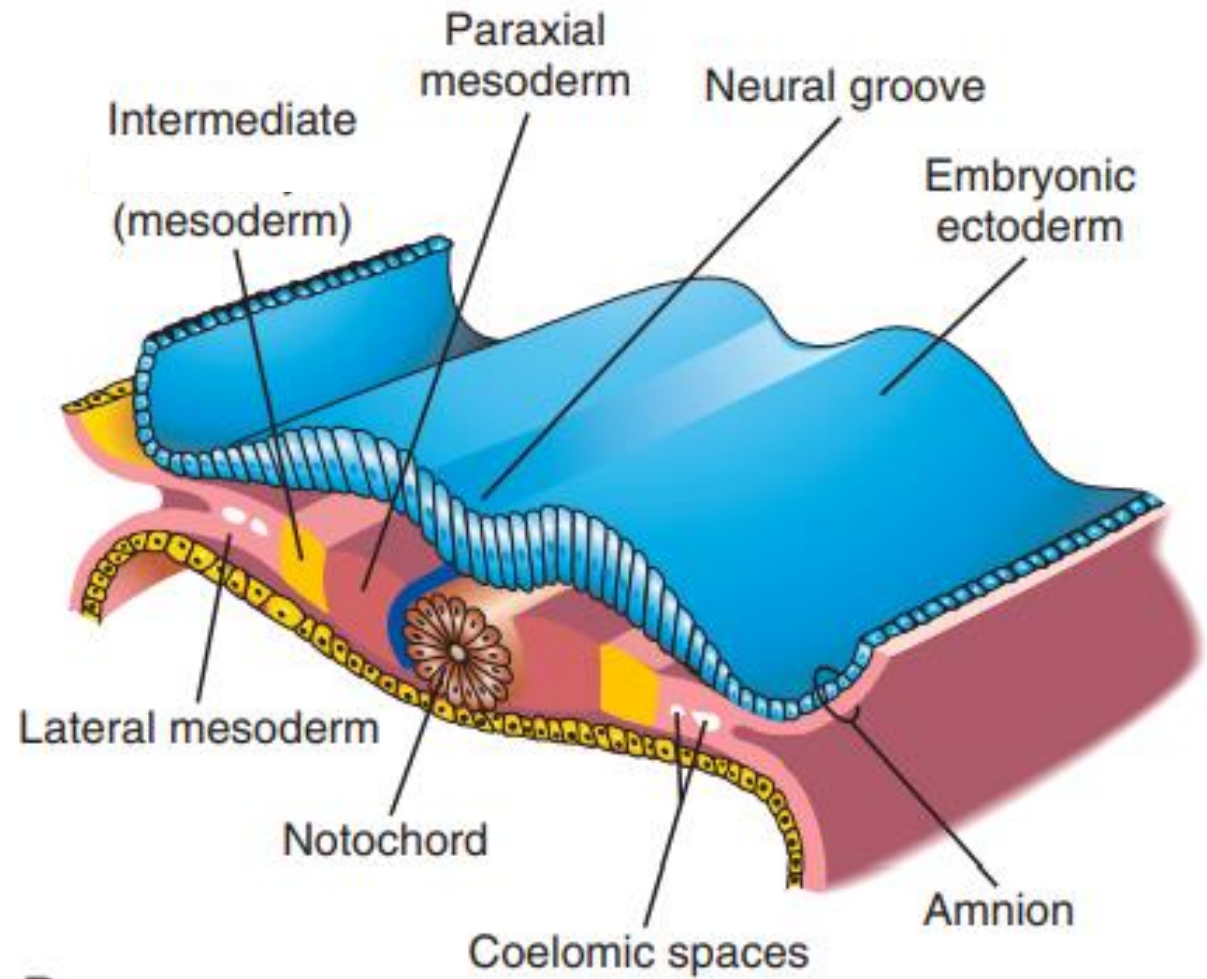


Přehled vývoje a teratologie močového systému

07.11.2024

Anna Mac Gillavry Danylevska

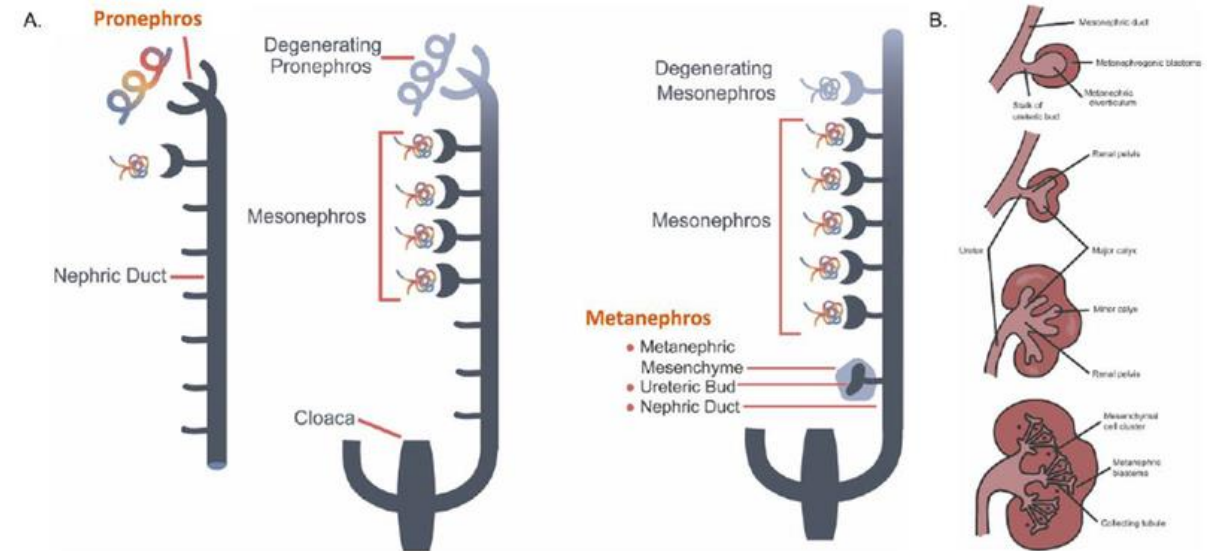


B

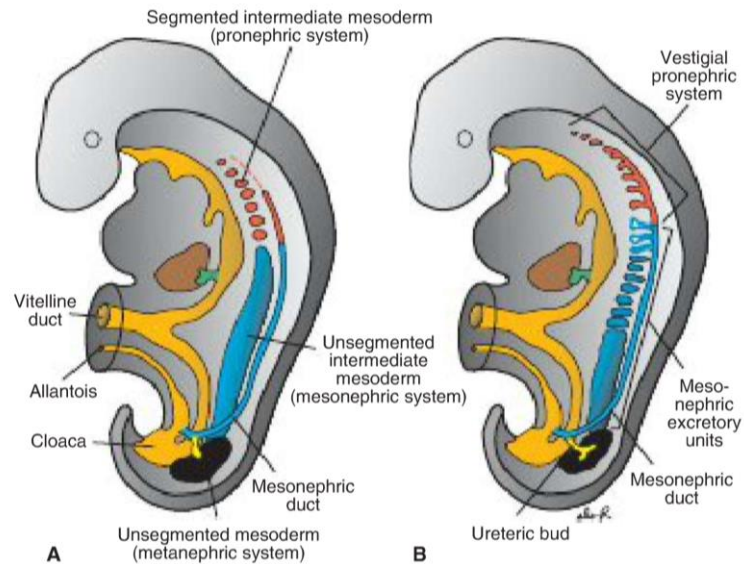
