



Þvagfærasjúkdómar barna

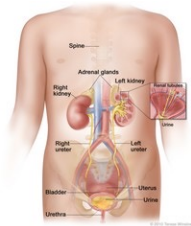
Þóra Sif Ólafsdóttir MD, FEAPU
 barnaskurðlæknir og þvagfæraskurðlæknir



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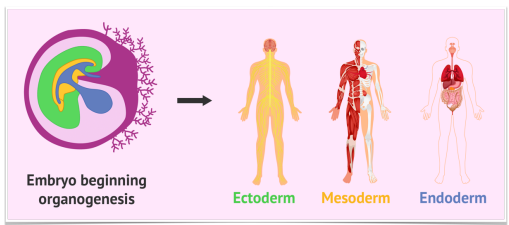
Hvað ætlum við að fjalla um?

- Antenatal Hydronephrosis
- UPJ
- PUV
- Megaureter
- VUR
- Hypospadiur



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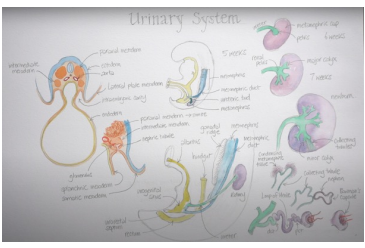
Embryology



Embryo beginning organogenesis → Ectoderm Mesoderm Endoderm

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Urinary System

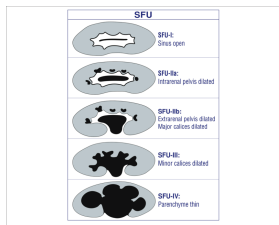


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Antenatal Hydronephrosis

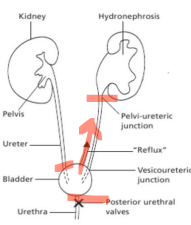
- 1% af öllum meðgöngum
- Uni vs bilateral
- Hydroureter
- Útítt blöðru
- Kyn
- Fósturvátn



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Etiologia ANH

Etiology	Incidence
Transient hydronephrosis	41–88%
UPJ obstruction	10–30%
VUR	10–20%
UVJ obstruction/megaureters	5–10%
Multicystic dysplastic kidney	4–6%
PUV/urethral atresia	1–2%
Ureterocele/ectopic ureter/duplex system	5–7%
Others: prune belly syndrome, cystic kidney disease, congenital ureteric strictures and megalourethra	Uncommon



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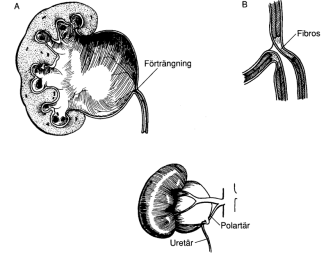
Greiningaraðferðir eftir fæðingu

- Ómskoðun
- Ísótóparannsókn
 - MAG 3 – Mercaptoacetyltriglycide
 - DMSA – Dimercapto succinic acid
- MUCG
- (Cystoscopy med retrograde skuggaefnisrannsókn)
- (CT/MR)

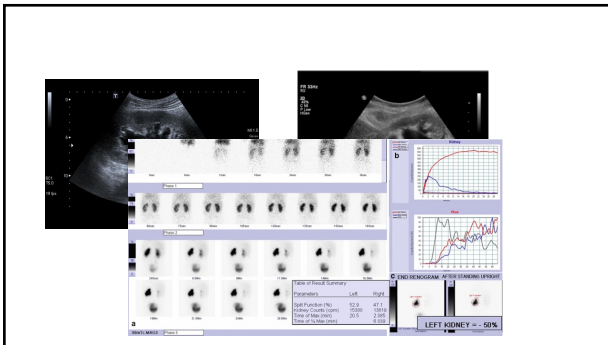
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Ureteropelvic Junction Stenosis

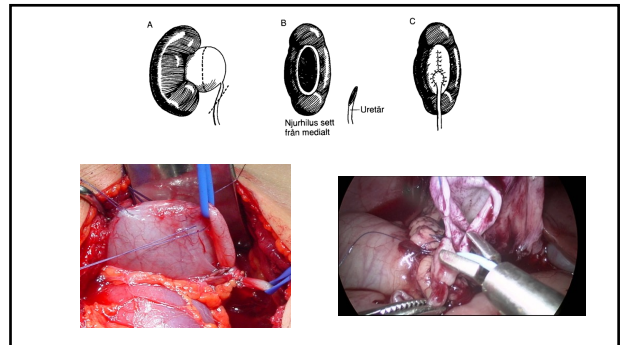
- 1:1500
- M>F = 2:1
- Intrinsic vs extrinsic
- Ómun
- MAG -3
- Einkenni +/-



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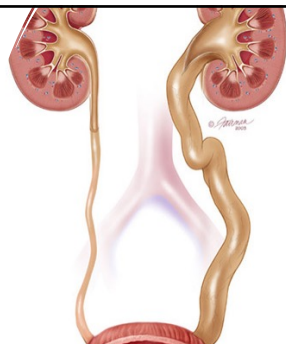
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Megaureter

