ACEU
Hydronéphroses et Méga-uretères

DIU Urologie Pédiatrique 2013

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Plan

1. Hydronéphroses

2. Méga-uretères
Hydronephrosis & PUJ Obstruction

Where do we stand?

SIU World meeting. BERLIN 2011

Prof. Marc-David LECLAIR
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What is hydronephrosis?

- Pathological dilatation of the renal pelvis and renal calyces
- A radiological sign

Several aetiologies:
- High urinary output
- VUR
- Developmental abnormalities of upper-tract
- Renal dysplasia
- Obstruction
Hydronephrosis: the two challenges

- Prove the presence of an obstruction
- Manage the population descended from the prenatal screening
What is Obstruction?

- Dilatation does not always mean Obstruction

- Dilatation may result from
  - morphological variations in conjunction with high urine output
  - dystrophia: sequellae of prenatal obstruction
  - active obstruction
What is Obstruction?

- « Some impediment to the flow of urine... which, if left untreated, would cause progressive damage to the kidney »

- Requires serial observations with both morphological and functional studies
PUJ obstruction

- Discuss
  - aetiologies
  - diagnostic procedures
  - management options

of unilateral isolated hydronephrosis

- suspected to be PUJO
- excluding bilateral hydronephrosis
- excluding ureteric dilatation
Aetiologies of PUJO

- **Intrinsic obstruction**
  - stenotic segment
  - hypoplastic adynamic segment (normal calibre)
    - decrease smooth muscle cells
    - increase collagen fibres
Aetiology of PUJO

- Intrinsic obstruction
- Extrinsic obstruction
  - Fibrous bands, kinks, ureteric folds
    - more frequent in older children?
    - spontaneous resolution with growth as ureter straightens?
Aetiology of PUJO

- **Intrinsic obstruction**

- **Extrinsic obstruction**
  - Fibrous bands, kinks, ureteric folds
  - Aberrant lower-pole crossing vessels
    - lying anterior to the pelvis
    - causing external anterior compression
    - variable incidence according to population selected
      - prenatal: 5%
      - symptomatic HN: 30-70%
The obstructive role of crossing vessels?

- CV can be associated with intrinsic PUJ obstruction
  ...but being barely obstructive by themselves
  - when performing surgery: relocate the CV posteriorly

- CV can be the only cause of obstruction

- CV could induce secondary intrinsic stenosis?
Imaging modalities - US

- Renal US is the cornerstone imaging modality in the diagnostic pathway of hydronephrosis

- SFU grading of hydronephrosis
  1. mild dilatation of the pelvis

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  1. Mild dilatation of the pelvis
  2. Large dilatation of the pelvis
     Pelvis remains intrarenal

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• SFU grading of hydronephrosis
  1. mild dilatation of the pelvis
  2. large dilatation of the pelvis pelvis remains intrarenal
  3. major dilatation of pelvis & calyces pelvis extrarenal

Imaging modalities - US

- Renal US is the cornerstone imaging modality in the diagnostic pathway of hydronephrosis

- SFU grading of hydronephrosis
  1. mild dilatation of the pelvis
  2. large dilatation of the pelvis
     pelvis remains intrarenal
  3. major dilatation of pelvis & calyces
     pelvis extrarenal
  4. major dilatation
     + parenchymal thinning

The most useful parameter is the antero-posterior (AP) pelvic diameter at the level of the renal sinus:
- 18th - 23rd WG > 7 mm
- 3rd trimester > 10 mm
- Birth > 12 mm

Abnormal
Imaging modalities - US

- The most useful parameter is the antero-posterior (AP) pelvic diameter at the level of the renal sinus
  - 18th - 23rd WG > 7 mm
  - 3rd trimester > 10 mm
  - birth > 12 mm

- The degree of dilatation of the calyces
Radiopharmaceutical agent: $^{99}$Tc-Mercaptoacetyltriglycerin
- high protein binding
- high tubular excretion
- low distribution in extra-vascular space

Serial dynamic acquisition: time / activity curve
- two distinct phases:
  - uptake phase: differential renal function
  - clearance phase: urine washout through the collecting system
Isotope imaging - Mag-3

Renal scintigraphy

Time activity curves
Regions of interest around the kidney with and without background subtraction

Rutland-Patlak plot: rate of uptake by the kidney from the 1st to the 2nd minute
Isotope imaging - Mag-3

- Poor washout: Obstruction or delayed emptying?
Drainage depends on:

- **How much urine is produced?**
  - renal function
    - a poorly functioning kidney will produce less urine than a normal kidney
    - a poorly functioning kidney will clear the isotope slower from the blood
  - hydration status
    - if dehydrated, less & more concentrated urine is produced