Pronephros: 4. týden

Mesonephros: 4. – 10. týden
(funkční 6. – 12. t.)

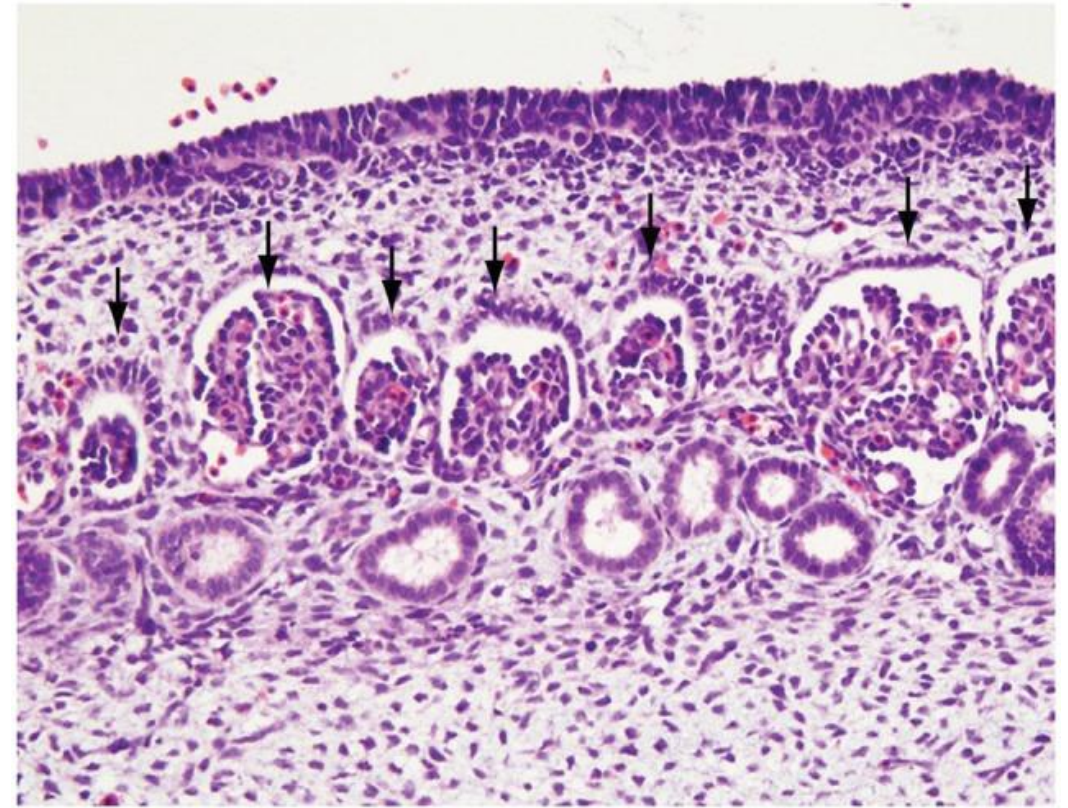
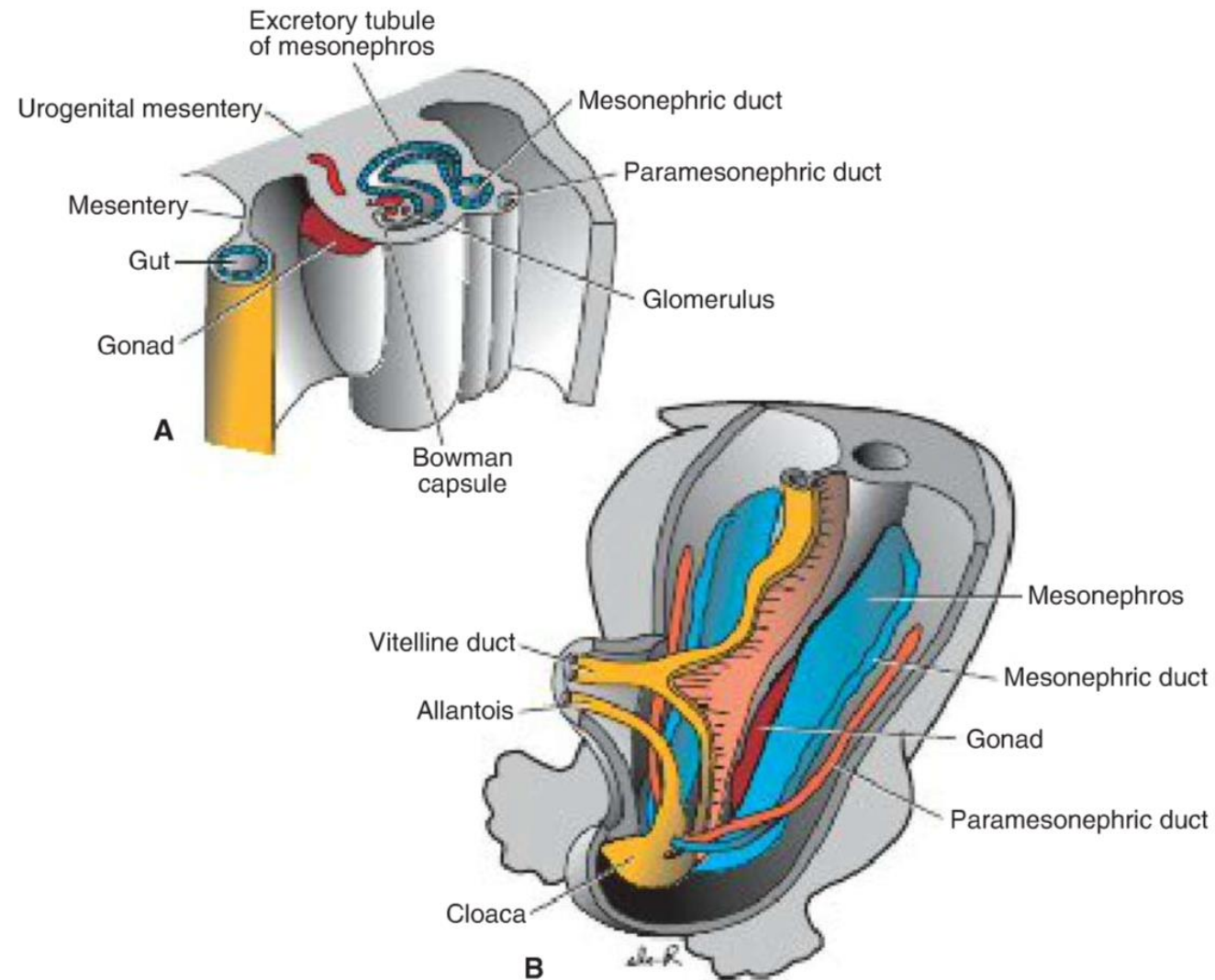
Metanephros: 5. - 9. týden
(funkční od 12. t.)



