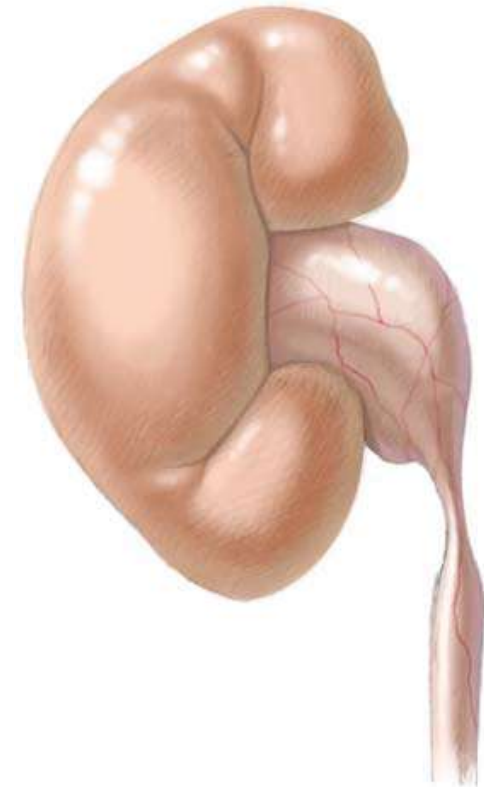


# Uretropelvic Junction (UPJ) obstruction



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# Outline

## Basics

- Embryology
- Etiology
- Pathophysiology

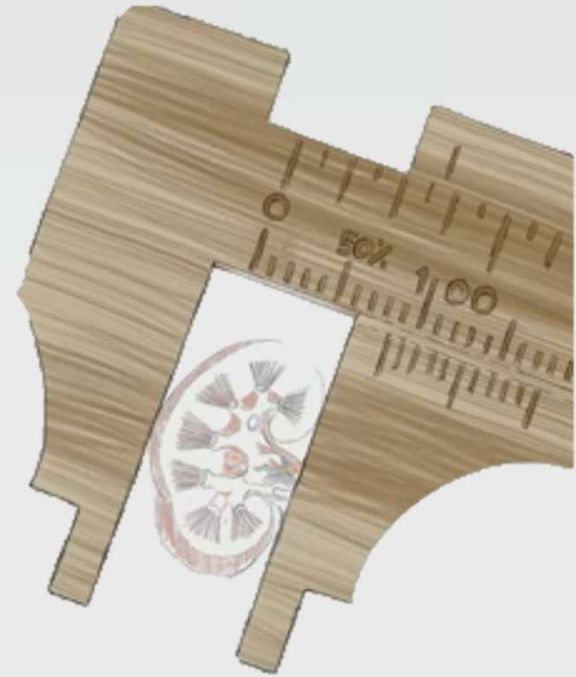
## Clinical evaluation

- Epidemiology
- Presentation
- Workup

## Pyeloplasty

- Indications
- Techniques
- Outcome

# 1 Basics



## Embryology and Morphological (Mal)Development of UPJ

Ali Avanoglu<sup>1\*</sup> and Sibel Tiryaki<sup>2</sup>

<sup>1</sup>Division of Pediatric Urology, Department of Pediatric Surgery, Ego University, Izmir, Turkey; <sup>2</sup>Gaziantep Maternity and Children's Hospital, Pediatric Urology, Gaziantep, Turkey

- UPJO is the most investigated anomaly from CAKUT
- Despite years of experimental/clinical research etiopathogenesis is unclear
- It involves both genetic and environmental factors.
- Main proposed mechanisms...
  - Failure in development of the renal pelvis
  - Failure in the **recanalization** of UPJ
  - Abnormal UPJ **innervation**
  - impaired **smooth muscle differentiation**
  - single gene mutations (AGTR2, BMP4, Id2)

# Etiology

## Intrinsic

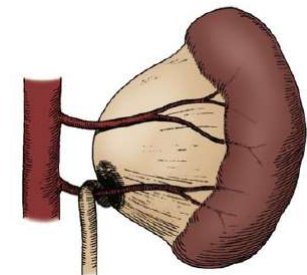
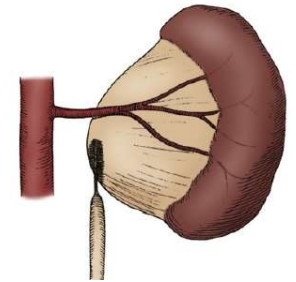
- Narrowing of UPJ with undeveloped circular muscle fibers
- Other – polyps & valves (Difficult to diagnose preop)

## Extrinsic

- Crossing vessels (accessory lower pole artery)- 20-30%
  - Presents in childhood and adolescence
  - May have concurrent intrinsic problem

## Secondary

- Severe VUR (tortuous ureter kinking of UPJ)
  - May improve with reimplantation or may need staged surgery



# Pathophysiology

- **Abnormal UPJ**
  - **ureter narrow** (variable length)
  - joins pelvis **above the expected dependent position**.
  - fibrosis & interruption of smooth muscle (**Peristaltic wave blocked**)
- **Functional obstruction** (Urine pass at low volume, but pelvis dilates as the flow increases)
  - inadequate drainage of urine from the renal pelvis,
  - hydrostatic distention of the pelvis and calyces (dilated pelvis may kink the ureter further)
  - Progressive damage to the kidney.
- May have **simultaneous VUR (10%) or VUJ obstruction** (but retrograde pyelogram & CUG isn't routine)

