ACEU

Hydronéphroses et Méga-uretères

DIU Urologie Pédiatrique 2013

Marc-David LECLAIR

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Plan

1. Hydronéphroses

2. Méga-uretères

Hydronephrosis & PUJ Obstruction

Where do we stand ?

SIU World meeting. BERLIN 2011

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CENTRE HOSPITALIER UNIVERSITAIRE DE NANTES

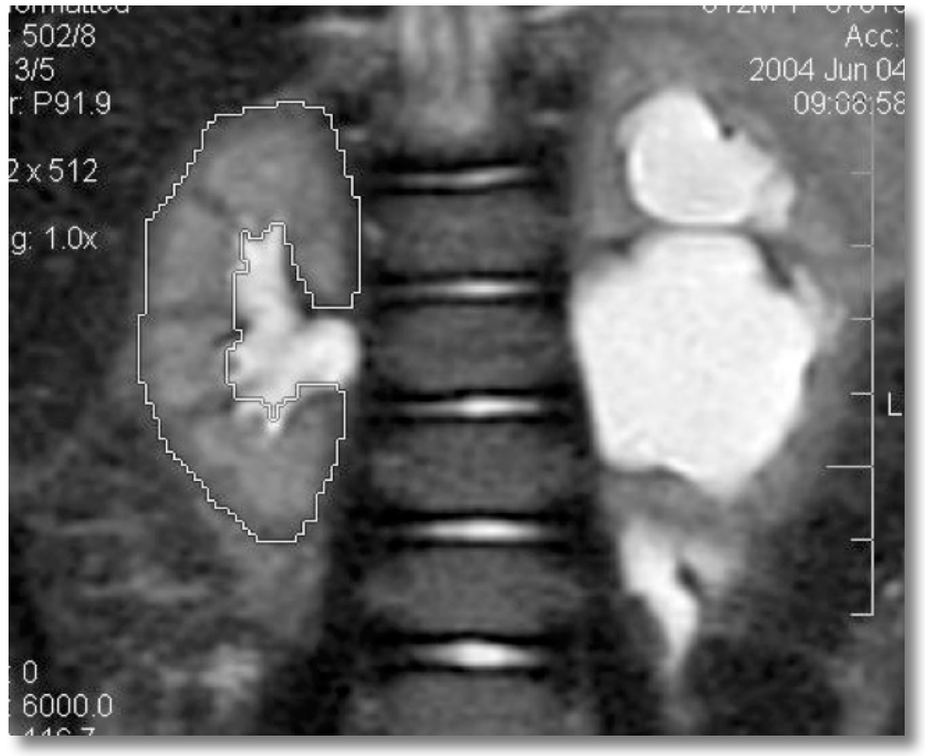
PEDIATRIC UROLOGY JUNIOR ORGANIZATION

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 outpu
  - 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

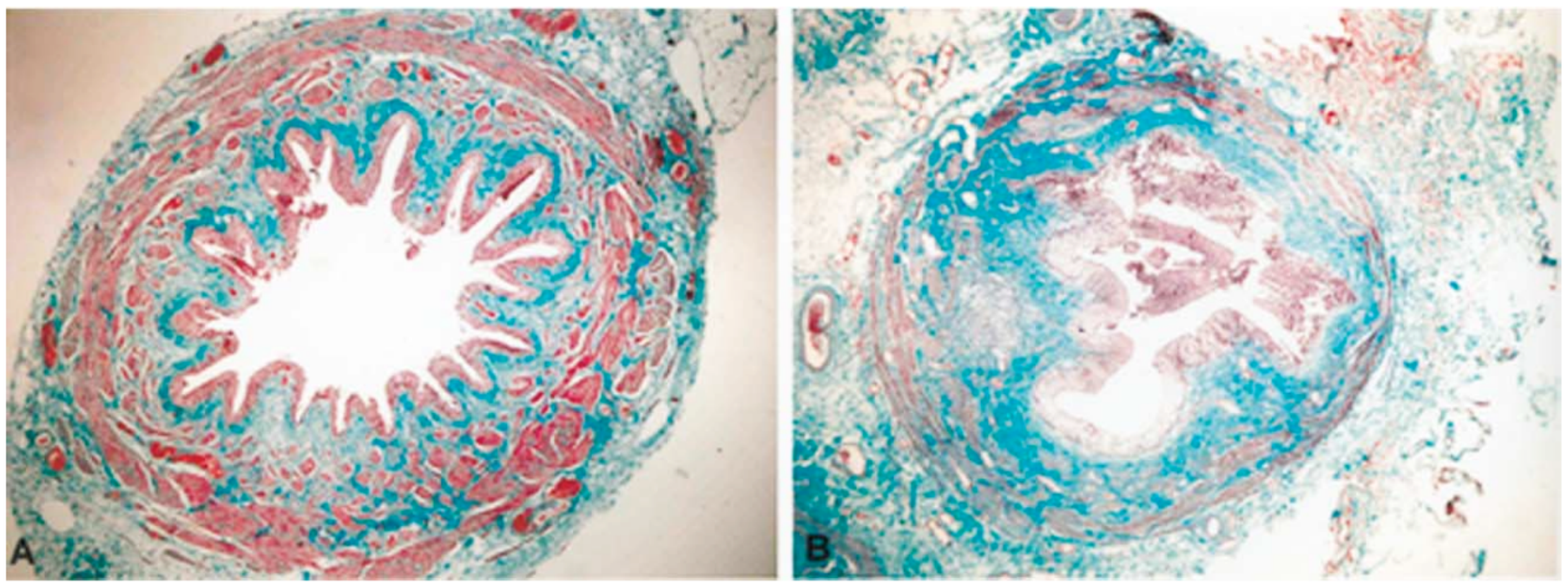
- **Discuss - aetiologies**
  - diagnostic procedures
  - management options

  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



Aetiologies of PUJO

- **Intrinsic obstruction**

- **Exstrinsic obstruction**
  - Fibrous bands, kinks, ureteric folds
    - more frequent in older children ?
    - spontaneous resolution with growth as ureter straightens ?

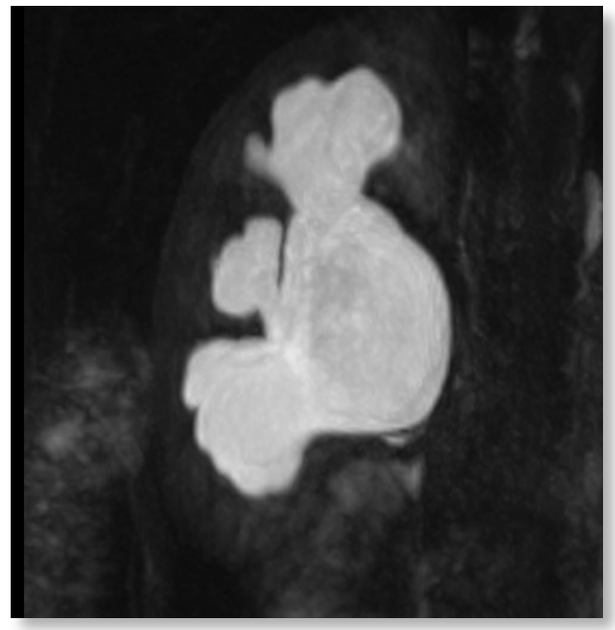
Aetiologies of PUJO

- **Intrinsic obstruction**

- **Exstrinsic 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

Maizels. *J Urol* 1992; 148:609

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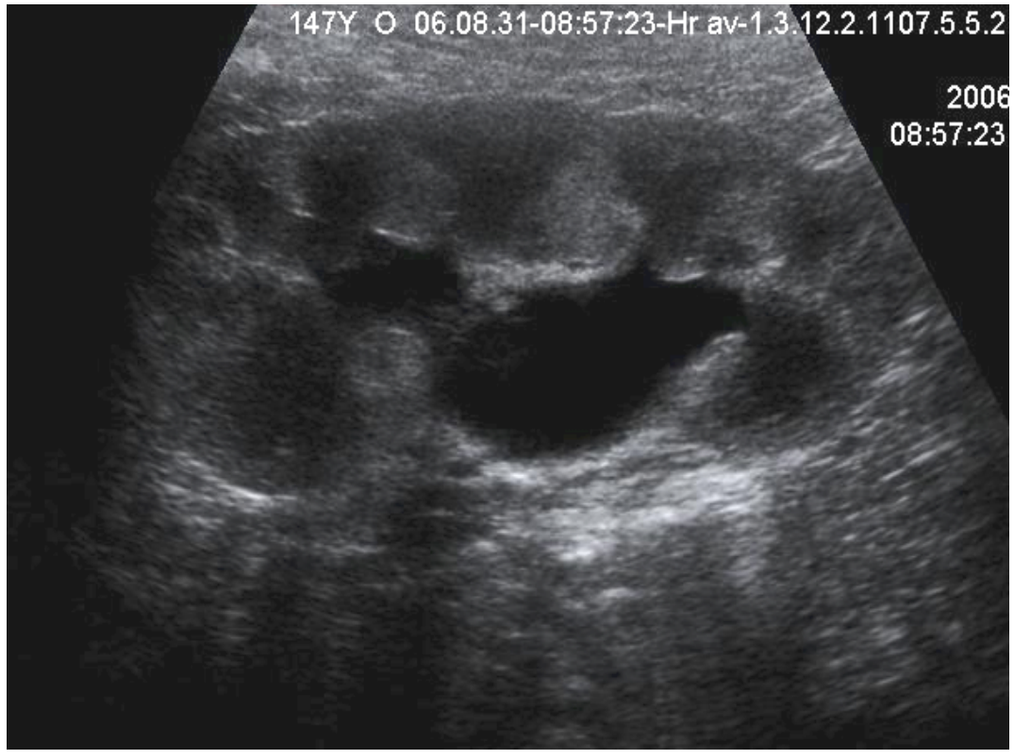
- **SFU grading of hydronephrosis**

  1. mild dilatation of the pelvis

  2. large dilatation of the pelvis
  pelvis remains intrarenal



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Imaging modalities - US

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

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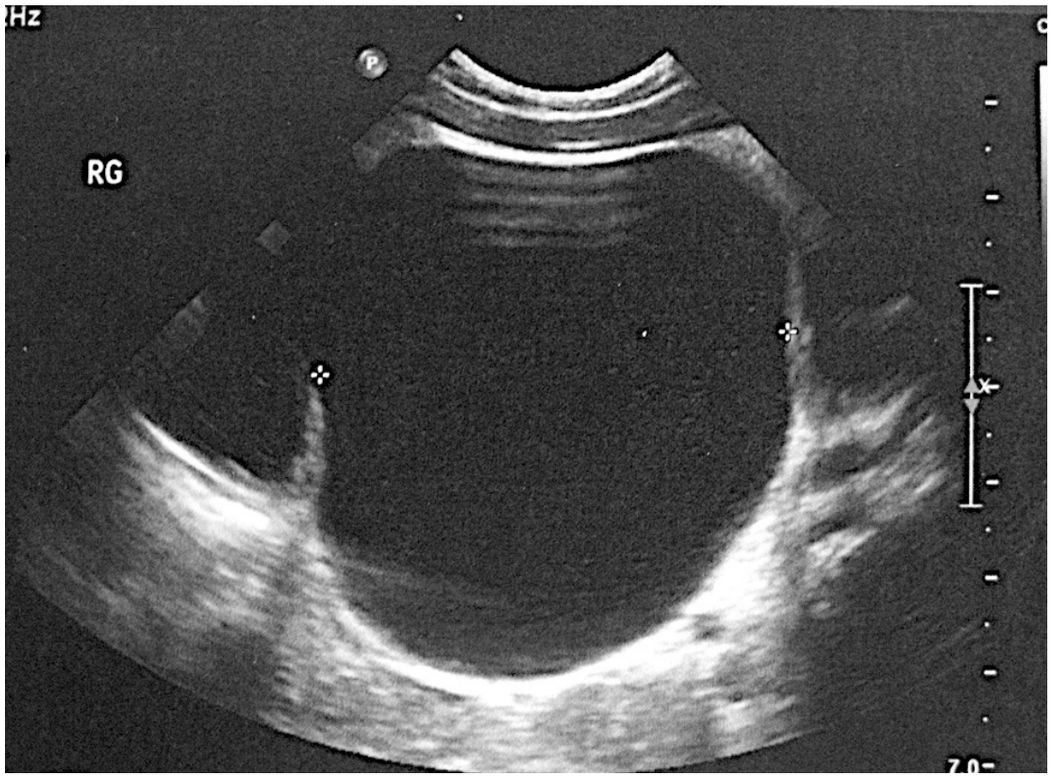
  2. large dilatation of the pelvis
     pelvis remains intrarenal
  3. major dilatation of pelvis & calyces
     pelvis extrarenal
  4. major dilatation
     + parenchymal thinning