Stonebrook E, Hoff M, Spencer JD. Congenital Anomalies of the Kidney and Urinary Tract: A Clinical Review. *Curr Treat Options Pediatr.* 2019;5(3):223-235. doi: 10.1007/s40746-019-00166-3. Epub 2019 Jun 11. PMID: 32864297; PMCID: PMC7451090.

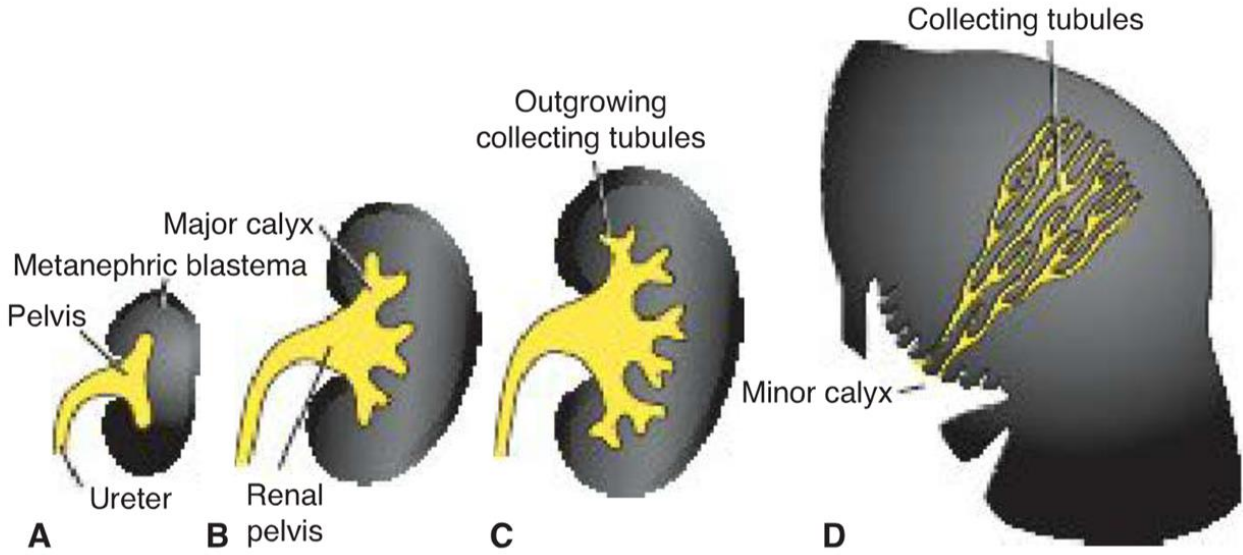
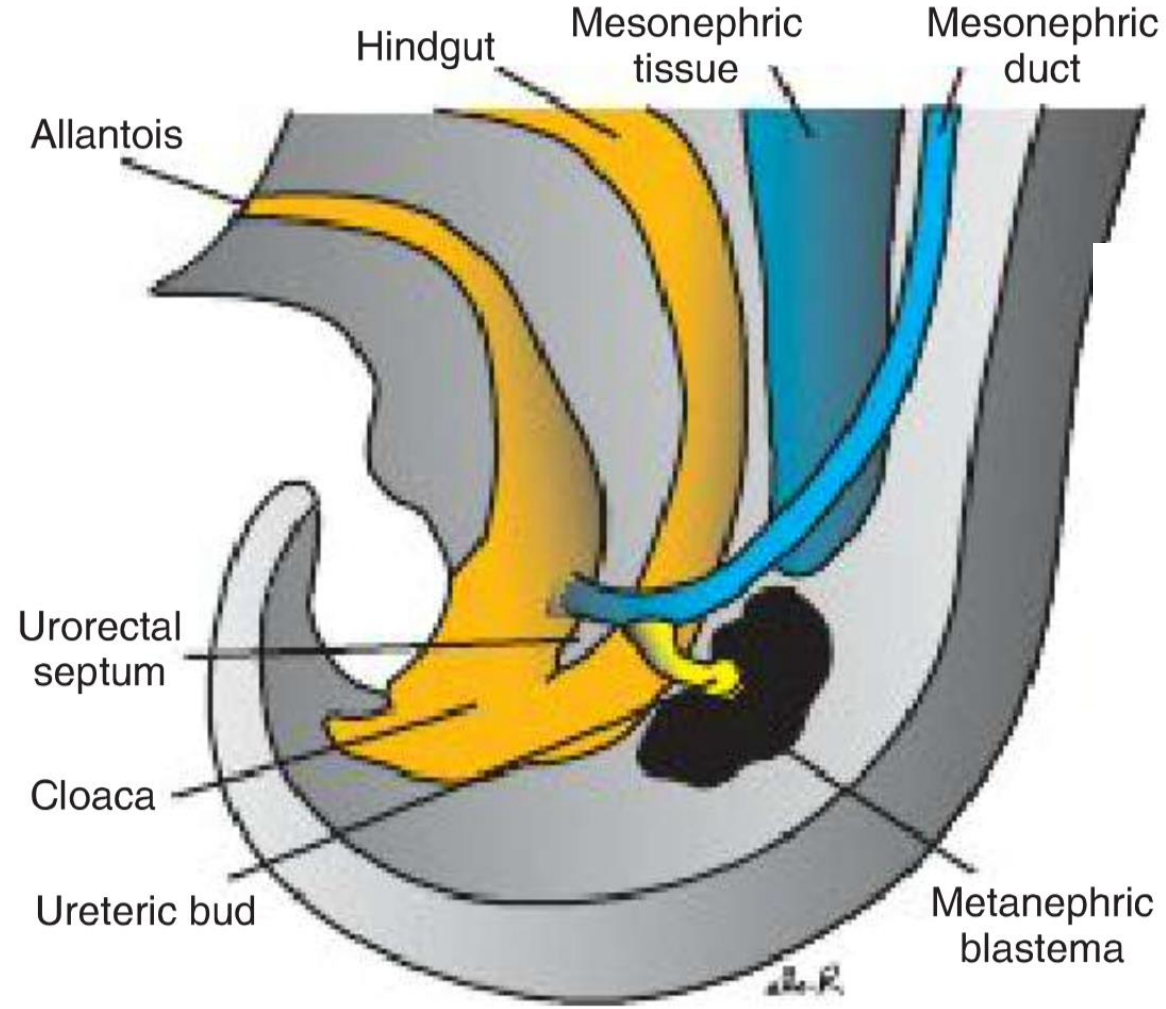


