Prognosis: viability depends on number of functional nephrons at birth.