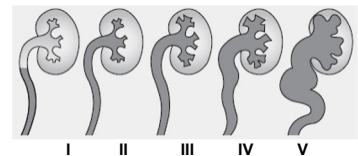
- Þvermál >7mm við blöðru
- Vesicoureteral junction stenosis (VUJ eða POM)
- Reflux
- Duplex með ectopy



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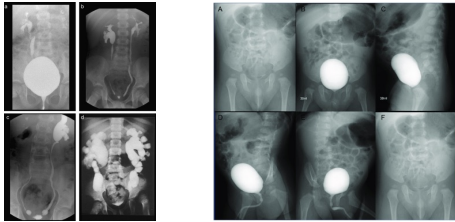
Vesikoureteral reflux (VUR)

- Tíðni 1-2%
- Orsök hjá ca 30% af börnum með UVI
- Mun algengara hjá stúlkum
- Þvag bakkar upp þvagleiðara til nýrna
- <4 árs álder
- Gráða 1-5 eftir MUCG



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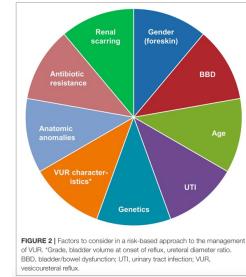
Micturitionsurethrocytogram



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Meðferð

- Observation
- Fyrirbyggjandi sjúkalyf
- Endoskopísk meðferð (Deflux) III-V
- Reimplantation (failed Deflux eða gr V)
- Aðalmarkmið er að koma í veg fyrir nýrmaskaða



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Vesikoureteral reflux

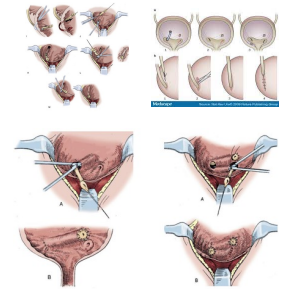


Deflux® (dextrankúlur í hyaluransýra) lækna VUR í 80% tilfella

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Re-implantation

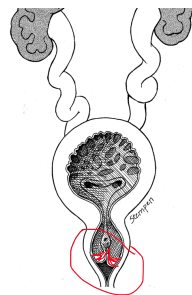
- Cross trigonal reimplantation
- Leadbetter politano
- Lich Gregoir



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Urethral valves - PUV

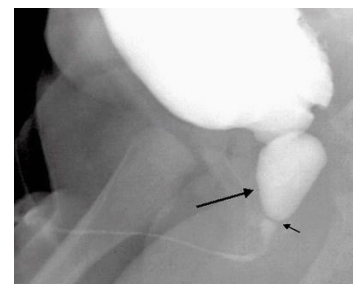
- 1:5000 drengjum
- Bladder outlet obstruction
- Bilateral hydronephrosis
- Nýrmaskaði/dysplasia
- Detrusor hypertrofia
- Oligohydramnion - Lungnávænþroski



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Posterior Urethral Valves - einkenni

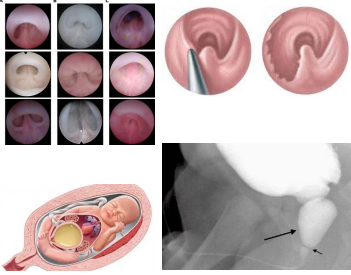
- Öndunarerfiðleikar
- Fyrirferð neðri hluta kviðar - Ascites
- Erfiðleikar við þvaglát - síðþ þuna
- Sýkingar
- Failure to thrive



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Bráð meðferð PUV

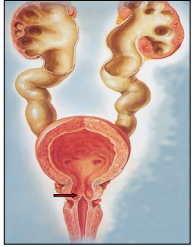
- Létta á þvagblóðru
- SP/þvagleggur
- Nephropyelostomia?
- Endoskopísk aðgerð
- Obs Polyuria



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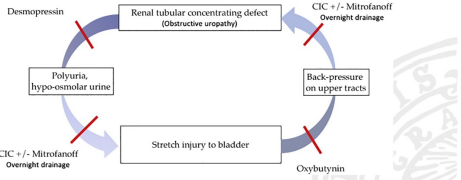
Snemmbúnir fylgikvillar PUV

- Þykkveggja blaðra leyfir ekki tæmingu ureters
- NP
- Ureterostomia
- Vesikostomia
- Pop off mekanismi – ónýtt nýra
- Sýkingar
- nefrektomí



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Síðbúnir fylgikvillar PUV



Hypothesized feedback loop in the deterioration of boys with PUV

A.E. Wright et al. / Journal of Ped Surgery 53 (2018) 316-320

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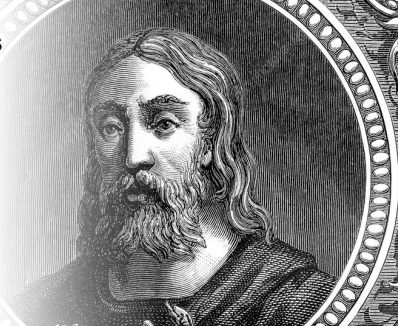
Eftirlit – síðbúin meðferð