Mesonephros



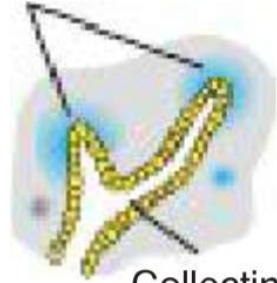
Jain S, Chen F. Developmental pathology of congenital kidney and urinary tract anomalies. Clin Kidney J. 2018 Dec 1;12(3):382-399. doi: 10.1093/ckj/sfy112. PMID: 31198539; PMCID: PMC6543978.

Metanephros



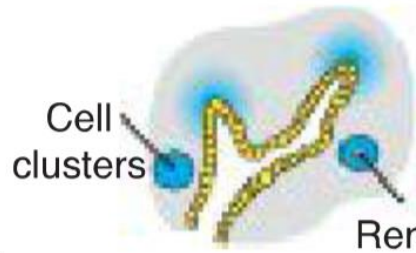
T.W. Sadler, Langman's medical embryology, 12th edition

Metanephric tissue caps



A

Collecting tubule



B

Cell clusters

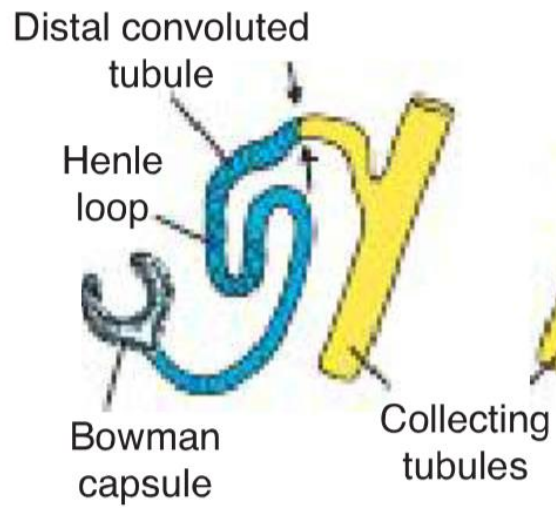
Renal vesicle



C

Nephron

Bowman capsule



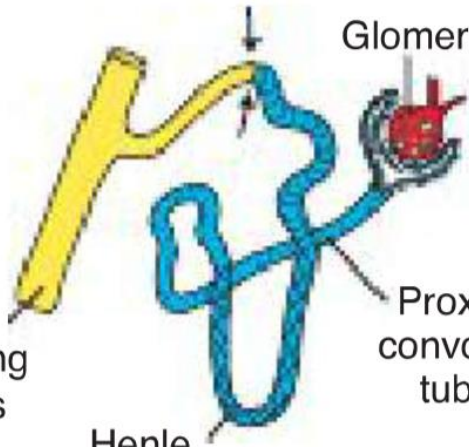
D

Distal convoluted tubule

Henle loop

Bowman capsule

Collecting tubules



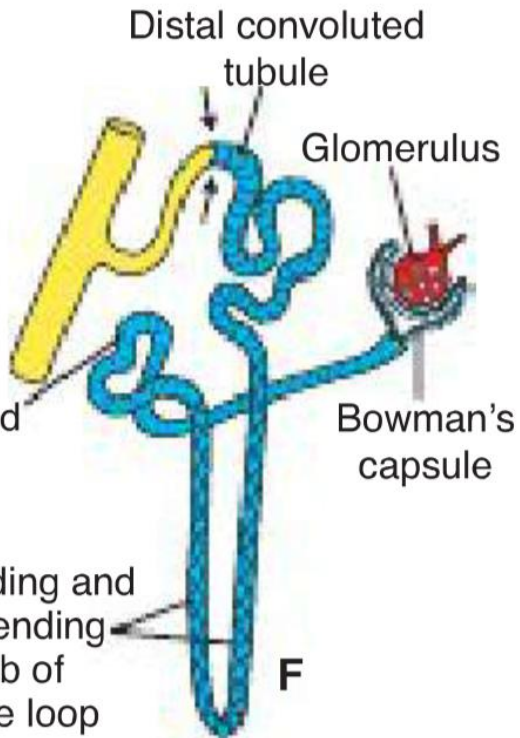
E

Henle loop

Glomerulus

Proximal convoluted tubule

Ascending and descending limb of Henle loop



F

Distal convoluted tubule

Glomerulus

Bowman's capsule

VVV ledvin

- Renální ageneze: unilaterální (1/1000 živě narozených) – častěji chybí levá, druhá podléhá kompenzační hypertrofii;
bilaterální (1/3000); 3:1 ♂ vs ♀ ; anuria -> oligohydramnion
-> hypoplasie plic, „Potter face“ ...

