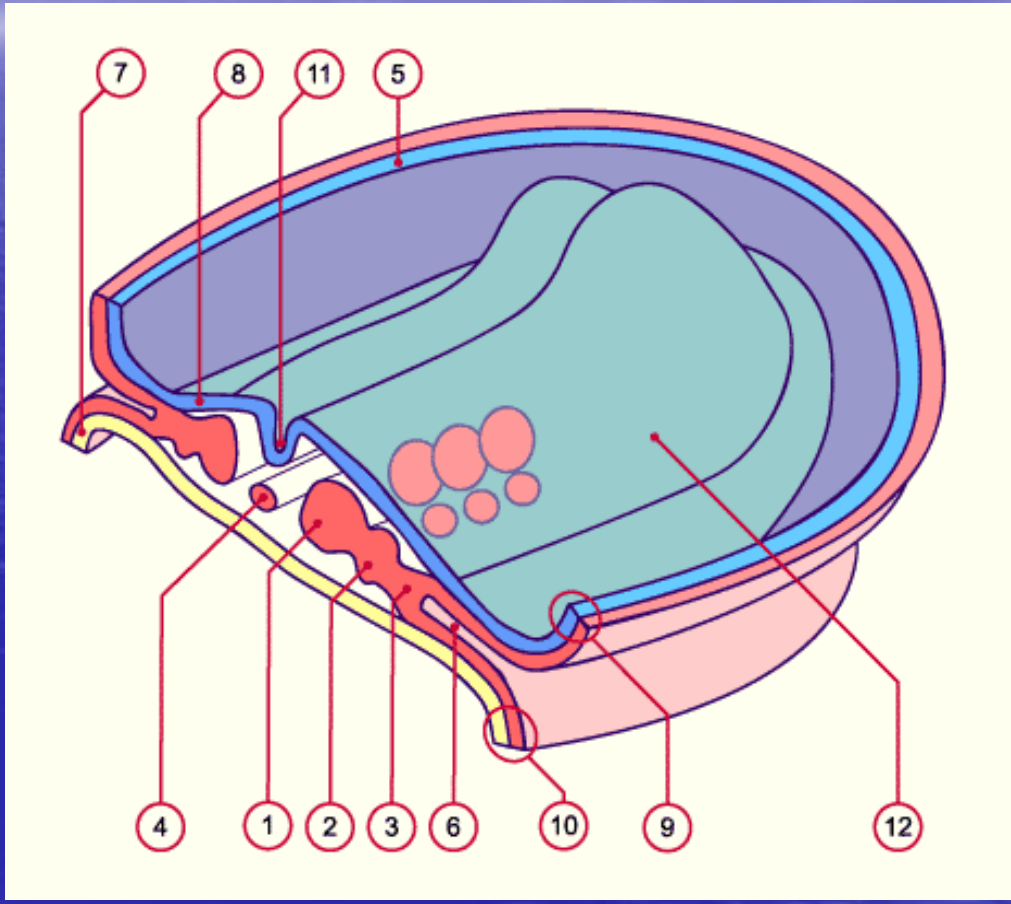
Urinary system

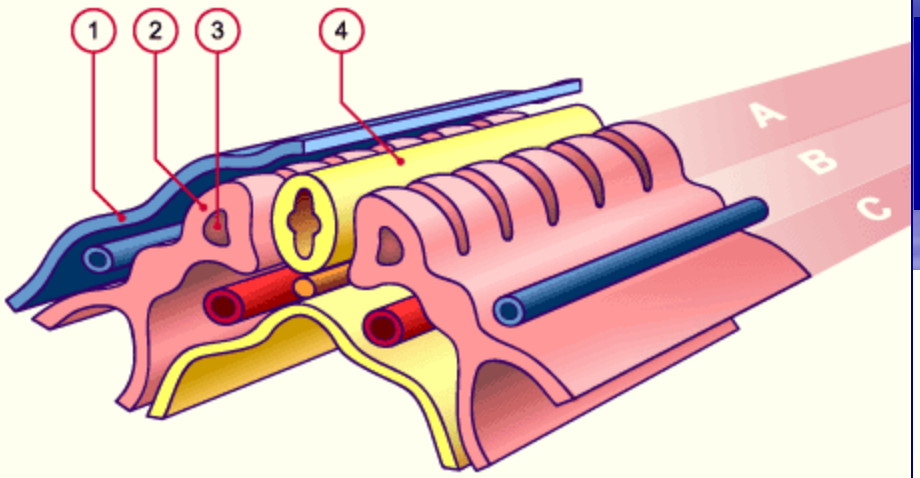
Development

Teratology

Intermediary mesoderm:

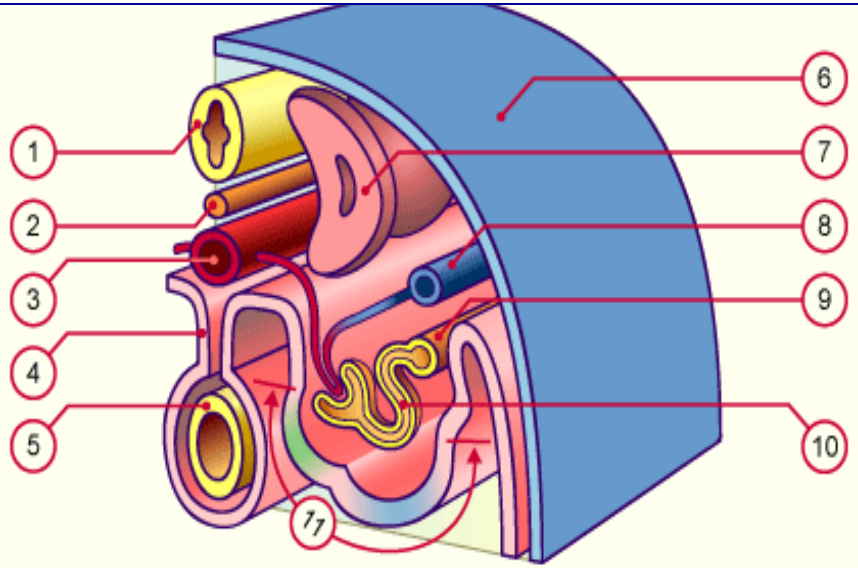
- **Pronephros**
  3rd week, C
  Ductus mesonephricus (Wolffi)
- **Mesonephros**
  4th week, C6-L3
- **Metanephros**
  5th week, L4-S