- **How big is the collecting system?**
  - a large pelvis will drain more slowly than a small

- **The anatomy & physiology of the ureter and the bladder**
Drainage & Obstruction

- Poor drainage at 20 min., in a child supine, does not necessarily mean obstruction
  - always get a post-micturition view

- Good drainage = no obstruction
- Obstruction =
  - Poor drainage AND decreased DRF or increased pelvic AP diameter
Imaging modalities - MRI

- Theoretically combines
  - detailed anatomical description
  - functional evaluation
Prenatally diagnosed HN
Prenatally diagnosed HN

- **Routine antenatal US screening**
  - urological abnormalities 1:500
  - hydronephrosis: 50%

- **Potential for spontaneous resolution of prenatally diagnosed HN**
  - a unique population of asymptomatic healthy newborns

  - dilatation increase
    - functional deterioration $\rightarrow$ Obstruction

  - dilatation stable
    - or improving with time $\rightarrow$ ???
75% of prenatal unilateral hydronephrosis will remain stable or improve if managed conservatively

Management strategy

- 75% of prenatal unilateral hydronephrosis will remain stable or improve if managed conservatively [Koff. J Urol 2000; 164: 1101]

- The risk of deterioration correlates with the initial degree of dilatation of pelvis and calyces

<table>
<thead>
<tr>
<th>AP diameter</th>
<th>surgery</th>
</tr>
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<tbody>
<tr>
<td>&lt; 20 mm</td>
<td>11 %</td>
</tr>
<tr>
<td>20-30 mm</td>
<td>40 %</td>
</tr>
<tr>
<td>30-40 mm</td>
<td>90 %</td>
</tr>
<tr>
<td>&gt; 40 mm</td>
<td>≈ 100%</td>
</tr>
</tbody>
</table>

Dhillon HK. BJU 1998; 81: 39
& Dhillon HK, unpublished data
Management strategy

- 75% of prenatal unilateral hydronephrosis will remain stable or improve if managed conservatively  

- The risk of deterioration correlates with the initial degree of dilatation of pelvis and calyces  
  Dhillon. *BJU* 1998; 81: 39

- It is safe to follow-up with renal US alone and perform MAG-3 if hydro worsens  
  Ransley. *J Urol* 1990; 144: 584
Management strategy [2]

- Management of hydronephrosis with impaired DRF is more debated:
  - if managed conservatively, a significant proportion will improved spontaneously
    Koff. *J Urol* 1994; 152: 593
  - risk of progressive deterioration
  - ultimately, surgery may not impact long term DRF
  - if managed conservatively, these children require serial MAG-3 follow-up
Management strategy [3]

Unilateral Hydronephrosis
- no ureter seen
- normal bladder

1 month
- renal US
- VCUG if ureters seen

3 months
- renal US
- Mag-3 renal scan

Impaired
differential renal function
(DRF < 40%)

Preserved
differential renal function
(DRF > 40%)
Management strategy [4]

- **DRF < 40%**
  - **pyeloplasty**
    - US @ 3 months postop
    - US + MAG-3 @ 1 year postop

Impaired DRF
Management strategy [4]

DRF < 40%
- pyeloplasty
  - US @ 3 months postop
  - US + MAG-3 @ 1 year postop

DRF < 10-15%
- repeat MAG-3 @ 9-12 months
  - DRF improved
    - follow-up
  - DRF low
    - nephrectomy
      systematic or if complications?
Pelvis AP diameter:
- 15-20 mm
- 20-30 mm
- > 30 mm
Management strategy [4]

If major calyceal dilatation or early prenatal diagnosis

pelvis AP diameter 15-20 mm

follow-up

Renal US @ 1 year
Renal US @ 2 years
Renal US @ 5 years
Then discharge

pelvis AP diameter 20-30 mm

pelvis AP diameter > 30 mm

Normal DRF
Management strategy [4]

pelvis AP diameter
15-20 mm

follow-up

renal US @ 1 year
renal US@ 2 years
renal US@ 5 years
then discharge

pelvis AP diameter
20-30 mm

If major calyceal dilatation
or early prenatal diagnosis

pelvis AP diameter
> 30 mm

pyeloplasty

Normal DRF
Management strategy [4]

pelvis AP diameter

15-20 mm

follow-up

renal US @ 1 year
renal US @ 2 years
renal US @ 5 years
then discharge

pelvis AP diameter

20-30 mm

follow-up

renal US @ 6 months
renal US + MAG-3 @ 1 year
renal US @ 18 months
renal US @ 2 years
renal US @ 3 years
renal US @ 5 years
renal US @ 7 years
renal US @ 10 years
renal US at 15 years
...then discharge

pelvis AP diameter

> 30 mm

pyeloplasty

follow-up

MAG-3 if hydronephrosis increase

Normal DRF
Symptomatic hydronephrosis

• **Characteristics**
  – relatively infrequent
  – observed in children with/without prenatal diagnosis
  – febrile UTIs, stones, abdominal/loin pain