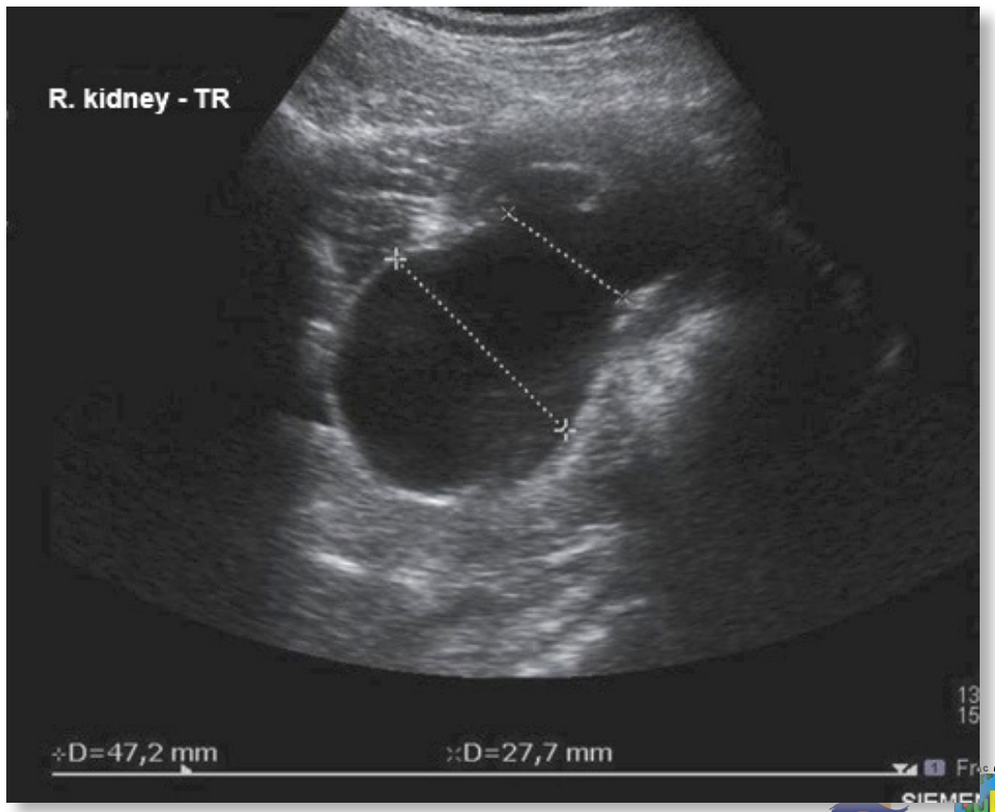
Maizels. *J Urol* 1992; 148:609



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

Abnormal



Imaging modalities - US

- **The most useful parameter is the antero-posterior (AP) pelvic diameter at the level of the renal sinus**
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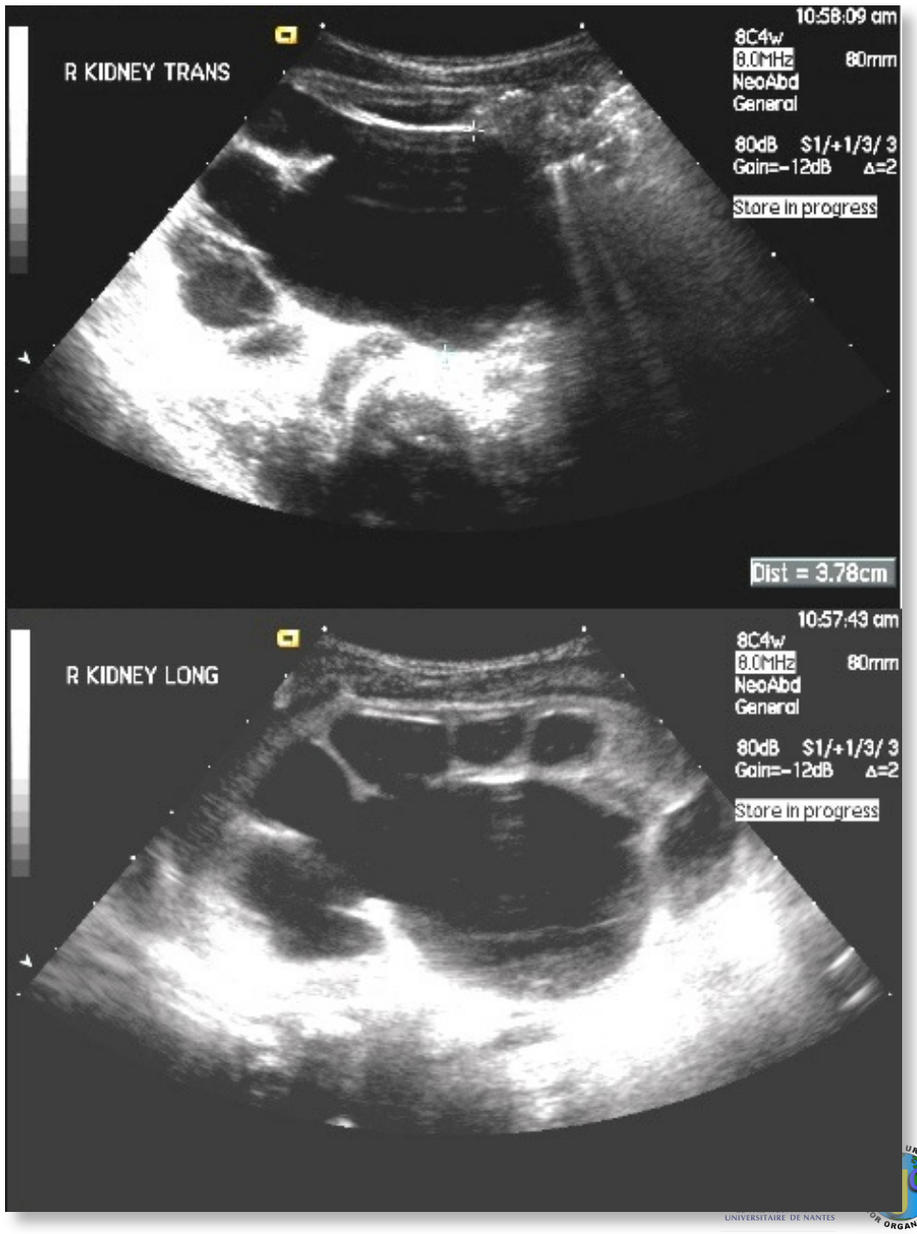
- **The degree of dilatation of the calyces**



Isotope imaging - Mag-3

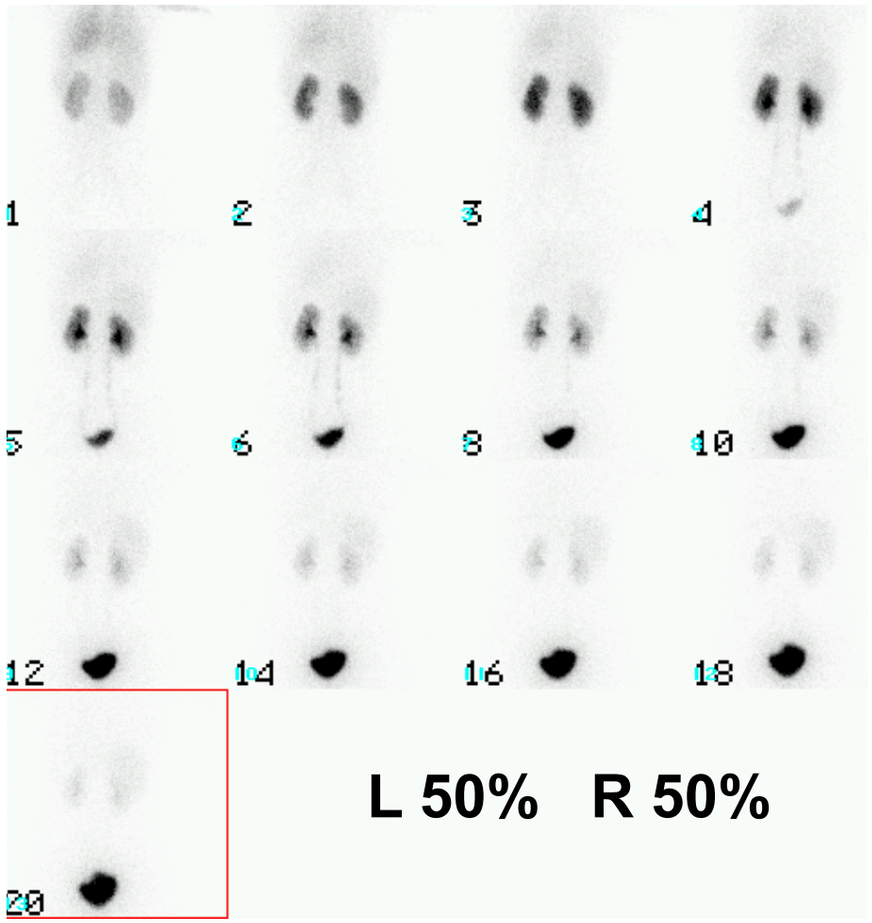
- **Radiopharmaceutical agent : ^{99}Tc-Mercaptoacetyltriglycerin**
  - high protein binding
  - high tubular excretion
  - low distribution in extra-vascular space

- **Serial dynamic acquisition : time / activity curve**
  - two distinct phases :

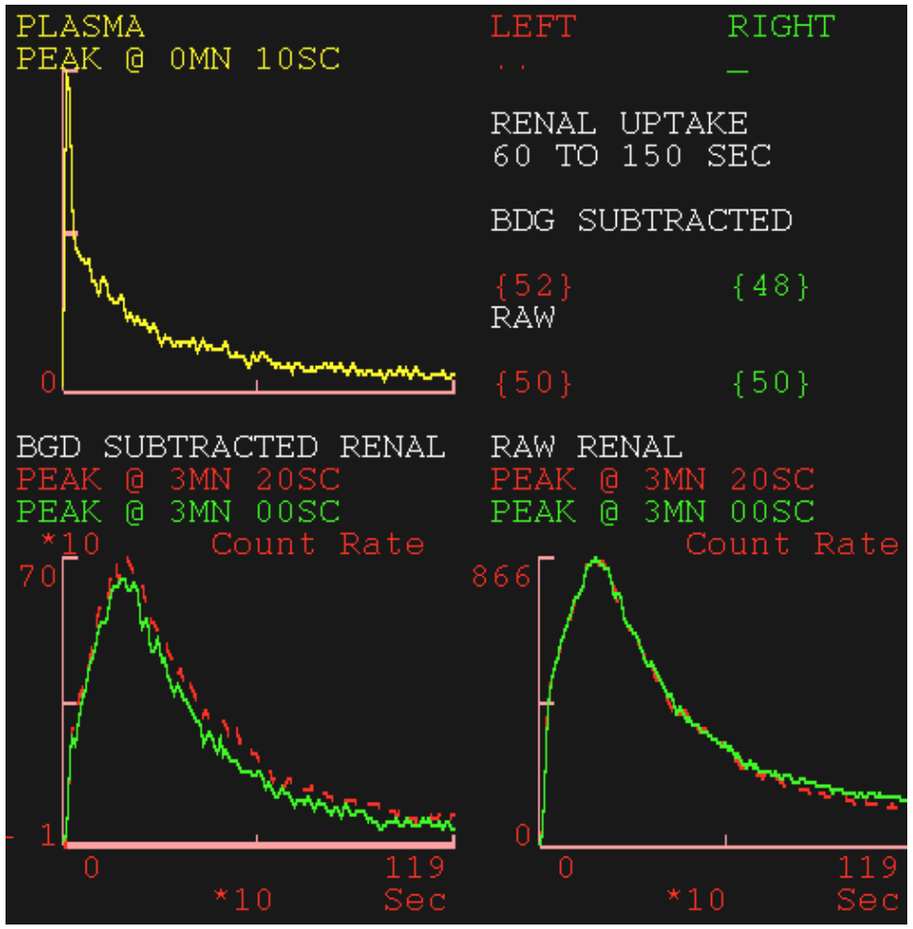
    uptake phase : differential renal funtion