# 2 Clinical Evaluation

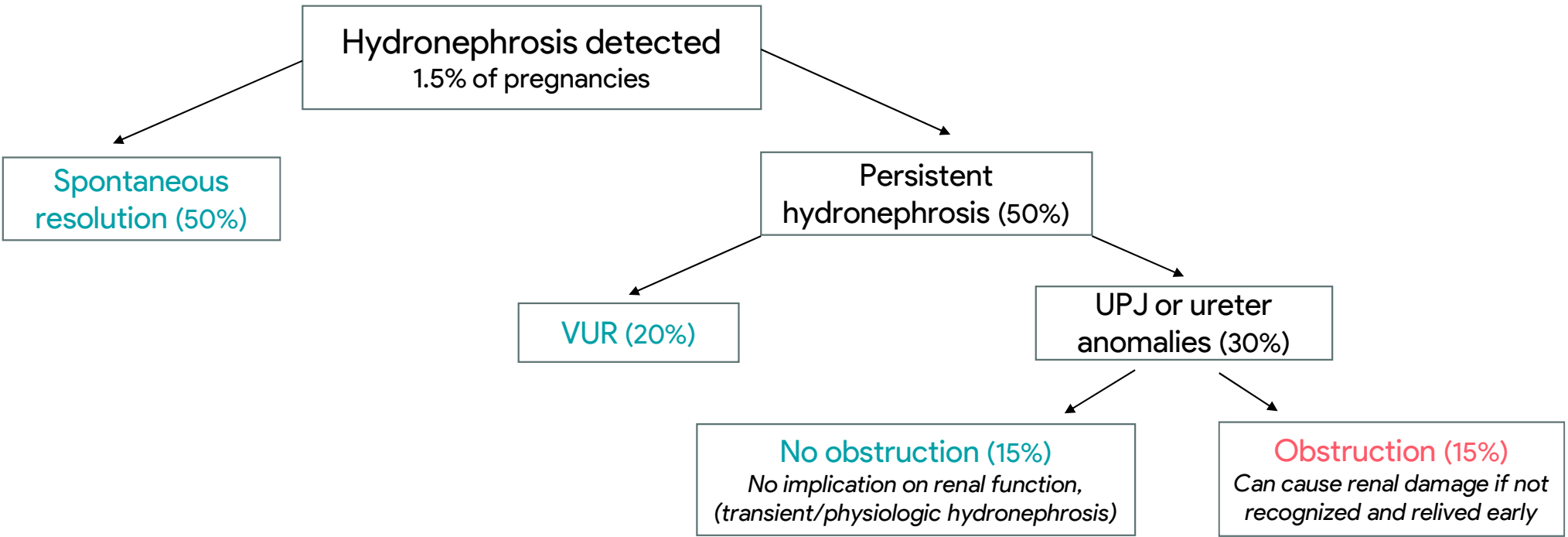


# Epidemiology

- The most common cause of antenatal hydronephrosis
- 1 in 5000 live births (inc with antenatal US)
- 1 in 1250 require surgery (constant)
- M:F = 2:1
- Left side in 60%
- Bilateral in 5-10%

# Presentation

- Antenatal Ultrasound
- flank mass (common presentation in the past)
- UTI
- Irritability, vomiting, feeding difficulties, constipation, FTT
- Dietl's crisis (intermittent abdominal pain, vomiting)
  - \*may not have dilatation on US during convalescence but thickening of pelvis is a clue
- Hematuria after trauma or exercise (rupture of mucosal vessels in dilated system)
- Associated anomalies (VACTERL)

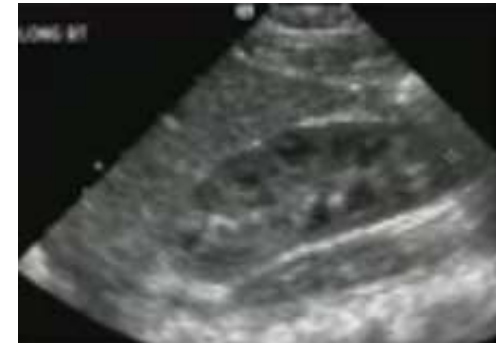


**Approach:** Bilateral PUJO is rarely associated with significant enough fetal obstruction to cause oligohydramnios and warrant antenatal intervention. Postnatal hydronephrosis (pelvic AP diameter >7mm in neonates/>10mm in children) is not necessarily due to obstruction and can often be explained by physiologic polyuria and natural kinks and folds in the ureter.

# Postnatal Ultrasound

**initial test:** evaluates distal ureter, bladder & contralateral system

- Timing (for antenatally diagnosed) - on 1-2 wk of life
- Finding - hydronephrosis without dilated ureters
  - \*pelvic APD - normal <4mm, severe HN > 15mm



UTD score	P1- low risk	P2- intermediate	P3- high risk
Caliceal dilatation	Central	Peripheral	
Renal pelvis	10-15	>15	
Ureter		Abnormal	
Cortical thickness			Abnormal
Parenchymal appearance			Abnormal
Bladder			Abnormal

SFU Grade	Dilatation	cortex
1	Slight splitting	
2	few calyces	
3	all calyces	
4		thinned

# Diuretic renography

**Confirmatory test:** provides functional information

- Types

- **DTPA** excreted by glomerular filtration
- **MAG3** excreted by tubular secretion (yields better image in those with impaired renal function and in infants)

*\*DMSA binds to proximal convoluted tubules and not excreted (parenchymal imaging not renography)*

- Timing (for antenatally diagnosed) - on 6-8wk of life

- Finding

- split renal function (early uptake first 1-2 min)
- rule out obstruction (washout augmented by diuretic)

- Pros and cons- Limited in bilateral obst, no anatomy, interpretation



# MRU

**Not widely available**, use limited to those with complex anatomy

- Timing – any age
- Finding
  - excellent anatomy independent of renal function (T2 weighted)
  - Differential renal function (gadolinium enhanced)
- Pros and cons
  - No radiation, non toxic contrast, good image
  - Costly and requires sedation

*\*has the potential to become a gold standard*



# IVU

**Historical**, indications have faded but still used in low resource settings

- Timing – after 4-6 wk of life
- Finding - anatomy + function (but limited)
- Pros and cons
  - Good anatomy if function is good, inaccurate in poor function
  - Radiation exposure (flouroscopy), nephrotoxic contrast,
  - Can't detect simultaneous VUJ obst. (MRU is better)

*\*Study from Iran found diuretic urography (IVU with lasix) has small difference in sensitivity from diuretic renography in those with good function*



Comparison Between Diuretic Urography (IVP) and Diuretic Renography for Diagnosis of Ureteropelvic Junction Obstruction in Children