• **Intermittent acute hydronephrosis**
  – a specific clinical picture
  – older child & adolescent
  – acute intermittent loin pain + vomiting
  – intermittent hydronephrosis
    (renal US may be normal between episodes)

  – high incidence of lower-pole vessels: US, MAG-3,…angio-MRI
Surgical techniques

• Anderson-Hynes dismembered pyeloplasty remains the gold standard
  – retroperitoneal open surgery: lateral or posterior lumbotomy
  – MIS: transperitoneal laparoscopy, retroperitoneoscopy, robotic
Surgical techniques

Transperitoneal approach

- Suspension of PUJ by stay sutures
Surgical techniques

Transperitoneal approach
Surgical techniques
Surgical techniques

- Anderson-Hynes dismembered pyeloplasty remains the gold standard
  - retroperitoneal open surgery: lateral or posterior lumbotomy
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- «Salvage» procedures should be known
  - Culp-De Weerd pyeloplasty
  - uretero-calicostostomy
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• « Salvage » procedures should be known
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  – uretero-calicostostomy

• Vascular hitch procedure: selected indications
  – intermittent acute hydronephrosis + lower pole vessels without intrinsic obstruction
    Gundeti et al. J Urol 2008; 180: 1832
  – excellent long-term outcomes
    55 cases (31 transp laparoscopy - 24 robotic-assisted)
    Follow-up: 31 months [12-84]
    2 failures

Villemagne-T et al. ESPU 2013. Genova
The vascular hitch technique

Illustrations: S. Spitzer, in Sakoda-A et al. BJU Int 2011:1364-8
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Indications

- **Infants with prenatally diagnosed HN**
  - Use sensible criteria and stick to it!
  - Impaired function & significant HN
  - Normal function + large HN (threshold ?)
  - Normal function + progressing HN
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  - Use sensible criteria and stick to it!
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  - Normal function + progressing HN

  - **Posterior lumbotomy** + classic Anderson-Hynes pyeloplasty
Indications - Which approach?

- Infants with prenatally diagnosed HN
- Infants with initially observed HN
  - the threshold for posterior lumbotomy is high
  - how low is the threshold for a beneficial laparoscopy?
Indications - Which approach?

- Infants with prenatally diagnosed HN
- Infants with initially observed HN
- Older children w/w.o prenatal diagnosis
  - transperitoneal laparoscopy!
  - if CV + no intrinsic obstruction: Vascular hitch?
  - in any other case: laparoscopic pteloplasty
Hydronephrosis does not mean Obstruction

- Prenatally diagnosed HN / symptomatic HN
- «Stable» HN results from an equilibrium
  - between urine output & outlet resistances
  - ...that may change with time
Megaureters

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Pathophysiology

- **Urologist’s Hirschsprung disease?**
  - but normal ganglia distribution in distal ureter

- **Abnormal muscle fibers**
Pathophysiology

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Pathophysicsiology

- **Urologist’s Hirschsprung disease?**
  - but normal ganglia distribution in distal ureter
  
  Leibowitz-S. *J Clin Pathol* 1963;16:342

- **Abnormal muscle fibers**
  - hypertrophy circular muscle layer instead of longitudinal
  - distal muscular dysplasia
  - excess collagen type III deposition

  Hanna-MK. *J Urol* 1976;116:725

- **Segmental maturational dvp**
  - could be explained by spont. downregulation TGF-β

  Nicotina-PA. *Br J Urol* 1997;80:946
Obstructive Megaureters

- **Primary**
  - adynamic ureterovesical segment
  - excessive pattern of collagen deposition
  - altered cell-to-cell junctions
  - disrupted myoelectric propagation and peristalsis

- **Secondary to bladder wall modifications**
  - neurogenic (or non-neuro-) bladder dysfunction
  - infra-vesical obstruction: PUV
  - chronic UTIs / cystitis cystica
  - others
Refluxing Megaureters

- **Primary & Secondary refluxing MGU**
  - replacement of ureteric smooth muscle by collagen type III
  - predominantly in the distal ureter

- **Obstructive and Refluxing MGU**
  - primarily due to ureteric orifice ectopia
Non-obstructive & Non refluxing

- **Primary**: typical in newborns
  - multifactorial causes
  - high prenatal urine output
  - dyscoordinated voiding
  - excess of elastin and collagen (over-expression TGF-β)

- **Secondary**
  - altered peristalsis due to bacterial endotoxins
  - high urine output: nephropathy, diabetes insipidus, polydipsia...
Imaging

• Ultrasound

• MAG-3 renal scan

• (functional) -MRI
Ultrasound

Retrovesical ureter diameter
- Longitudinal -

Retrovesical ureter diameter
- Transversal -
Therapeutic options

- **Conservative : watchfull waiting**
  - under ATB-prophylaxis ?