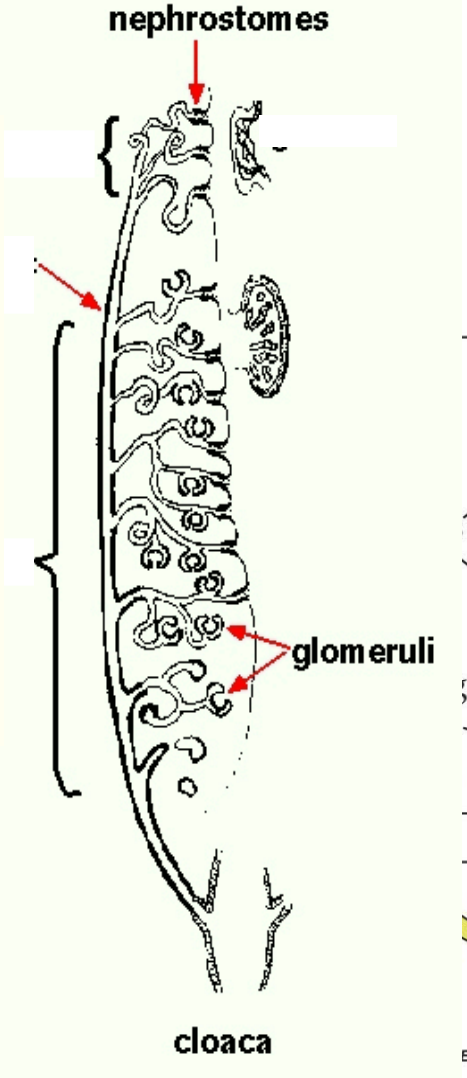


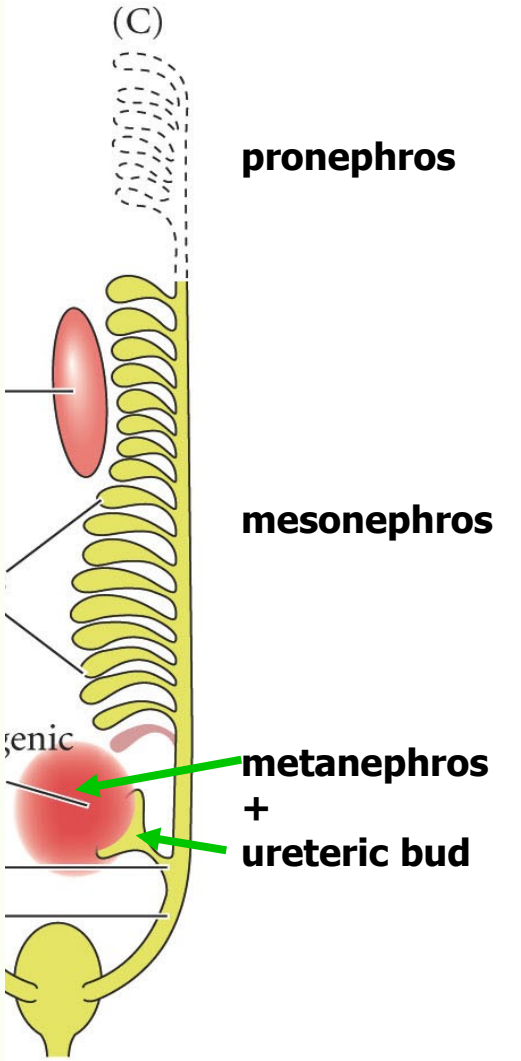


- paraaxial
- intermediary
- lateral

mesoderm







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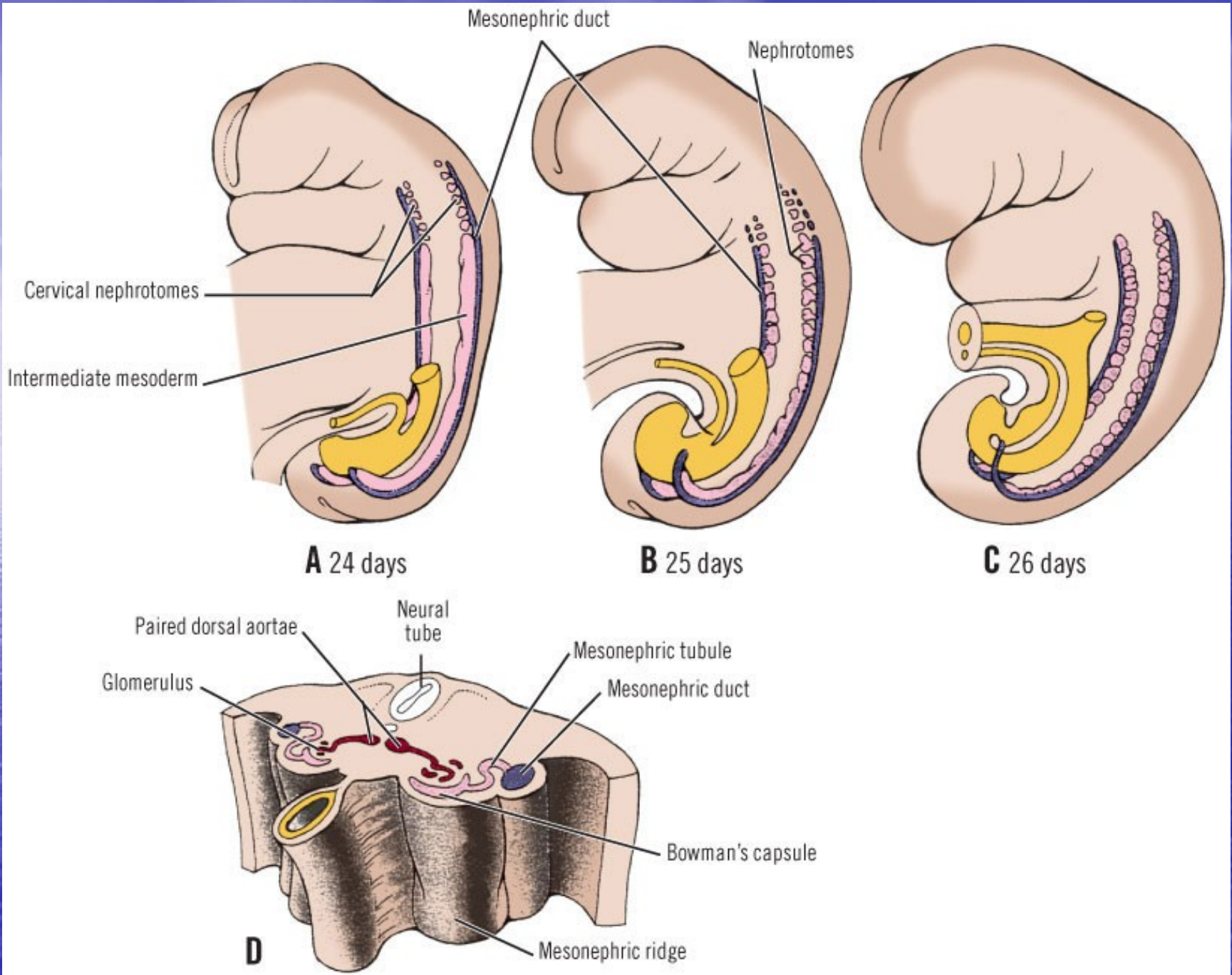
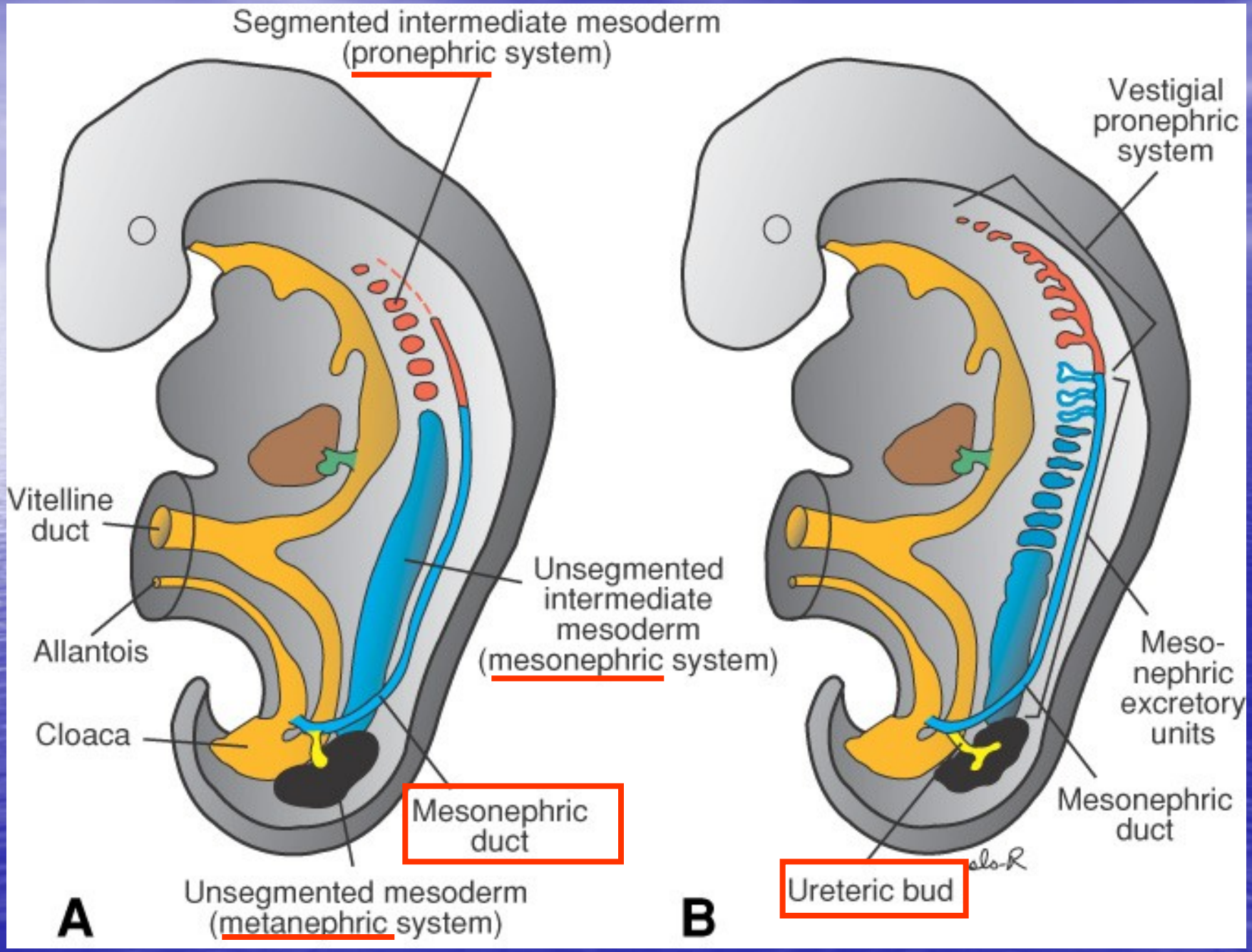
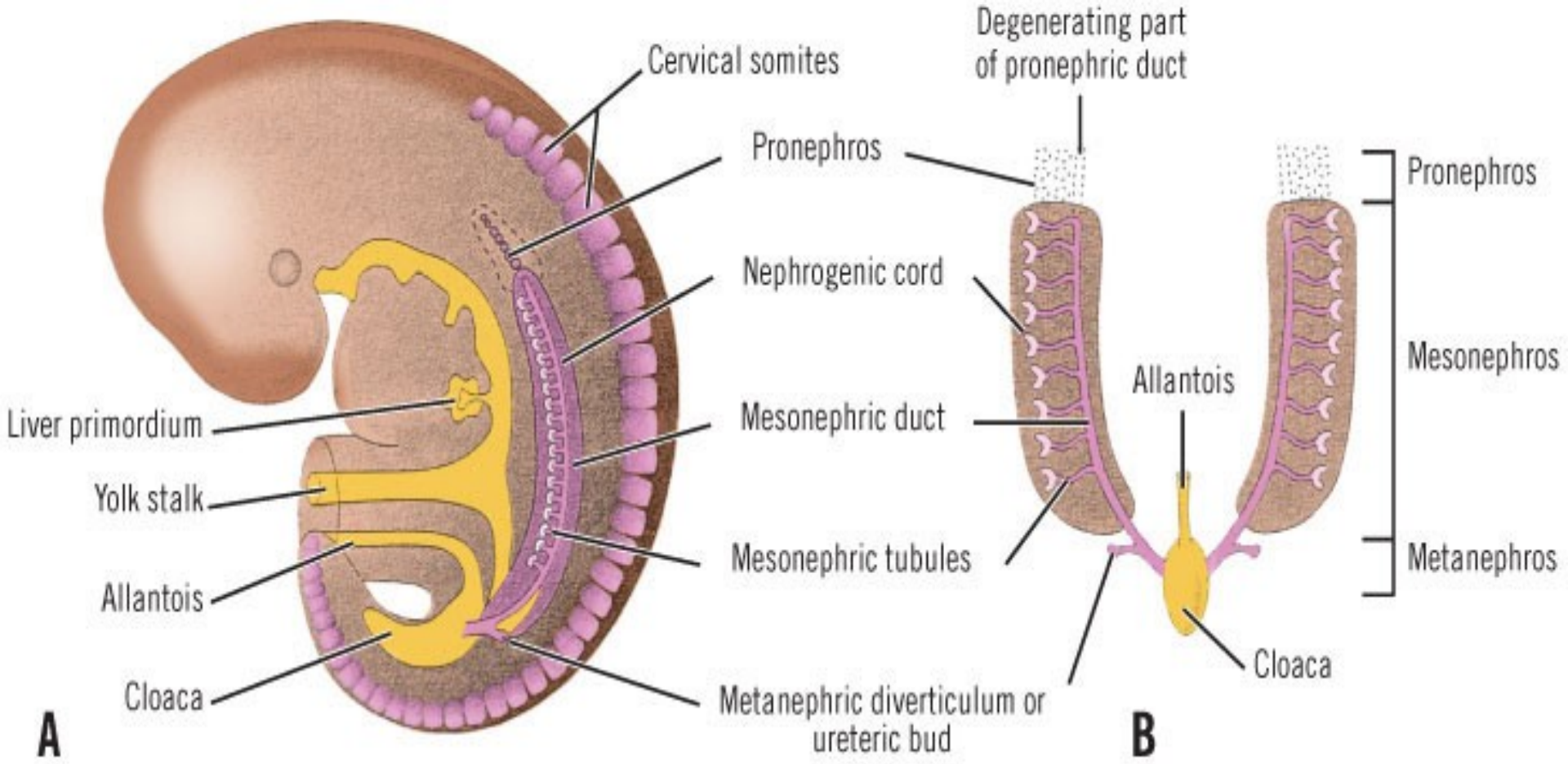
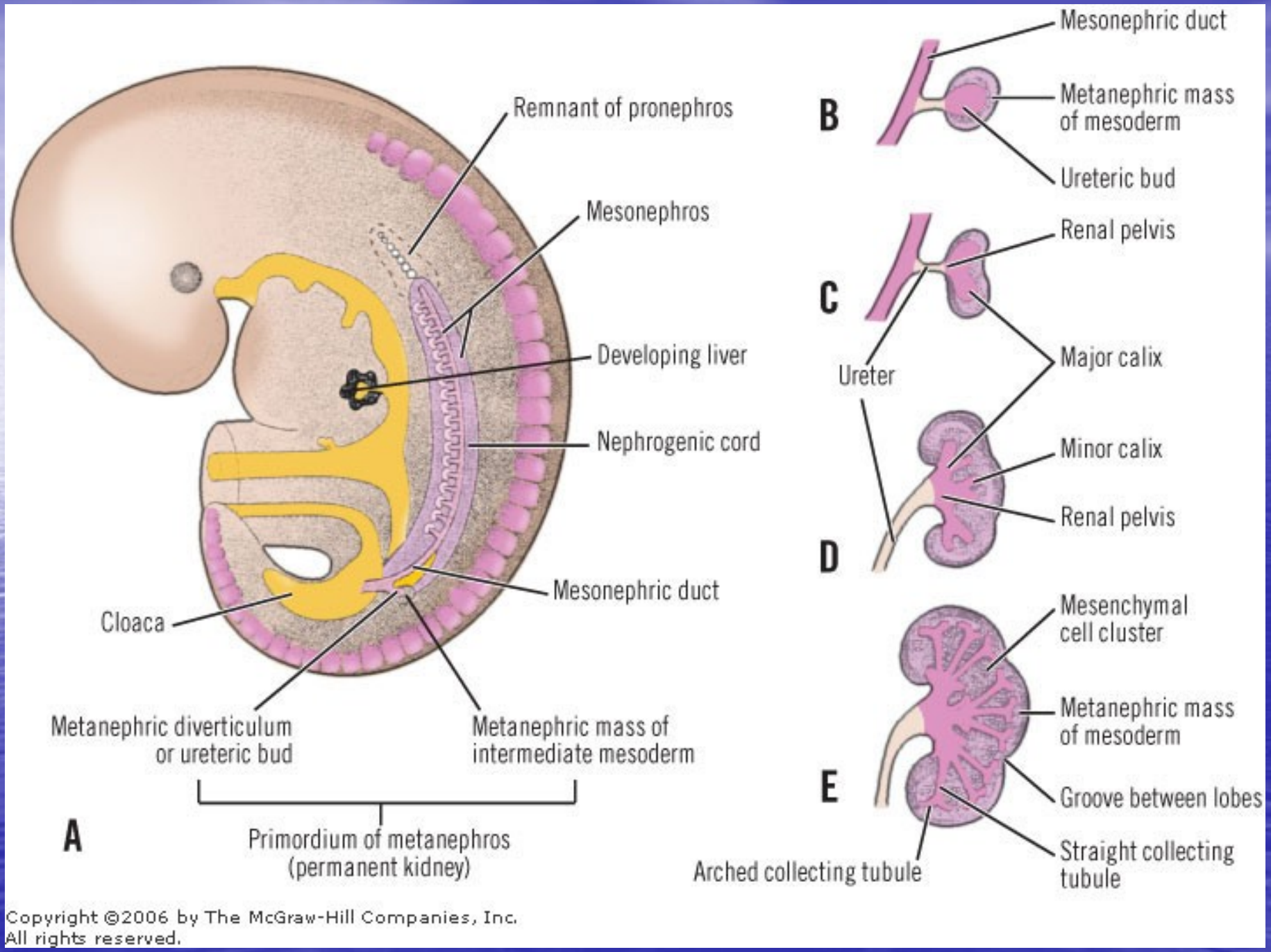
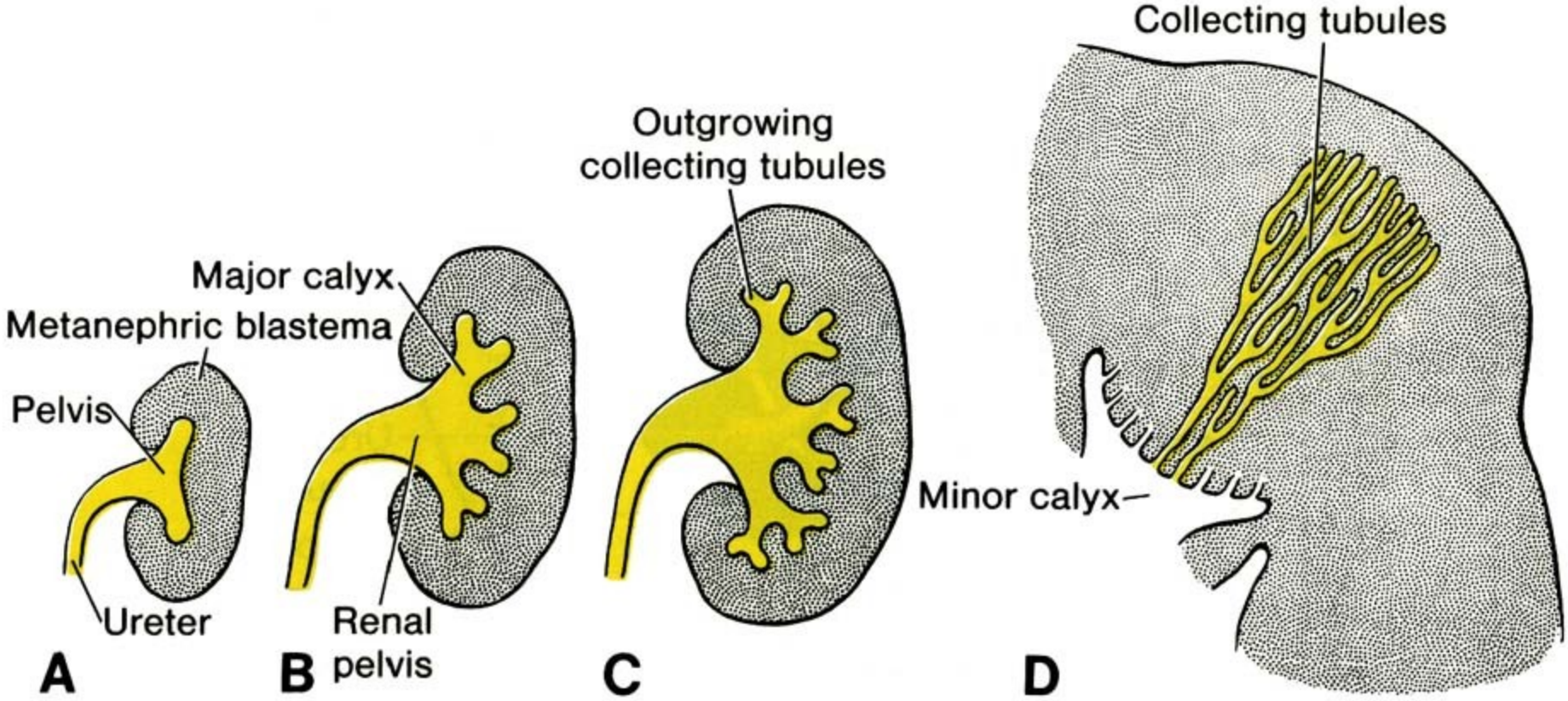



Segmented intermediate mesoderm
(pronephric system)
Vestigial
pronephric
system
Vitelline
duct
Allantois
Cloaca
Unsegmented
intermediate
mesoderm
(mesonephric system)
Meso-
nephric
excretory
units
Mesonephric
duct
Mesonephric
duct
Unsegmented mesoderm
(metanephric system)
Ureteric bud
A
B