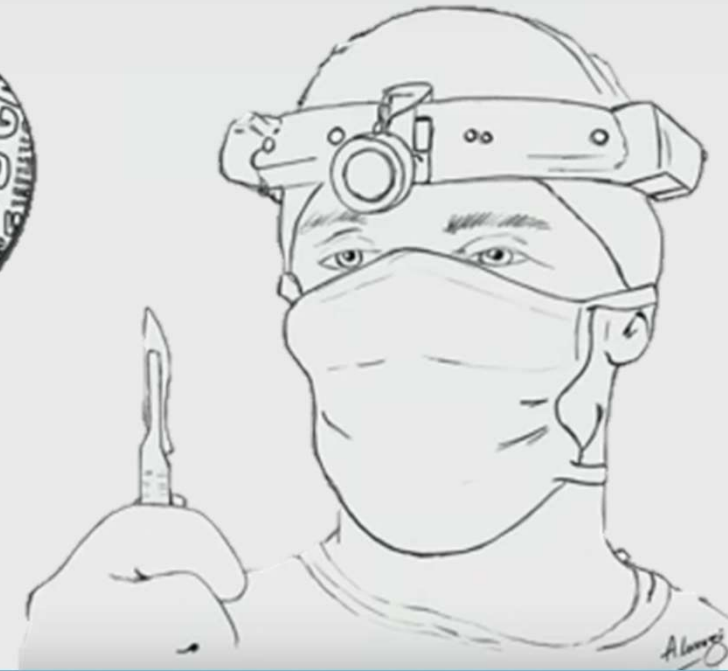
#### AUTHORS

Mohammad Esmaili<sup>1</sup>, Marjan Esmaili<sup>2,\*</sup>, Fatemeh Ghane<sup>1</sup>, Ali Alamdaran<sup>3</sup>

#### ARTICLE INFORMATION

Iranian Journal of Pediatrics: 26 (1): e4293  
Published Online: January 30, 2016

# 3 Pyeloplasty



# Conservative vs Surgery

- Only approximately 1/3 will need surgery
- no tool adequately predicts need for surgery
  - Ultrasound - doesn't correlate with renal function
  - radionucleide imaging- repeated radiation
  - MRU – repeated anesthesia
- No universal agreement on indications
  - Threshold varies among hospitals/surgeons
  - non-clinical factors are also involved

# Threshold for pyeloplasty

- Symptoms
  - Flank pain
- Ultrasound
  - persistent high Degree dilatation (hydronephrosis, APD)
  - worsening of hydronephrosis
  - Cortical thickness < 3.5 mm
- Diuretic Renal scan
  - Obstructed curve (tracer half life >20min)
  - Absolute differential function <35-40%
  - Decrement in differential function >5-10%
- Other factors
  - Family preference/anxiety
  - History of UTI
  - Stones
  - Contralateral kidney function
  - Comorbidities



Journal of Pediatric Urology  
Volume 16, Issue 1, February 2020, Pages 116-120



How I do it

## Feed and wrap magnetic resonance urography provides anatomic and functional imaging in infants without anesthesia

Sila Kurugol <sup>a</sup>, Catherine M. Seager <sup>b</sup>, Hatim Thaker <sup>b</sup>, Jaume Coll-Font <sup>a</sup>, Onur Afacan <sup>a</sup>, Reid C. Nichols <sup>a</sup>, Simon K. Warfield <sup>a</sup>, Richard S. Lee <sup>a,1</sup>, Jeanne S. Chow <sup>a, b, 1</sup>



Journal of Pediatric Urology (2019) 15, 242.e1–242.e9

## The role of renal biomarkers to predict the need of surgery in congenital urinary tract obstruction in infants

D. Kostic <sup>a,\*</sup>, G.P.N.S. Beozzo <sup>b</sup>, S.B. do Couto <sup>a</sup>, A.H.T. Kato <sup>a</sup>, L. Lima <sup>c</sup>, P. Palmeira <sup>c</sup>, V.L.J. Krebs <sup>b</sup>, V. Bunduki <sup>d</sup>, R.P.V. Francisco <sup>d</sup>, M. Zugaib <sup>d</sup>, F.T. Dénes <sup>e</sup>, W.B. de Carvalho <sup>b</sup>

**Potential future predictors:** MRU might be widely used with the feed and wrap technique which combined with dynamic radial VIBE and compressed sensing image reconstruction showed to be a promising anesthesia free modality to obtain MRI in infants. Renal biomarkers such as TGF- B1, MCP-1, ca 19-9 are also in investigational phase

# Pyeloplasty vs Nephrectomy

- nephrectomy is rarely performed in infants with PUJ
- No test accurately predicts recovery of renal function
  - If differential function  $<10\%$ , insert nephrostomy/J stent to see if kidney recovers
- Indications
  - Intraoperative finding of gross dysplasia
  - frozen section analysis of dysplasia

# Timing

- If acute pain or infection, wait 1-2 wk for inflammation to resolve
  - Febrile infection are often sufficiently treated with antibiotics
  - Avoid stent b/c develop inflammation (use nephrostomy if diversion required)

# Objective

- Achieve PUJ which is ...
  - Dependent , funnel shaped
  - Adequately calibrated
  - Watertight
- Principles
  - Redundant Pelvis should be excised to avoid stasis
  - Narrow ureter should be wide enough to have open anastomosis,
  - tissue handling – minimize manipulation (use traction sutures) to prevent edema
  - Anastomosis – make sure it is not twisted

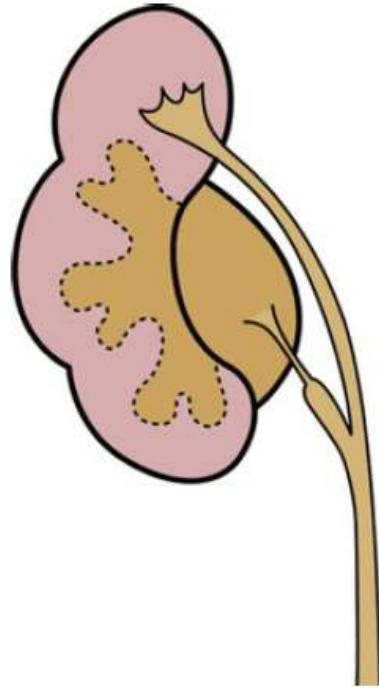
# Types

- **Dismembered technique** ([Anderson Hynes pyeloplasty](#)) suitable for the majority, high success



- complete excision of PUJ (adynamic ureter)
- Correction of high insertion of the ureters
- reduction of renal pelvis
- straightening of lengthy / tortuous proximal ureters,
- transposition of PUJ if obstruction is secondary to an aberrant vessel.