- **Diversion**
  - Ureterostomy
  - Ureteral drainage : JJ stenting

- **Endoscopic treatment**
  - pneumatic VUJ endoscopic dilatation
    ± ureteric stenting ?

- **Ureteral reimplantation**
  - intra- / extra-vesical
  - with / w.o tapering
Conservative approach

- **Conservative management of primary obstructive megaureters**
  - high rates of spontaneous regression
  - low rates of complications

- **observational study**
  50 ureters, long-term follow-up: 50% **resolution rate**
  - 26 >10mm: 76% 60 months [18-204]
  - 24 >10mm: 17% 102 months [42-210]
  12% complications: 3 recurrent UTIs + function deterioration
    - 2 stones
    - 1 hypertension
    - all in group B

• Ureterostomy has long been a standard of treatment of severe complicated MGU in infants

Lettgen-B. *Br J Urol* 1993;72:826
Gearhart-J. *Br J Urol* 1994;74:133
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- What is the place for ureteric internal stenting?
Diversion

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  Gearhart-J. *Br J Urol* 1994;74:133

- What is the place for ureteric internal stenting?

  - 10 newborns / infants
  - 50% open insertion!
  - 70% complications (UTIs)...
  
  but (only) 50% open reimplant

  Castagnetti-M *Urology* 2006;68:870
Diversion

- Ureterostomy has long been a standard of treatment of severe complicated MGU in infants
  
  Lettgen-B. Br J Urol 1993;72:826
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- What is the place for ureteric internal stenting?

  - 19 ureters in infants
    - 66% open insertion!
    - 31% complications (UTIs)...
    - 56% drainage improved
  

  6/19 reimplanted
Diversion

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  Lettgen-B. *Br J Urol* 1993;72:826
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• What is the place for ureteric internal stenting?

  – *38 ureters in infants* 66% resolution
    Carroll-D *Urol Ann* 2010;2:114-8

  – *12 children* stented prosp. compared to 15 observed equivalent final outcome (50% reimplantation) 41% stent-related complications
    Barbancho-DC. *Cir Pediatr* 2008;21:32
Diversion

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• What is the place for ureteric internal stenting?

  - This strategy requires severe prognosis indicators to be defined
  - At best, it can be viewed as a method to temporize a surgical treatment
Endoscopic Treatment

- **Endoscopic pneumatic balloon dilatation**
  - 3-5mm, 2-10 min., 10-18 bar, ± JJ stenting 4-6 wks, double stenting?
Endoscopic Treatment

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  - **Barcelona experience**
    Romero-RM. ESPU 2012. Zurich
    29 children, F-up 47 months ±24 (all >18 months)
    5/29 secondary VUR
    5 reimplantations required (VUR :2, early comp : 2, persistent obstruction :1)

  - **Feasible in infants**
    Torino-G. *J Endourol* 2012;26:325
    5 infants - 3F catheter (4mm), 4.7F JJ 6-8 wks
    all improved on US
    no secondary VUR
Endoscopic Treatment

- **Endoscopic pneumatic balloon dilatation**
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    - all improved on US
    - no secondary VUR

  - **Personal series**
    - 14 primary obstructive MGU
    - 3/14 failures of insertion - high rate of stent related complications
    - 6/9 improved at >12 months
Surgical reimplantation

- Intra-vesical reimplantation: INFRA-hiatal
  - cross-trigonal Cohen reimplant
  - often requires ureter tapering or plication
Surgical reimplantation

• Supra-hiatal reimplant
  – Leadbetter-Politano + ureteral tapering

  – Psoas-hitch reimplant +++
    93 patients primary refluxing or obstructive MGU
    ureteral tapering 17/93
    98% improved hydro-ureteronephrosis
    16% postop VUR, 7% breakthrough UTIs

Rod-J. unpublished data
Indications

• Less and less interventional

• Symptomatic MGU
  – decreasing function
  – stones ?
  – urinary sepsis, breakthrough UTIs ?

  – Remember high rates of spontaneous resolution / low complication rates

• Symptomatic infants : buy time
  – circumcision, JJ stenting, dilatation ?
  – ureterostomy in (very) severe situations
So What?

- Few obstructive megaureters require surgical intervention
- Potential spontaneous resolution
- Endoscopic management successes may only reflect a (spontaneous) favourable outcome
- The place for endoscopic dilatation as a first-line treatment remains to be defined