- Venja af bleiju snemma - fylgjast vel með þvaglátum
- 4 t þvagláta obs
- Flæðimælingar/restþvag
- 20-30% nýrnabilun
- Lyfjameðferð: antikolinergika, alfablocker
- CIC/þvagleggur yfir nótt
- Mitrofanoff kanall
- Mickeyhnappur til tæmingar

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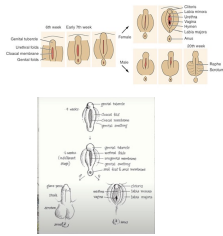
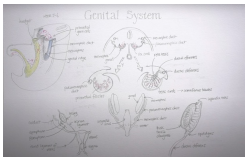
Hypospadias - Neðanrás

- Gríska
- "Hypo" och "spadon"
- Galen 130-199 AD



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Myndun ytri kynfæra

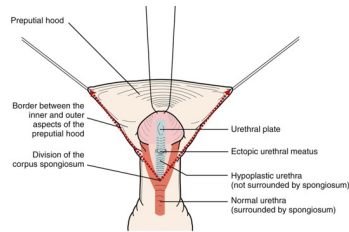



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Anatómía

- Opin forhöð með hettu dorsalt
- Klofinn glans
- Urethraplata opin
- Ventral kurvatur



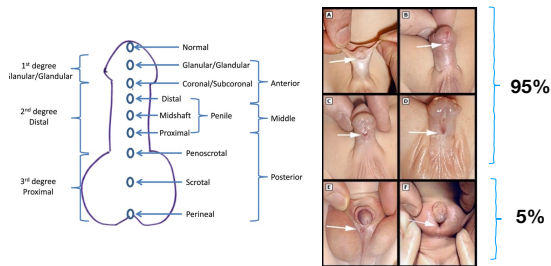
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Incidens och etiologi

- 1/125 nyfödda pojkar i Sverige
Nordenfjord et al. Population-Based Nationwide Study of Hypospadias in Sweden, 1973-2009. Pediatrics and Child Health. 11(2015): 167-170-6
- 300 per år i Sverige
- Risk faktorer:
 - Närsläkt med hypospadi
 - Låg födelsevikt eller liten placenta
 - Tvillingar
 - Hög blodtryck eller pre-eclampsia i mor
- Endocrine disrupting factors?
- Vissa syndromer t.ex.
 - Optiz B
 - Denys-Drash
 - VACTERL

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Flokkun



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General principles of treatment

- Timing of surgery
- Penil hypoplasia – hormon behandling?
- Associerade anomalier – retentio testis
- Kirurgisk teknik



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Surgical principles

- Correction of ventral curvature
- Reconstruction of the missing urethra with adequate calibre
- Glanuloplasty and sometimes spongioplasty
- Skin covering: elongation or reconstruction of the short or missing ventral skin

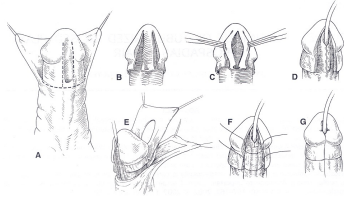
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Operationer

- Förhudsplastik/circumcision*
- MAGPI*
- TIP – Snodgrass*
- Mathieu
- Onlay Island flap
- Tubularized preputial Island flap (Duckett)
- STAG – Bracka*
- Koyanagi – Nonomura
- Hadidi
-
-

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TIP - Snodgrass



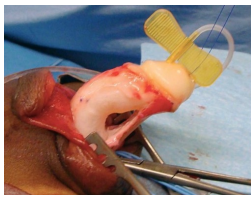
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TIP - Snodgrass



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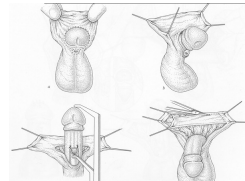
Erektionsprov



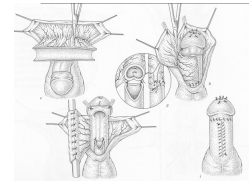
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Island flap or free transplant

STAG



Duckett



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STAG - Bracka



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HOPE/HOSE/(PPS)

- Valideruð instrument
- Post op kontroll:
- Meatus lega
- Glans meuffit
- Húð á skaffi
- Framsveigja

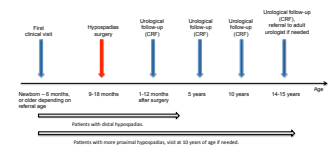


Figure 14: The Swedish national surveillance program for hypospadias. Adapted from Swedish Healthcare program for hypospadias.

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Take home message

Bilateral hydronephrosa hjá dreng – PUV?
Þvagbuna?, Þvagleggur og MUCG skjótt

Unilateral hydronephrosa – hvernig lífa
þvagleiðarar út? UPJ vs POM eða VUR

VUR – einstaklingsmiðuð meðferð, kir
meðferð III-V, meðhöndla BBD samtímis

Löng hypospadia og retentio testis –
merki um undirviriliseringu, DSD?