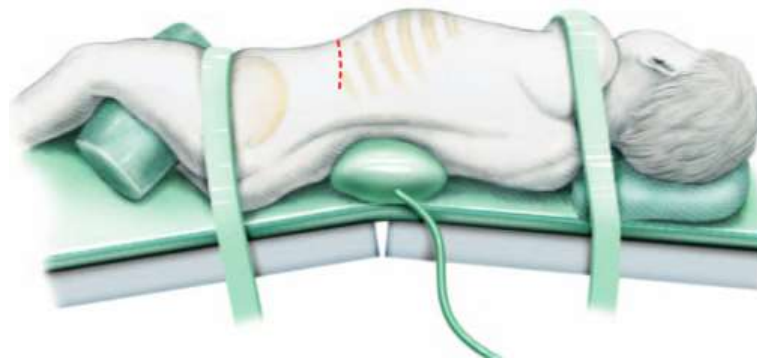
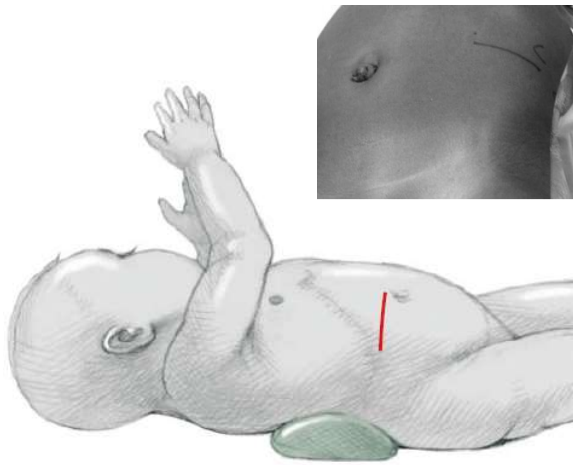
- **Flap techniques** ([Foley Y-V](#), [Spiral](#)) – for UPJ with long stenosis , malrotation, fusion anomalies
- **Endopyelotomy** (ureteral cutting balloon catheter) – success in 70%, inc use in reoperation (>95% success)
- **Ureterocalicostomy** – unusually small intrarenal pelvis, significant scar on reoperation
- **Vascular Hitch**– for crossing vessel with normal ureter and UPJ with peristalsis



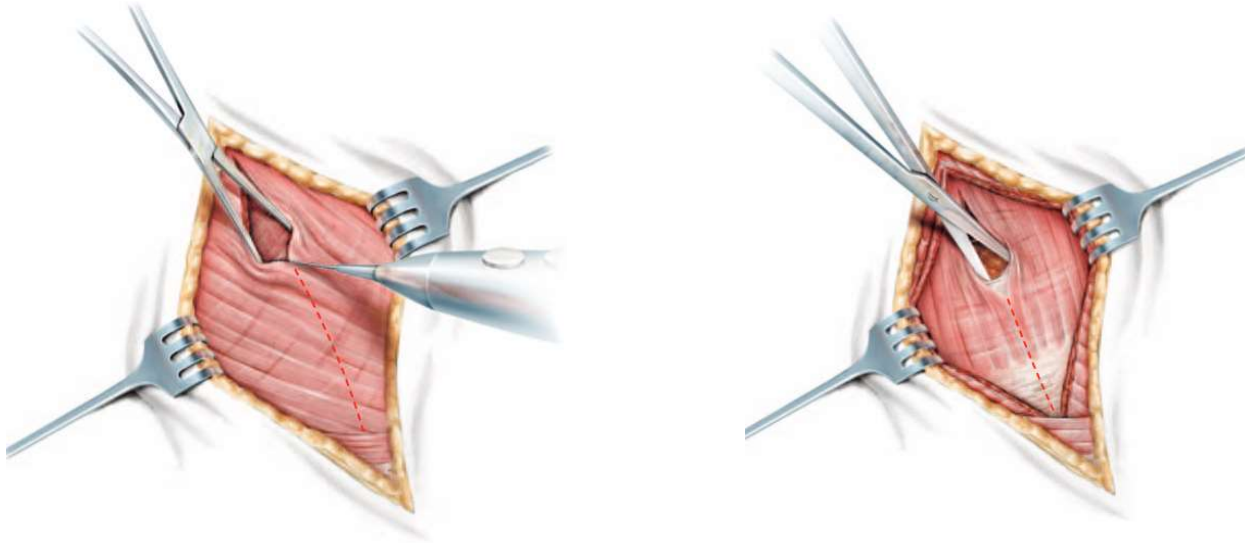
**\*Procedures for UPJO in duplex system** : the lower pole is mostly affected b/c upper pole lacks true renal pelvis. Standard **dismembered pyeloplasty** is preferred if there is good length of the lower ureter. Otherwise a **V-plasty** (anastomosing end of pelvis to the side of upper ureter) will solve the problem. However if the lower moiety's function is low (<5-10%) it should prompt **partial nephrectomy**.

# Approach

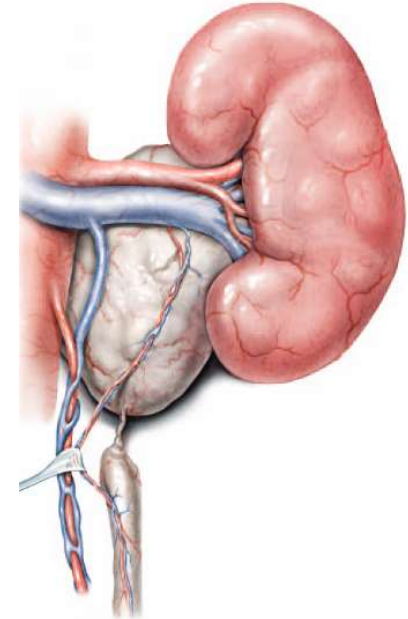
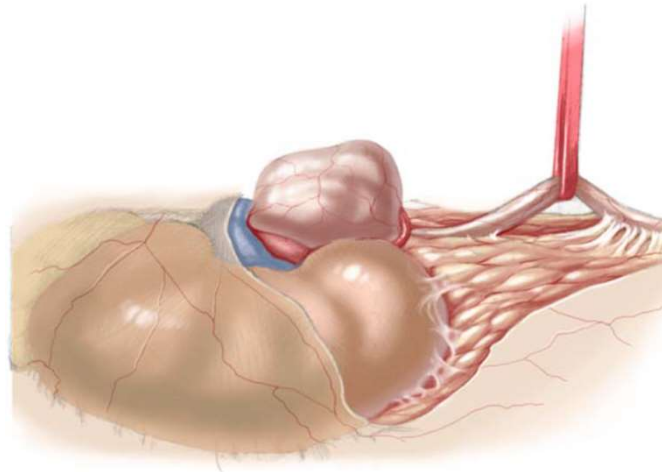
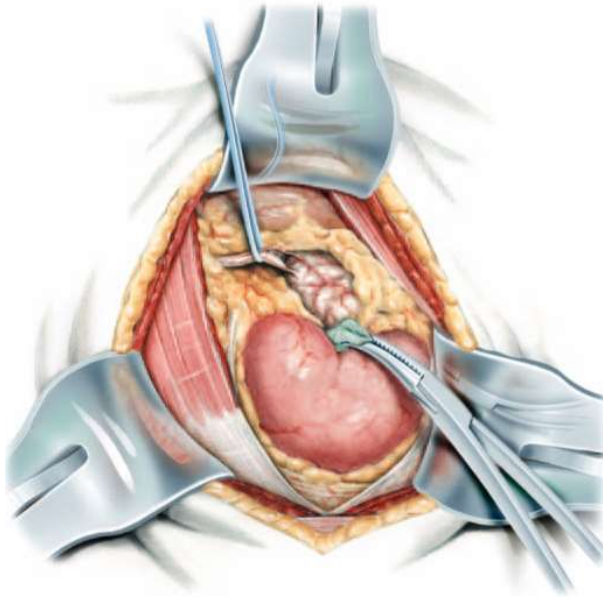
- **Lateral flank**
  - excellent exposure but painful incision
- **Anterior subcostal (extraperitoneal)**
  - for younger children with large renal pelvis
  - Difficult in obese
- **Dorsal lumbotomy**
  - Cosmetic, minimally invasive (muscle splitting) , access other kidney without reposition
  - Requires precise localization (not for malrotated, small, high or intrarenal pelvis and reoperations)
- **Anterior subcostal (transperitoneal)**
  - for reoperations (to avoid scarring)
- **Laparoscopy / Robotic surgery**



