Posterior urethral valve:

management from antenatal period to puberty

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How well can you manage PUV?

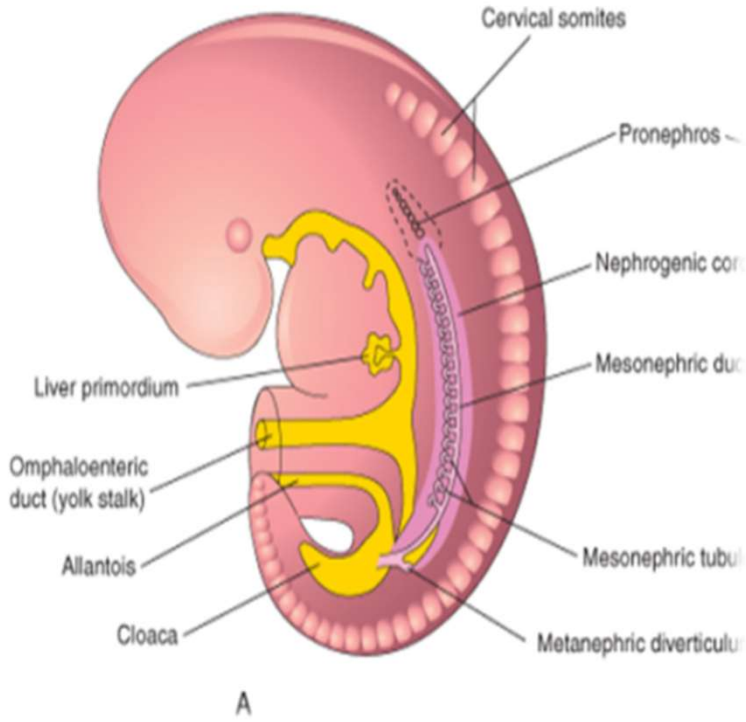
- You are consulted to see a **12 year old** boy who underwent valve ablation at 1 year of age. Cystoscopy has been done and incomplete ablation has been ruled out. He is approaching **ESRD** and has been counseled for renal transplant. His family also complains of persistent diurnal urinary **incontinence** and **recurrent UTI**.

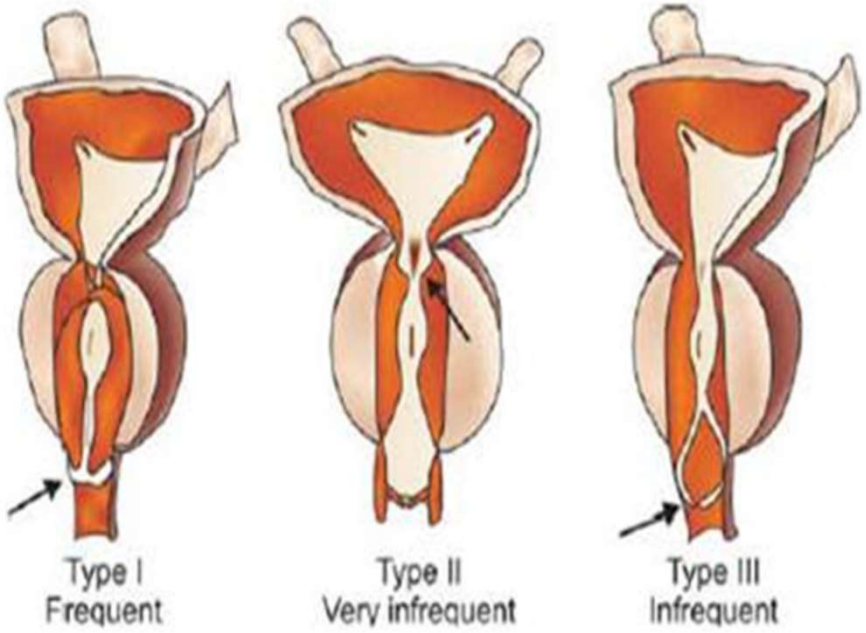
1. *What could be the problem of this child? And why?*
2. *What leads to this problem?*
3. *How would you approach this boy's further evaluation? (first step)*
4. *What investigation can help treat his disorder?*
5. *What treatment would you recommend?*

References

1. Hollcomb and Ashcraft pediatric surgery, 2020
2. Kelalis-king-belman pediatric urology , 2019
3. obstetric imaging, fetal diagnosis and care, 2018
4. Pediatric endourology. 2014
5. Pediatric urogenital radiology, 2008

But first…What is PUV / COPUM ?





Introduction

- A common anomaly in pediatric urology and a common cause of CKD in children
- antenatal cases are far more than those that are actually delivered live.
- irony of relatively simple diagnosis and surgical intervention but significant long-term consequences
- *No studies done in Ethiopia*

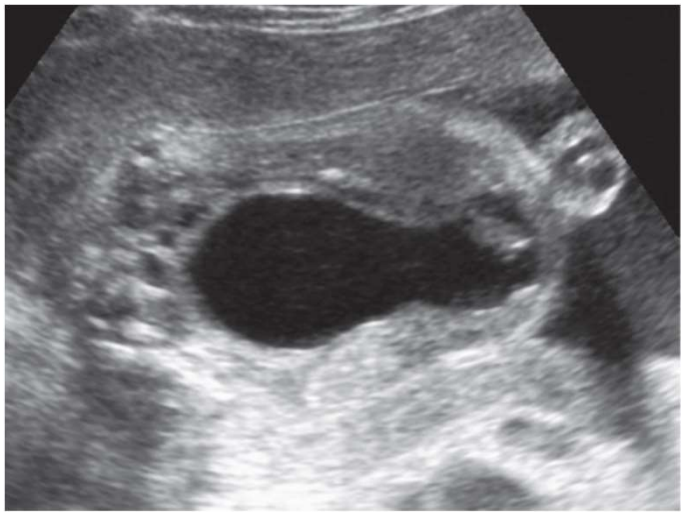
Outline

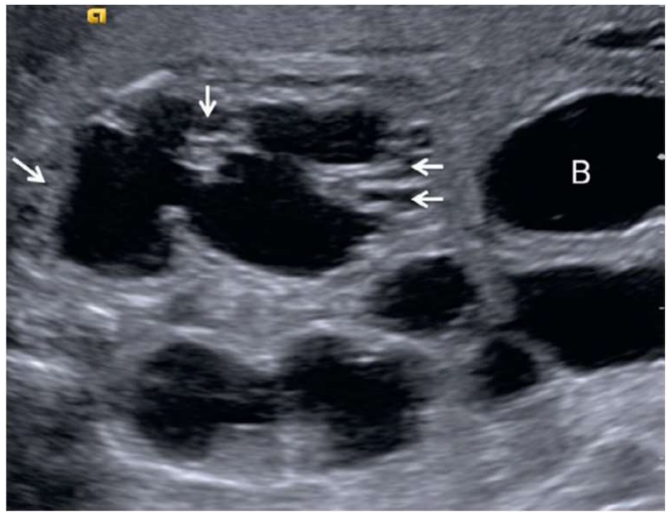
1. Antenatal Management
2. Postnatal Diagnosis
3. Initial Surgery
4. Follow up
5. Transition to adult urology/nephrology