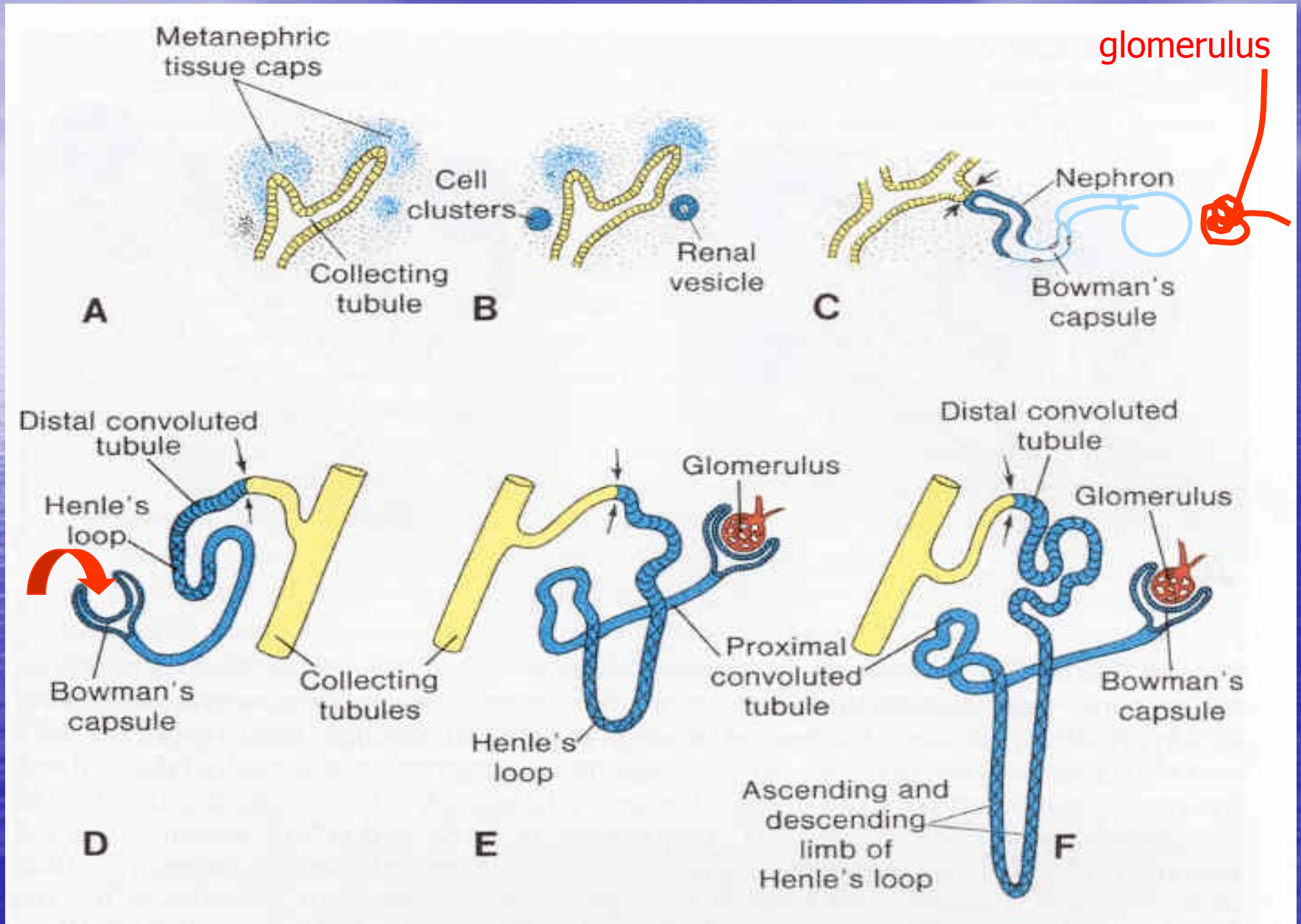




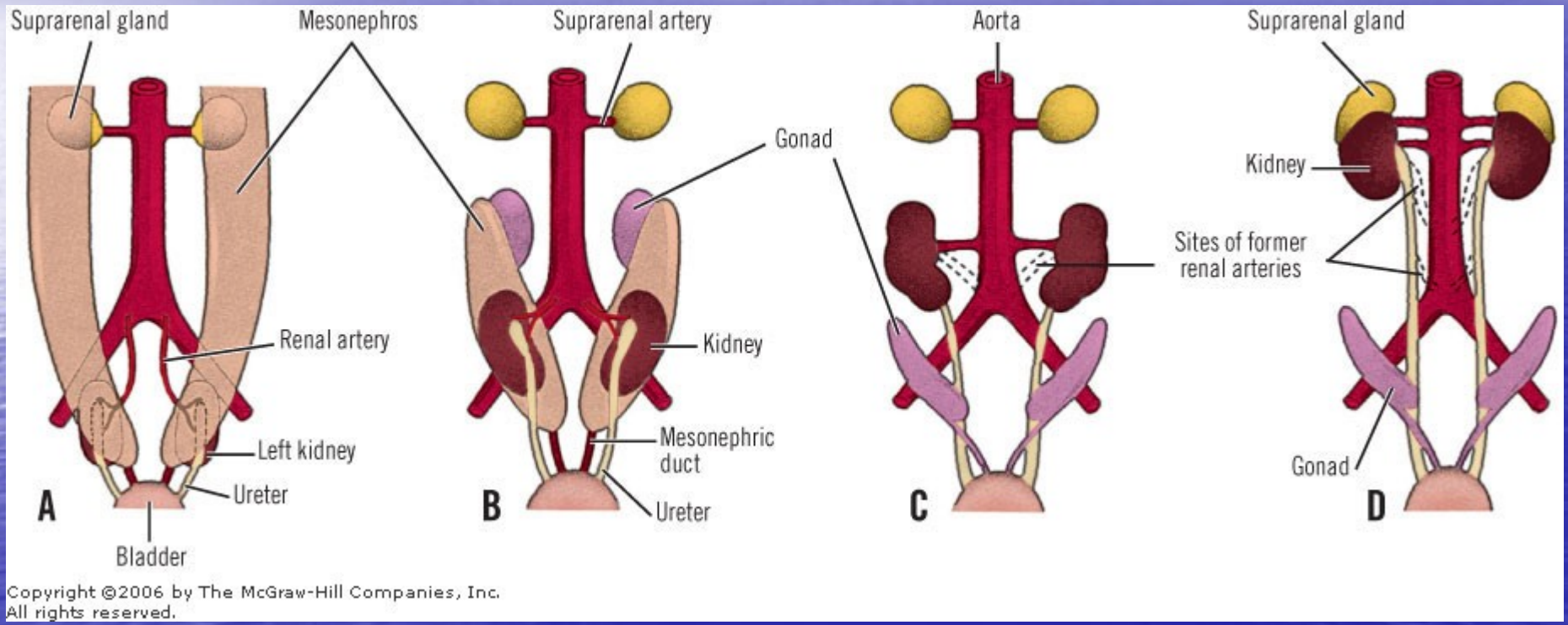
Kidney development



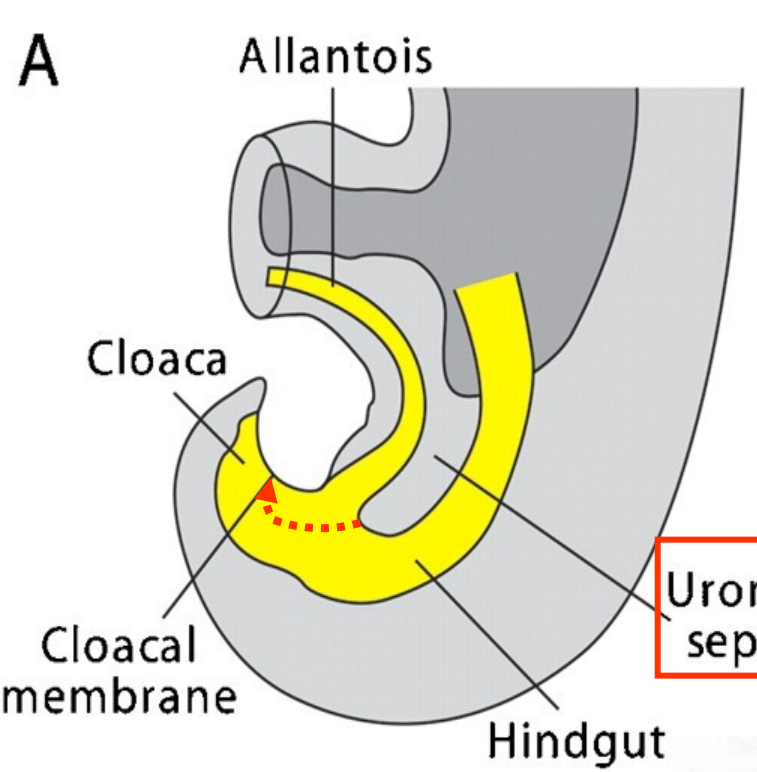
Nephron development



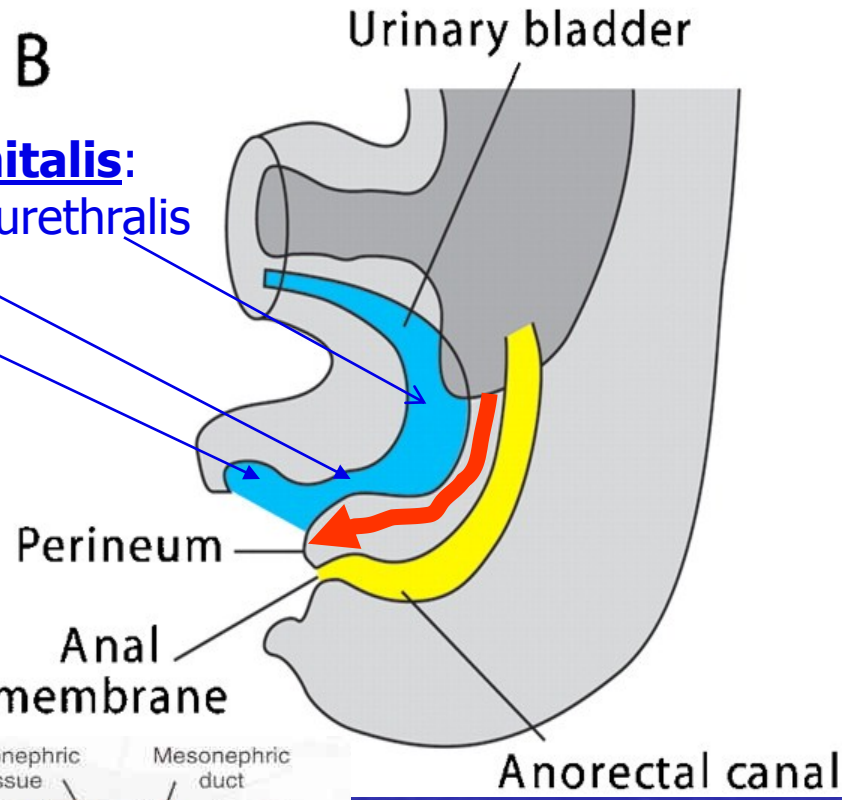
Ascensus renis

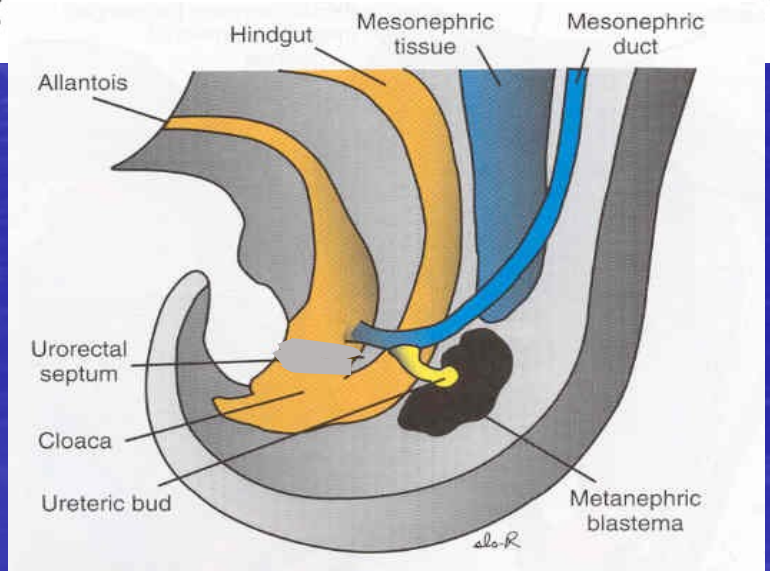


Cloaca development



Sinus urogenitalis:
-canalis vesicourethralis
-pars pelvina
-pars phalica

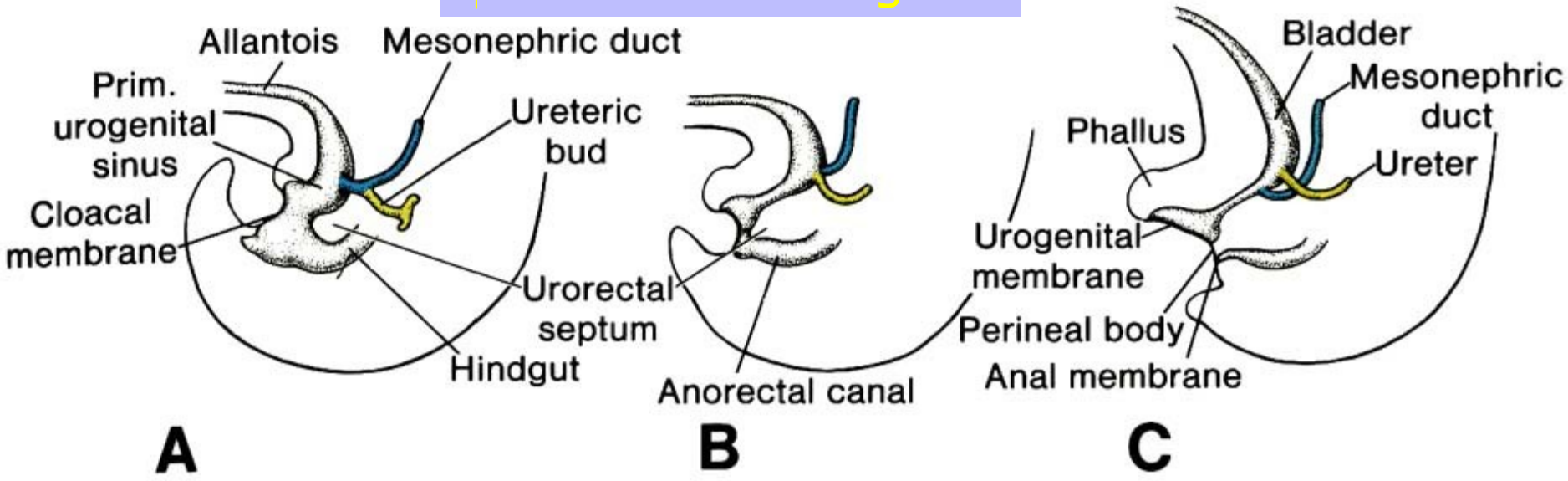




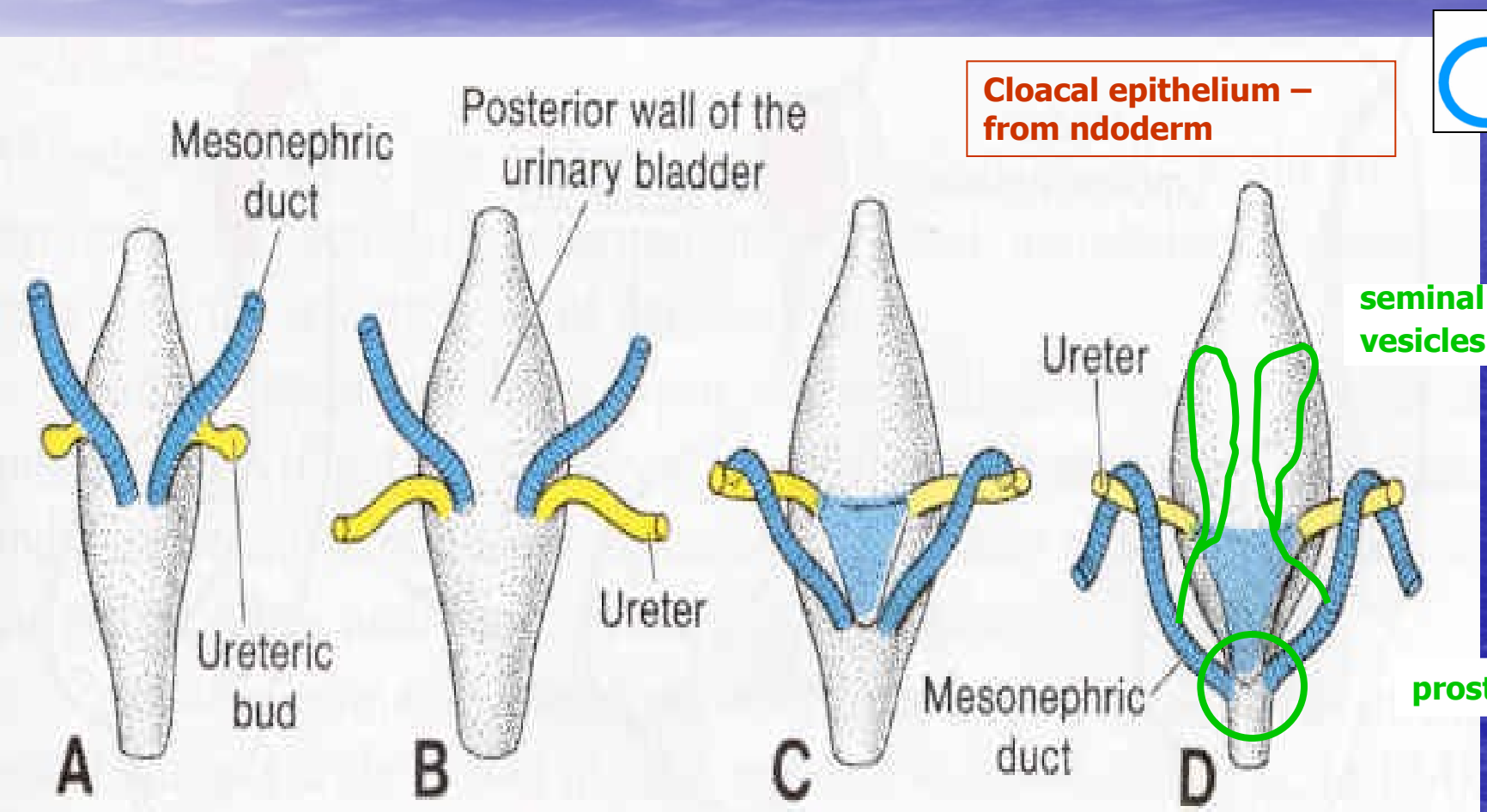
Cloaca

- Septum urorectale – sinus urogenitalis + rectum
  (cloacal membrane – urogenital m. + anal m.)
- Sinus urogenitalis
  - canalis vesicourethralis ⇨ urinary bladder, ♀ uretra
  - pars pelvina ⇨ ♂ pars prostatica et diaphragmatica uretrhrae
  - pars phalica ⇨ ♂ pars phalica urethrae