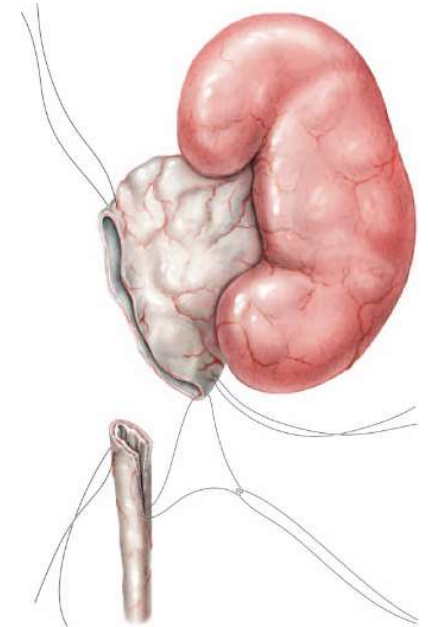
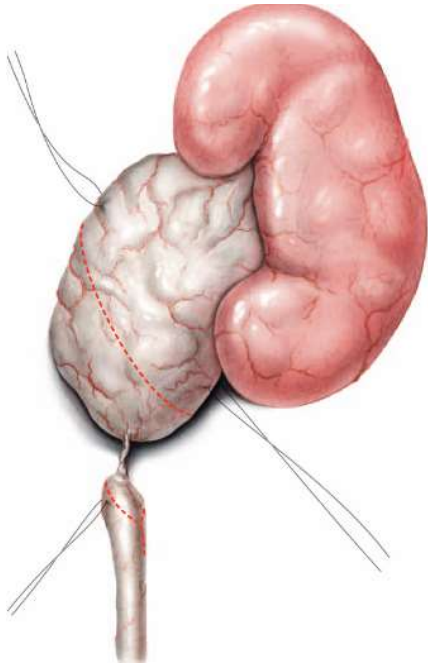
**Incision:** elevate affected side on a roll to expand subcostal space. **Anterior approach** (Horizontal incision from just below tip of 12<sup>th</sup> rib extending anteriorly to edge of rectus). **Flank approach** (incision is made anterior to the tip of the 12th rib, or if necessary, a supracostal 12th rib incision is made). **Dorsal lumbotomy** (from the angle of the 12th rib down to the iliac crest at a point one-third of the distance from the ASIS to the spinal processes)



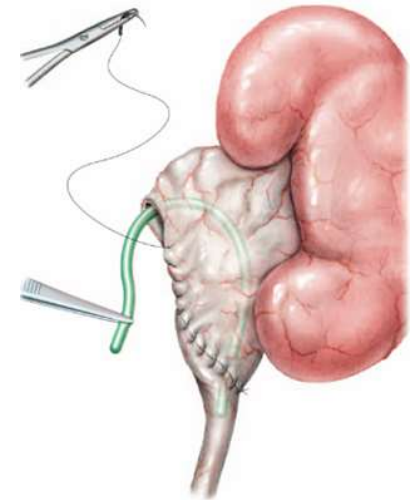
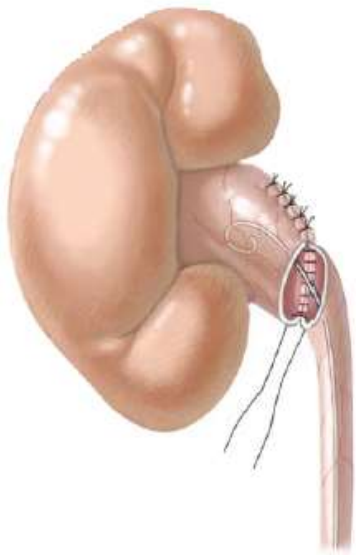
**Entering abdomen:** oblique muscles are cut extending to latissimus dorsi (muscle splitting in infants). Transverse abdominis is split bluntly and lumbodorsal fascia is opened. Retroperitoneum is entered at the rib tip and extended posteriorly along superior edge of the rib.