1. Antenatal Management

* *fetal surveillance*
* *fetal interventions*

Counseling: Poor prognostic factors





- Fetal ultrasound: dilated thick walled bladder, keyhole sign
- Fetal ultrasound: renal dysplasia

Counseling: Poor prognostic factors

- Younger gestational age at delivery
- Lung hypoplasia
  - Oligohydraminos
- Renal Dysplasia
  - Renal cortical cysts
  - * *echogenic kidneys, loss of cortico medullary differentiation*
- Loss of renal function
  - Elevated fetal urine electrolytes and β_2-microglobulin

1.1. antenatal observation
(no intervention)

- Applicable for most cases of PUV
- Communication between the **obstetrician and the pediatric urologist** to establish a clear plan of action for the family
- antibiotic prophylaxis for the neonate before postnatal imaging
- Families may also opt for termination of pregnancy. (In severe cases)

1.2 percutaneous vesicoamniotic shunt (VAS)

- **Prerequisite**: **Karyotype** first to rule out chromosomal abn (12%)
- **Indication**: Normal amniotic fluid that dec to **oligohydraminos** on follow up (previously for oligohydraminios when urine is hypotonic) and especially before **24 wk gestation**
- **Aim**: To restore amniotic fluid for **pulmonary development**
- **Outcome**: doesn't allow bladder to cycle so may not prevent **ESRD**
- **Risk**: 5% fetal loss, 70% preterm, urinary ascites (20%), additional procedure if shunt blocked/displaced, gastroschisis (10%)

1.2. percutaneous vesicoamniotic shunt (VAS)

- Play Video => Vesicoamniotic shunt is placed under US guidance using a double-pigtail catheter inserted through a narrow trocar.

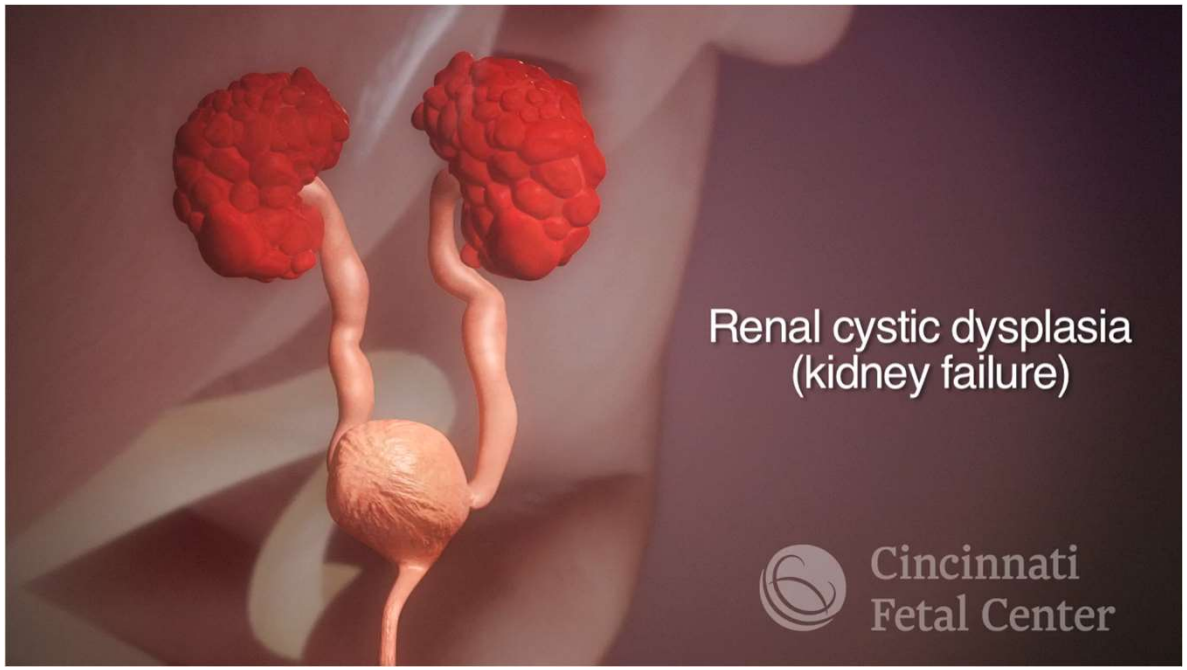
PLUTO trial (Percutaneous Shunting in Lower Urinary Tract Obstruction)

- Initiated due to **lack of evidence** regarding bladder drainage
- **International RCT** comparing VAS versus no intervention
- study **closed early** due to low recruitment
- 16 pregnancies in the VAS group and 15 in the conservative
- *Data show **improved perinatal survival** with intervention but **no difference in ESRD***

1.2. fetal cystoscopy (percutaneous endoscopic ablation)

- Perinatal survival advantage but not more than VAS
- using a 1 mm fetoscope in antegrade fashion and valve incised through bladder neck
- **Nd: YAG laser** is used (previously hydroablation & mechanical)
- Complications include fetal mortality, urethrocutaneous and urethrorectal thermal fistulas

1.2. other



- Play Video => Amnio-infusion for fetal renal failure

2. Postnatal Diagnosis

* *Voiding cystourethrogram (VCUG)*

Imaging in PUV

- Ultrasound - initial
- VCUG - definitive
- Renal scan – if abnormal parenchyma or high grade VUR
- MRU (dynamic, contrast enhanced) – alternative to renal scan

