Normal and abnormal development of the urogenital tract

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Normal development of the urogenital tract

- anatomy of development
- regulating genes involved in the development of the urogenital tract
- research of the congenital anomalies
- to get known and understand the embiogenesis
- stopped development in mice by driven "nil" mutation

Beginning of life and role of the urological tract

- intrauterine life (human fertilization)
- human gestation (fusion of spermatozoon, oocyta)
- development of organs and form of the foetus (10 weeks)
- expansion of the embryo, enlargement, maturation(40th week)
- role of the upper urinary tract
 - regulate secretion of nitrogenous waste products
 - homeostasis of water and electrolytes
 - acid base balance
 - the production of hormones

<u>Human embryonic disc</u>



P.M. Cuckow, P. Nyirády: Embryology and Pathophysiology of the Kidneys and Urinary Tracts in Pediatric Urology, W.B. Saunders, 2001.

Development of head and buccopharyngeal, tail and

<u>cloacal membrane</u>



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Embriogenesis of the kidney (nephrogenesis) I.

- three paired `kidneys' in the mammalian embryo: (intermediate mesoderm of the dorsal body wall) arises sequently
- the pronephros (22nd day)
- the pronephros and mesonephros degenerate during mammalian fetal life
- whilst the metanephros develops into the adult kidney
- in contrast, the pronephros: functioning kidney in the adult hagfish
- in some amphibians the mesonephros is the excretory organ in adult lampreys, some fishes and amphibians

AGENESIA, HYPOPLASIA RENIS

Embriogenesis of the kidney (nephrogenesis) II.

- 26th day pronephric duct arises and elongates (caudally) and reaches cloacal membrane arises
- this duct is renamed as Wolffian duct
- at 24th day of gestation <u>mesonephros</u> develops
- mesonephric tubules develops with "mesenchymal to epithel" transformation (40 tubules); glomeruli, proximal and distal collecting tubules (up to 6-10. weeks)
- cranial part (epigenital), caudal part (paragenital)

5th week



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Embriogenesis of the kidney (nephrogenesis) III.

- metanephros (consist of only 2 cell types)
- ureteric bud (epithelial cells)
- metanephric mesenchym (mesenchymal cells)
- at 28th day ureteric bud arises (Wolffian duct's postero-medial

aspect of mesonephric duct as a diverticulum)

- 32nd day reaches the metanephric blastema (elongates and tip)
- series of reciprocal interactions to branch multiple times

HYPOPLASIA RENIS, REN MULTICYSTICA

Embriogenesis of the kidney (nephrogenesis) IV.

induction of the ureteric bud: 4 major calyces, 6-8 generation

fuse to form minor calyces, final 8 definitive collecting duct system

mesenchymal induction: proximal convoluted tubes, loop of

Henle, distal convoluted tubes (glomerulogenesis: 8-9. week)

- nephrogenesis 40. week
- angio-, vasculogenesis (arteria iliaca)

HYPOPLASIA, REN DEGENERATIONEM MULTICYSTICA VASCULARIS ATYPIA

<u>5. hét</u>



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Ascend of the kidney and filtration

- Iobulated kidney ascends from its pelvic position during the 6-9.
 weeks
- blood supply from the aorta (lower branches degenerates)
- pelvic kidney faces anteriorly and rotates medially
- foetal urine is produced from the 10th week (plasma filtrations little modified)
- tubular function develops after the 14th week
- the foetal kidneys provide over 90% of the amniotic fluid

(adequate volume to move freely, lung and skeletal development)

UNROTATED KIDNEY, DYSTOPIC KIDNEY, OLIGOHYDRAMNION

8th week



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<u>Stephen- Mackie rule</u>



HYPOPLASIA v. DYSPLASIA

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<u>Weigert-</u>

<u>Meyer rule</u>

REN DUPLEX

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Hypoplasia Re

Ren dystopicus sacralis





Ren arcuatus



Pyeloureteral obstruction Praehydronephrosis





Normal development of the urinary bladder I.