Syndrom Potterové

Pulmonary hypoplasia

Oligohydramnios

Twisted skin

Twisted face

Extremities defects

Renal agenesis

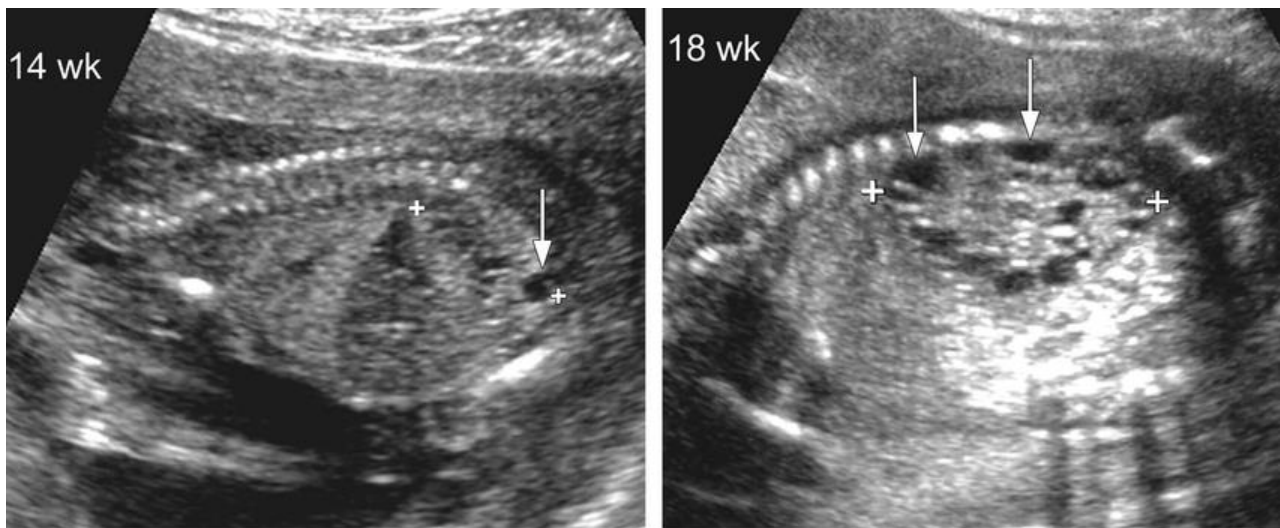
Table 1. Subtypes of Potter's syndrome [4,5].

Classic Syndrome	Bilateral Renal Agenesis
Potter's syndrome type 1	Autosomal recessive polycystic kidney
Potter's syndrome type 2	Renal dysplasia
Potter's syndrome type 3	Autosomal dominant polycystic kidney
Potter's syndrome type 4	Obstruction either in kidney or ureter leading to kidney disease

Kostov, Stoyan & Slavchev, Stanislav & Dzhakov, Deyan & Strashilov, Strahil & Yordanov, Angel. (2020). Discordance for Potter's Syndrome in a Dichorionic Diamniotic Twin Pregnancy—An Unusual Case Report. *Medicina*. 56. 109. 10.3390/medicina56030109.



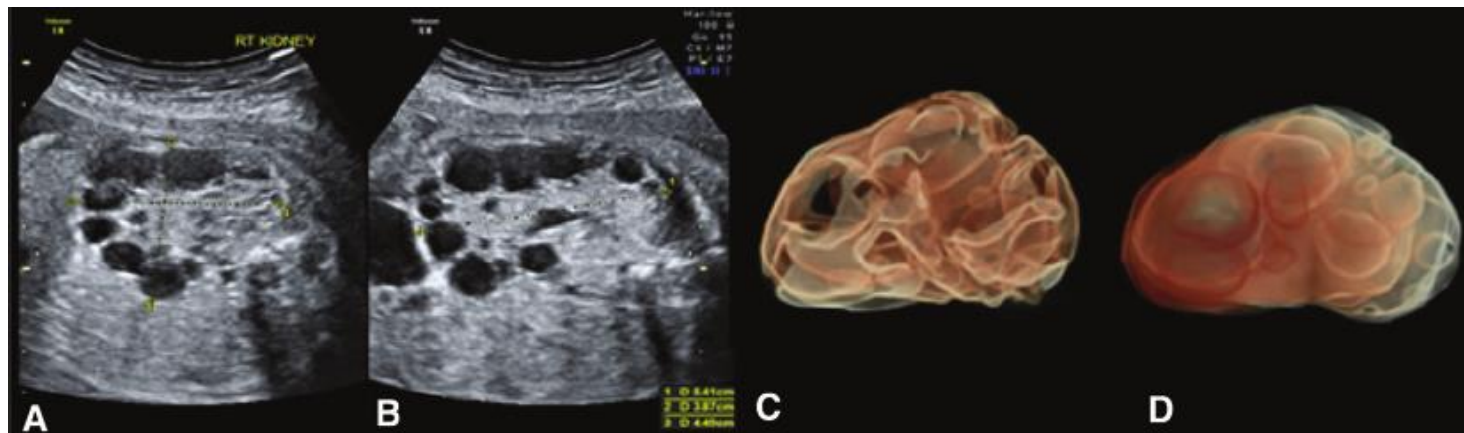
- Renální dysplasie
 - renální multicystická dysplasie (MCDK): unilaterální vs bilaterální



a. Fong, Katherine & Toi, Ants & Salem, Shia & Hornberger, Lisa & Chitayat, David & Keating, Sarah & McAuliffe, Fionnuala & Johnson, Jo-Ann. (2004). Detection of Fetal Structural Abnormalities with US during Early Pregnancy1. Radiographics : a review publication of the Radiological Society of North America, Inc. 24. 157-74.

b.