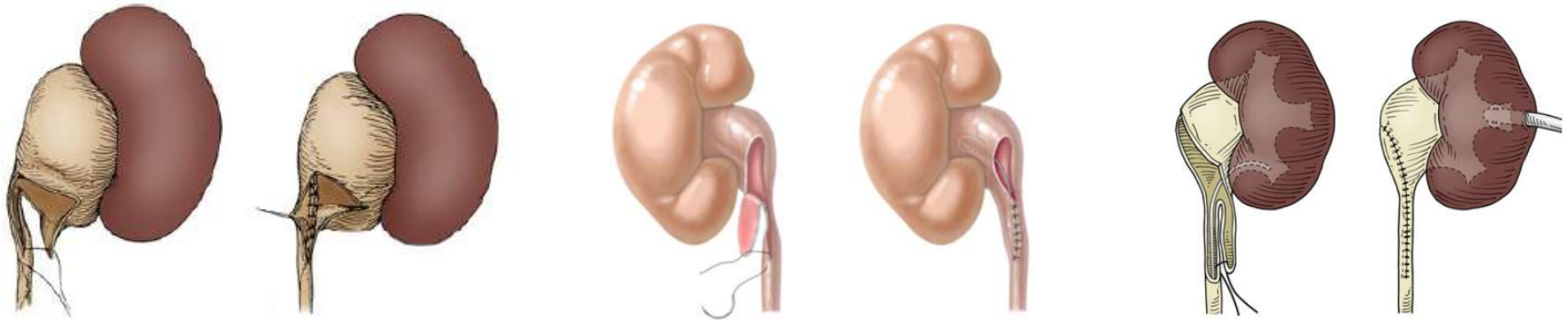
**Exposure:** peritoneum is retracted medially to expose gerotas fascia. Self retaining retractor is placed. gerota's fascia is opened longitudinally. Lower pole is mobilized and kidney retracted laterally to identify pelvis and ureter. Avoid traumatizing blood supply to upper ureter. Kidney doesn't need mobiization(anastomotic torsion occurs when kidney is exteriorized). Ureter & pelvis can be delivered. If the pelvis is dilated, it can be decompressed to facilitate identification of the UPJ



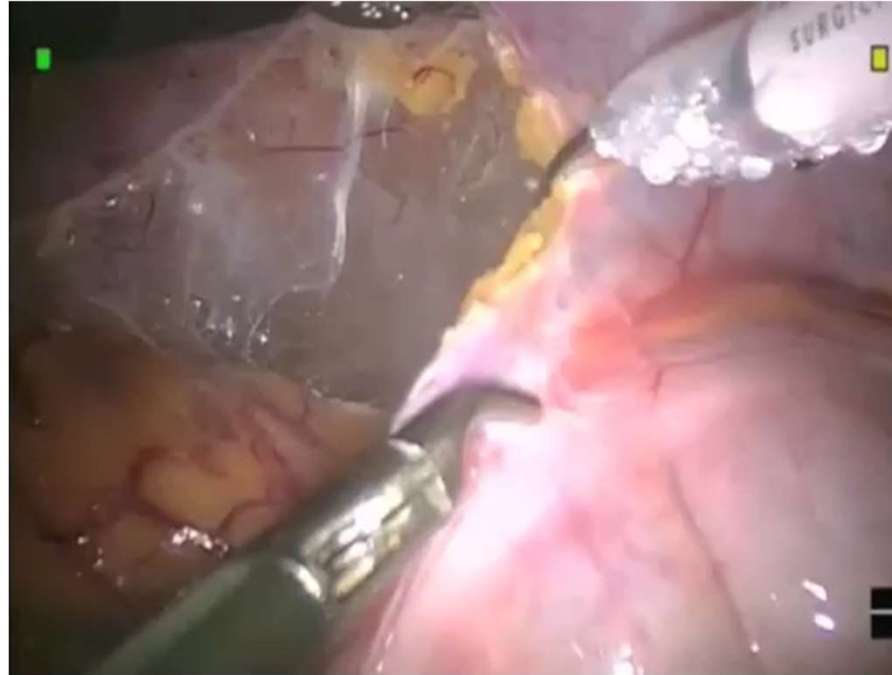
**Resection:** Three absorbable 6/0 sutures are placed (1) at the superomedial aspect of the pelvis, (2) at the inferolateral aspects of the pelvis, and (3) on the ureter about 5 mm below PUJ. Ureter is divided obliquely from anterior to posterior, leaving the medial blood supply undisturbed. The redundant pelvis is trimmed. Ureter is spatulated for 2-3mm posterolaterally.



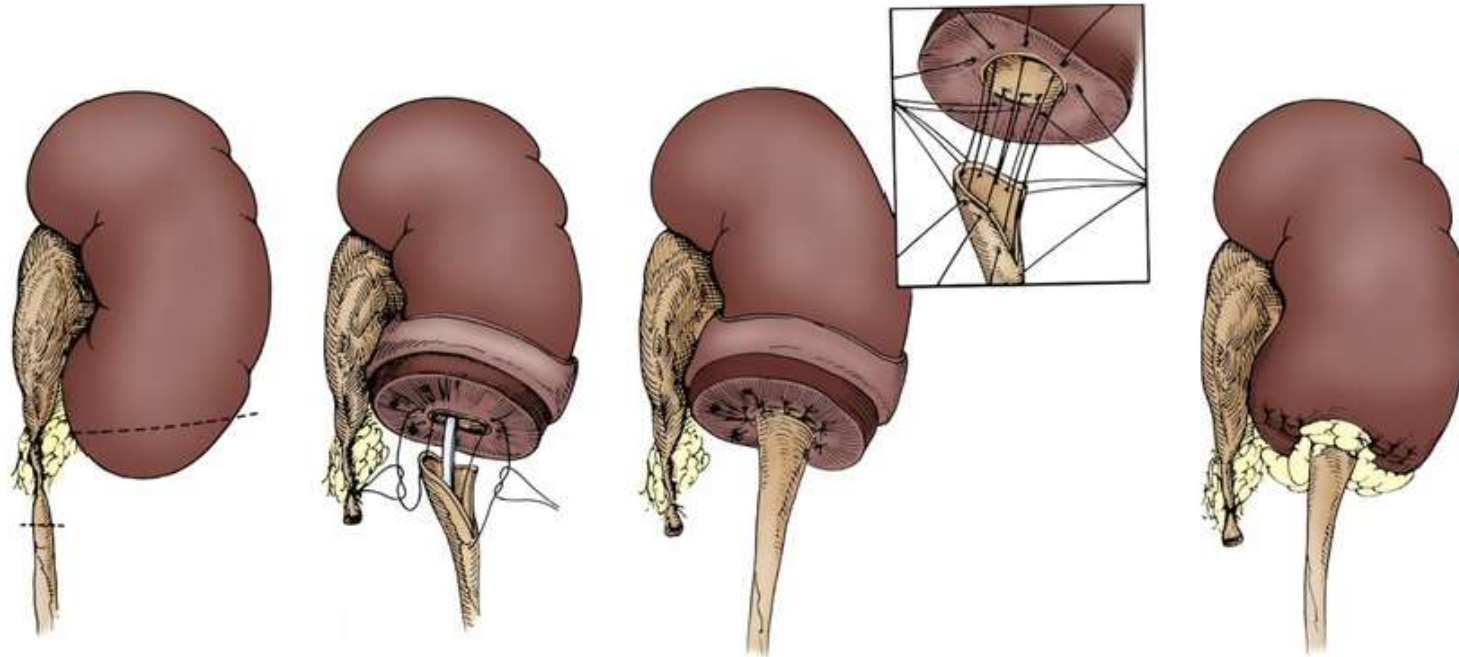
**Anastomosis:** Start from lower margin of ureter to renal pelvis with 6/0 absorbable. Continue interrupted sutures on dependent portion then advance up the anterior wall with interrupted or running sutures. Knots should be outside the lumen. Sutures should include 1mm tissue on each side and be 1.5-2mm apart. If aberrant renal artery, anastomosis should be anterior to the vessel. A ureteric stent may be utilized or a temporary stent might be used to prevent inadvertent picking up and inclusion of the posterior wall. Temporary stent is removed at end of procedure and drain is left near the anastomosis. If VUJ cant be passed with fine feeding tube or wire, consider VUJ obstruction and place a nephrostomy.