- ingrowth of mesoderm point of confluence of the allantois and the hindgut
- partitioning of the cloaca: (around 28th day) by an advancing septum
- septum aided by the in-growth of lateral Rathke's folds
- cloaca is divided into an anterior primitive urogenital sinus that receives the mesonephric ducts, and a posterior rectum
- septation is completed urorectal septum reaches cloacal membrane (6.w)
- urogenital membrane breaks down (7th week)
- continuity between the developing urinary tract and the amniotic cavity.
- primitive urogenital sinus: between the allantois and the mesonephric

ducts called the vesicourethral canal form the definitive bladder.

5th week



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Normal development of the urinary bladder II.

- inductive interactions between endoderm of the urogenital sinus
 endoderma: urothelium
- mesenchyma: detrusor smooth muscle and adventitia
- Growth of the anterior abdominal wall between the allantois and urogenital membrane is accompanied by an increase in size and capacity
- at 13th week circural and longitudinal muscles are visible on trigone
- at 16thweek: continentia? (inner and outer longitudinal and circular detrusor smooth muscle)
- 21st week urothelium is developed

8th week



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Development of the ureterovesical junction

- ureteric orifices are incorporated into the developing bladder
- become separated from the mesonephric duct orifices
- further incorporation of more proximal mesonephric ducts the ureteric orifices move superolaterally relative to the mesonephric duct orifices
- These stay close to the midline and descend into the developing posterior urethra
- 9th week smooth muscle of the ureters develop (urine production)
- 18th week PU and UV obstruction

Ureterocele







Malformations of the urinary bladder

US: bladeer 11-12nd week Dg.: (extrophia 20th week)

Agenesia: hydronephr., dysplasia

<u>Hypoplasia:</u>

<u>Megacysta - megaureter</u> VUR

Duplex: obstruction?

<u>complete:</u> separate urethra (penis, uterus, vagina)

partial: 1 urethra

Malformations of the urinary bladder II.

Urachus: opened, cysta, sinus, diverticulum.



D

Malformations of the urinary bladder III.

Extrophia: epispadiasis complete reconstruction. symphysealysis fecal incontinenece tersticular retention



Exstrophia vesicae urinariae





bladder exstrophia

Cloacal exstrophia

Development of the external male genitals



Circumcision piramis from Sakara (b.c. around 2200)



Medici Katalin, II. Henrik és Diana de Poietiers

<u>Chordee</u>

- Galen (130-199) doctor of the gladiators
- "Hypospadias": infertility, curvature of the penis (1500 years)
- Oribasus (325-403) penile curvature sexual intercourse is impossible
- Mettauer (1842) U.S. : "succession of subcutaneous incisions until the organ is liberated" - 1967 D.R. Smith

Urethral reconstruction

- Antyl (I. sz.) és Paul of Aegina (625-690) amputation
- (1000 years)
- more than 300 methods

E. Durham Smith: The history of hypospadias, Pediatr Surg Int (1997) 12: 81-85

Development of the external male genitals



Development of the penis



Anatomy of the normal penis



Penile congenital anomalies

Meatal stenosis: obstruction: acute indication for operation

Frenulum breve:

Phimosis: - obstruction

- after the age of 2 years, opus

Penile curvatura:

hypospadiasis sine hypospadia Prostaglandin probe fascia-dyspl.

chordectomia corporoplastica

short urethra?

<u>Urethral problems II.</u>

Hypospadiasis penis

- place of the meatus
- chordee (adolescent)
- estetical lookeing
- Acute operation: meatal

stenosis!

Dg.: curvature

Time: 1/2 - 3 years



<u>Hypospadiasis</u>







Duplex urethra

Megalourethra: no corpus spongiosa

Paraurethr. cysta:

Cowper, utriculus prost.

Endoscopic incision



Urethra duplex



Epispadiasis: gland episp.

1:100.000





- agenesia
- aplasia
- hypoplasia
- retentio testis
- intraabdominal abdominoinguin. inguinal inguinoscrotal ingahere - fusional disorder of

testis and epididymis

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Developmental disorder of the testes II.



THANK YOU FOR YOUR ATTENTION!