    clearance phase : urine washout through the collecting system

Isotope imaging - Mag-3

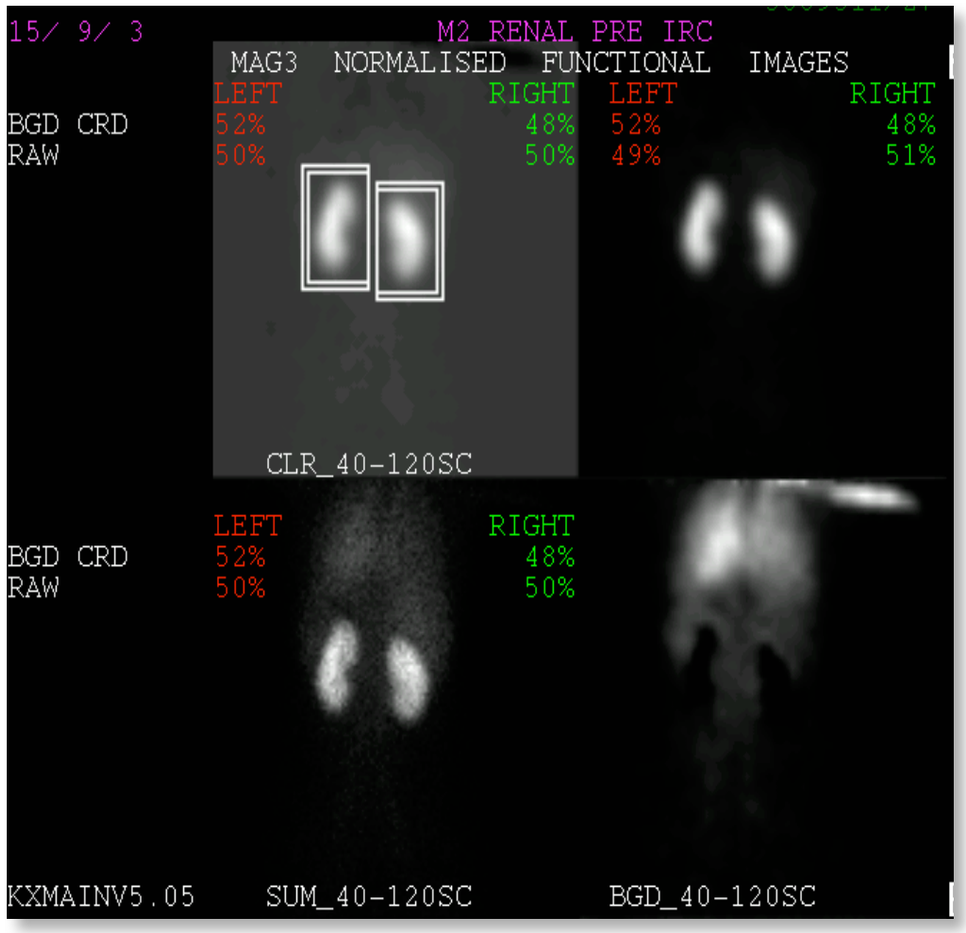


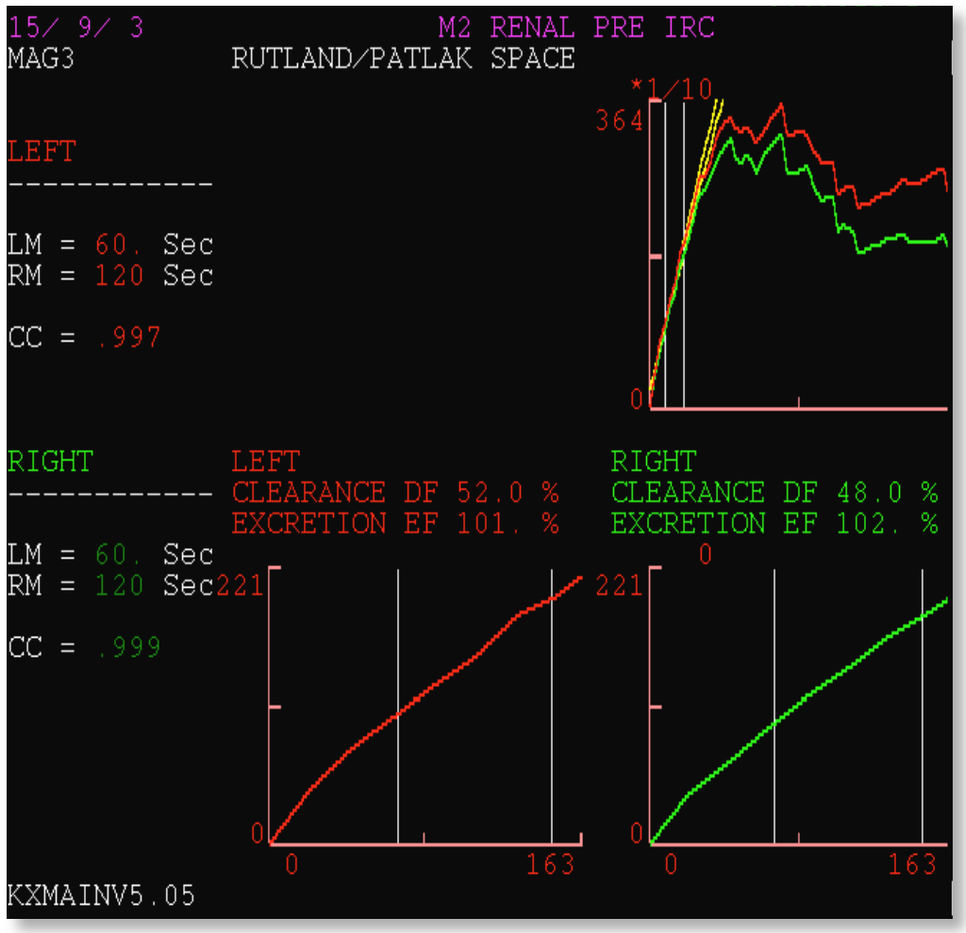
Renal scintigraphy



Time activity curves

Isotope imaging - Mag-3



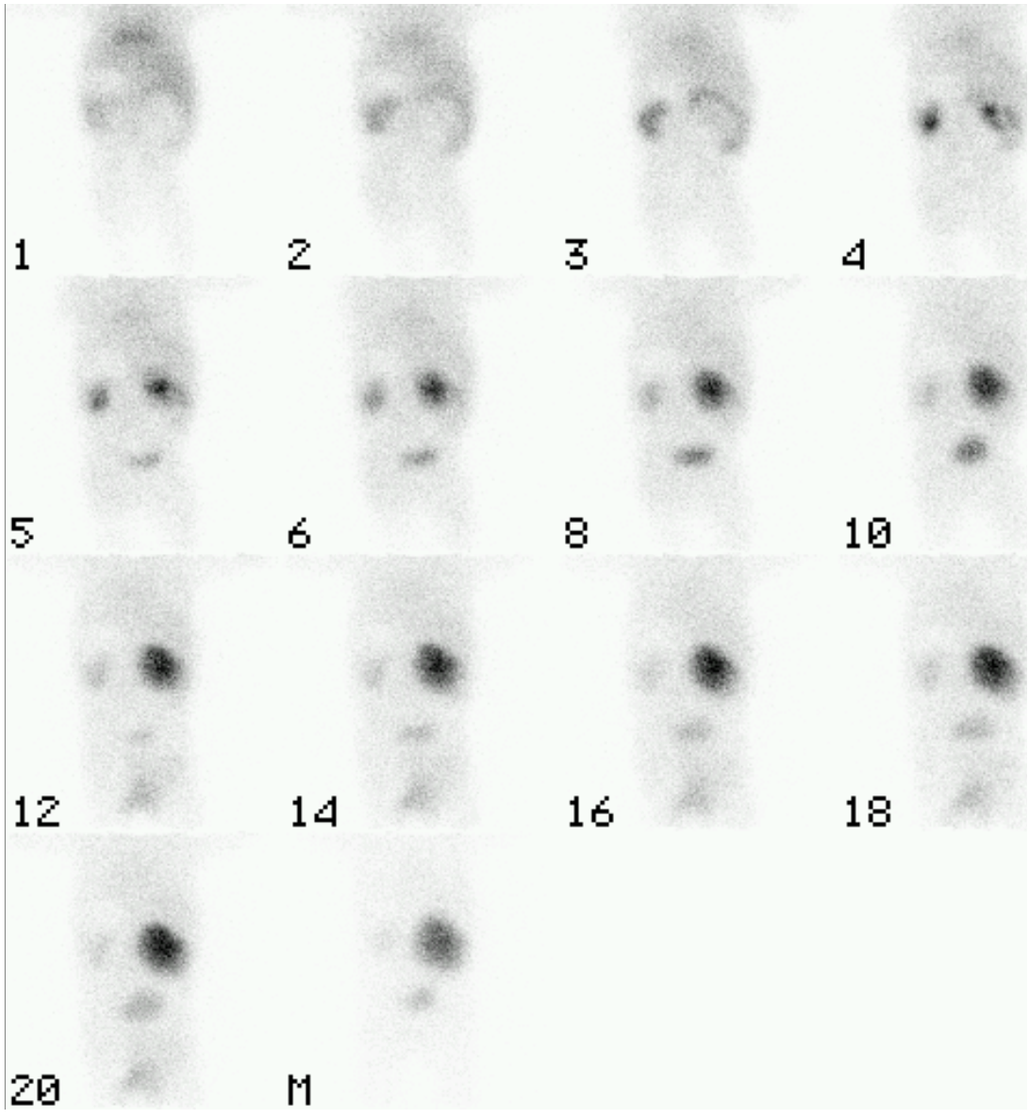


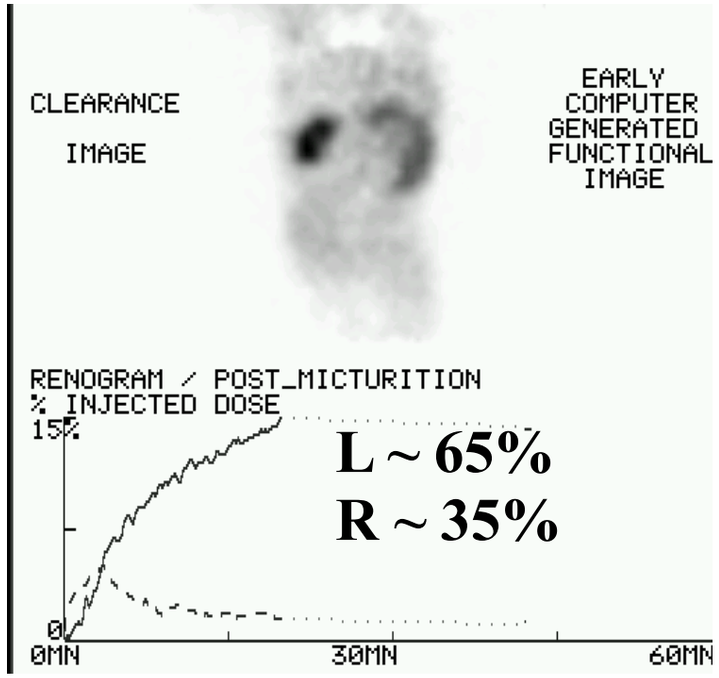
Regions of interest
around the kidney
with and without
background subtraction

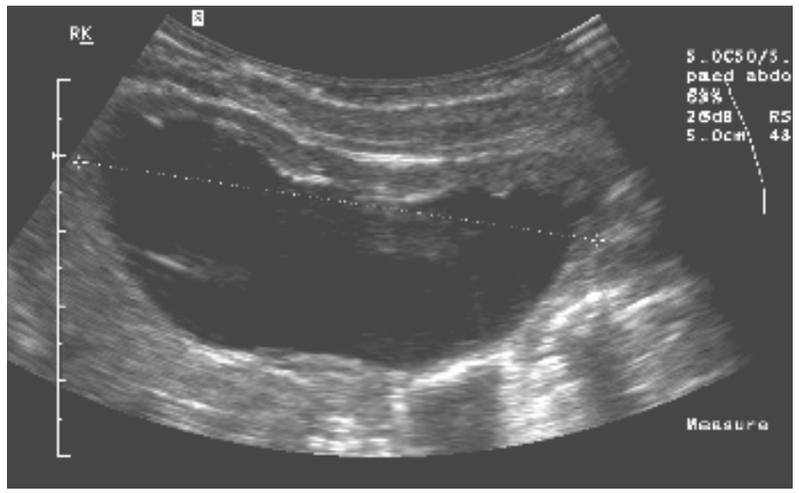
Rutland-Patlak plot:
rate of uptake by the kidney
from the 1st to the 2nd minute

Isotope imaging - Mag-3

- **Poor washout : Obstruction or delayed emptying ?**







Isotope imaging - Mag-3

Drainage depends on :

- **How much urine is produced ?**
  - **renal function**
    - a poorly functionning kidney will produce less urine than a normal kidney
    - a poorly functionning kidney will clear the isotope slowlier 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

0022-5347/03/1695-1828/0
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DOI: 10.1097/01.ju.0000062640.46274.21

IMPAIRED DRAINAGE ON DIURETIC RENOGRAPHY USING HALF-TIME OR PELVIC EXCRETION EFFICIENCY IS NOT A SIGN OF OBSTRUCTION IN CHILDREN WITH A PRENATAL DIAGNOSIS OF UNILATERAL RENAL PELVIC DILATATION

J. AMARANTE, P. J. ANDERSON AND I. GORDON

From the Great Ormond Street Hospital for Children, London, United Kingdom

- **Poor drainage at 20 min., in a child supine, does not necessarilly means 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 → Obstruction
  - dilatation stable
    or improving with time → ???