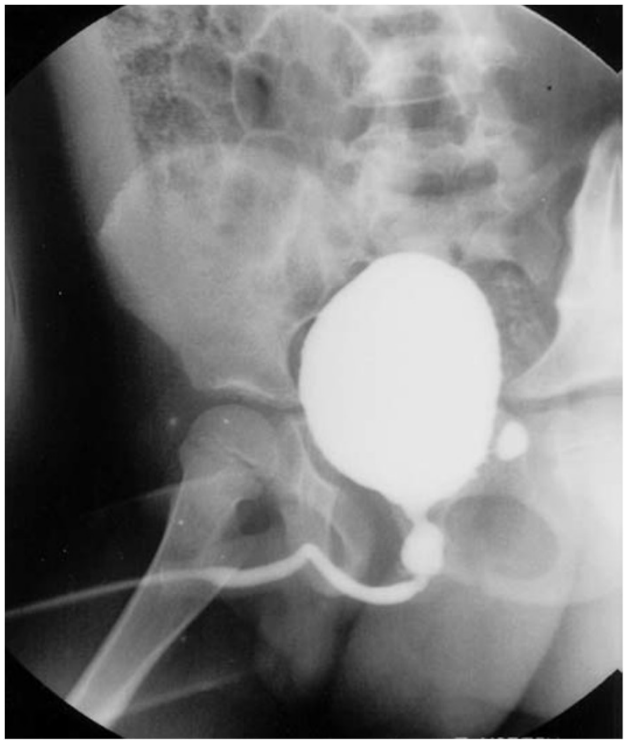
When to request for a VCUG

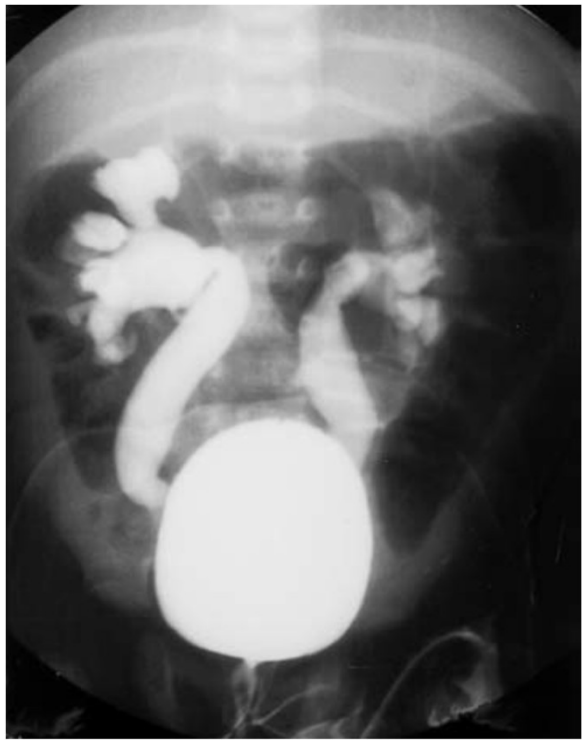
- Timing with regard to age: after ultrasound shows abnormalities, in the neonatal period
- Timing with regard to infection: debatable issue
  - ? During infection
  - As soon as clinical symptoms have disappeared
  - ? After 1 week
  - ?After 2 weeks

Considerations to conduct CUG

- **Catheterization**
- **Bladder capacity**
  - **Infants =** 7 x Wt in kg
  - **Older children =** (age +2) x 30 (koff)
- **Urografin** (dilute contrast – 120/5ml at 30 cm H20 pressure)
- **Digital Flouroscopy**

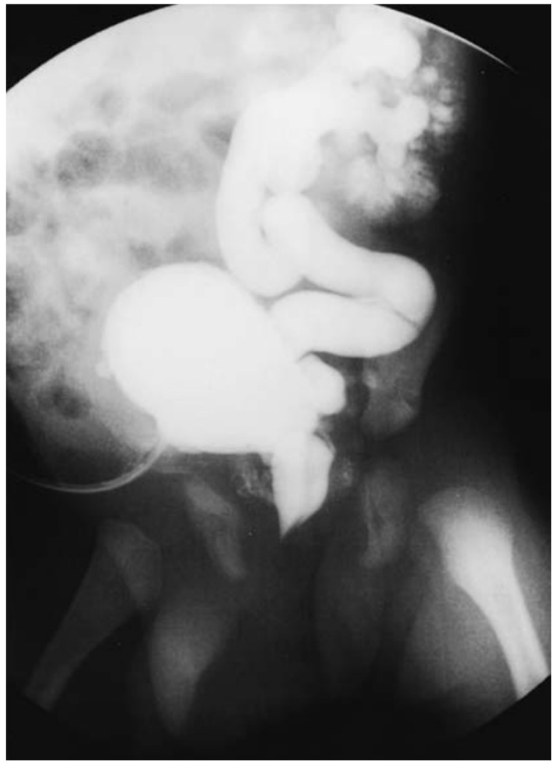
Interpreting the VCUG





- VCUG: bladder diverticula, passive reflux

Interpreting the VCUG



- VCUG: Valves seen (“sail in the wind” sign), intrarenal reflux

3. Initial Surgery

* *cystoscopic valve ablation (TUR valve)*
* *vesicostomy*

Initial Surgical intervention for PUV

- Valve ablation
- Temporary diversion
  - Vesicostomy
  - *Supravesical diversion (ureterostomy/pyelostomy)*
- **management of pop off mechanisms*
- ***Circumcision

3.1. Cystoscopic valve ablation

Preop optimization

- 5-8 F feeding tube
- Cr = 0.4 or stabilize
- Electrolyte
- Prophylactic antibiotics

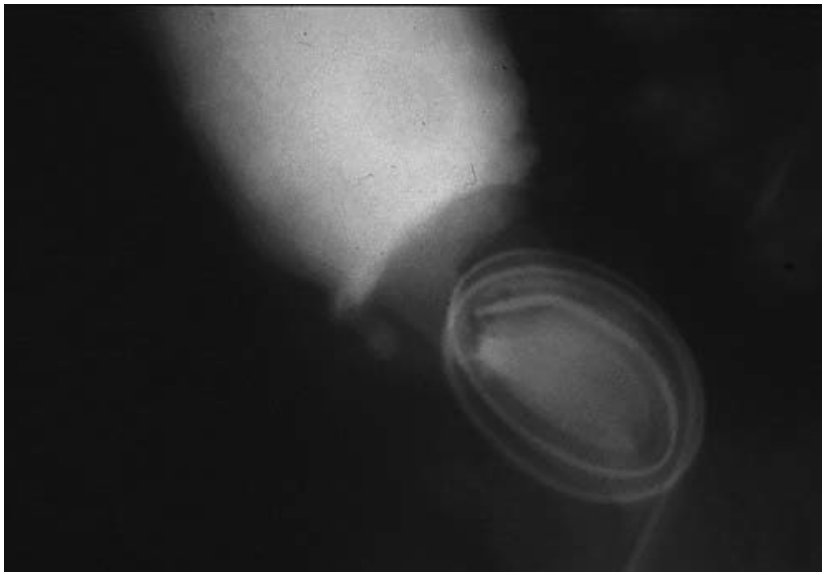
Complications

- Incomplete ablation (<10%)
- UTI
- Bleeding
- Stricture

Postop instructions

- Feeding tube 1-2 days
- Prophylactic antibiotics
- VCUG after 3mo
- ?scope after 2-4 weeks
- Long term follow up