10.1148/rg.241035027.



Spalldi Barišić, Lara & Stanojevic, Milan & Kurjak, Asim & Porovic, Selma & Gaber, Ghalia. (2017). Diagnosis of fetal syndromes by three- and four-dimensional ultrasound: is there any improvement?. Journal of perinatal medicine. 45. 10.1515/jpm-2016-0416.

- Cystická onemocnění ledvin:

- polycystická choroba ledvin autozomálně dominantního typu (PCHLAD; ADPKD): 1/500-1000; cysty vznikají z libovolných částí nefronů, k selhání ledvin dochází nejčastěji až v dospělosti.

5000 pacientů v ČR.

- polycystická choroba ledvin autozomálně recesivního typu (PCHLAR; ARPKD): 1/5000-20000 cysty vznikají ze sběrných kanálků, k selhání ledvin dochází ve fetálním období nebo brzy po narození.

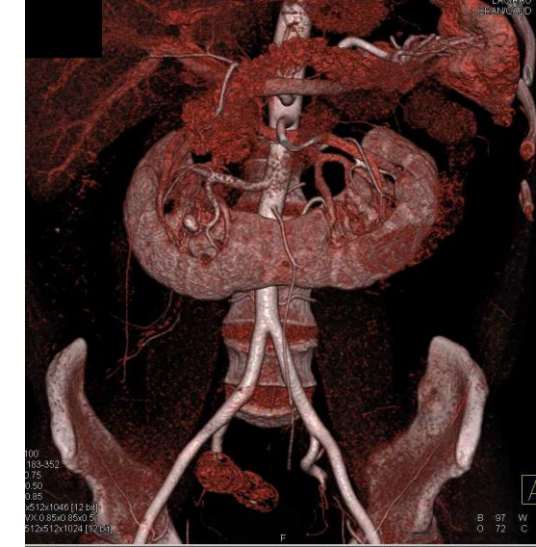
- Abnormální poloha:

- ektopická ledvina

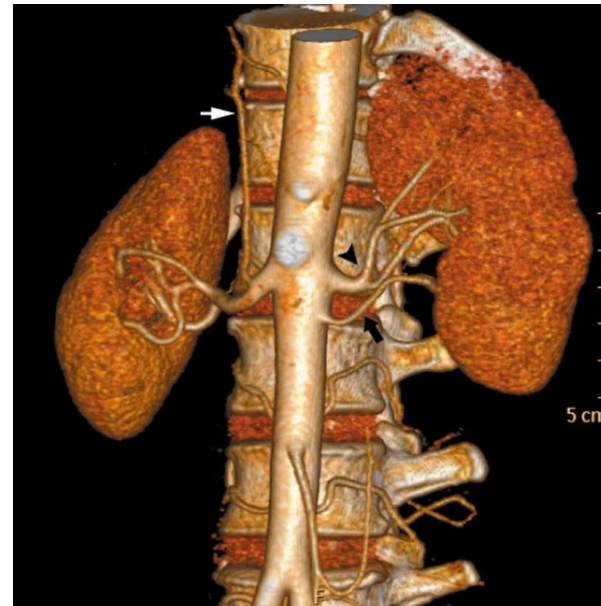
- *ren arcuatus* (1/600)

- *ren sigmoideus* ...

- nadpočetné renální arterie – ¼ ledvin má 2 až 4 arterie



[Horseshoe Kidney with Calculus but Normal Function - Kidney Case Studies - CTisus CT Scanning](#)



- **Nádory ledvin:**

Wilmsův tumor – postihuje děti do 5. roku života včetně fetálního období

WAGR syndrom (mikrodelece na chromzomu 11 *WT1* and *PAX6* geny)