Management strategy

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

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**

AP diameter	surgery
< 20 mm	11 %
20-30 mm	40 %
30-40 mm	90 %
> 40 mm	≈ 100%

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 — Koff. *J Urol* 2000; 164: 1101
- 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**

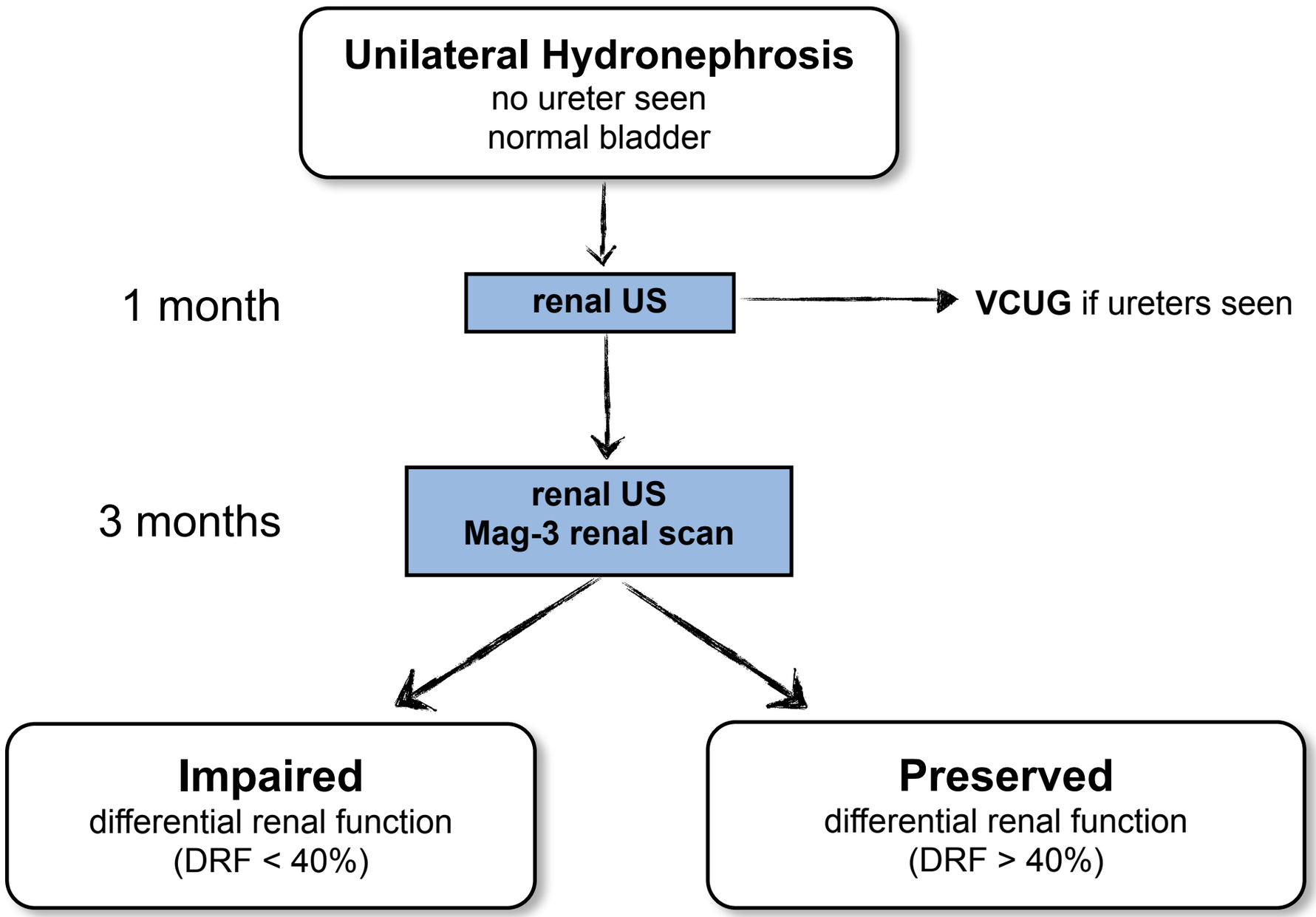
Management strategy [2]

- **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

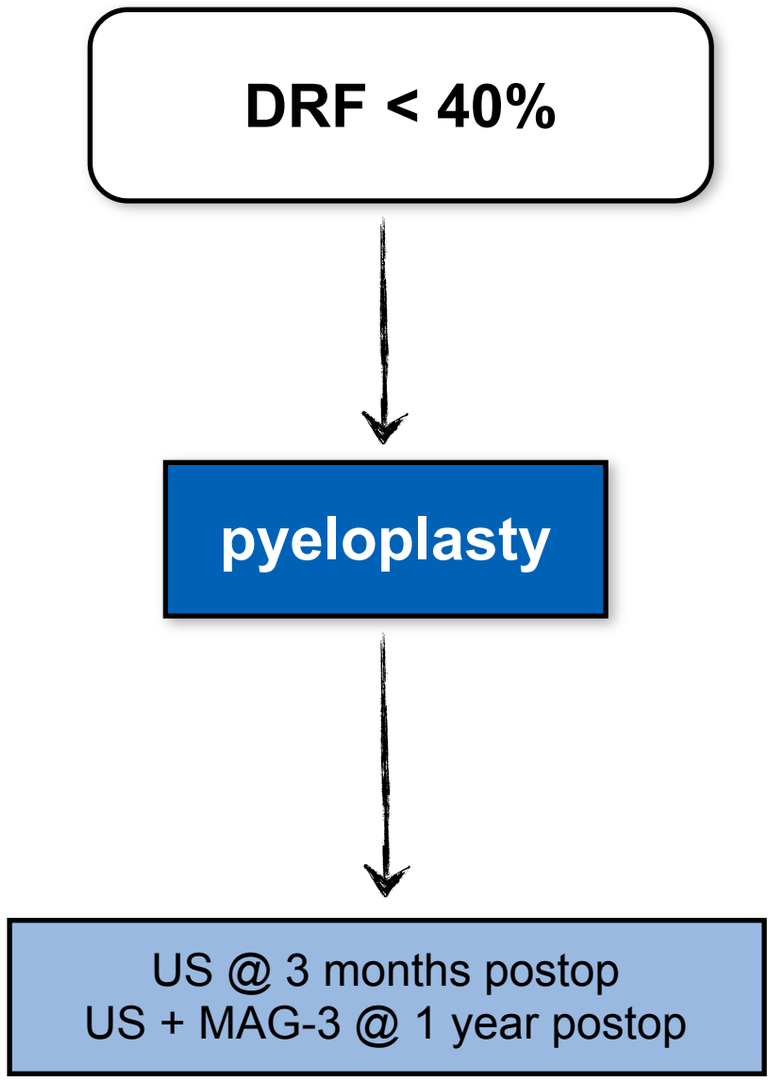
  - ultimately, surgery may not impact long term DRF
  - if managed conservatively,
    these children require **serial MAG-3 follow-up**

Management strategy [3]



Management strategy [4]

Impaired DRF



Management strategy [4]

Impaired DRF

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 ?

Management strategy [4]

Normal DRF

pelvis AP diameter
15-20 mm

pelvis AP diameter
20-30 mm

pelvis AP diameter
> 30 mm

Management strategy [4]

Normal DRF

pelvis AP diameter
15-20 mm

pelvis AP diameter
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pelvis AP diameter
> 30 mm

If major calyceal dilatation
or early prenatal diagnosis

follow-up

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

then discharge

Management strategy [4]

Normal DRF

pelvis AP diameter
15-20 mm

pelvis AP diameter
20-30 mm

pelvis AP diameter
> 30 mm

If major calyceal dilatation
or early prenatal diagnosis

follow-up

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

then discharge

pyeloplasty

Management strategy [4]

Normal DRF

pelvis AP diameter
15-20 mm

follow-up

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

then discharge

If major calyceal dilatation
or early prenatal diagnosis

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

MAG-3 if
hydronephrosis increase

pelvis AP diameter
> 30 mm

pyeloplasty

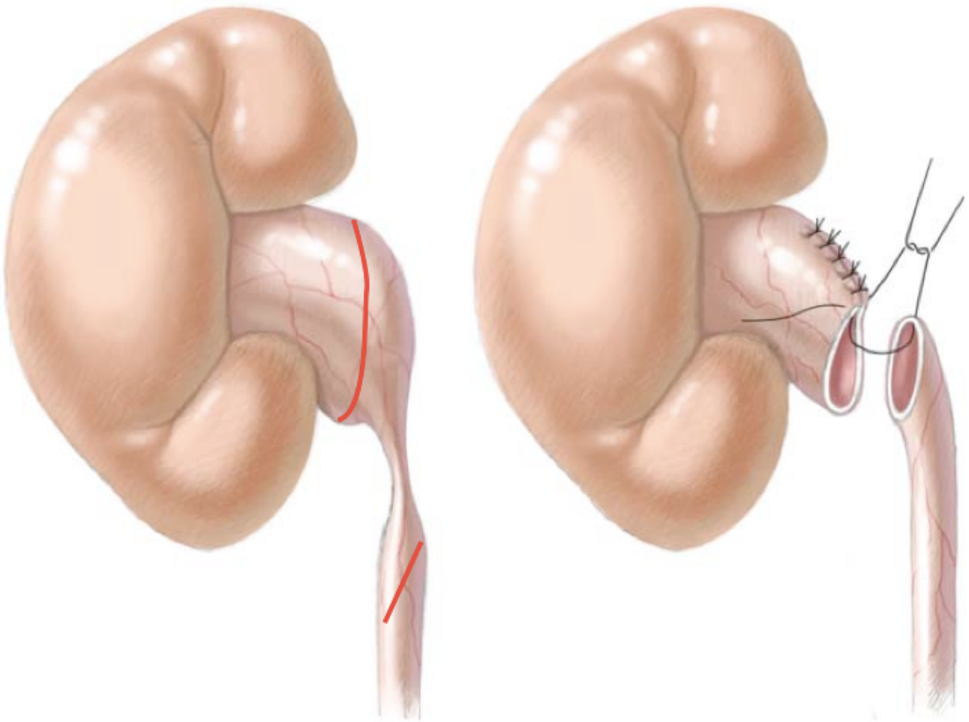
Symptomatic hydronephrosis

- **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

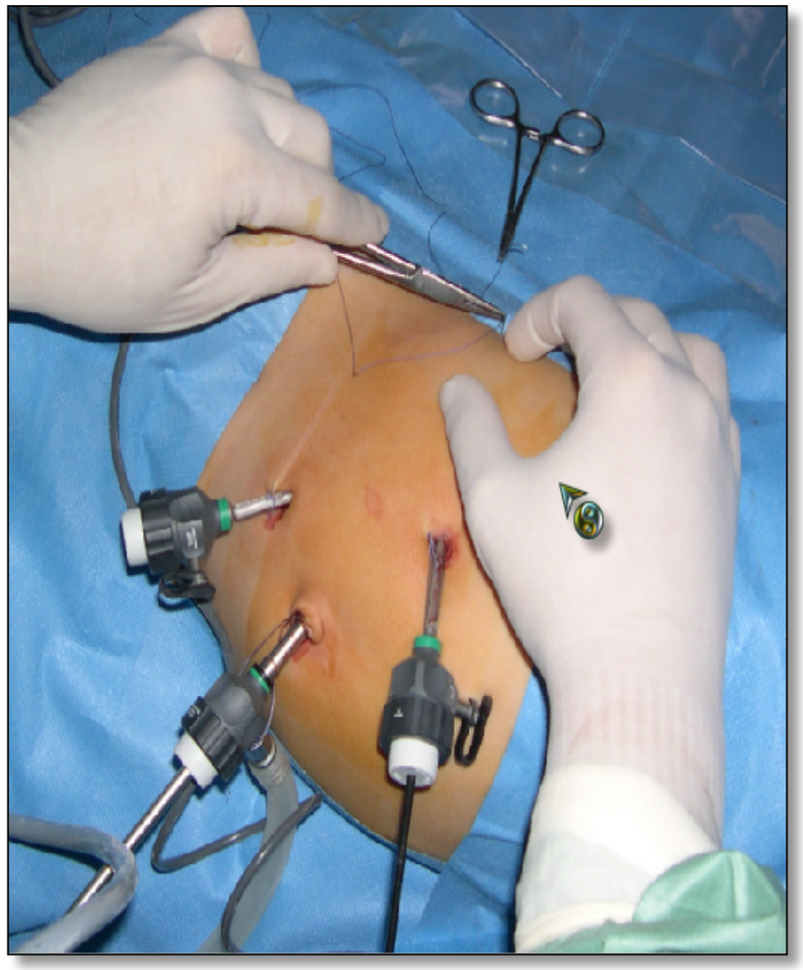
- **Anderson-Hynes dismembered pyeloplasty remains the gold standard**
  - retroperitoneal **open** surgery : lateral or posterior lumbotomy
  - **MIS** : transperitoneal laparoscopy, retroperitoneoscopy, robotic