Initial Surgical intervention for PUV



- Sometimes it is difficult to pass an 8F feeding tube b/c it coils in dilated prostatic fossa. coudé-tip catheter allows passage over hypertrophied bladder neck

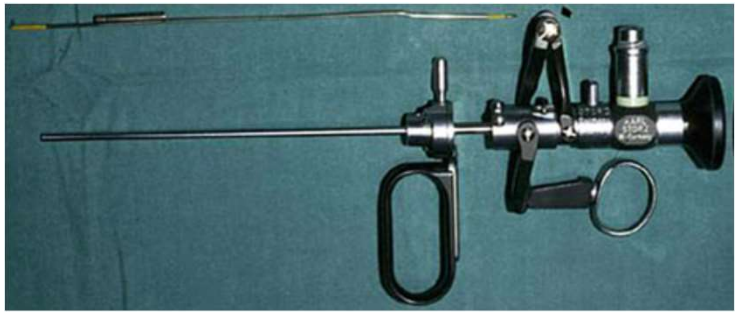
3.1. Cystoscopic valve ablation

Pediatric cystoscope (6-7.5 Fr)

- 3F bugbee electrode

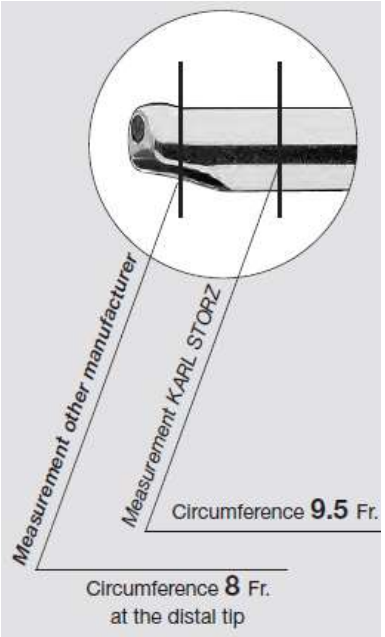
Pediatric resectoscope (11 Fr/ ?9.5)

- -- cold knife



Other methods - *Fogarty balloon, Nd:YAG laser, Hot loop*

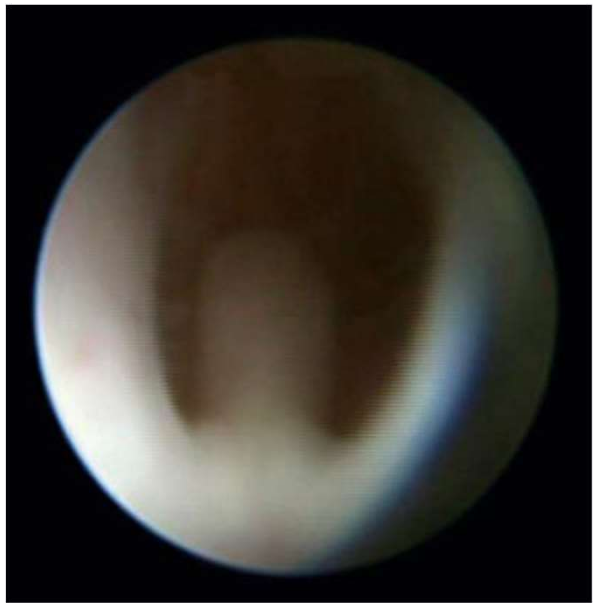
3.1. Cystoscopic valve ablation



8F Pediatric optical urethrotome and accessories

(From Karl-storz pediatric surgery catalog 2010)

3.1. Cystoscopic valve ablation



Cystoscopy: type 1 PUV (incision at 12 o'clock or at the 5 and 7 oclock).

** Valve incision requires only disrupting the integrity of the valve; overzealous attempts to fully excise, raises the risk of stricture formation

3.2. Vesicostomy

Indications

- VLBW (cant accommodate scope)
- After ablation/catheterization => Impaired renal function, high bladder volume and upper tract deterioration