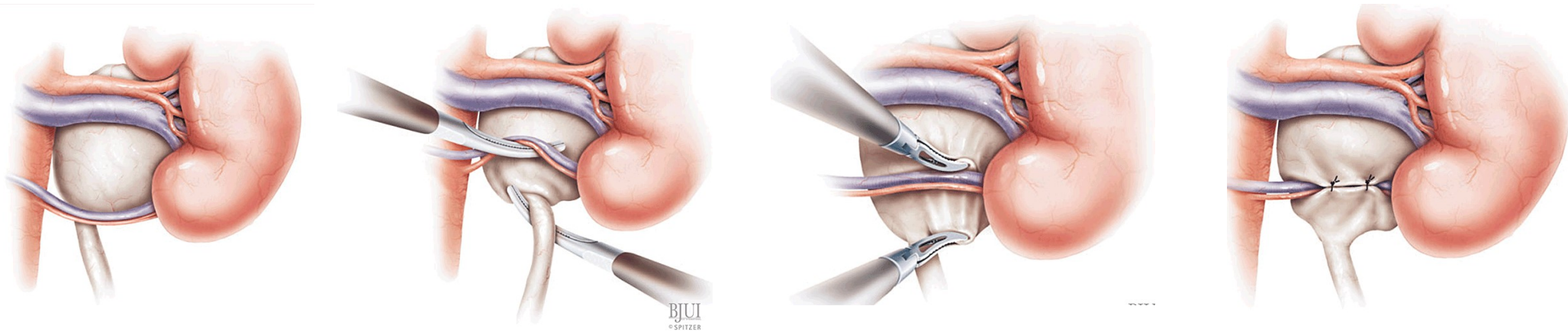
**Flap techniques:** incision on ureter must be adequate covering stenotic area. Flap of equal length from pelvis is based on a broad base. Posterior layer of ureter and flap is sutured. A **Foley Y-V plasty** is best applied to a ureteropelvic junction obstruction associated with a high insertion of the ureter. **Vertical flap** technique may be used when a dependent ureteropelvic junction is situated at the medial margin of a large extrarenal pelvis. A **spiral flap** may be indicated for relatively long areas of proximal ureteral obstruction when the ureteropelvic junction is already in a dependent position.



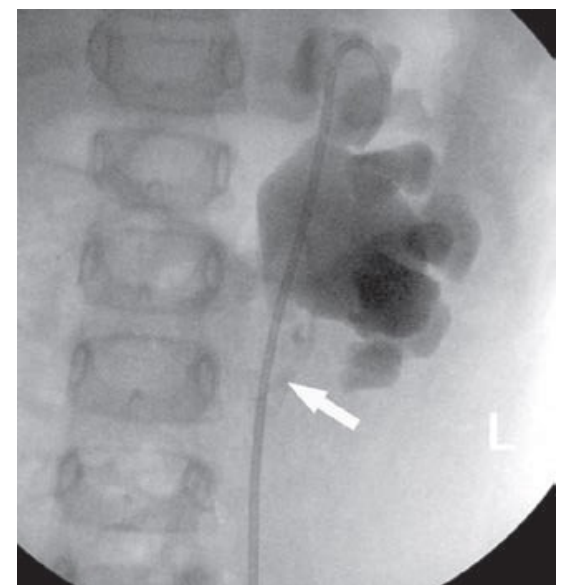
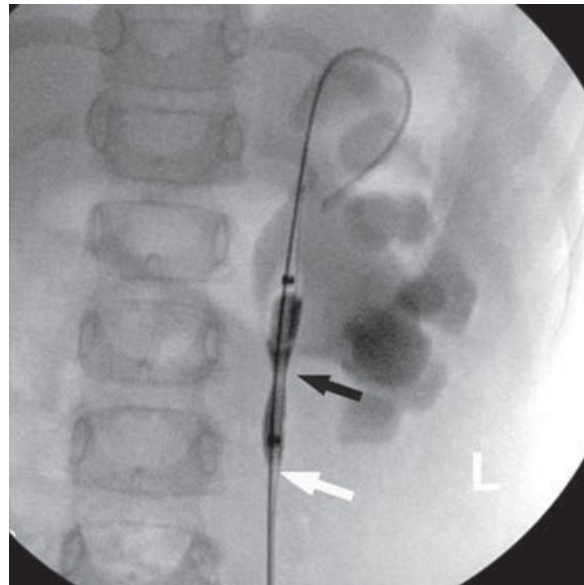
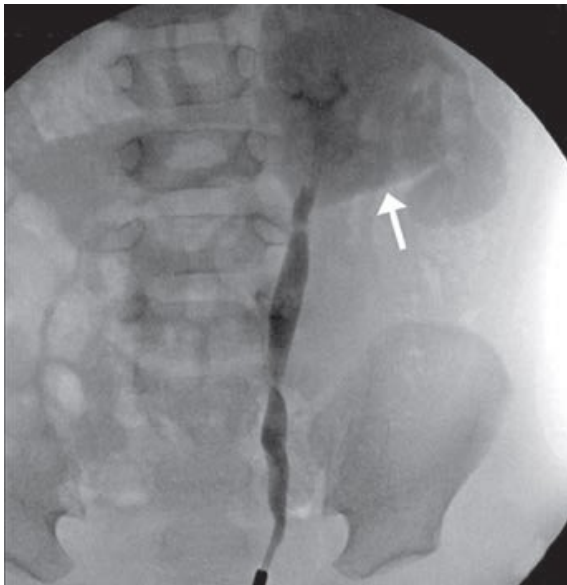
**Laparoscopy and robotic surgery (video)** : laparoscopy is reported in children as young as 2 months. Introduction of robotic surgery has made suturing easier and more precise. Success is similar to open.