♀ - vestibulum vaginae



Wolffian duct (ductus mesonephricus) and ureteric bud



Cloacal epithelium – from ndoderm

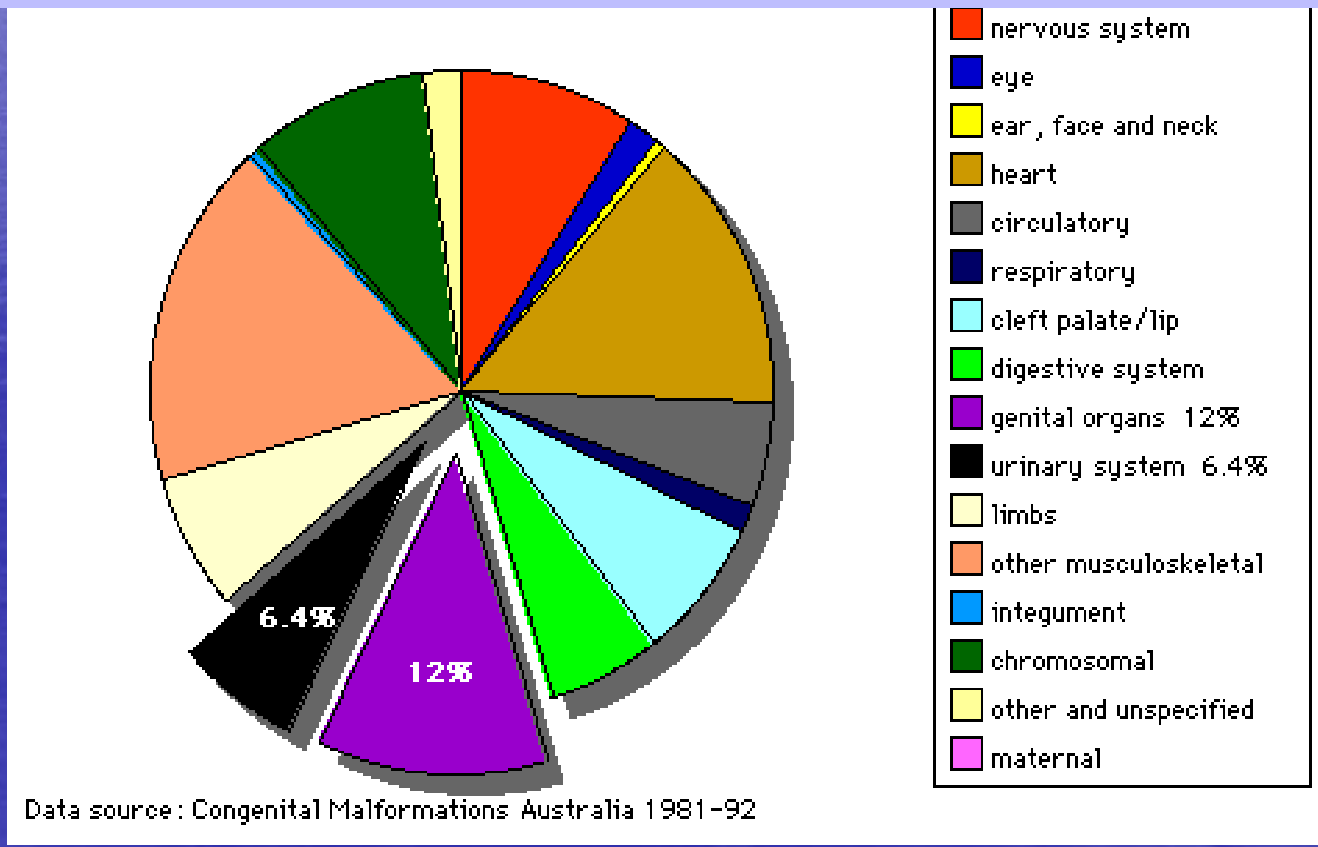
seminal vesicles

prostate

Epithelium of trigonum vesicae – from mesoderm

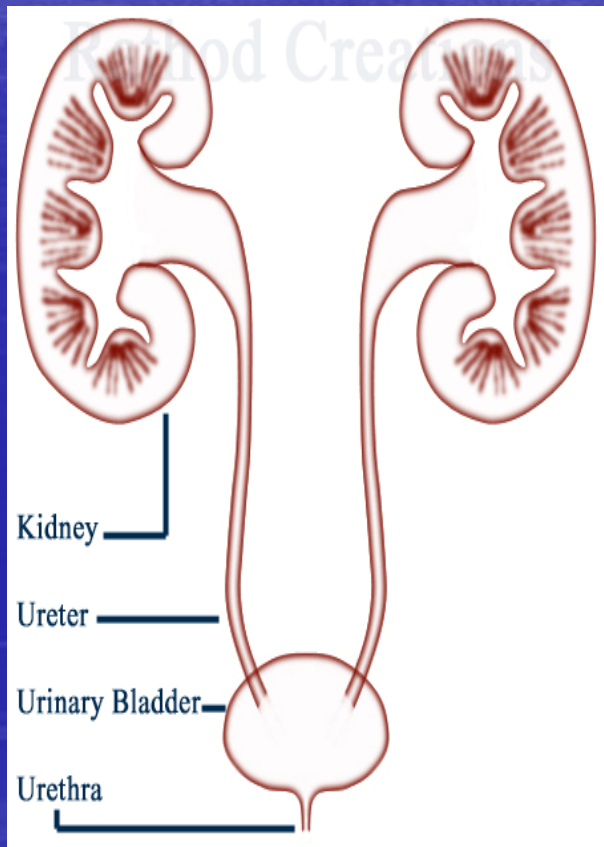
Congenital malformations of urinary system

Hypoplasia – small kidney
Reflux –
Hydronephrosis – urine stasis



Congenital malfromations (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 postion (ectopia)
- anomalies of parenchyma (nephrodysplasia)
- anomalies of vessels

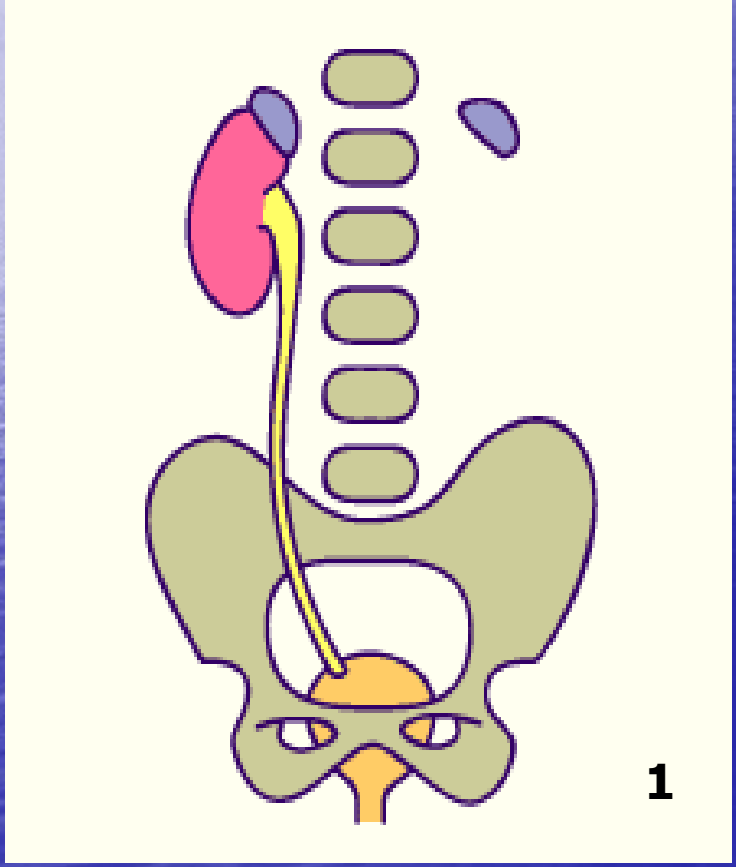
Kidney malformations arrise at the begining of development (*development of metanefros isn't induced by ureteric bud or both kidney are closely together – before week 6*) or later (*during incomplete ascensus renis – after week 8*).

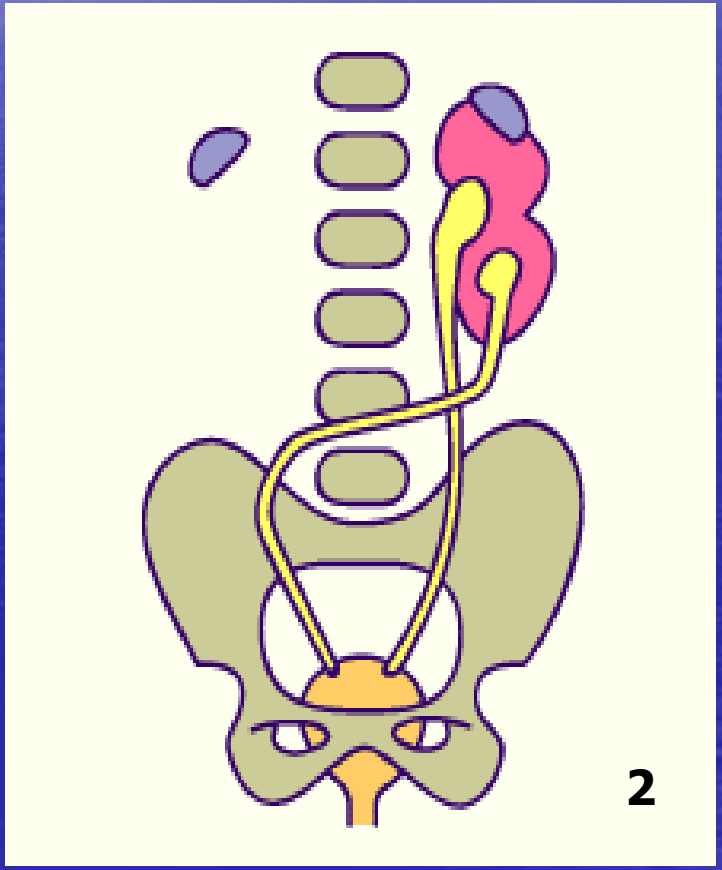
Agenesis renis

- bilateral (1 : 3000; prenatal dg. – oligohydramnion, hypotrophic fetus,
  skeleton deformities and lung hypoplasia due to fetus oppresion) - *(death by uremia and drespiratory distress)*
- unilateral (1 : 1500) + agenesis 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 – kidnye agenesis

2 – kidnye agenesis + cross ectopia of ureter

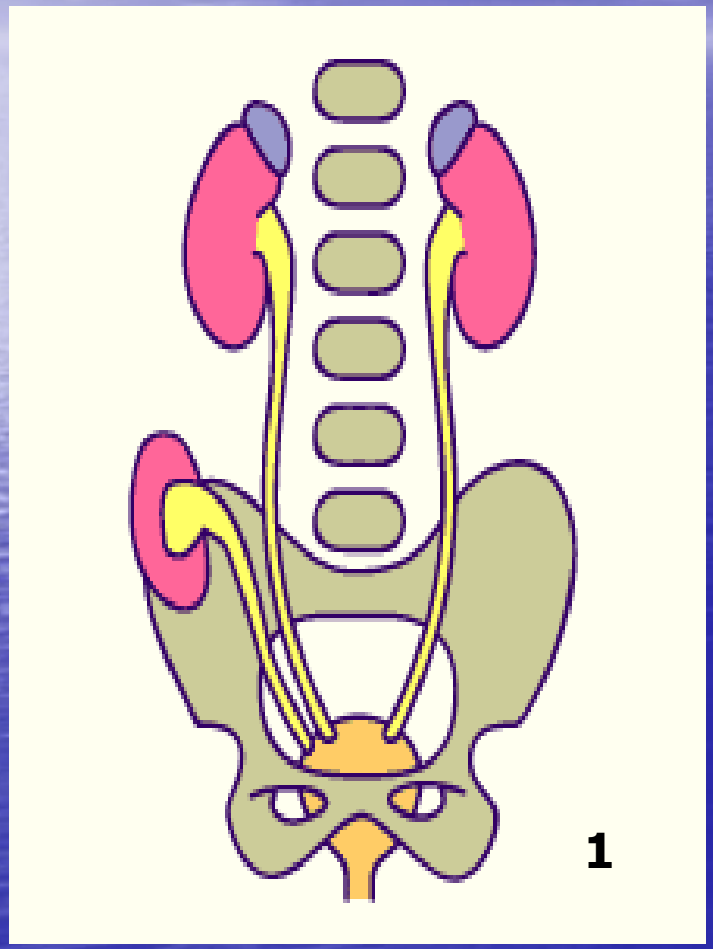


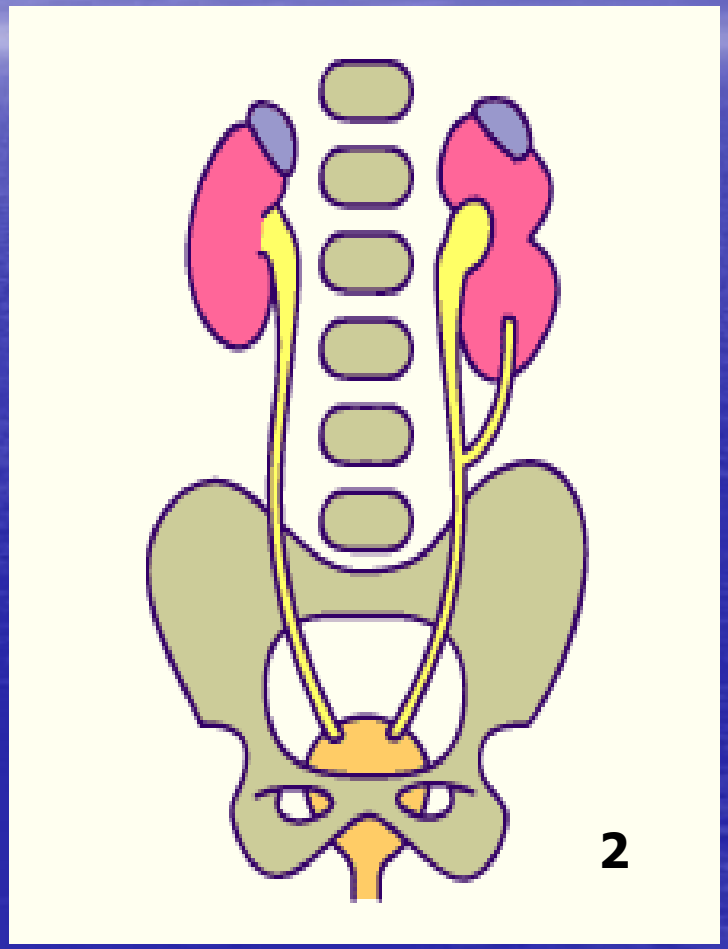


Supernumerary kidney (2-3 % newborns)
Ren duplex

- unilateral or bilaterl
- + pelvis duplex and partially or completely ureter fissus or ureter duplex
- etiology: 2 ureteric buds or branching of one bud in proximal end