Wilms tumor

Aniridia

Gonadoblastomas

Retardation

Denys-Drash syndrom - 90% pravděpodobnost vzniku Wilmsova tumoru

Anomálie močovodů

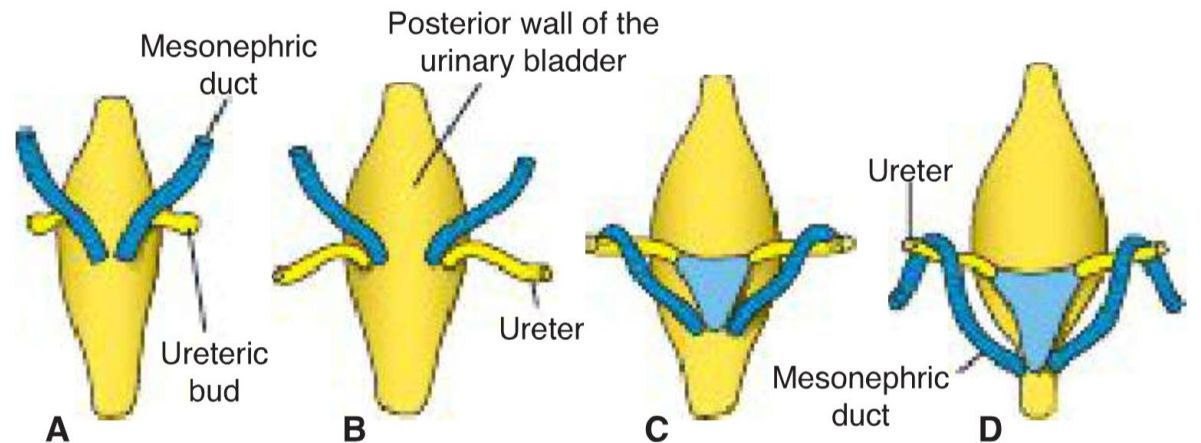
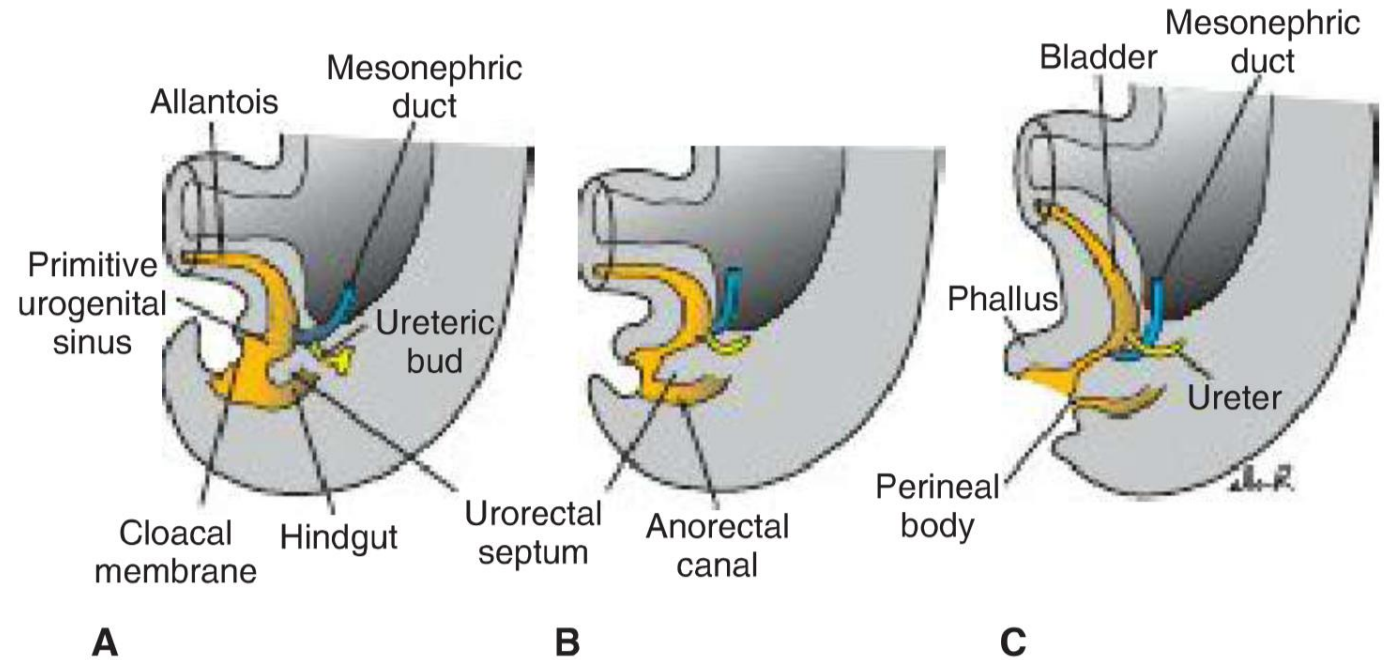
- *Ureter duplex* – rozštěpem ureterového pupenu
- Ektopický ureter – vznikem 2 ureterových pupenů
- Megaloureter (megaureter, hydroureter): < 7mm; 1:2 ♀ vs ♂; 1:2 p vs l
 - obstrukční
 - s VUR
 - VUR bez obstrukce
 - primární



Močový měchýř a urethra

Sinus urogenitalis:

- *pars vesica*
- *pars pelvica*
- *pars phallica*

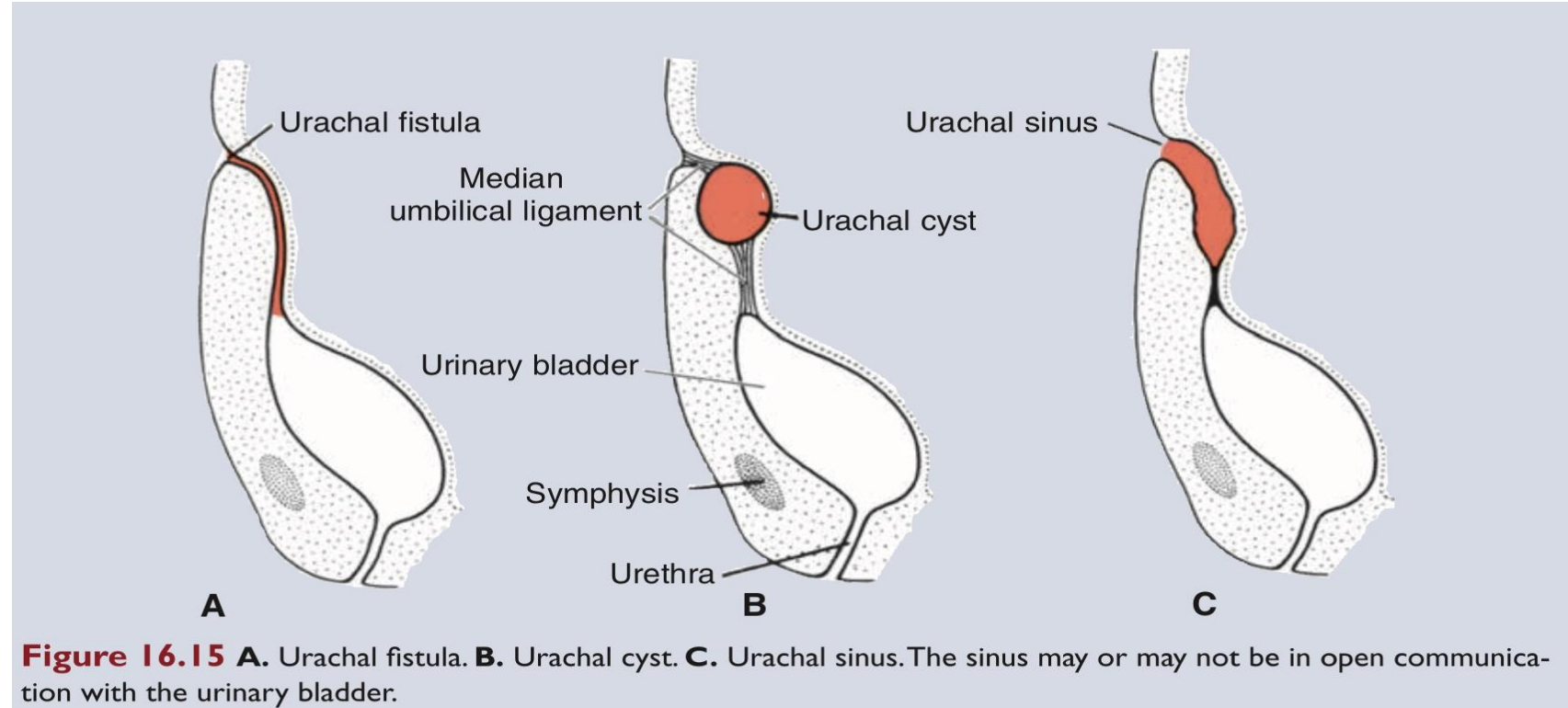


- Anomálie urachu

- fistula

- cysta

- sinus



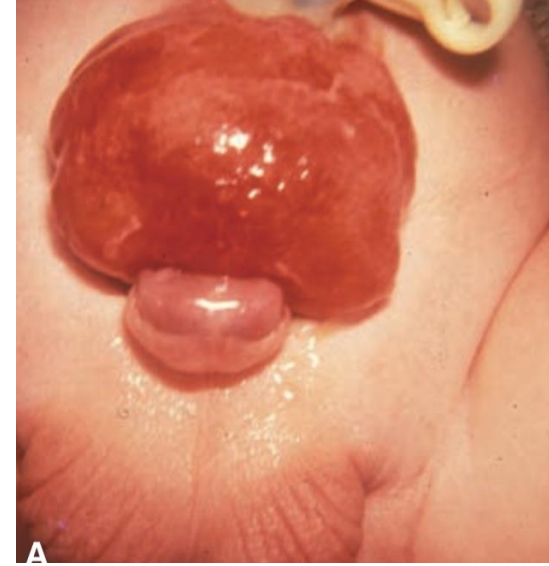
T.W. Sadler, Langman's medical embryology, 12th edition

- Anomálie rozdělení kloaky

- kolonovaginální fistula

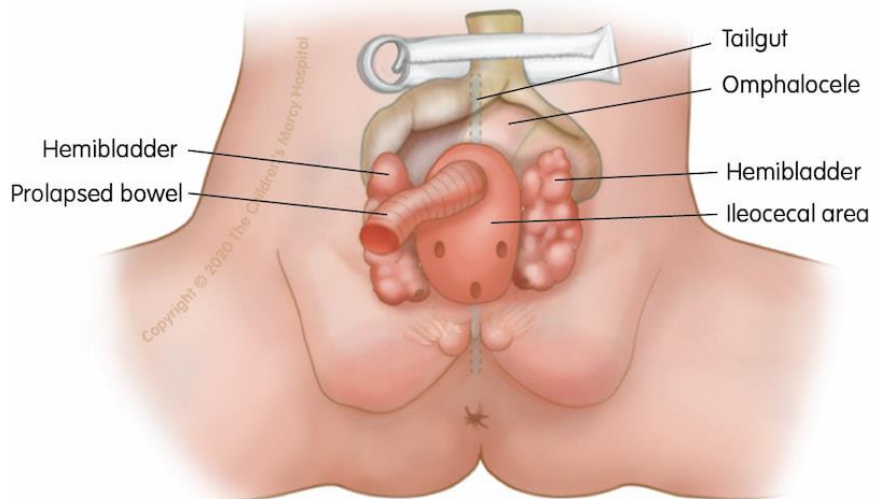
- kolonovezikulární fistula

- Defect přední tělní stěny
- extrofie močového měchýře: 1/50000, u ♂ s epispadii
- extrofie kloaky: 1/200000



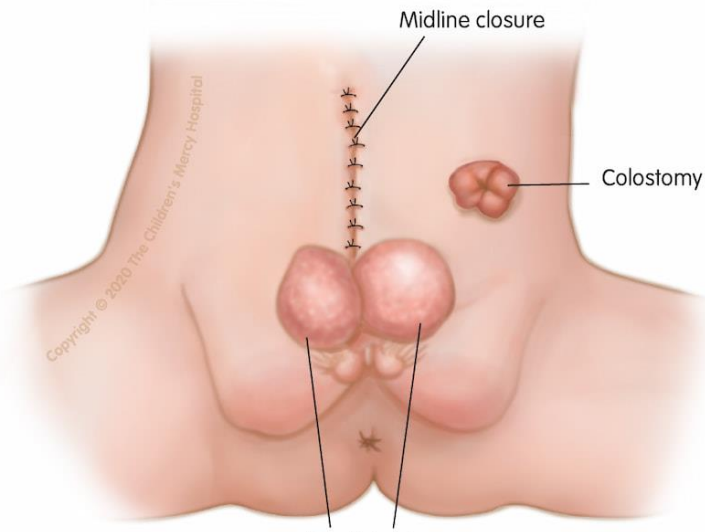
T.W. Sadler, Langman's medical embryology, 12th edition

CLOACAL EXSTROPHY



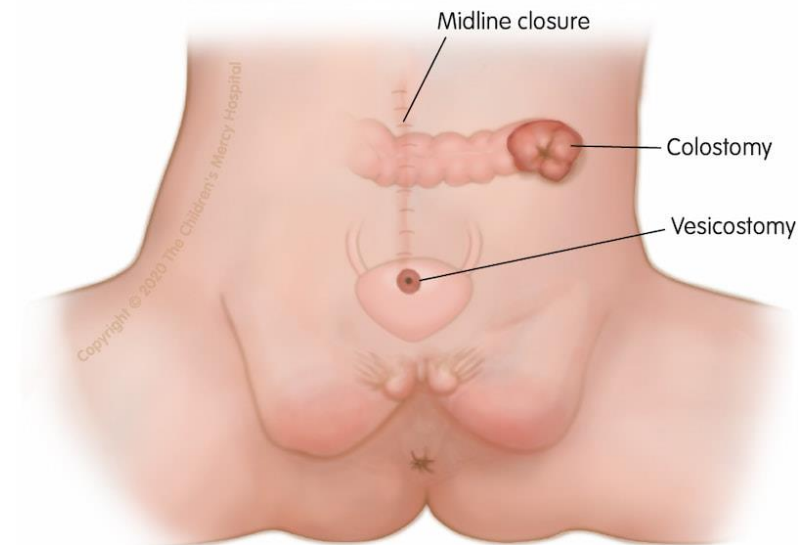
COMPREHENSIVE COLORECTAL CENTER

PRIMARY OMPHALOCELE CLOSURE



COMPREHENSIVE COLORECTAL CENTER

COMPLETE CLOSURE WITH COLOSTOMY AND VESICOSTOMY



COMPREHENSIVE COLORECTAL CENTER