Aim

- For Bladder to cycle and grow

Postop care

- Prophylactic antibiotics

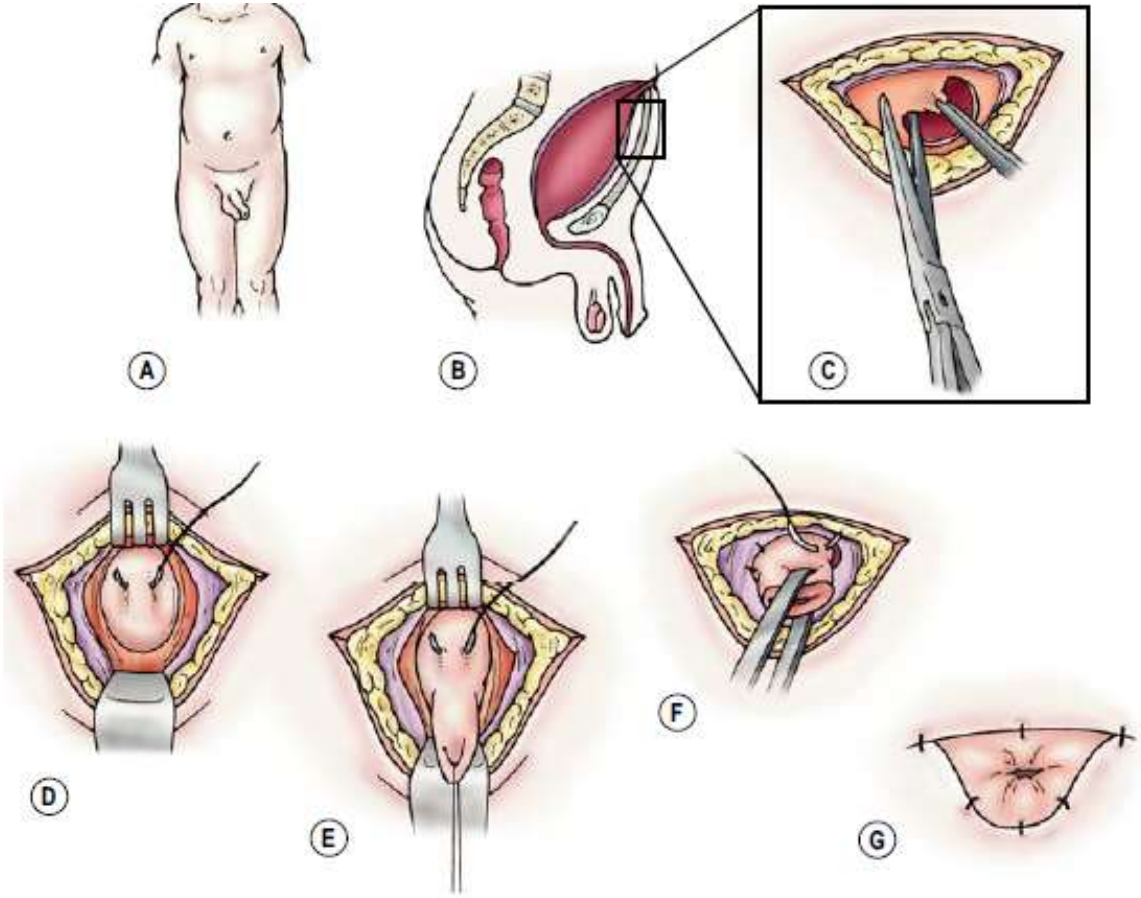
Complications

- Stenosis
- Prolapse

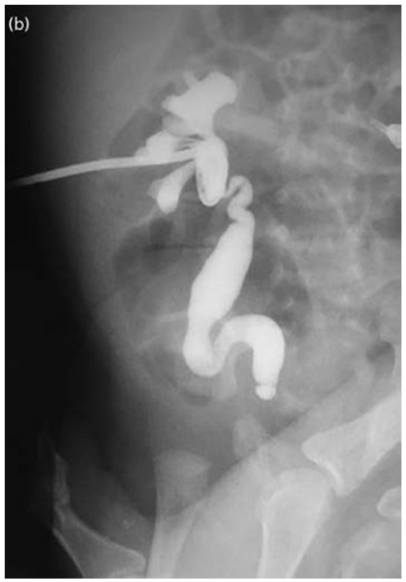
When to close

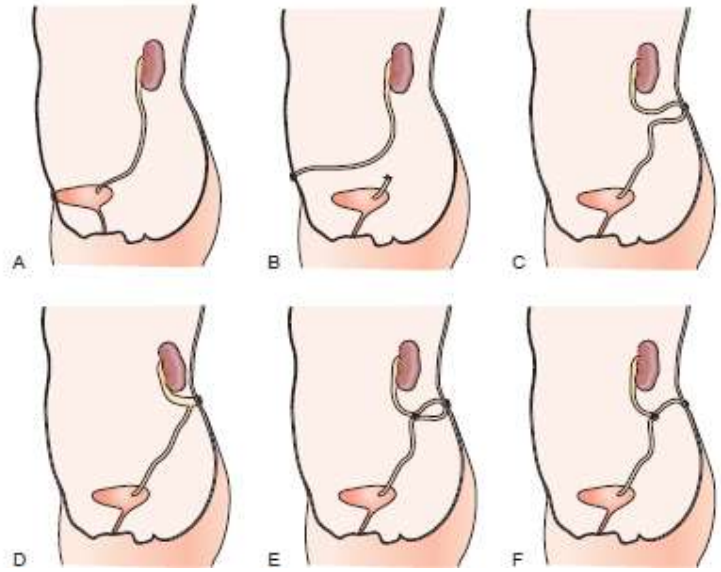
- Upper tract stabilized and child old enough for simultaneous ablation (1-3yrs)

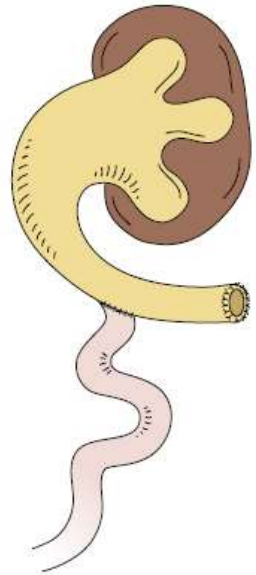
3.2. Vesicostomy



3.3 Supra vesical diversion

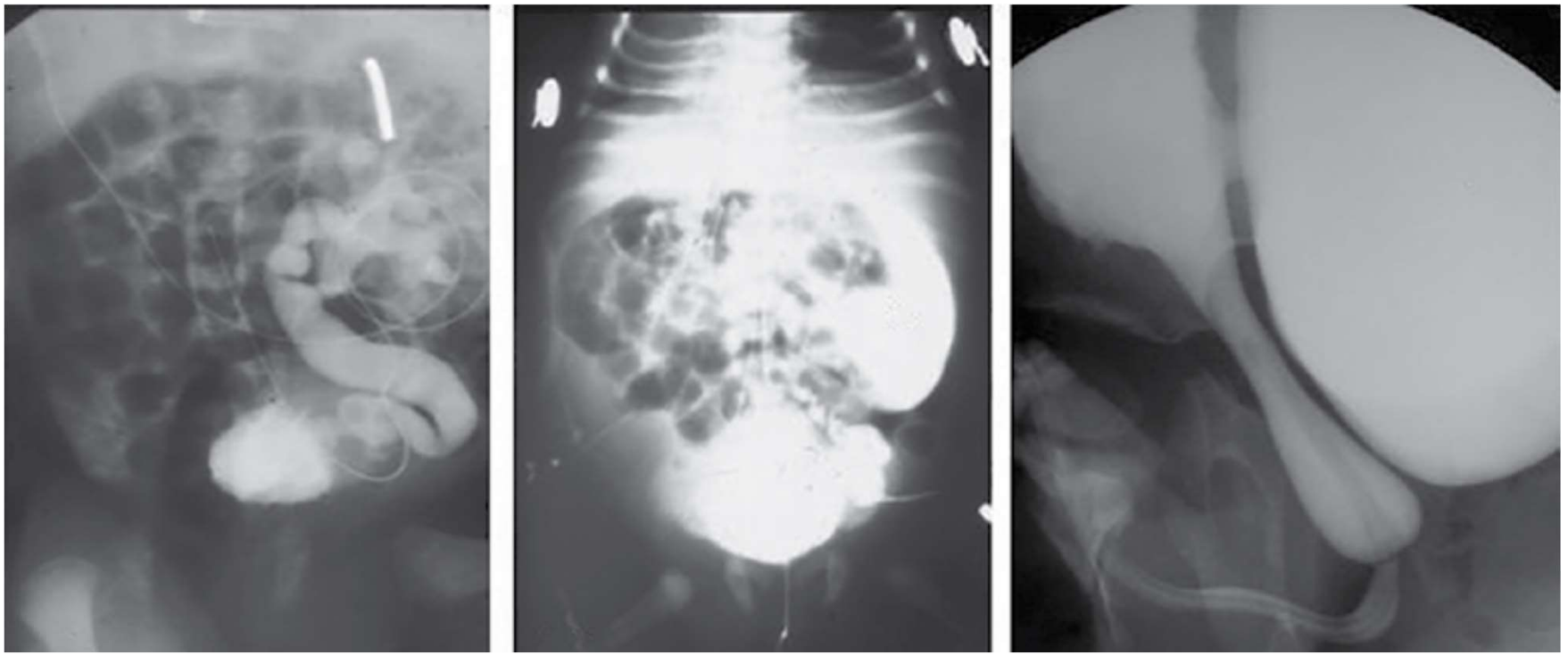






Long standing controversy. *reserved for the rare cases where ablation and vesicostomy fail to improve upper tract drainage. Sober-en-T ureterostomy prefered