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





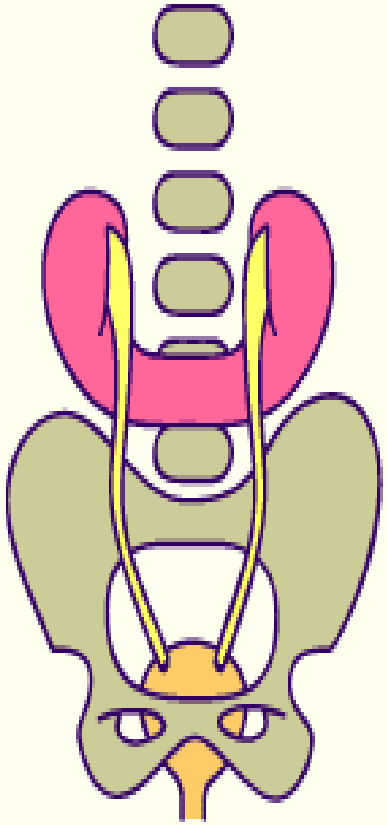
Shape malformations of kidney:

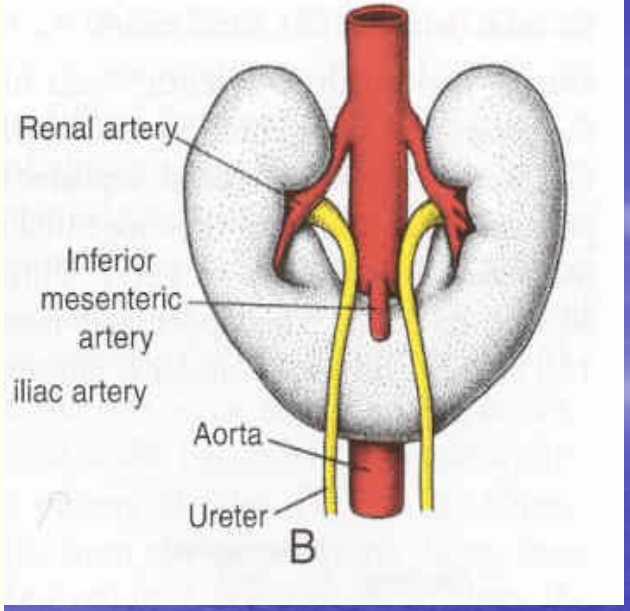
Horse-shaped kidney (ren arcuatus)

1:500

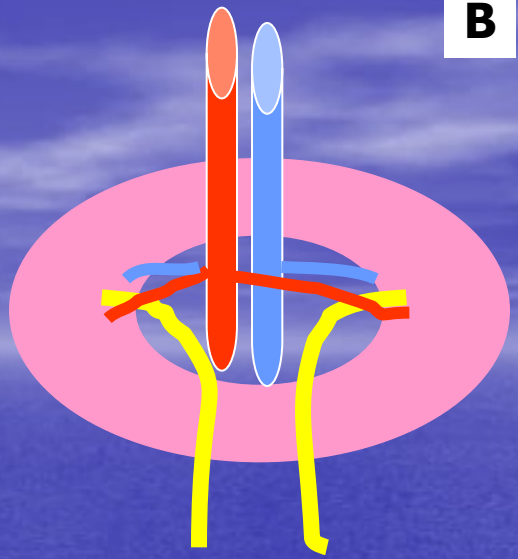
- etiology: fusion of lower pole of both metanephros in front of large vessels (aorta + v.cava inf.)
- fused parenchyma = isthmus „brakes" ascensus renis bellow detachment of a. mesenterica inf. (+position anomaly - ektopia) and rotation (+ malrotation; hilus – ventrally), ureters run in front of isthmus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

A



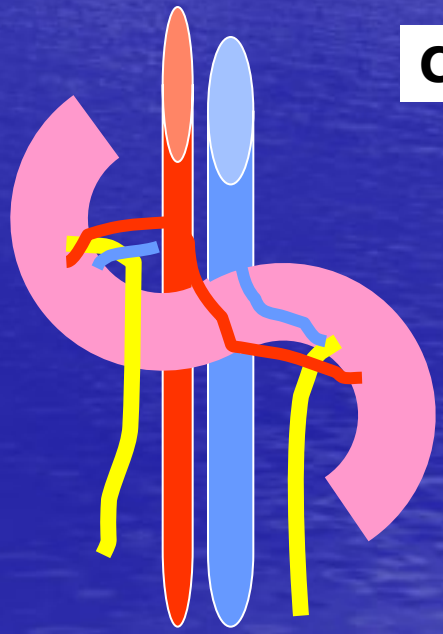


B



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

C



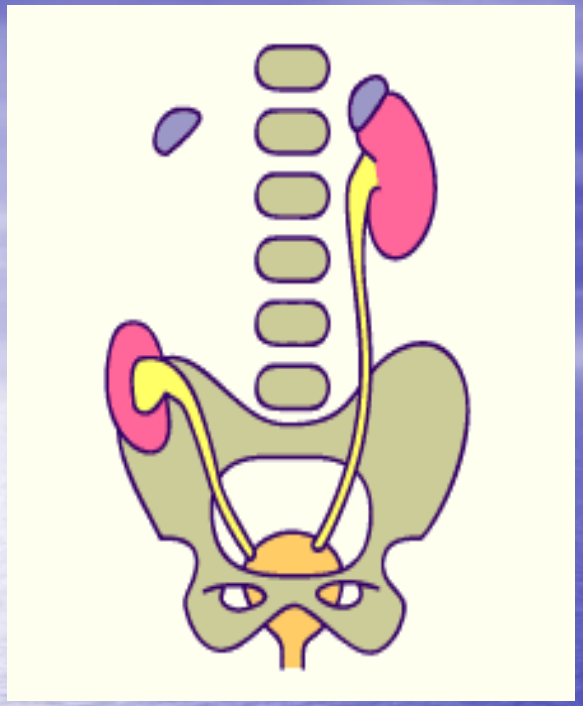
<u>Anomaly of the shape</u> + ektopia:

+ [urine stasis – hydronefrosis
vesicaureteric reflux
secondary infections]

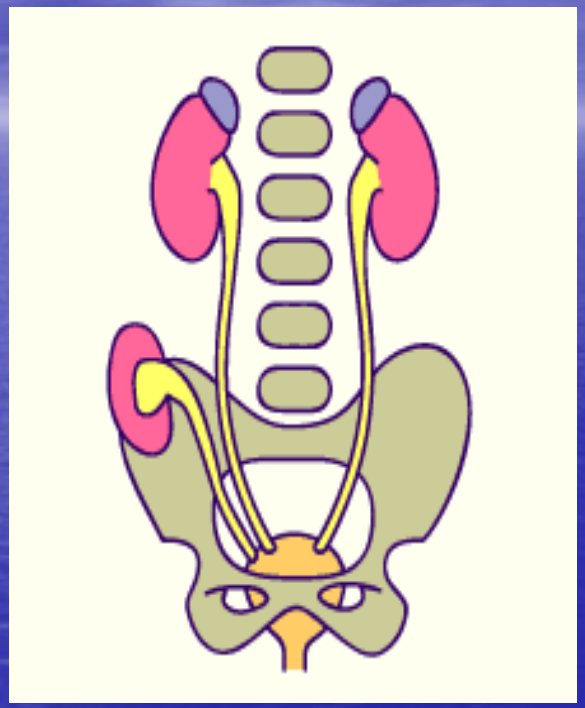
Position anomalies:

Ectopia of kidney uni-, bilat.

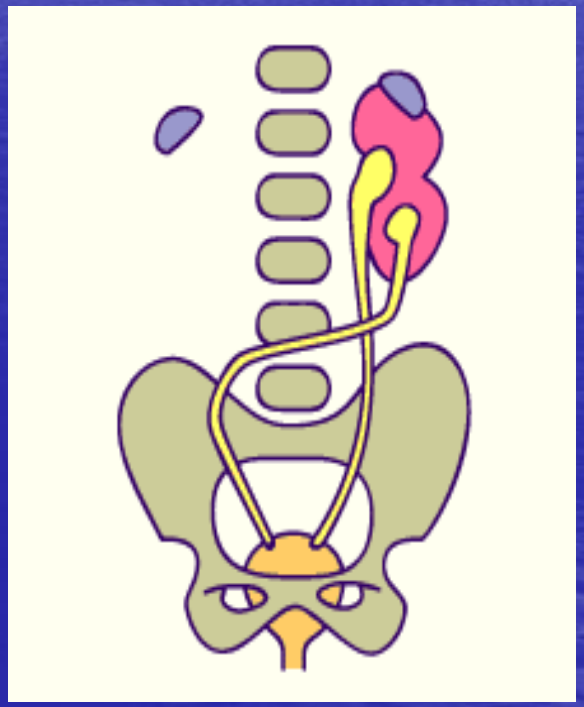
- - ren pelvicus (ren sacralis, ren lumbalis): retention of kidney during ascensus renis
- - cross ectopia: both ureters grow into metanephros on one side or during ascensus renis one kidney transfers on the oposit side and fuse with the other kidney



Ren pelvicus

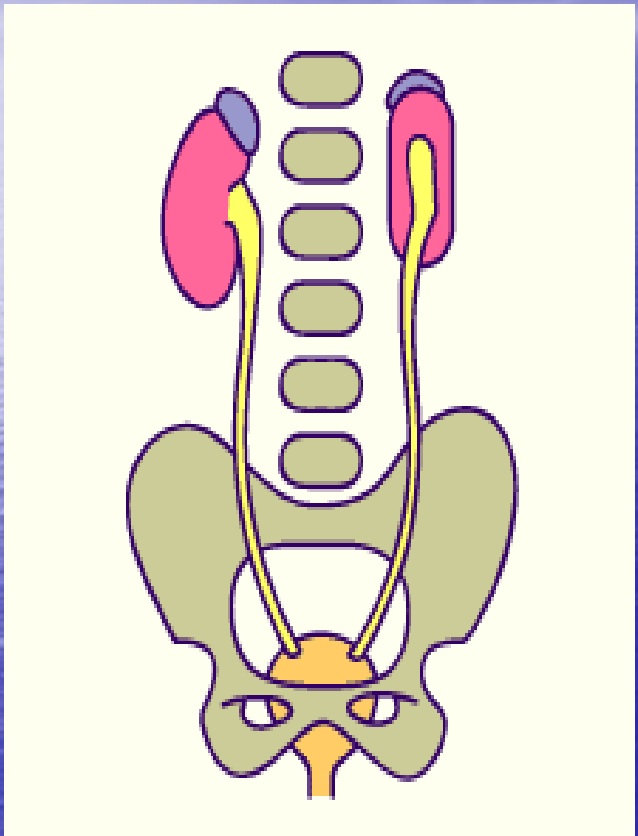


+ ren + ureter duplex



Cross ectopia

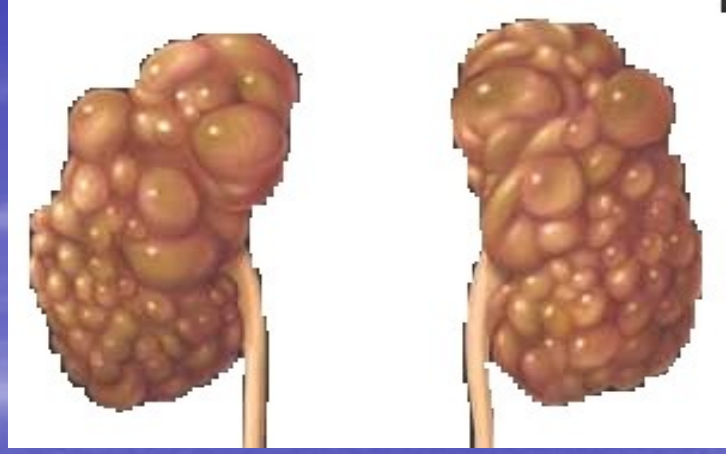
Malrotation (or hyperrotation) **of kidney**



- is connected with ectopia or anomaly of kidney shape
- hilus – ventrally (embryonic position) or dorsaly

- *Notice:(normal adult position of hilus is medial)*

Defekts of parenchyma:
Polycystic kidneys
nephrodysplasia polycystica



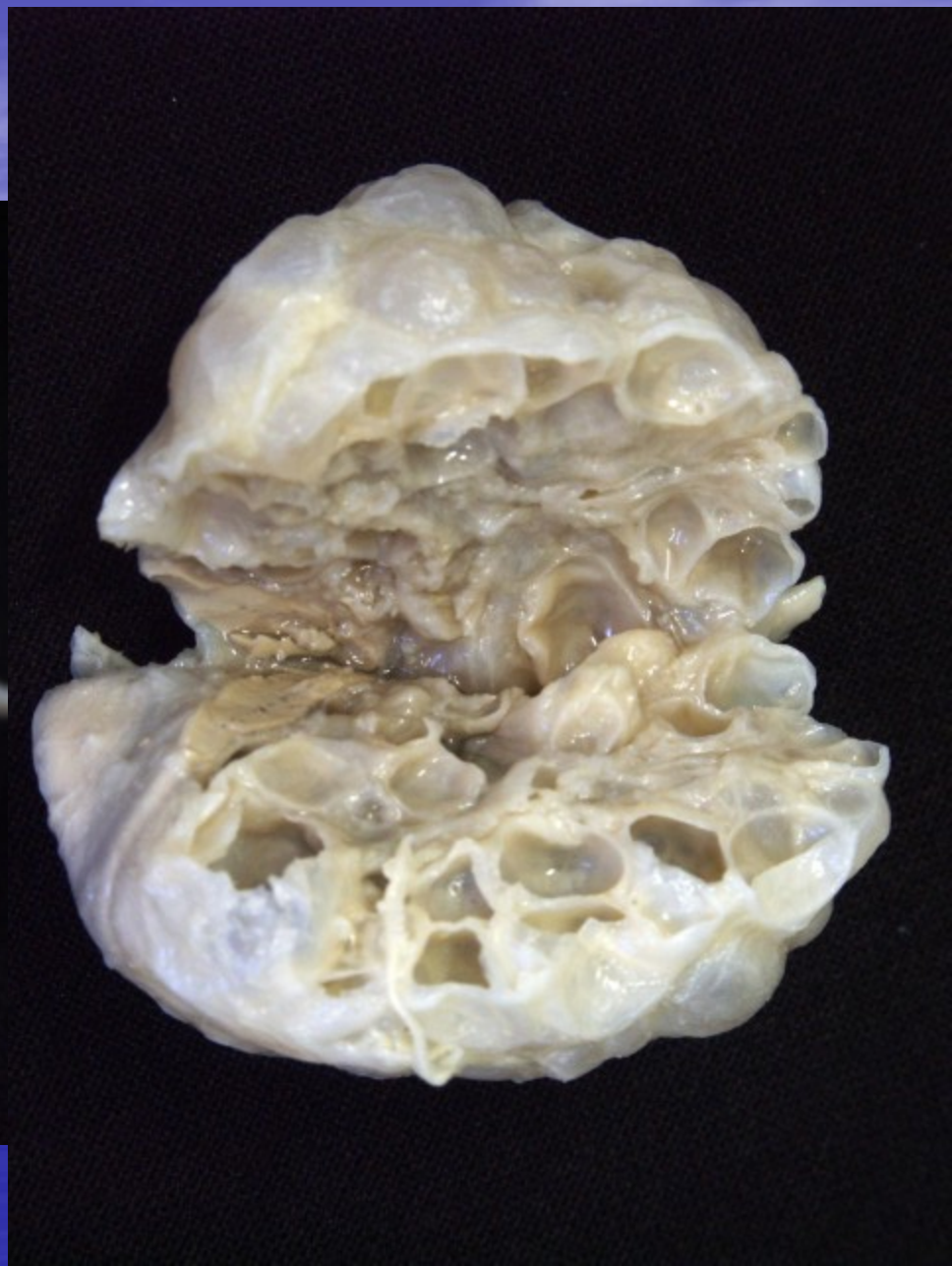
- diffuse cystic malformation (always bilat.)
  – cystic degeneration of kidney
- 2 forms of polycystic disease:
  - autosomally dominant type
    „adult" macrocystic form
  - autosomally recesive type
    „infantil" microcystic form