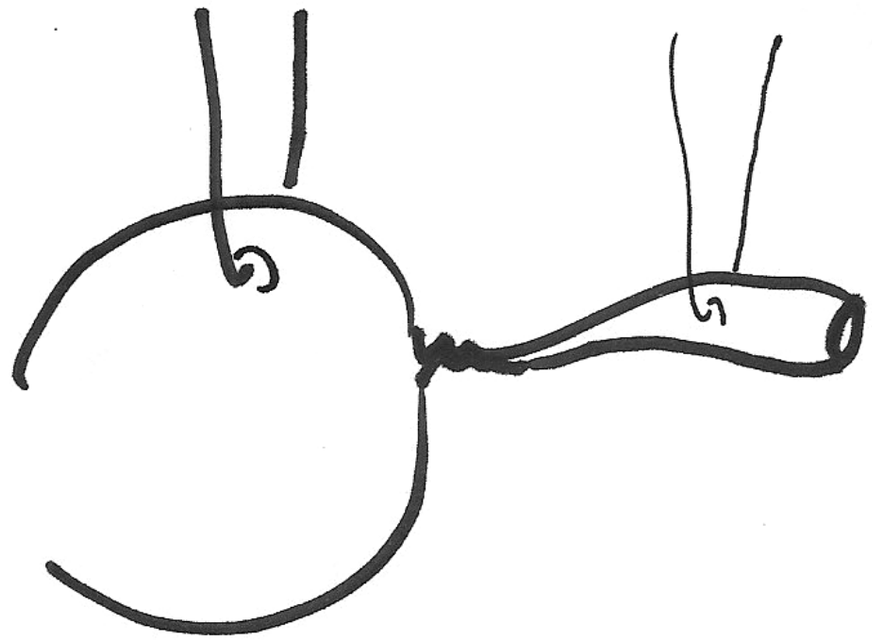


Surgical techniques

Transperitoneal approach

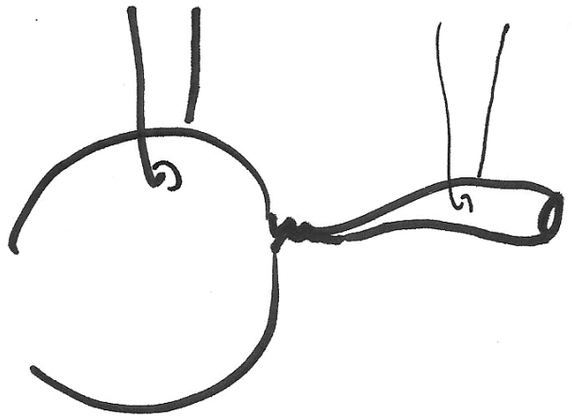
- **Suspension of PUJ by stay sutures**

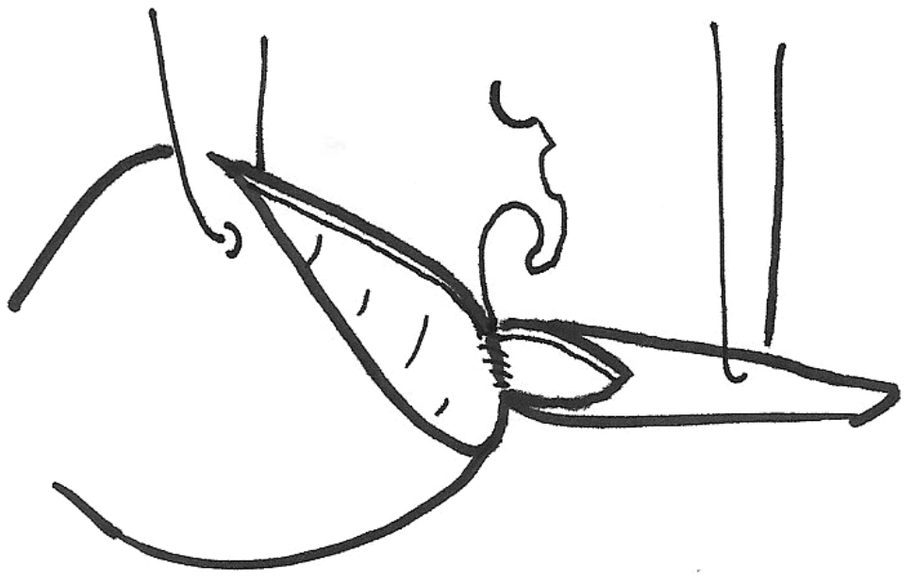




Surgical techniques

Transperitoneal approach





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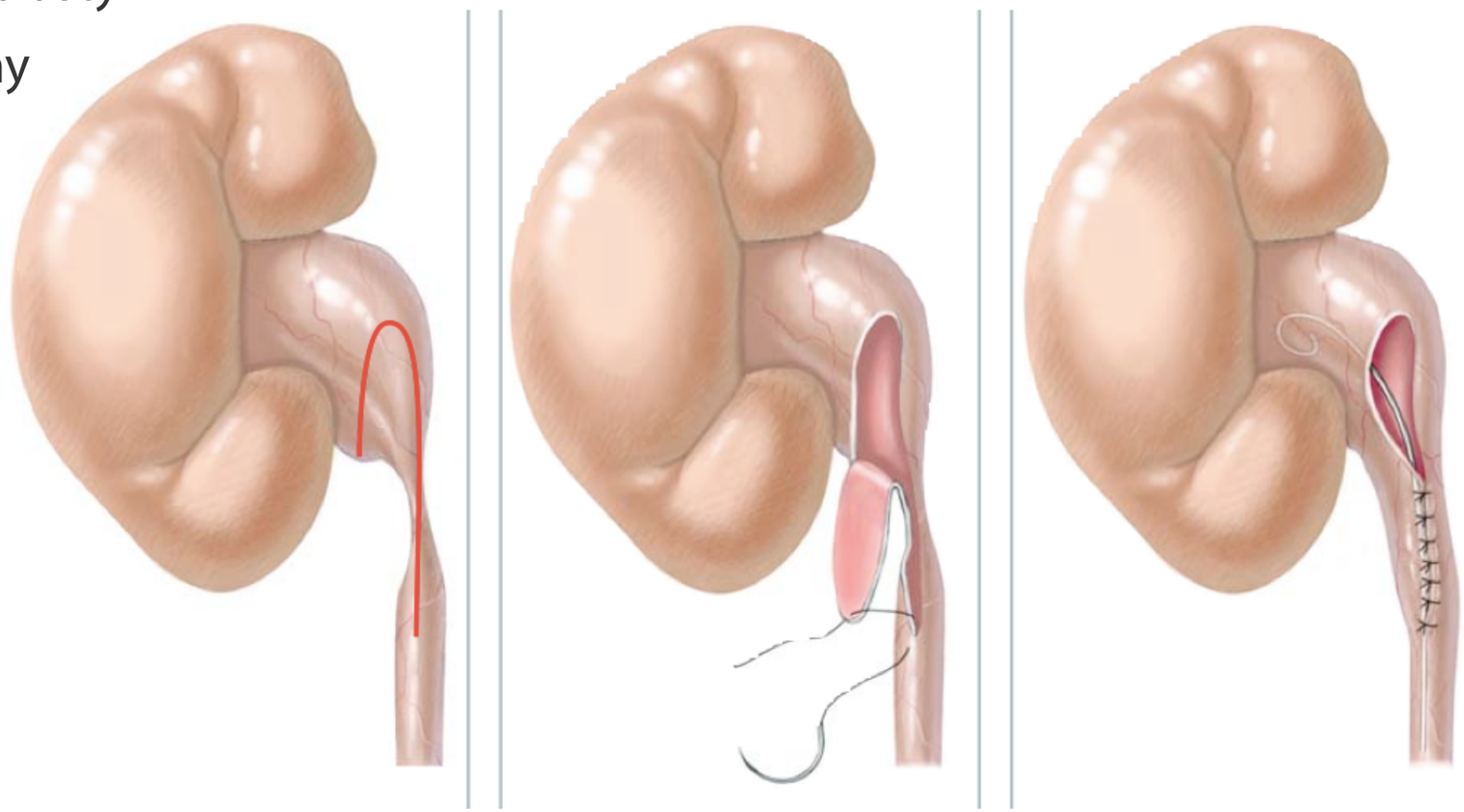
- **« Salvage » procedures should be known**
  - Culp-De Weerd pyeloplasty
  - uretero-calicostostomy



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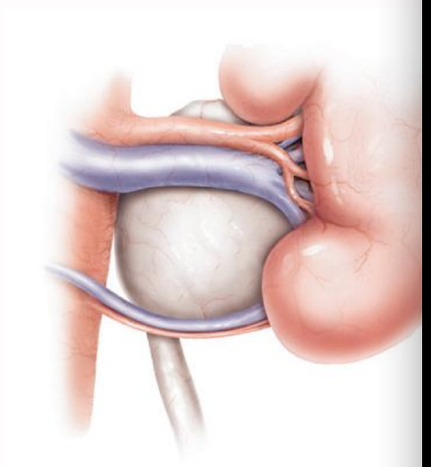
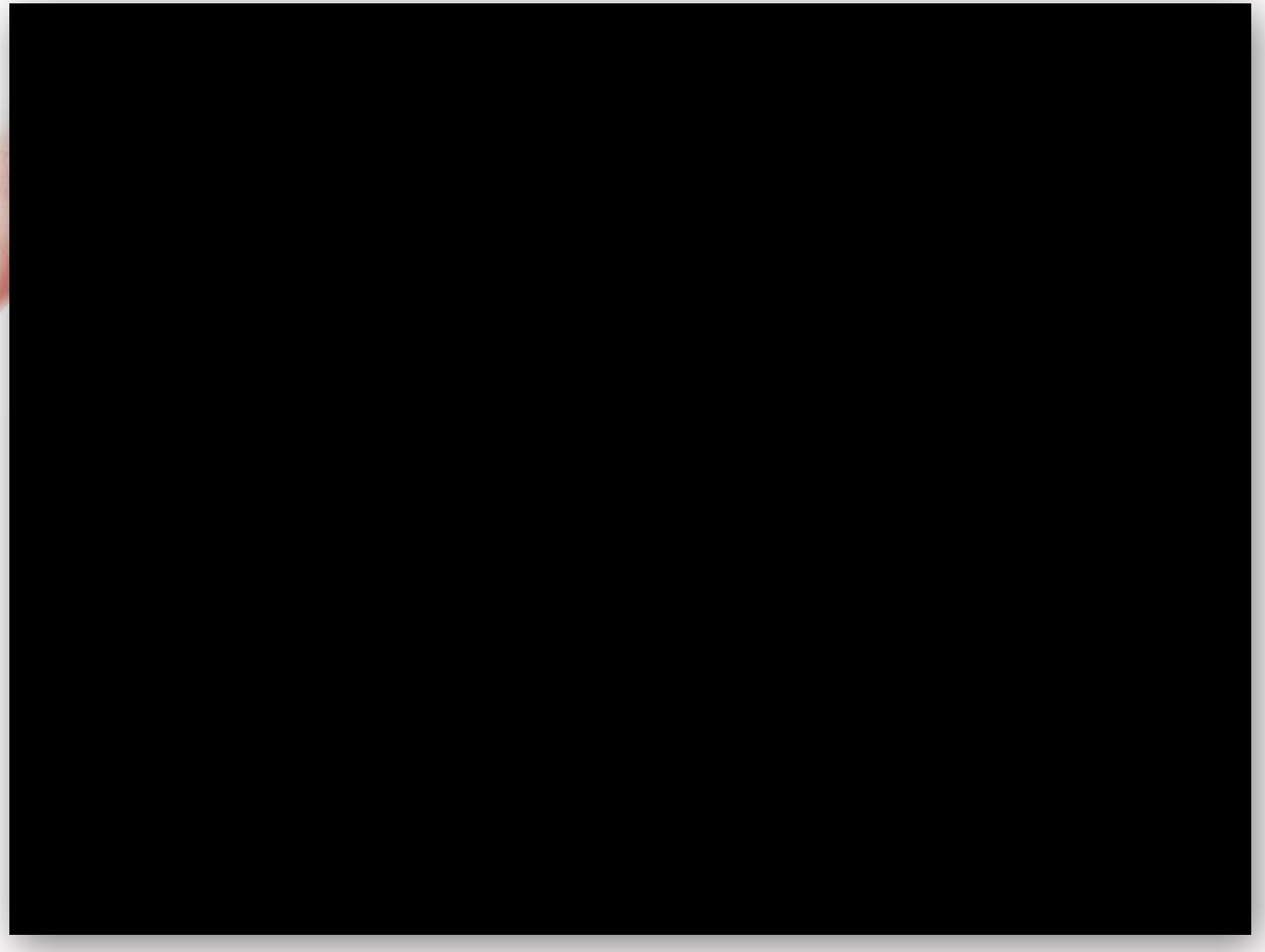
- **« Salvage » procedures should be known**
  - Culp-De Weerd pyeloplasty
  - uretero-calicostostomy