3.4 Pressure “pop-off” mechanisms



- VURD , urinoma, diverticula

3.4 Pressure "pop-off" mechanisms

- **Calcyceal rupture** with Urinoma / urinary ascites (5-15%)
  - **Feeding tube** may decompress the bladder and upper urinary tract sufficiently to permit the fornix to seal
  - **Percutaneous drainage** if extravasation and Cr continue to inc, resp compromise, infection, hypertension or parenchymal compression
  - **Paracentesis** for urinary ascites (correct fluid-electrolyte & distention)
- **VURD** (valves, unilateral reflux, renal dysplasia)
  - **? Nephroureterectomy** should be considered at some point
  - **Consider preserving the ureter**
  - ** remaining kidney should be monitored carefully
  - Indication for pretransplant nephrectomy are uncontrollable HTN & polyuria
- **Bladder diverticula**
  - **Do not excise** b/c they provide low pressure storage capacity
  - Intervene if complicate with urinary retention or infection

4. Follow up

- VUR
- Incontinence
- * valve bladder syndrome

Prognosis

- Depends on
  - status of kidney and bladder at **diagnosis**
  - method of **bladder management** as child grows
- ESRD/CKD occurs in 40% of the boys
- A majority of these boys have **voiding dysfunction**
  - Initial overactivity >> low contractility >> high capacity bladder
  - Problems manifest as incontinence and/or persistnet hydronephrosis

4.1. Vesicouretral reflux

- **50% of PUV** have VUR (Unilateral 25%, bilateral VUR in 25%)
- With successful ablation a majority will improve
- Follow up with antibiotics, periodic upper tract imaging, VCUG
- Antibiotic prophylaxis is continued until the upper tract dilation improves
- VUR may not resolve for as long as 3 years after initial treatment

4.1. Vesicouretral reflux

- VUR should be corrected if **breakthrough infections** occur or if it **remains high grade**
- Focus on managing the bladder (reimplant rarely offered)
- Some argue management based on urodynamic data (If bladder function is optimal, then reimplantation)
- High complication 15–30% (persistent reflux or ureteral obstruction, compromise bladder compliance, worsen the failing kidney)
- Endoscopic treatment & transureteroureterostomy into the nonrefluxing ureter is also an option.

4.2. urinary incontinence in PUV

- Self reported incontinence varies 0%-70% (mean 19%)
- 55% urodynamic abnormalities may persist after ablation
- 50% have ongoing incontinence into childhood
- Boys with PUV **achieve urinary continence later** than age matched controls (6 to 7 yrs in less severe valves)

4.2. causes of urinary incontinence in PUV

*Ironically, child **withholds urine** in an attempt to be dry and aggravates the situation*

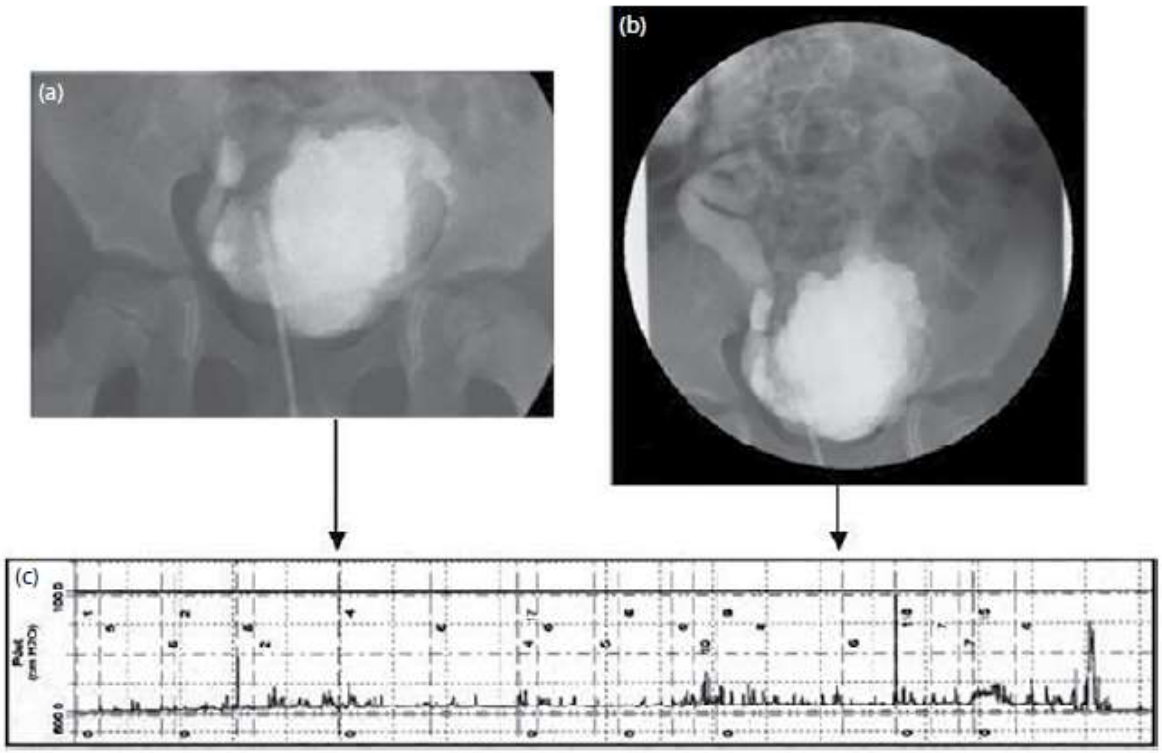
- **Incomplete ablation** (high pressure voiding)
- **Detrusor abnormalities** (hypercontractile or myogenic failure)
- **Detrusor sphincter dysinergia- DSD** (incomplete emptying)
- **Renal tubular damage** (polyuria)
- **Valve bladder** (poorly compliant bladder with inc pressure causing secondary uretral obstruction)