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 funcions have to be compensated (hemodialysis, peritoneal dialysis, transplantation)

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

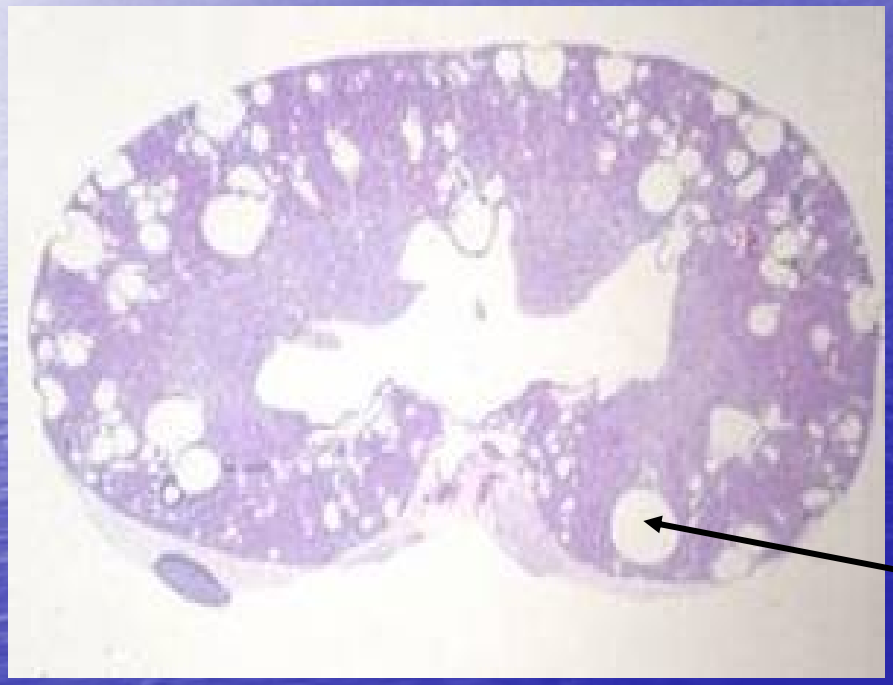


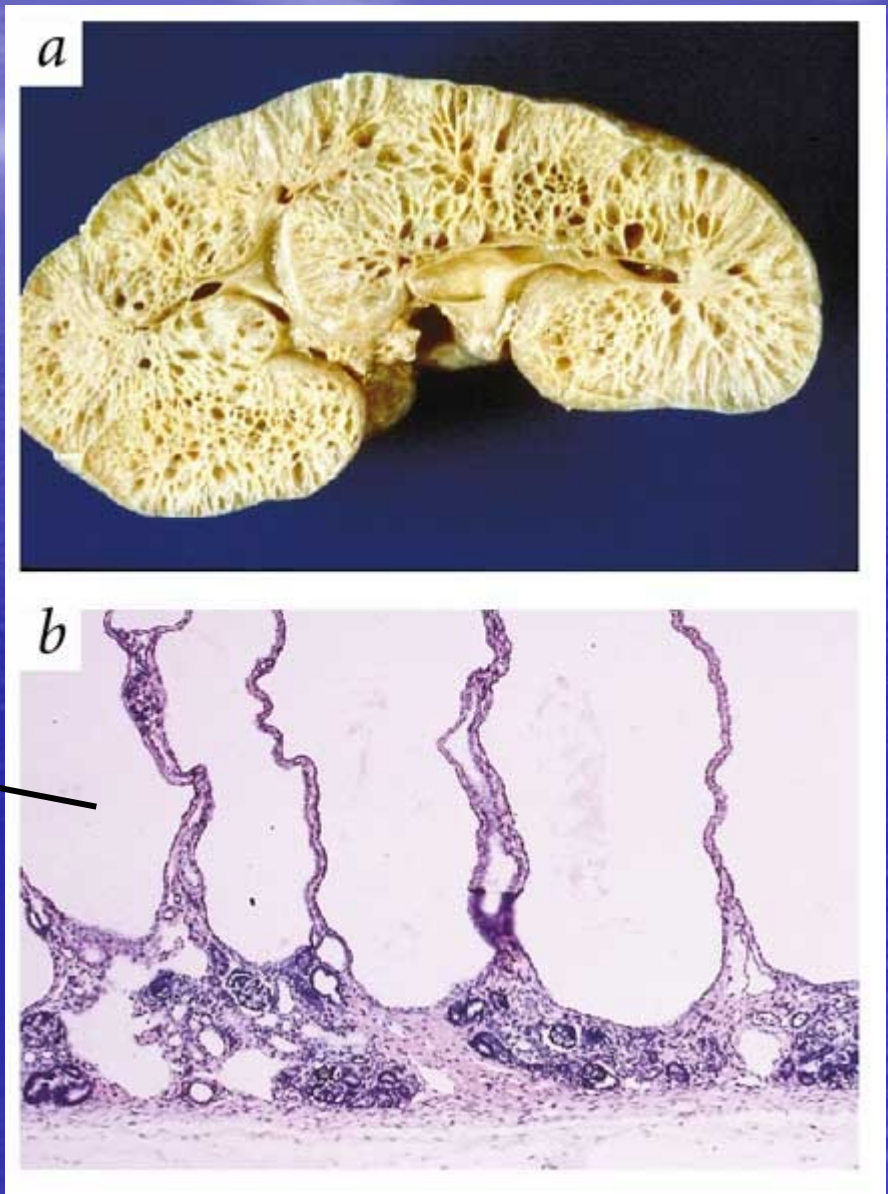
autosomally recesive type

IPCD - ″infantil″ form

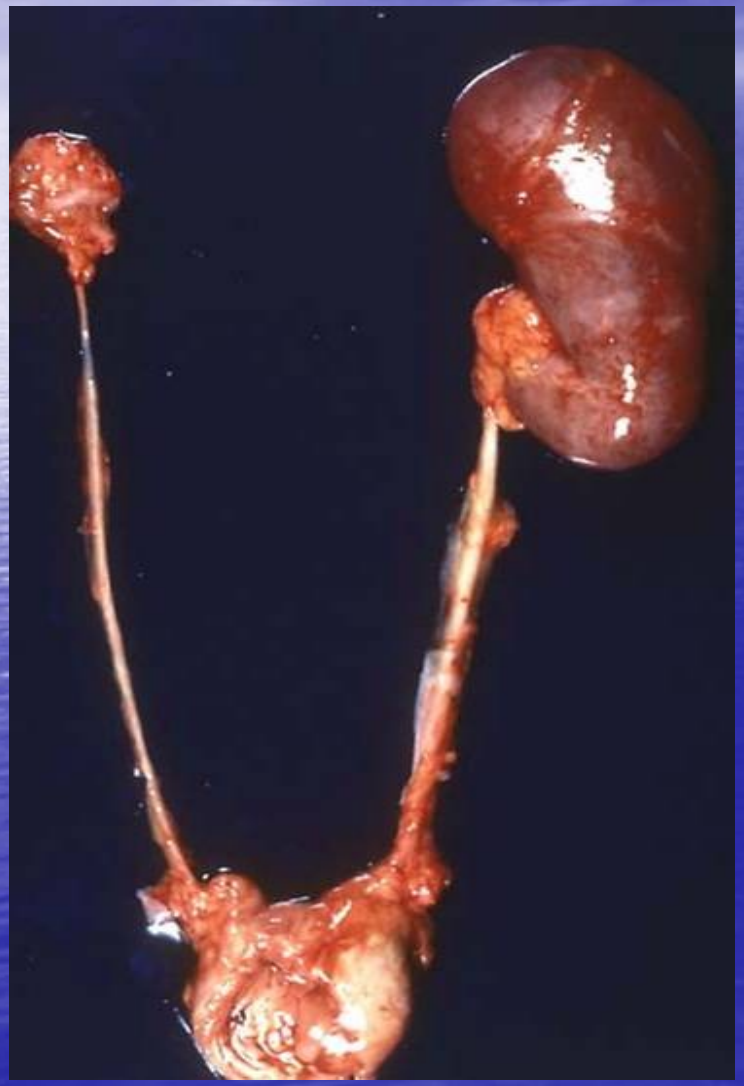
- 1 : 40.000, probability of transmission to offspring - 25 % children of healthy parents „disease carriers″;
- + anomalies also in liver, spleen, lungs,
- etiol.: unclear - defect of ureter development (nephrons are not connected with collecting ducts)
- Klinic manifestation: bilat. enlarged kidneey, 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*

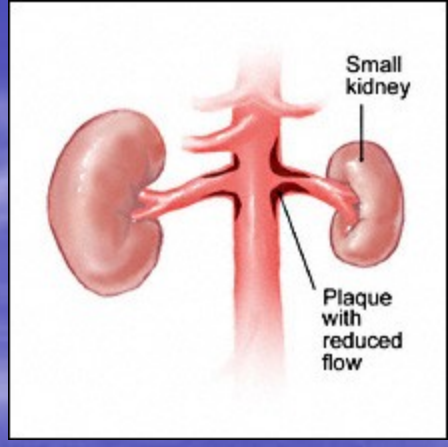
Polycystic kidney – cysts are not seen on the surface of kidney



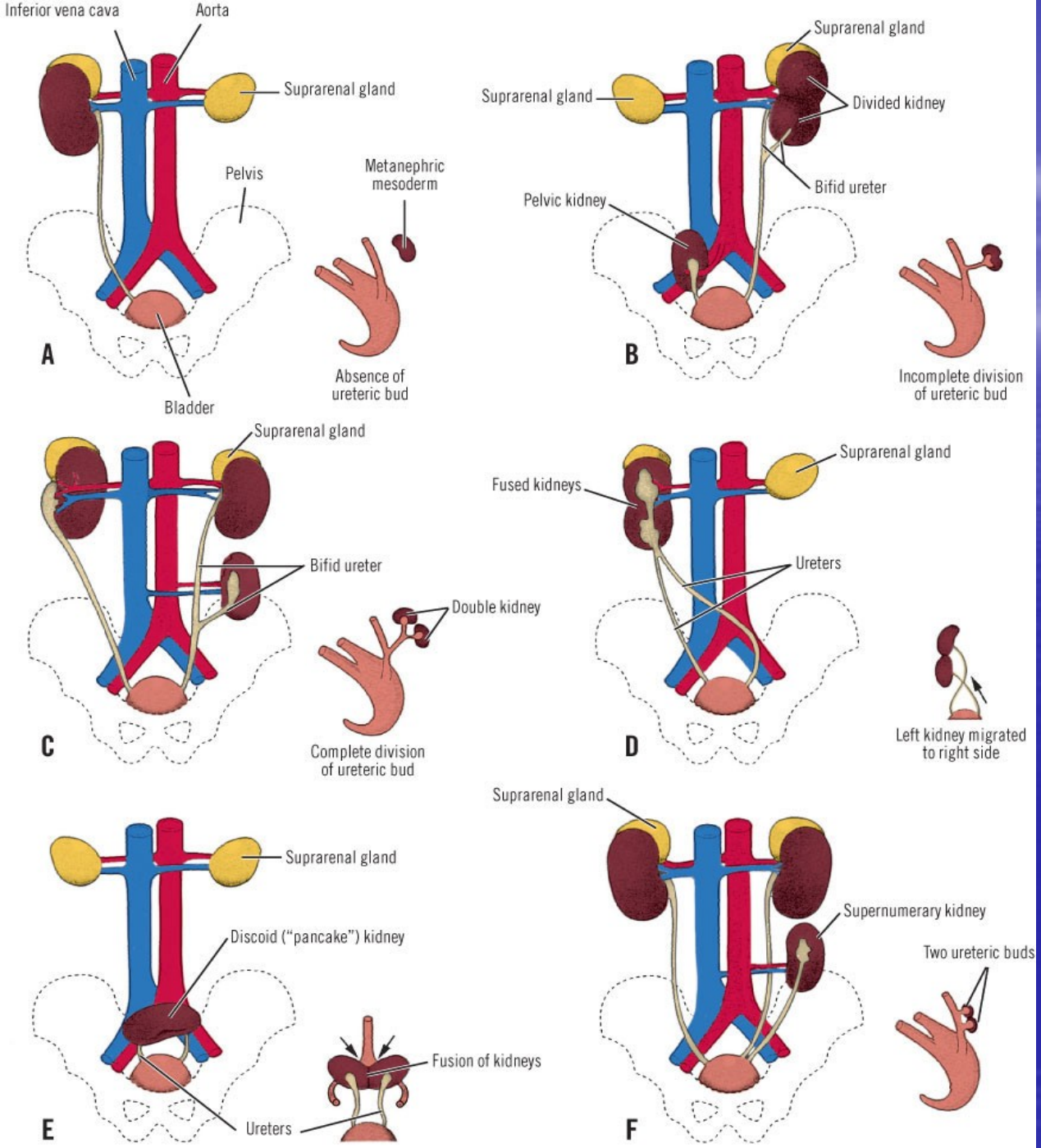


Hypoplasia renis





- Insuficiently developed kidney – small, small amount of histologically normal and functional nephrons
- usually unilateral
- compenzational hypertrofia of the other kidney



A – unilateral renal agenesis
B – pelvic ren + ureter bifidus

C – kidney malrotation + ren duplex
D – crossed ectopia

E – „pancake" kidney

F – supernumerary kidney

Wilms' tumor (nephroblastom)

- The most frequent type of tumors in chidren under 5 years, rare in adulthood
- 90% treatment succes, also in case of greater distribution (metastasis)
- familiar occurrence – tumor contais 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*