- **Vascular hitch procedure : selected indications**
  - intermittent actute hydronephrosis + lower pole vessels without intrinsic obstruction — Gundeti *et al. J Urol* 2008; 180: 1832
  - excellent long-term outcomes — Villemagne-T *et al. ESPU* 2013. Genova
    55 cases (31 transp laparoscopy - 24 robotic-assisted)
    Follow-up : 31 months [12-84]
    2 failures

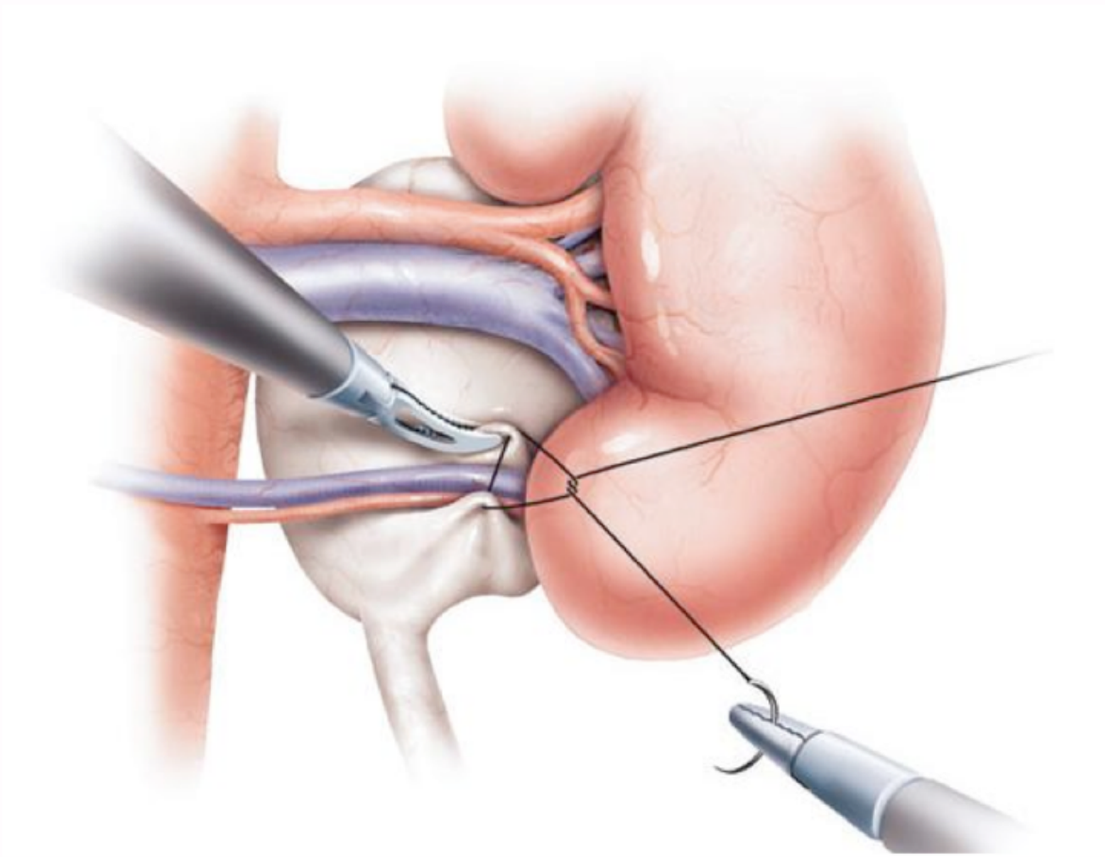
The vascular hitch technique

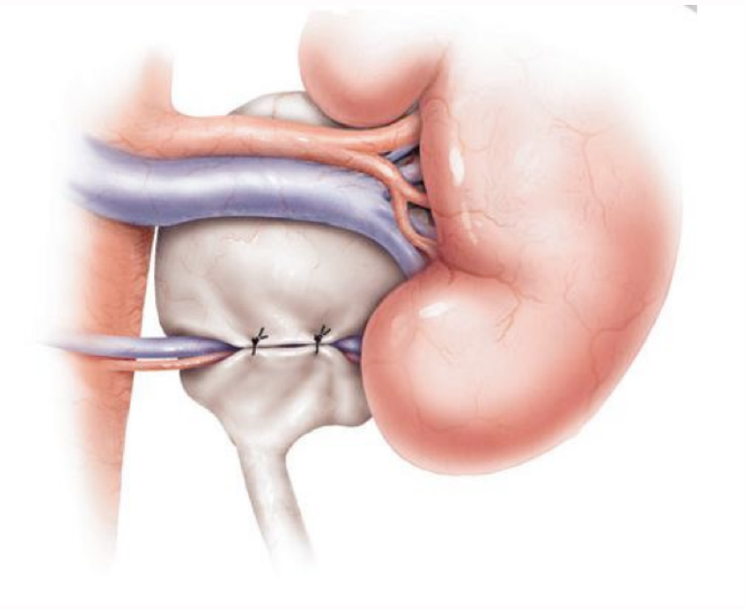




Illustrations : S. Spitzer,
in Sakoda-A *et al.* BJU Int 2011:1364-8

The vascular hitch technique





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The vascular hitch technique

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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

Indications - Which approach ?

- **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

  - **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

What did he say ?

- **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

Prof. Marc-David LECLAIR

Hôpital Mère-Enfant. NANTES. FRANCE

Pathophysiology

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

Leibowitz-S. *J Clin Pathol* 1963;16:342

- **Abnormal muscle fibers**

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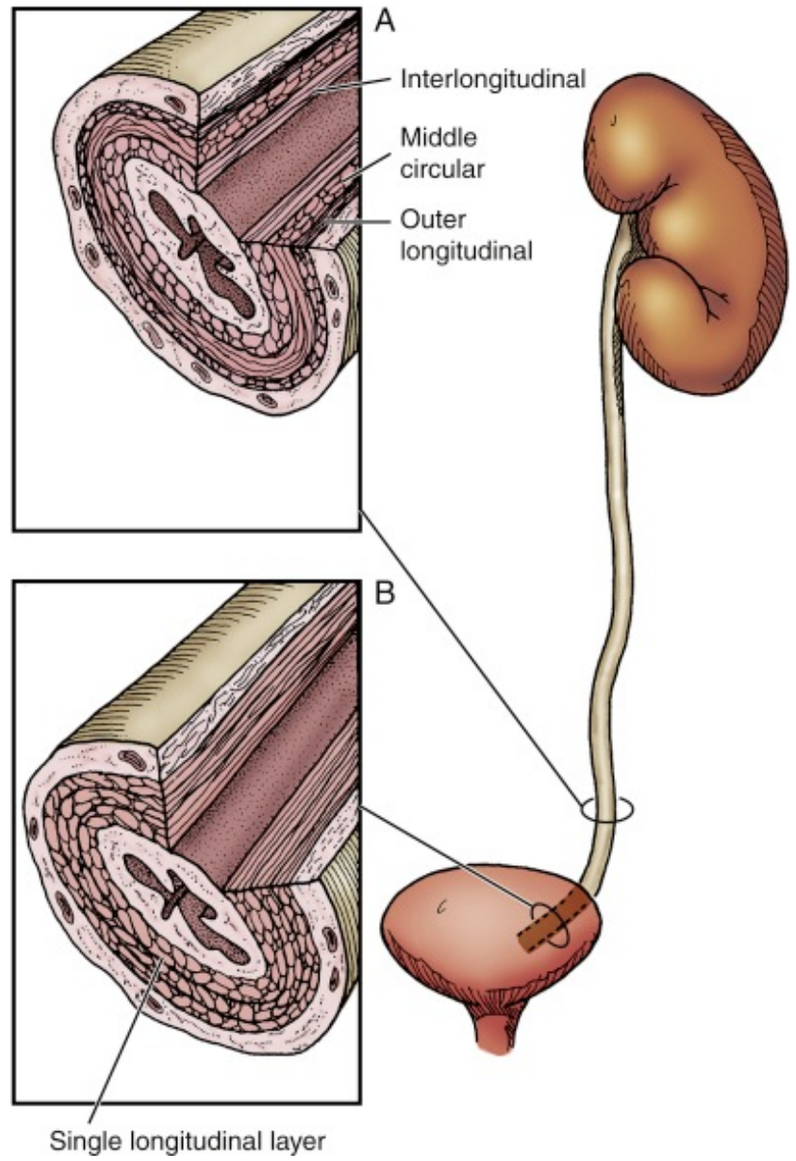


Pathophysiology

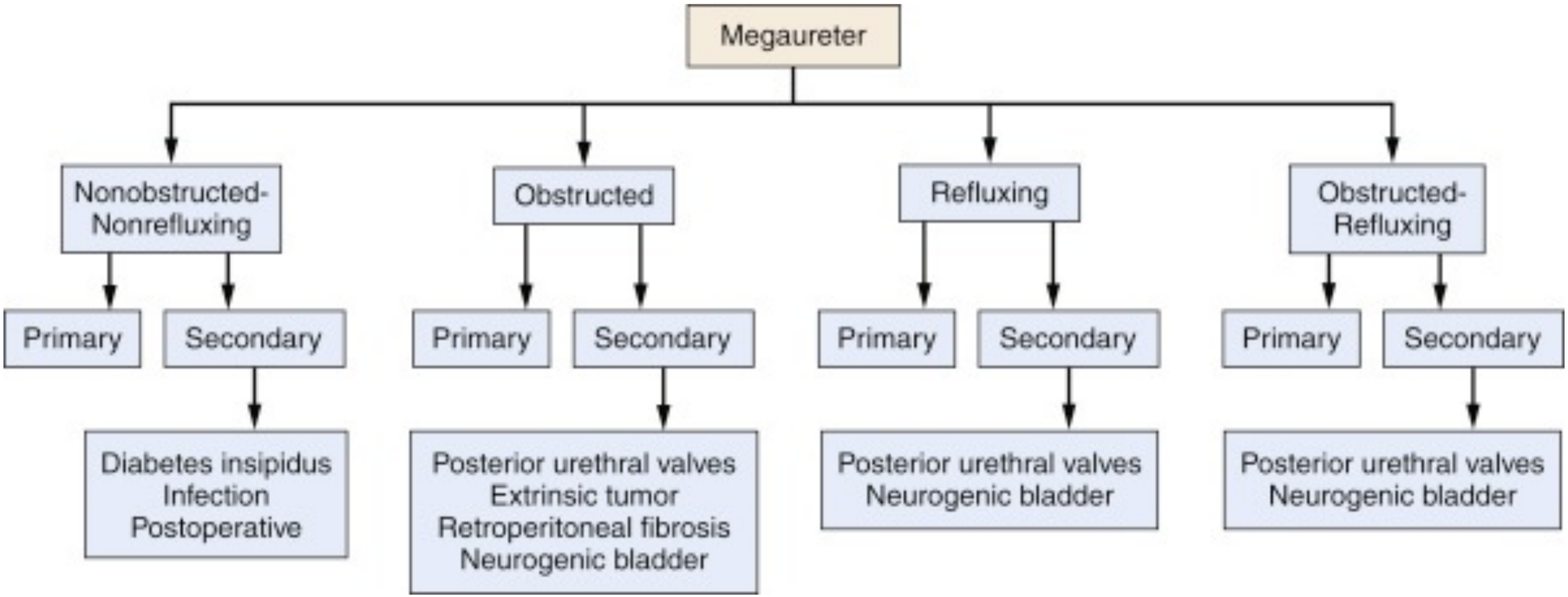
- **Urologist's Hirschsprung disease ?**
  - but normal ganglia distribution in distal ureter — Leibowitz-S. *J Clin Pathol* 1963;16:342