4.2. treatment of incontinence in PUV

a critical time to follow a boy with valves closely is around the time the child begins to express an interest in toilet training

- *Voiding Diary* (urine output, fluid intake)
- regimen of timed voiding is effective for **most**
- double voiding in those with **upper tract diltation**
- overnight catheter drainage and CIC will provide continence, **lower storage pressures**, and preserve **renal function** for most
- For persistent urinary incontinence, *or* approaching ESRD, ***videourodynamics*** *offer an invaluable tool to optimize*

4.2. treatment of incontinence in PUV



- Video urodynamic study

4.3 Valve bladder syndrome

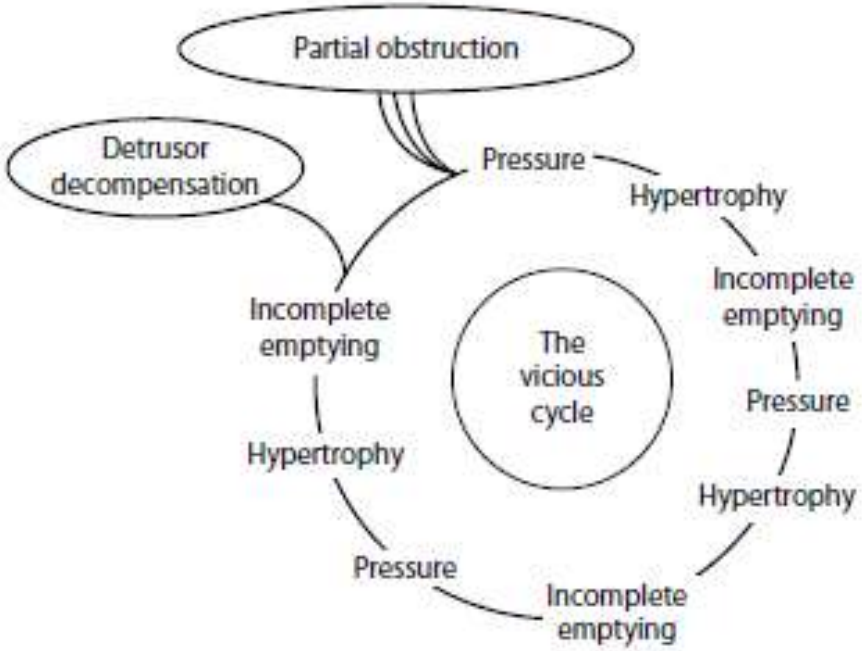
In periodic imaging on followup, Renal deterioration without infection may be a sign of bladder dysfunction.

- 3 basic components
  - chronically distended bladder
  - Dilated refluxing ureters
  - dysfunctional renal units
- **3 contributing factors**
  - Poor bladder compliance & high voiding pressure
  - Residual urine volume
  - Polyuria
- Lower tract evaluation with videourodynamics is important.

4.3. The Valve Bladder Syndrome

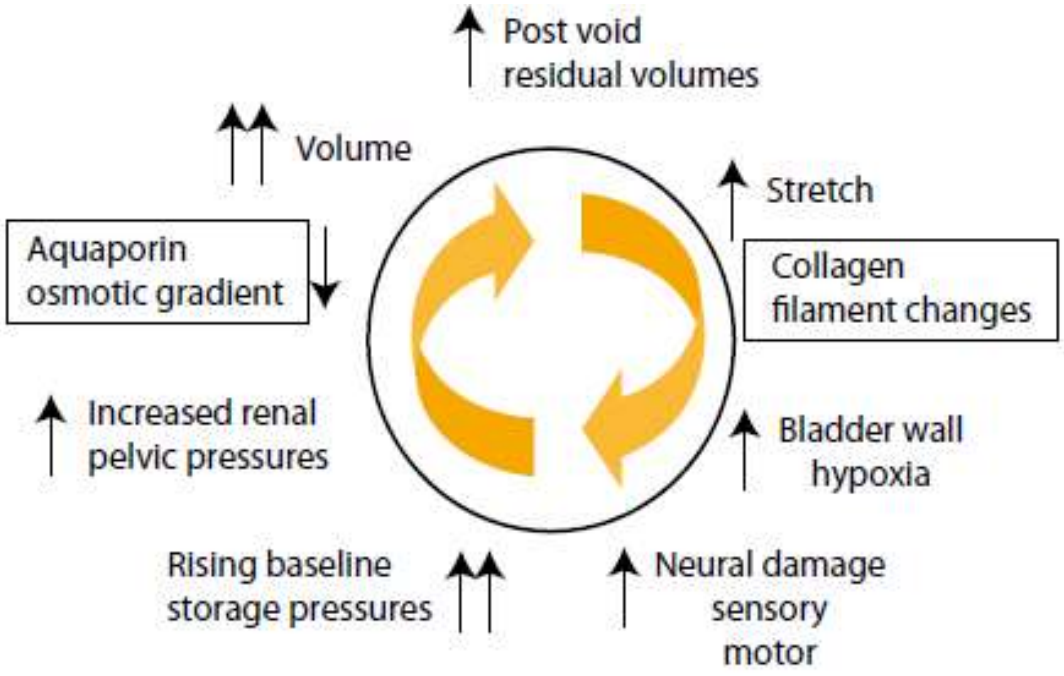
Damaged organ	Complication	Natural history
Obstructive uropathy	Reversible renal failure	- Improve with initial treatment - recur with bladder dysfunction
Renal Dysplasia	Irreversible renal failure	- progresses, Limit growth, HTN
Tubular injury	Nephrogenic diabetes incipidus -Renal tubular acidosis	- Progresses, rapid bladder filling (hydronephrosis, incontinence) - Impaired growth, bone demineralization
Ureteric diltation	-Inability to coapt	- improve initially but HN persists
Bladder dysfunction	- Fibrosis (obst, infection, surgery) - Low volume, low compliance - Poor sensation to high pressure -hypercontractile>>Myogenic failure - Bladder neck hypertrophy	- Life long problems (change with age) - incontinence, poor emptying - High bladder pressure (obstruction of reimplant, progressive upper tract damage)

4.3 Valve bladder syndrome



*schematic concept of how **partial outlet obstruction** affects the **bladder cycle** and over time alters the bladder