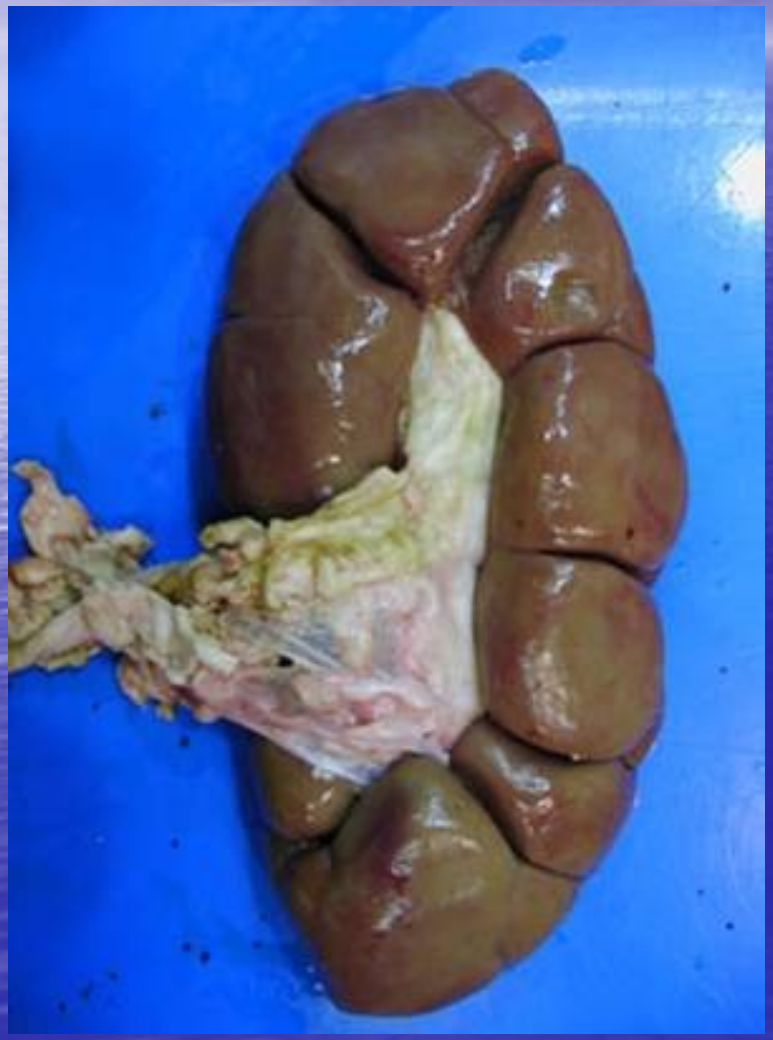
- Anderson-Fabry disease (storage of cerebrosides *= neutral sphingolipids*),
- von Gierke disease (storage of glycogen),
- Gaucher disease (storage of glukocerebrosides),
- Fanconi sy. (storage of cystine; **cystinóza**, **cystinurie**)
- **Primary hyperoxaluria** – cong. defect of glykooxalates production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

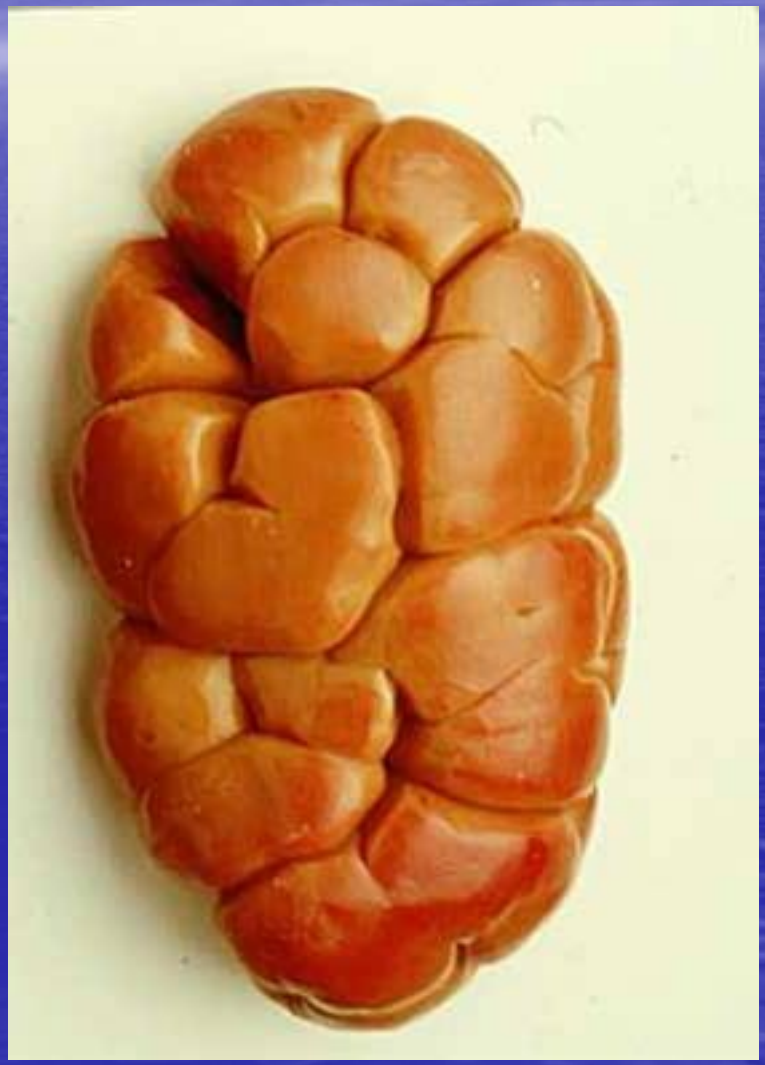
Anomalies of renal vascularization

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

- accesory arteries – 25 %, veins - 12,5 %

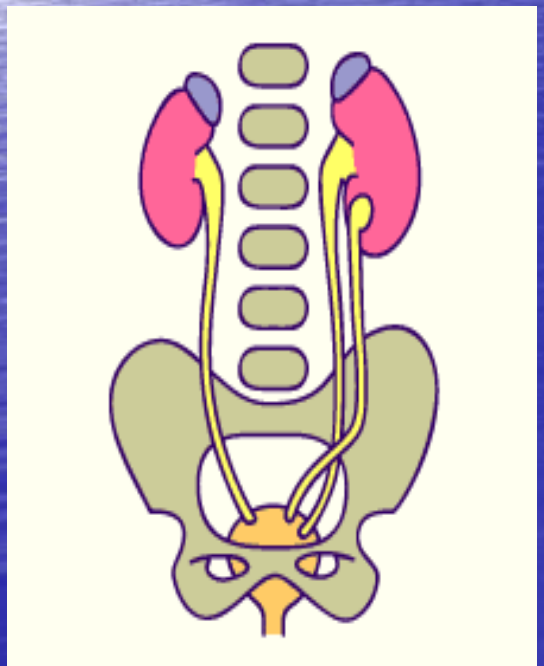
Renal renculi

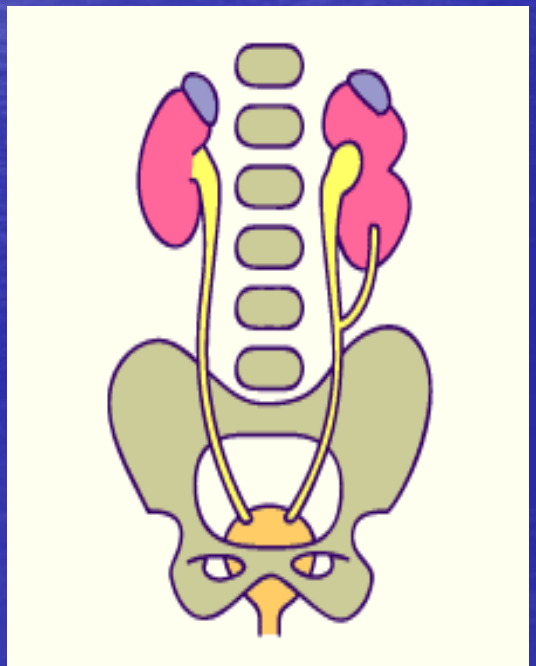


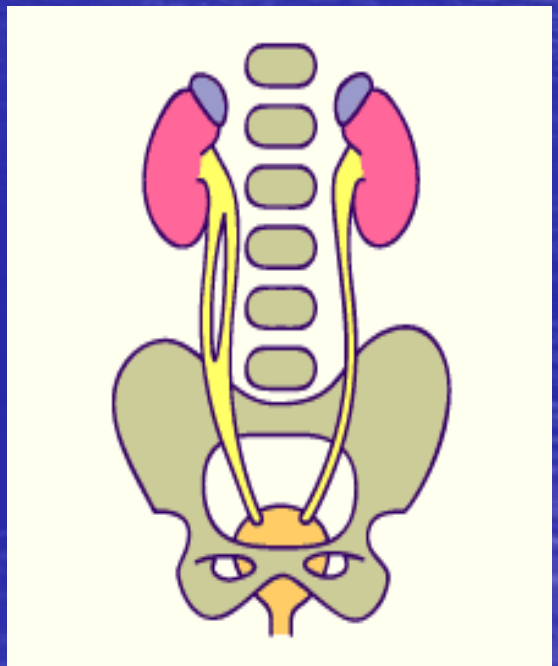


2. CM of pelvis and ureter

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



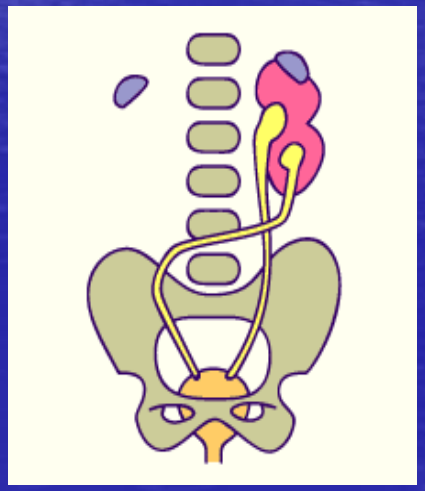


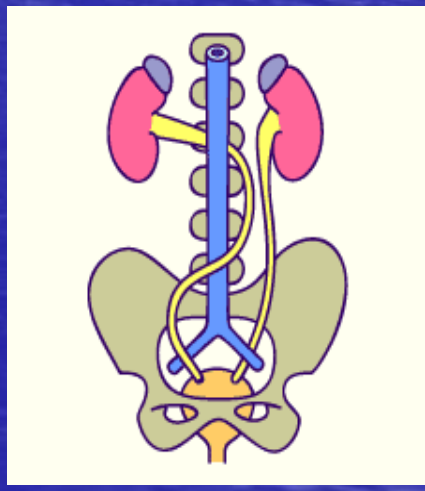


Ectopic defects of ureter

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

- cross ectopia of ureter, „retrocava" ureter –

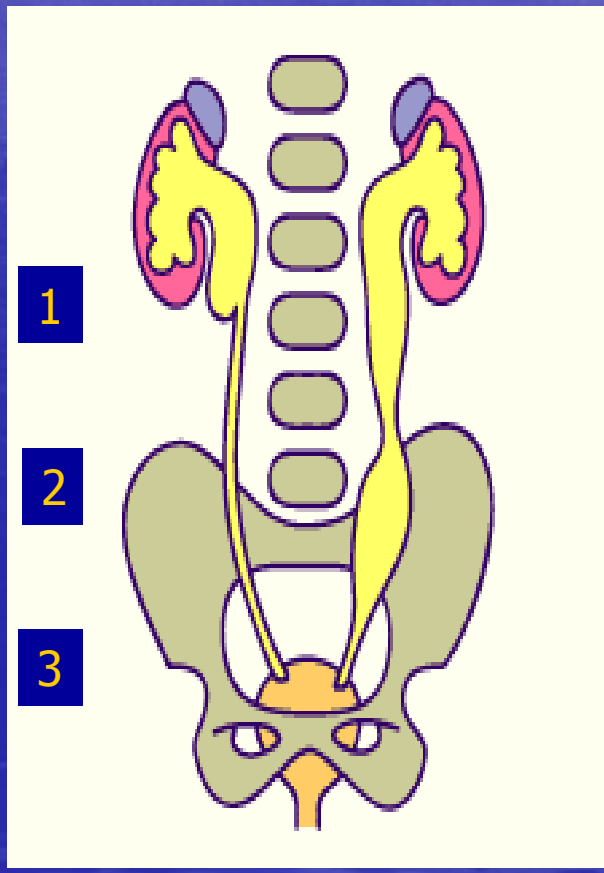




Congenital stenosis, obstructions, atresis

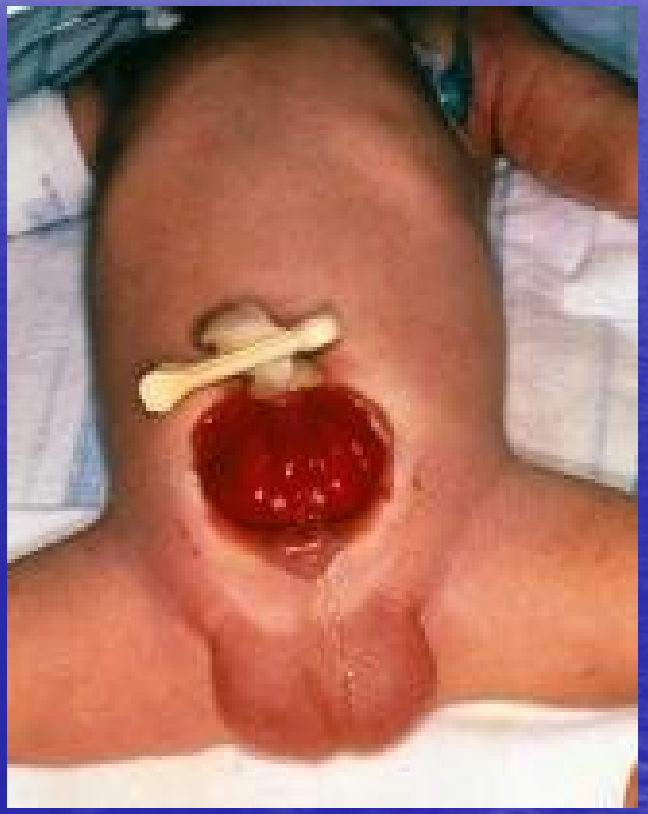
Physiol. ureter narrowings:

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

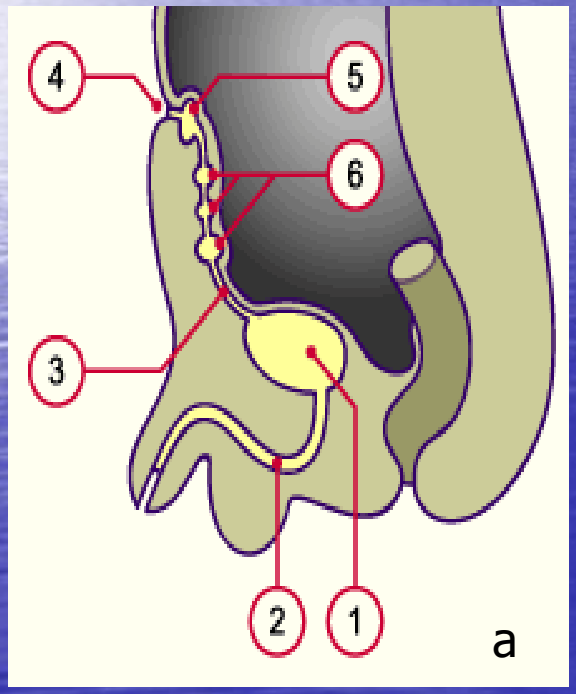
- etiol.:  defect of mezenchyme migration between ectoderm of abdominal wall and cloaca in week 4
- Reconstruction of the wall (24 - 48 h after birth), epispadia (about 2nd year).