- **Abnormal muscle fibers**
  - hypertrophy circular muscle layer instead of longitudinal — MacKinnon. *J Urol* 1970;103:134
  - 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-ß — Nicotina-PA. *Br J Urol* 1997;80:946

King's Classification

King-LR. *J Urol* 1980;123:222



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

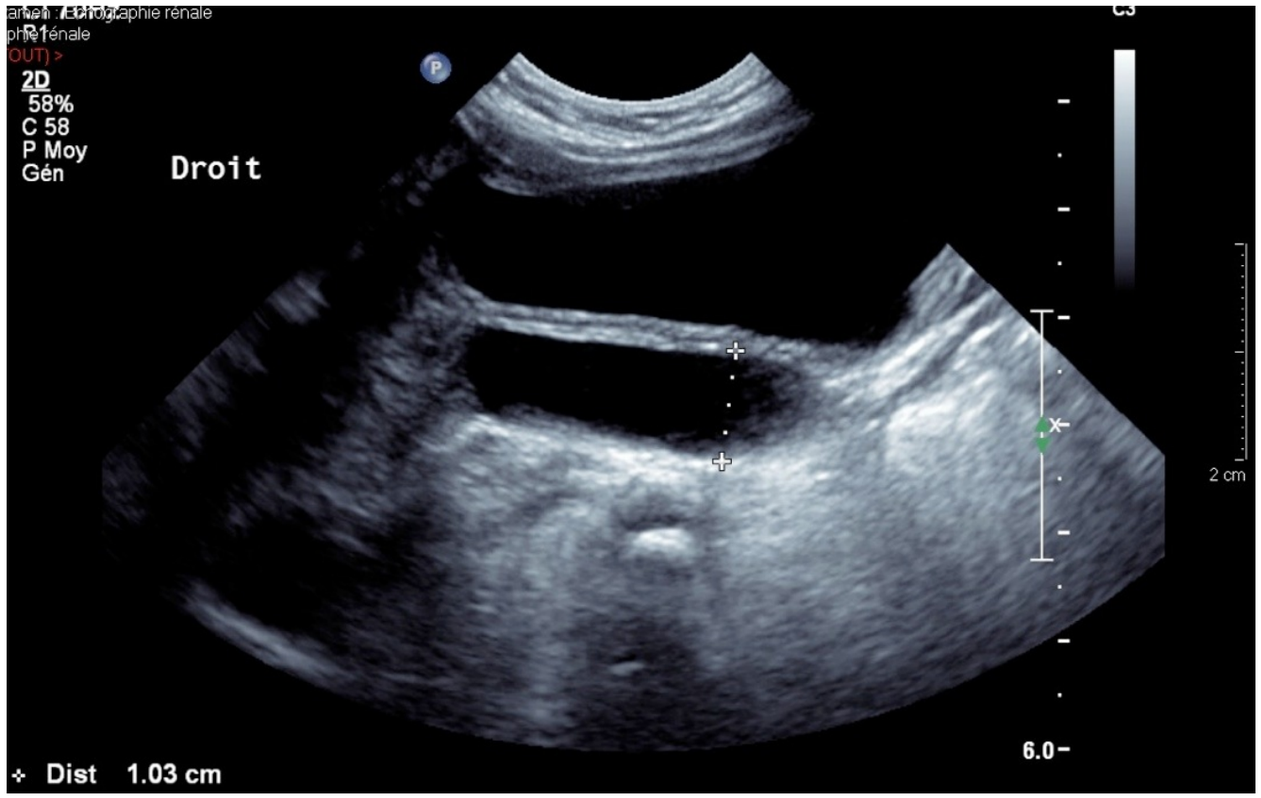
- **Primary : typical in newborns**
  - multifactorial causes
  - high prenatal urine output
  - dyscoordinated voiding
  - excess of elastin and collagen (over-expression TGF-ß)

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

Imaging

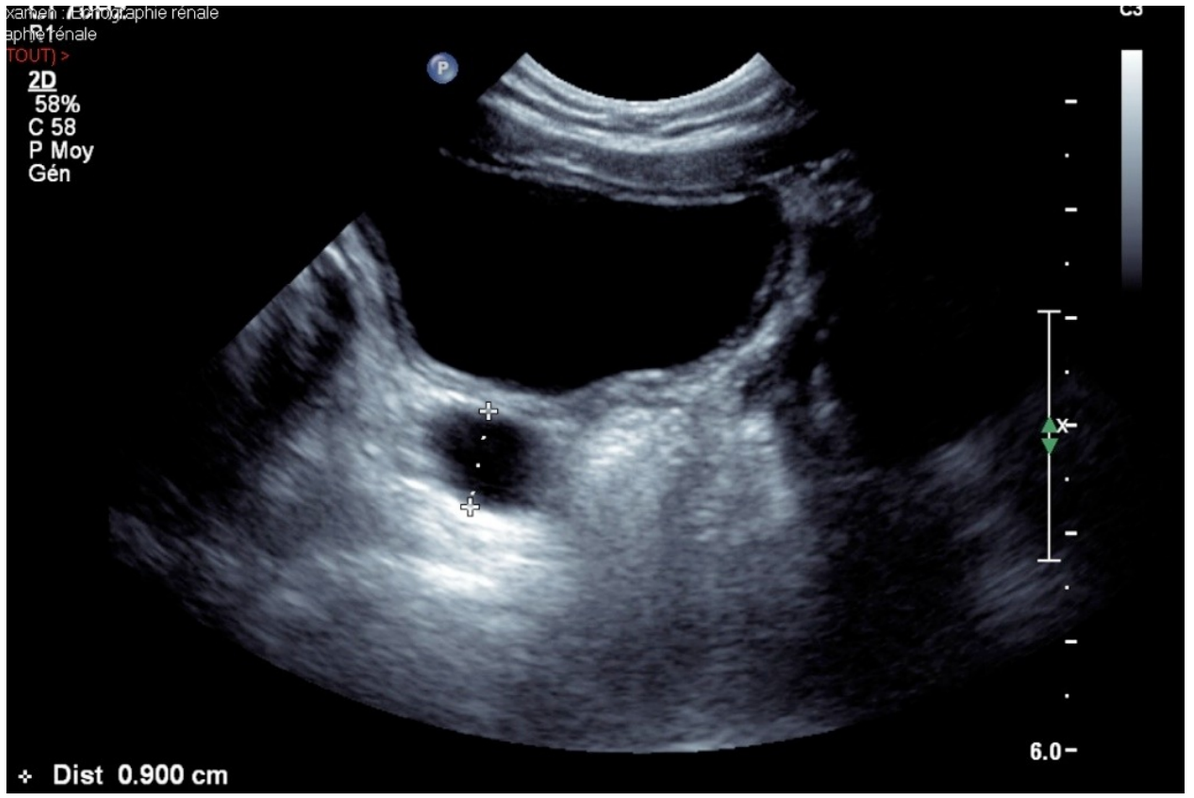
- Ultrasound
- MAG-3 renal scan
- (functional) -MRI

Ultrasound



Retrovesical ureter diameter

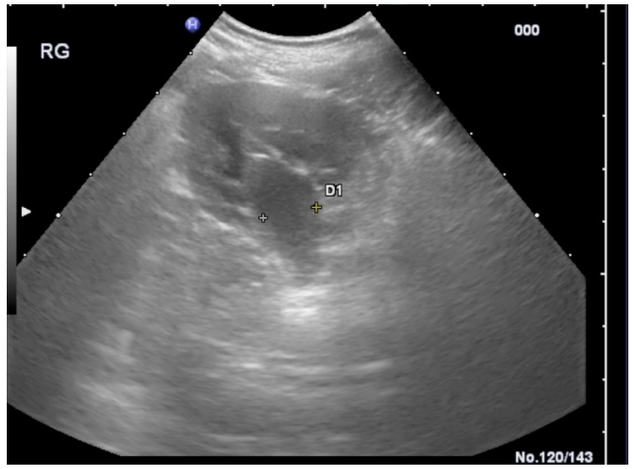
- Longitudinal -

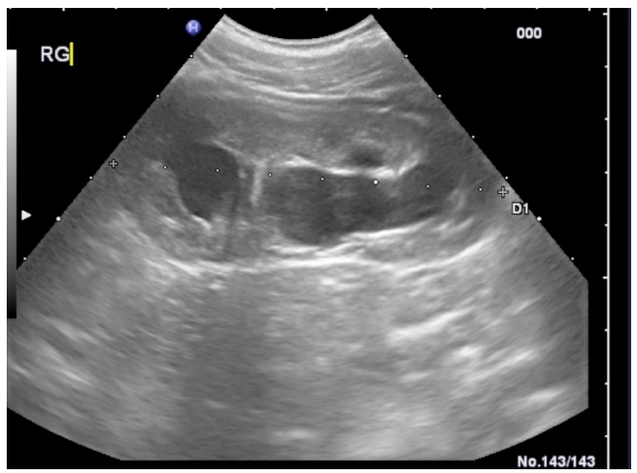


Retrovesical ureter diameter

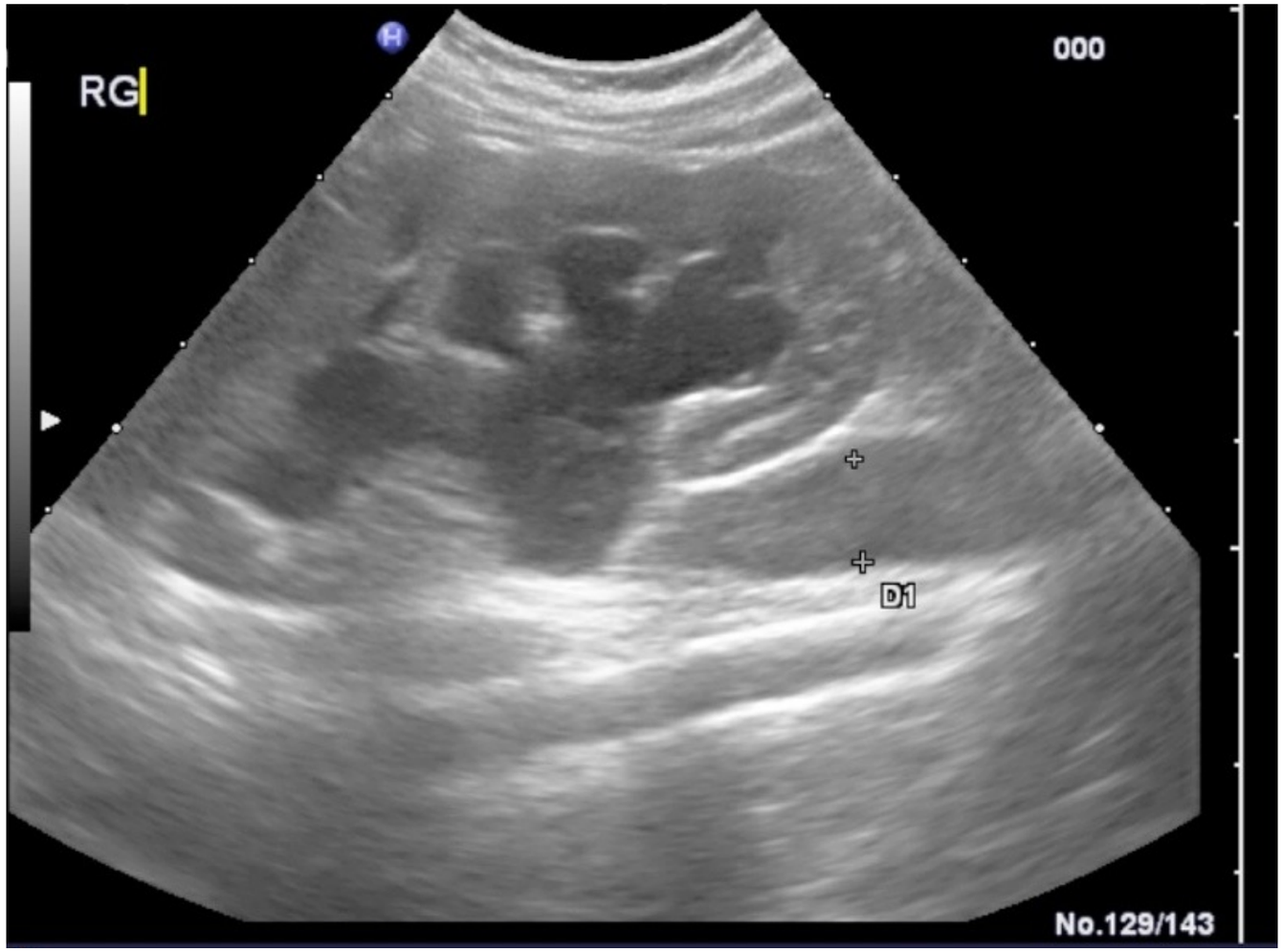
- Transversal -

Ultrasound





Pelvicalyceal dilatation



Lumbar ureter diameter

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** Ranawaka. *J Pediatr Surg* 2013;48:380