4.3 Valve bladder syndrome



*schematic showing **integration of upper and lower urinary tract dysfunction**

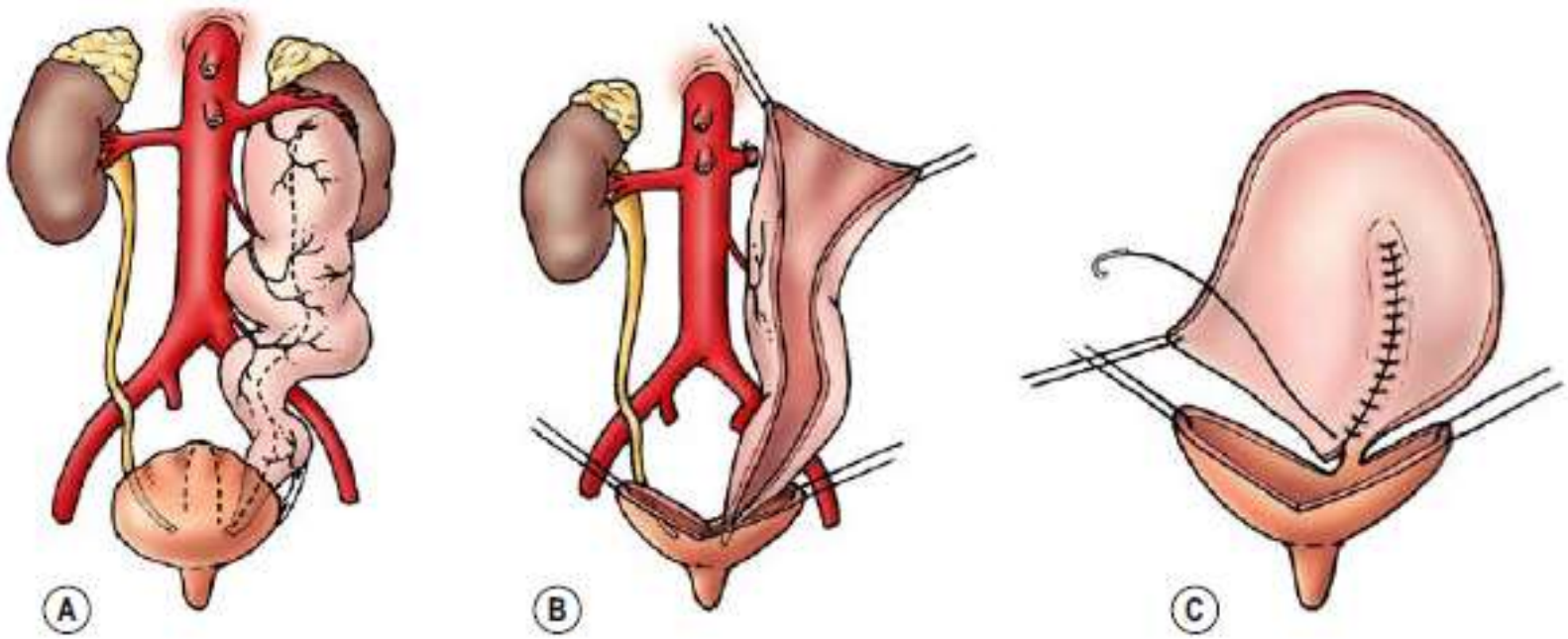
4.3 Valve bladder syndrome

- **Bladder management**
  - **Urodynamics** - Monitoring PVR, flow rate, voiding pressure;
  - **Ultrasound –** dangerous bladder dysfunction, response to therapy
  - **Behavior modification** - Timed voiding, double voiding
    - Other = biofeedback, pelvic floor exercise
  - **Overnight bladder drainage** = inturruptes the viscious cycle
  - **CIC** – when progress to myogenic failure
  - **Appendicovesicostomy** / mitrofanoff (for CIC)
  - **Bladder augmentation** – rarely needed

4.3 Valve bladder syndrome

- **Anticholinergic and alpha blockers** – role is unclear
  - **Alpha blockers =** BNH and external sphincter hyperreflexia
    - *? BNI = concern with retrograde ejaculation, impact on fertility
  - **Oxybutynin =** bladder wall thickening with collagen deposition (*?detrusor overactivity)*
    - Early start of oxybutin is beneficial (non randomized trial)
    - Helpful as long as the bladder is emptying
- **Other adjuncts:** managing CKD, vitamin D deficiency and PTH.

4.3 Valve bladder syndrome



- **Augmentation cystoplasty**

5. Transition to adult care

- *Renal transplantation
- Sexual function and fertility
- LUTS

5.1 progression to ESRD

- **Impaired renal function**
  - may be stabilized during childhood
  - might require dialysis/transplant during adolescence
- Poor outcome of renal transplant if bladder function is not addressed prior
- Higher UTI in those with bladder augmentation on immunosuppresion but still had higher graft survival

5.1 early progression to ESRD

- Every effort should be made to optimize renal function and delay transplant
- If occur early and affect growth– GH, ca, Pth, bicarb
- **Change in dogma**
  - Augmentation not necessary before transplant
  - Can transplant in vesicostomy
  - Can grow to the age of augmentation and CIC

5.1 progression to ESRD

- Heikkilä et al. from Finland (193 patients, 1953-2003)
  - Life time risk of ESRD 28.5%
  - Of those who progressed to ESRD, 68% were <17 years
  - No ESRD after mid 30s
  - ESRD was associated with early presentation, bilateral VUR and recurrent UTI following ablation
  - Time to progression to ESRD correlated with Nadir Cr

5.1. progression to ESRD: factors

- Nadir creatinine (lowest cr at frist year after diagnosis)… >0.8
- Creatinine velocity (first 5 days after decompression)…rising>3/d
- Failure to achieve diurnal continence by age of 5 (bladder function)
- Renal changes (discussed on antenatal ultrasound)
- Presence of bilateral VUR
- Pressure "pop off" mechanism…is it beneficial?
- Late presentation….beneficial or detrimental?

5.2 Sexual function and fertility

- **Prostate function** may be affected due to high urethral pressure **during development** and **ongoing voiding dysfunction**
- Reflux into seminal vesicles and ejaculatory ducts
- ** no increase in ejaculatory problems in those men who had undergone both PUV ablation and BNI in recent report
- ** ?no erectile dysfunction
- **Azoospermia was uncommon, and associated with CKD

5.3 LUTS

- **** 2-3x inc symptoms than controls in recent study**
  - mild hesitancy, weak stream, incomplete bladder emptying, and straining

Summary

- **Emphasize on detecting and treating bladder dysfunction**
  - observation
  - clinical history
  - urodynamics (Routine followup after toilet training)
- Successful management requires Education of parents and growing children

Thank you!