**Ureterocalicostomy** : kidney is mobilized as much as necessary to gain access to the lower pole. lower pole arterial supply is ligated and lower pole nephrectomy performed, removing as much parenchyma as necessary to widely expose a dilated lower calyx. Anastomosis of ureter to calyx performed in an open fashion, placing each suture circumferentially but not securing them until the anastomosis has been completed. Stent is left and consideration given to leaving a nephrostomy. The renal capsule is closed over the cut surface of the parenchyma. Anastomosis should be protected with a graft of perinephric fat or with a peritoneal or an omental flap.



**Vascular Hitch** : originally entailed cephalad translocation of crossing vessel and suturing it to tissues around the kidney. Chapman's modification recommended fixing the vessel in a tunnel made of the pelvis, away from the UPJ. Recently has regained acceptance by laparoscopic surgeons because it is easier to perform than pyeloplasty.



**Endopyelotomy** : Guidewire across the UPJ. The Accucise cutting balloon catheter is introduced over the wire in an antegrade or retrograde fashion (percutaneous/ cystoscope) balloon is inflated to identify the site of narrowing which is cut laterally and stent is left for 4-6wk. The need of fluoroscopy makes this option less attractive in pediatrics. CT/MRI is also required preoperatively to screen for accessory vessels unless already documented in a previous surgery.

# Postop care

- **Foley catheter** can be removed after 1 day
- **External drain** is usually removed after 1 day
- **J Stent** can be removed in 2-6wk
  - *Intraoperative **antegrade** stenting can be **difficult to pass VUJ** in infants*
  - ***retrograde** stenting at time of pyeloplasty **adds significant time** to procedure*
  - *consigns patient to another **anesthesia for stent removal** (\*unless extraction string is placed)*
- **Nephrostomy / nephro-uretric stent** can be removed 1-2wk
  - *\*has added benefit of doing **antegrade study** before removal*

Journal of  
**Pediatric  
urology**

FULL LENGTH ARTICLE | VOLUME 14, ISSUE 5, P450-E1-450-E6, OCTOBER 01, 2018

**Outcomes of externalized pyeloureteral versus internal ureteral stent in pediatric robotic-assisted laparoscopic pyeloplasty**

D.I. Chu • D. Shrivastava • J.P. Van Batavia • ... D.A. Weiss • A.R. Shukla • A.K. Srinivasan • • •  
Show all authors

Published: May 07, 2018 • DOI: <https://doi.org/10.1016/j.jpurol.2018.04.012> • Check for updates

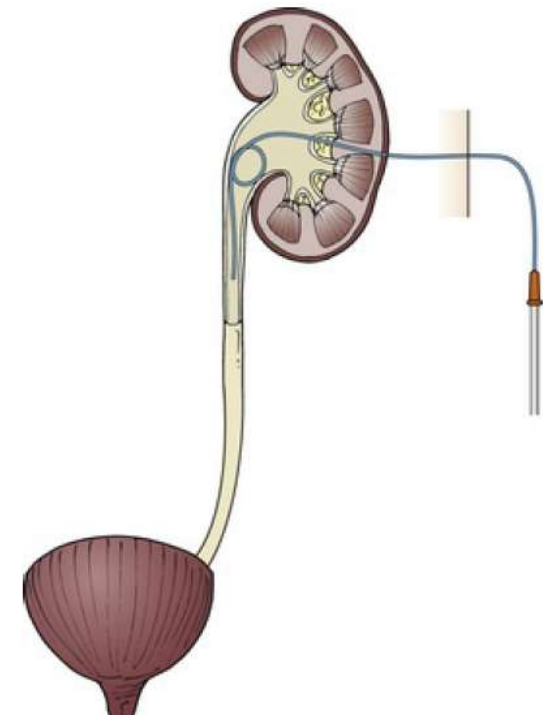
**UROLOGY**

PYELOPLASTY | VOLUME 101, P38-44, MARCH 01, 2017

**Dismembered Pyeloplasty in Infants 6 Months Old or Younger With and Without External Trans-anastomotic Nephrostent: A Prospective Randomized Study**

Farouk M. Nasser • Ahmed M. Shouman • Mohammed S. ElSheemy • • ... Waleed Ghoneima •  
Hany Morsi • Hesham Badawy • Show all authors

Published: September 28, 2016 • DOI: <https://doi.org/10.1016/j.urology.2016.09.024> • Check for updates



**Drainage tubes** : Only few authors recommend stentless anastomosis which mandates extra careful tissue handling to prevent edema resulting in leak and obstruction. Stenting is strongly indicated if contralateral kidney is not good and if procedure is redo surgery. **Nephrostomy** can be used in small infants if can't pass stent. Type of stent is a matter of choice that differs among surgeons (**DJ stent** or **nephro-uretric stent**: brought out through a middle calyx/upper part of anastomosis). The duration of stent between 1-4 weeks has no difference. **External drainage** (penrose) is usually not indicated. **Foley catheter** also not indicated unless we expect VUR as a result of DJ stent insertion.

# Outcome

- **Nephrectomy** <2%
- **Failed pyeloplasty** (Success >95%)
  - Increasing pain, dilatation, infection
- **Prolonged extravasation**
  - Can be sent home with drain
  - If leak significant insert stent or PCN-
- **Delayed drainage** through anastomosis (when nephrostomy used)
  - Patience b/c 30% open in 3mo
- **Recurrence** <1%
  - scarring or fibrosis
  - nondependent anastomosis
  - Ureteral angulation/malrotation
  - Uretric narrowing distal to anastomosis

# Follow-up

- **Ultrasound**- to ensure that the hydronephrosis is improving
  - First - 6-8wk
  - Last - 1yr (imaging beyond this point is debatable)
- **Renal scan** – if there was marked disparity in renal function preop (70%/30%)
  - First - 3 mo
  - Second - 1-2yrs after

# References

- **Hollcomb and Aschcraft pediatric surgery** 7<sup>th</sup> E (2020)
- **Kelalis King Belman textbook of pediatric urology** 6<sup>th</sup> E (2020)
- **Campbell Walsh Urology** 12<sup>th</sup> E (2020)