    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

Diversion

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

Castagnetti-M *Urology* 2006;68:870

Diversion

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- **What is the place for ureteric internal stenting ?**

  - **19 ureters in infants** Farrugia-M *J Pediatr Urol* 2011;7:198

    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

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

- **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

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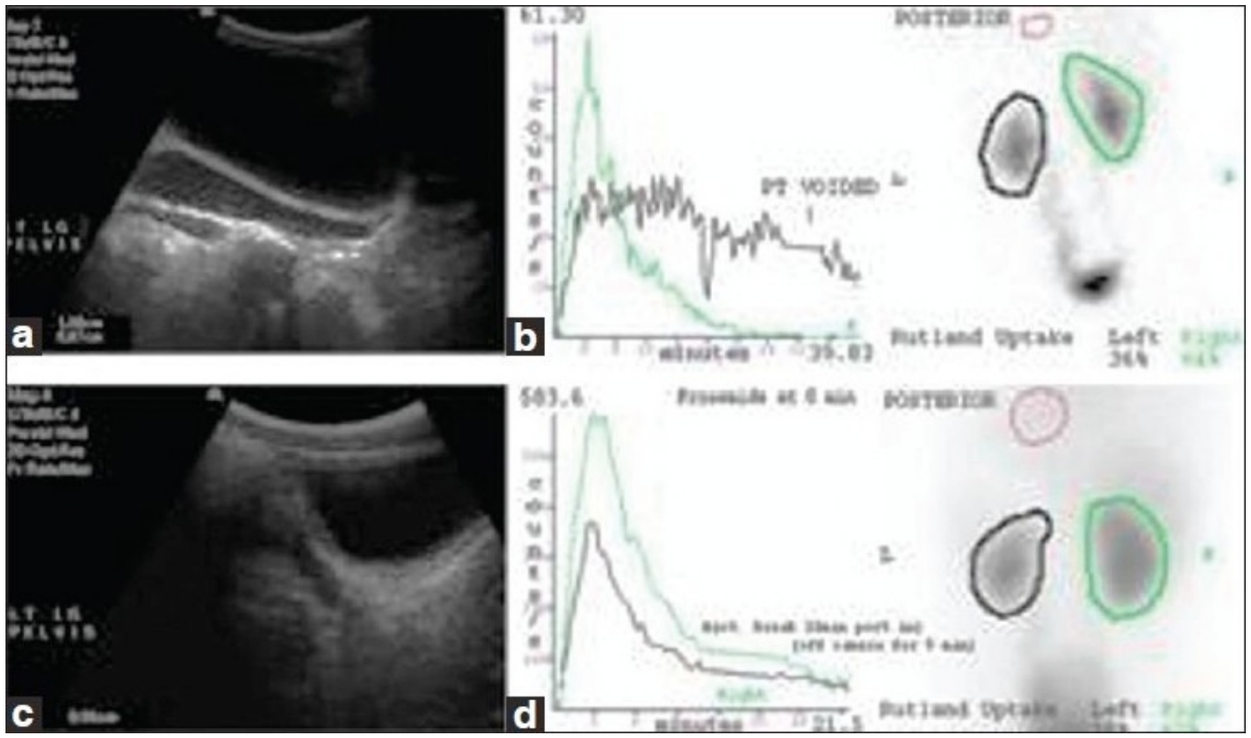

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- **What is the place for ureteric internal stenting ?**

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

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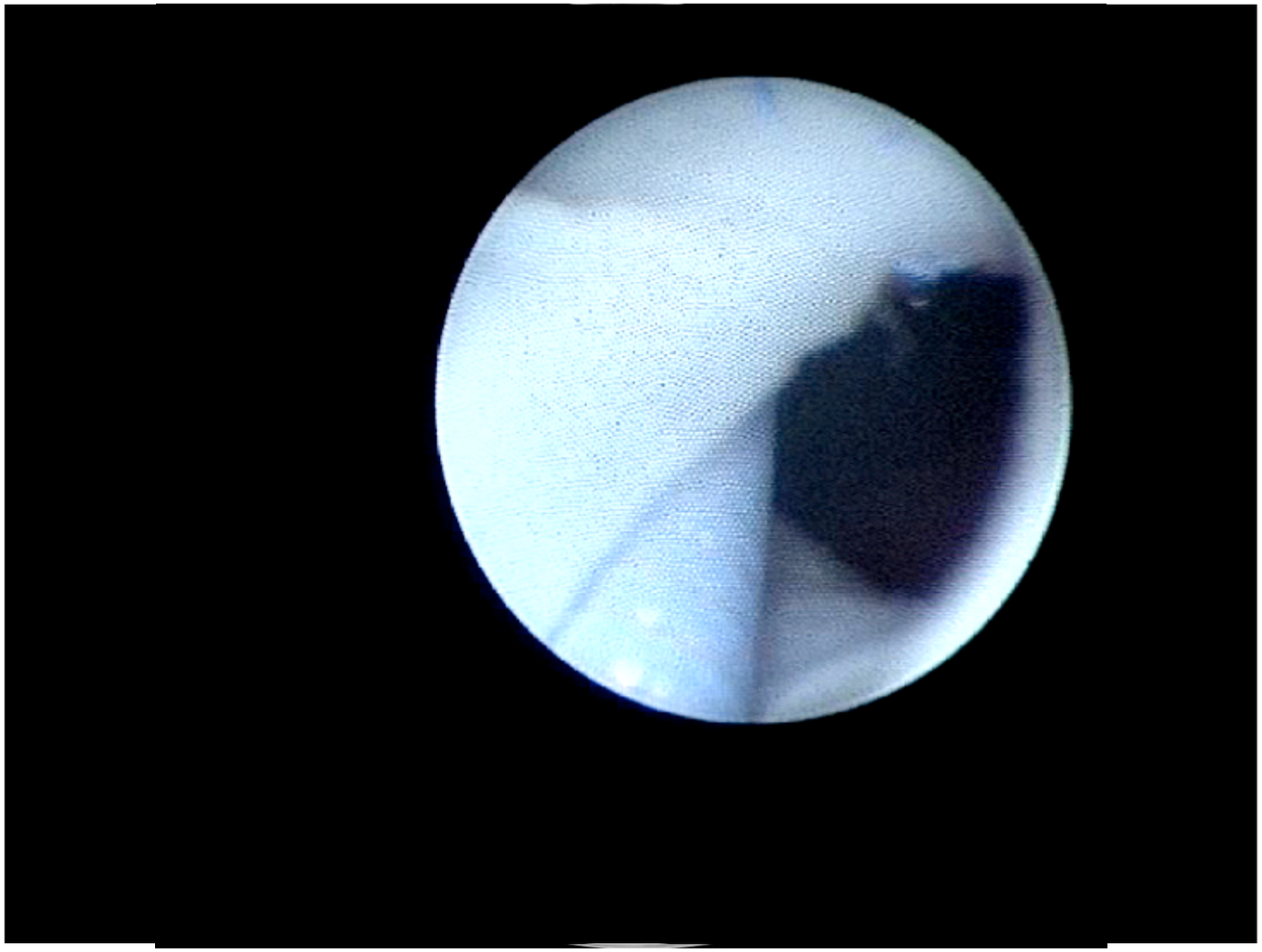
Endoscopic Treatment

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



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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

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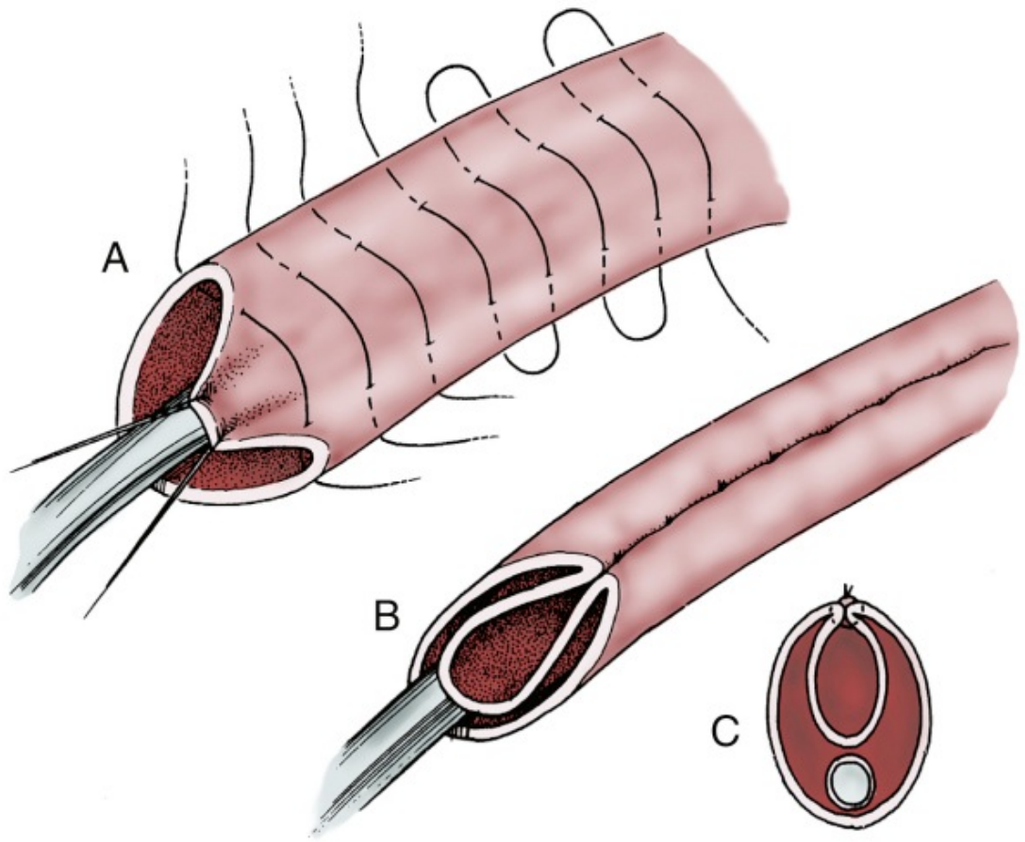
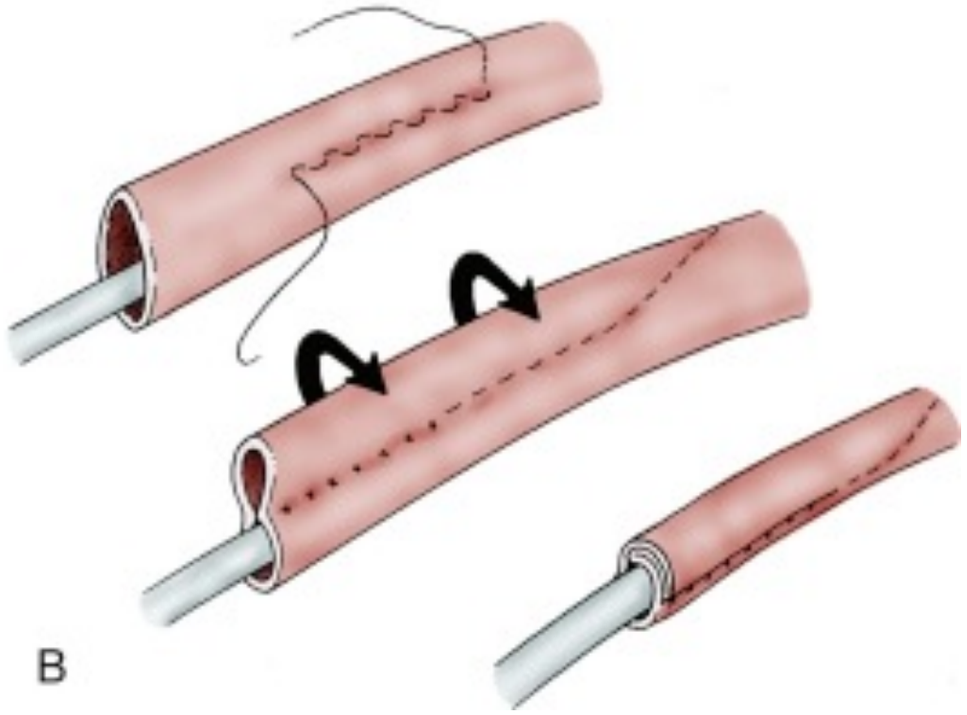
  - **Personnal series** Clermidi-P. *unpublished data*
    14 primary obstructive MGU
    3/14 failures of insertion - high rate of stent related complications
    6/9 improved at >12 months

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**