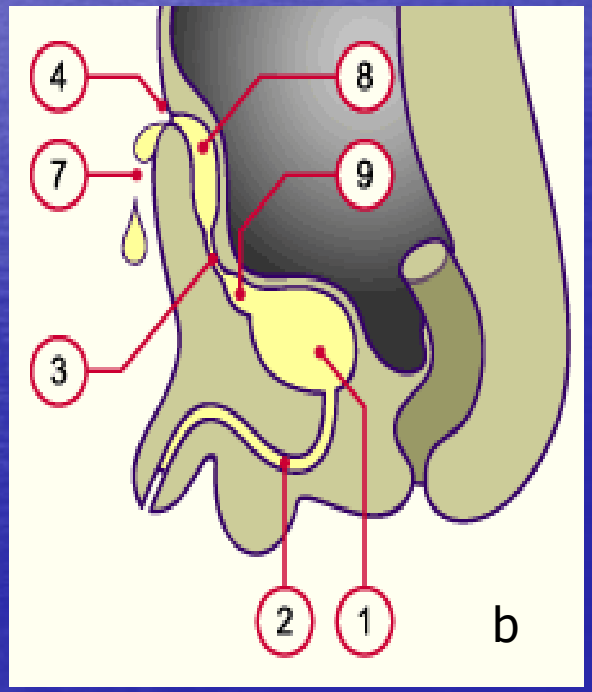
Sy. prune – belly

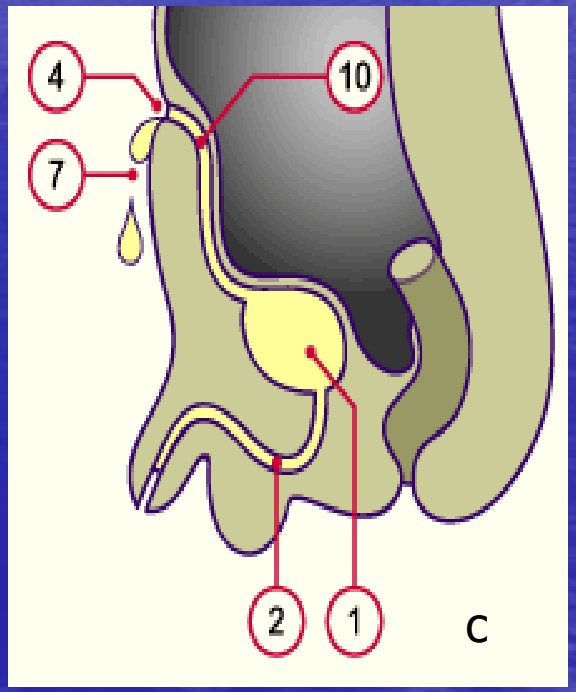
- 1 : 40 000
- Trias of syndromes:
  1. agenesis of muscles in abdominal wall
  2. obstruction of urinary bladder in region of internal urethral ostium (dilatation of bladder, megaureters and hydronephrosis – above obstruction)
  3. bilateral kryptorchism
- Etiol.:  unclear
  hormonal treatment (estrogens)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

defect obliteration of ductus allantoideus –

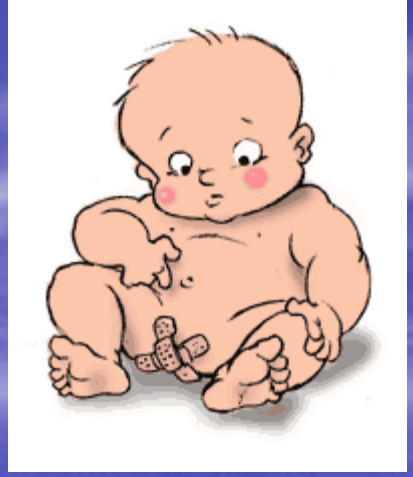
- urachal cysts and fistulae (a, b)
- urachus patens (c)







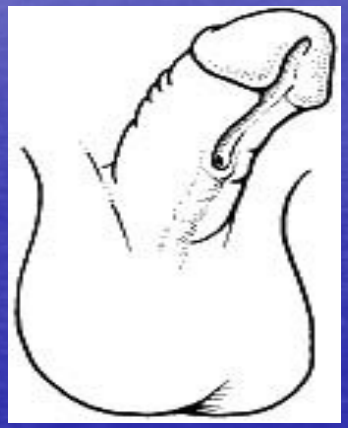
4. CM or urethra



- **Clefts of urethra:**

Hypospadia
insufficient fusion of plicae genitales

Epispadia
see extrophia



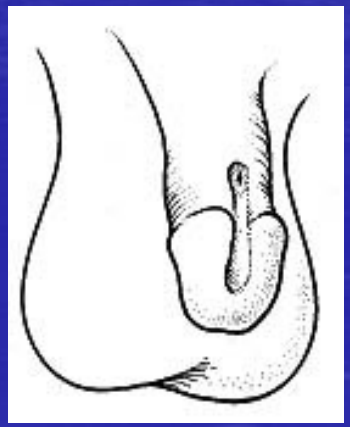


Figure 1. Various locations of the urethral opening or meatus

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.

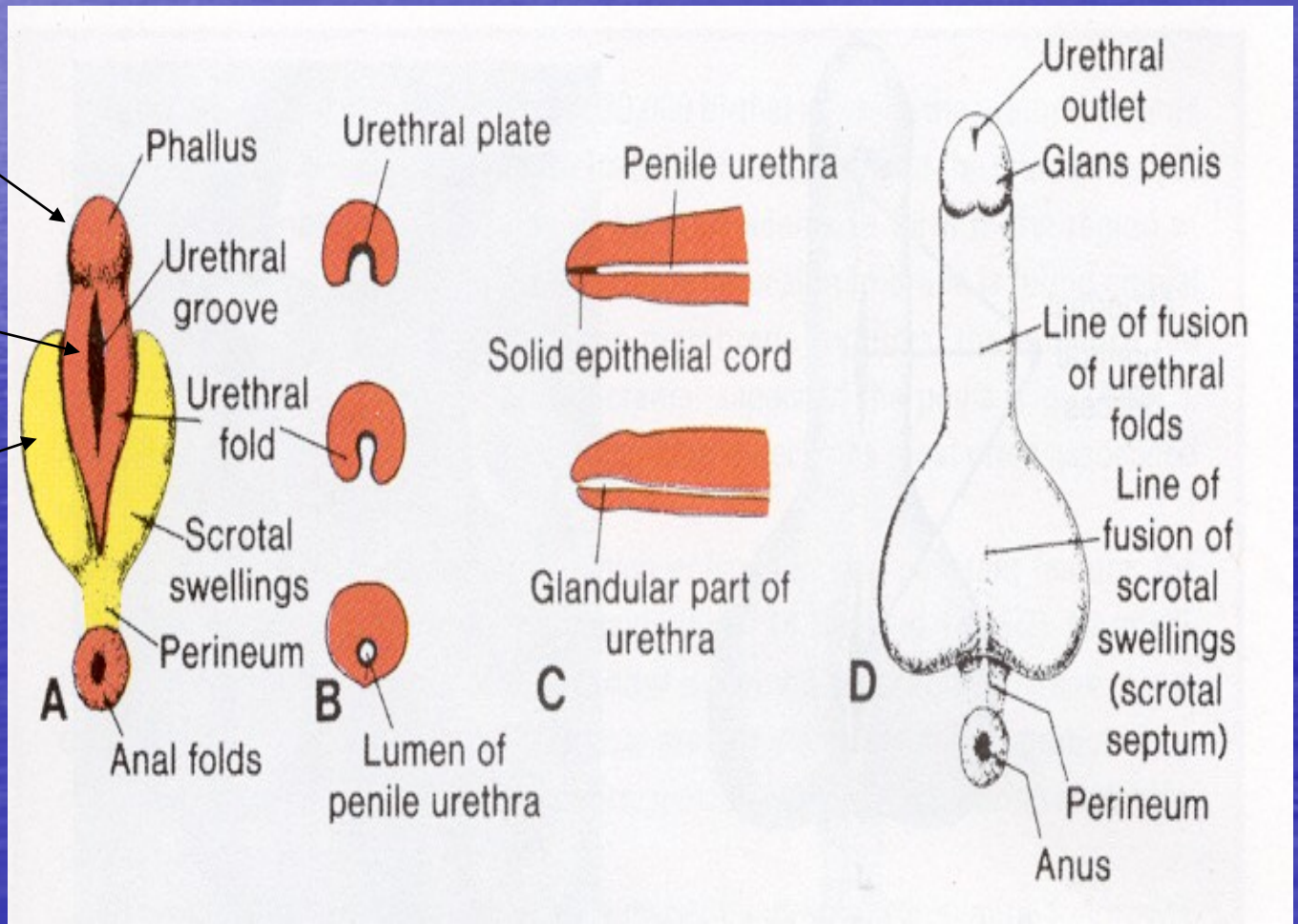
Vývoj zevního genitálu a uzavření uretry ♂

Zdroje pro vývoj zevního genitálu:

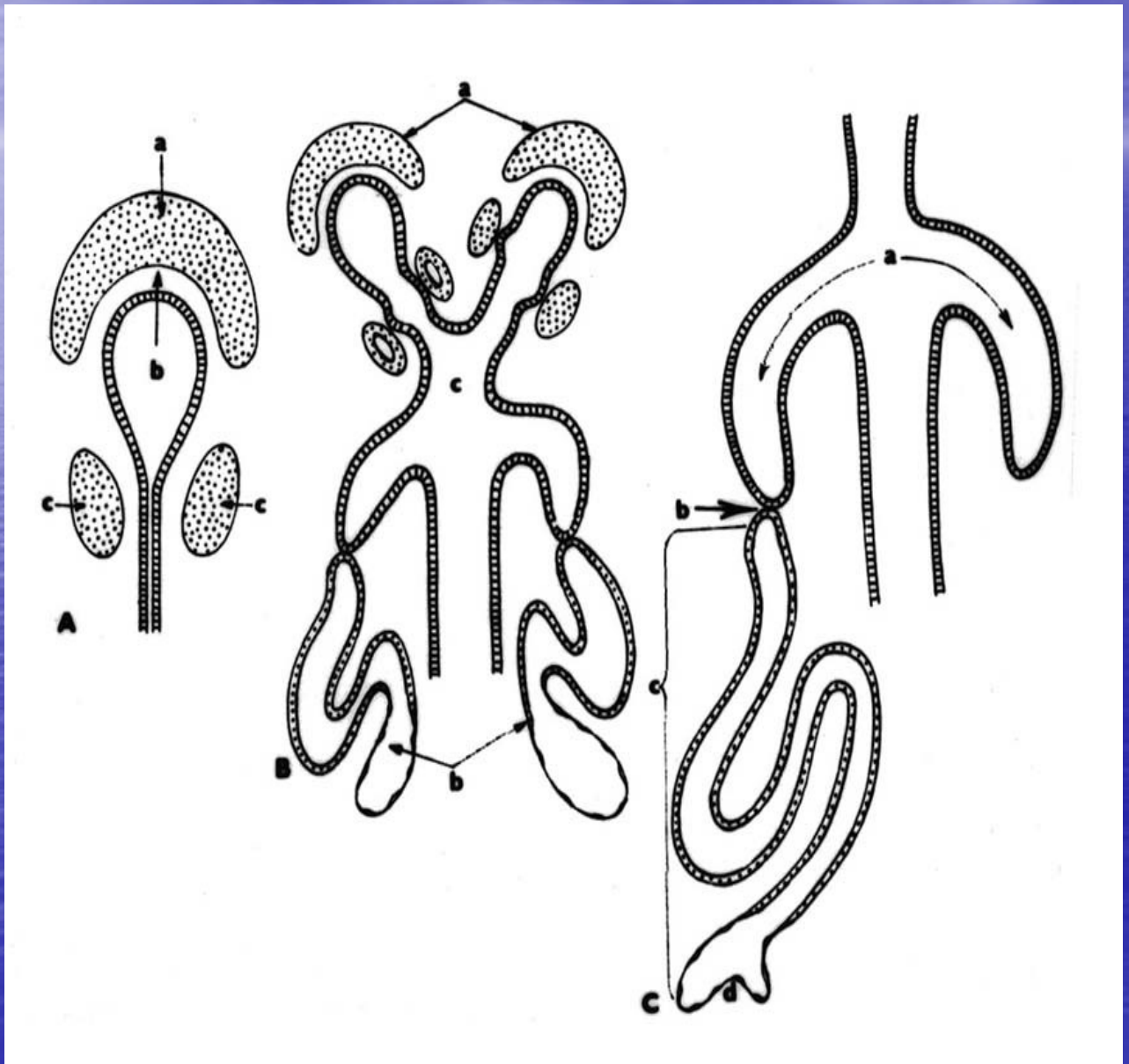
Tuberculum genitale

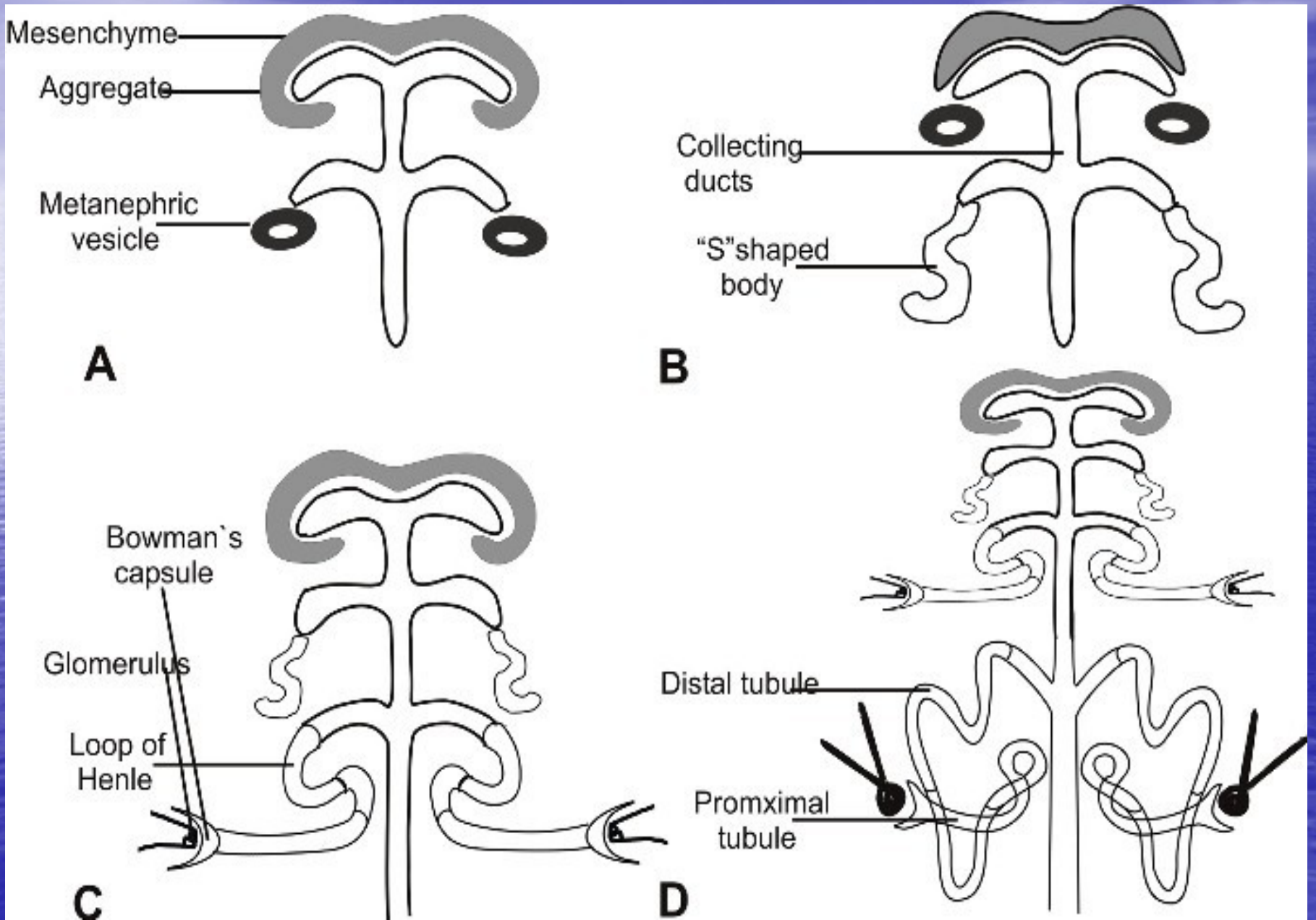
Plicae genitales (urethrales)

Tori genitales (plicae labioscrotales)



Vývoj nefronu




Mesenchyme
Aggregate
Metanephric vesicle
A
Collecting ducts
"S"shaped body
B
Bowman`s capsule
Glomerulus
Loop of Henle
C
Distal tubule
Promximal tubule
D

Močový měchýř, ureter