POSTERIOR URETHRAL VALVE

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Incidence

• The most common cause of lower urinary tract obstruction (LUTO) in male infants
• 1 : 5000 live births \(^1\)
• 1/1250 fetal ultrasound screenings \(^2\)
• The few children that survive do poorly, with over 50% progressing to ESRD in 10 years \(^4\)

ETIOLOGY

- Not clear
- Genetic factors are poorly understood
CLASSIFICATION

• Young classification 1919

- Type I
  - Obstructing membrane that extends distally from each side of the verumontanum towards the membranous urethra where they fuse anteriorly

- Type II
  - Described as folds extending cephalad from the verumontanum to the bladder neck

- Type III
  - Represent a diaphragm or ring-like membrane with a central aperture just distal to the verumontanum
  - Thought to represent incomplete dissolution of the urogenital membrane
95% more serious
• Normal bladder
  • Store urine at low pressures
  • Empty effectively
  • continent

• PUV bladder
  • Pressure work to empty
  • Normal cycling does not occur
  • bladder wall thickened & non-compliant
  • Intra vesical pressure
    – Ureteral dilatation
    – Urinary stasis
    – Parenchymal damage or primarily abnormal (primary renal dysplasia)

Fetuses with the highest grades of obstruction often do not survive in utero
OR
Birth with progressive renal failure and inadequate bladder function
PUV damage the entire urinary tract proximal to the valve

Pulmonary hypoplasia is the most common cause of mortality in valves patients

Most renal damage occurs early in fetal life

Bladder dysfunction is usually a life long problem resulting in incontinence and poor emptying
Pathophysiology

PUV

Obstruction

↓ urine in fetus

- bladder
  - detrusor ms hypertrophied
  - Poor compliance
  - Poor sensation
  - Hypercontractility

- ureters
  - hydrenephrosis & hydroureter

- kidneys
  - Glomerular
  - tubular

Oligohydramnios

Pulmonary Hypoplasia

Poor sensation

Prolonged obst

Failure to

Chronic dilated systems

Acidify urine

Reversible

↓ GFR, ↓ renal perfusion

Reversible renal impairment

Irreversible renal dysplasia
Effects of PUV on Urinary Tract

1. RENAL DYSPLASIA
2. RENAL FUNCTION
3. RENAL TUBULAR FUNCTION
4. HYDRONEPHROSIS
5. VUR
6. VESICAL DYSFUNCTION/VALVE BLADDER
1-Renal Dysplasia

• Cause?
  1- high pelvic pressure during nephrogenesis
  2- primary embryologic abnormality from abnormal position of ureteric bud

• Severity will determine ultimate renal function
2-Renal Function

- Children with PUV may demonstrate gradual loss of renal function over time
- Causes
  - Renal parenchymal dysplasia
  - Incomplete relief of obstruction
  - Parenchymal injury
    - UTI, Hypertension, progressive glomerulosclerosis from hyperfiltration, Obstruction

ESRD
- Occurs in 25% - 40%
- 1/3 soon after birth
- 2/3 during late teenage
3-Renal Tubular Function

• 50% of patients with PUV have impairment concentration ability

• Persistently high urinary flow rate regardless of fluid intake/state of hydration

• severe dehydration and electrolyte imbalance

• ureteral dilatation and high resting vesical pressure
4-Hydronephrosis

- Significant urethral obstruction  variable degree of ureteral dilatation

- After relief of obstruction
  - gradual but substantial reduction of hydronephrosis
  - If not reduced we have to rule out:  
    1- High intravesical pressure 
    2- ureteral muscle weakness 
    3- UVJ obstruction-functional
5-Vesicoureteral Reflux

- 50% VUR at time of diagnosis
- Primary or Secondary
6-Vesical Dysfunction/Valve Bladder

- Usually secondary to irreversible changes in organization and function of the smooth muscle from outlet obstruction
- Present as urinary incontinence (20%)
- Bladder dysfunction persist in 75 % after valve ablation
• May cause deterioration of renal function

• Persistent hydronephrosis

• Three groups of dysfunction were described
  Detrusor – hyperreflexia (29%)
  Hypertonic and poor compliant bladder (31%)
  Myogenic failure and overflow incontinence (40%)
<table>
<thead>
<tr>
<th>Good</th>
<th>Poor</th>
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<tr>
<td>Presentation after 1 y</td>
<td>Prenatal diagnosis on or before 24 wk gestation</td>
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<tr>
<td>Nadir serum creatinine less than 0.8 mg/dL at 1 y</td>
<td>Respiratory distress at birth</td>
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<tr>
<td>Nadir serum creatinine less than 0.8 mg/dL after 4-5 d of initial drainage</td>
<td>Presentation before 1 y</td>
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<td>Identification of corticomedullary junction</td>
<td>Bilateral vesicoureteric reflux</td>
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<td>Presence of pop-off mechanism</td>
<td>Persistent serum creatinine higher than 1.0 mg/dL after initial therapy</td>
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<td>Persistent incontinence after 5 y</td>
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<td>Echogenic kidney on ultrasonogram</td>
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<td>Subcapsular renal cyst</td>
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<td>Proteinuria</td>
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<td>Hypertension</td>
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Diagnosis-Clinical Presentation

- Varies by degree of obstruction
- Symptoms vary by age of presentation
- 70% of PUV by antenatal presentation
• Antenatal

  – Bilateral hydronephrosis
  – Distended and thickened bladder
  – Dilated prostatic urethra
  – Oligohydramnios - accounts for co-presentation of pulmonary hypoplasia.
- Newborn

  - Palpable abdominal mass
    - Distended bladder, hydronephrotic kidney
    - Bladder may feel like a small walnut in the suprapubic area

  - Ascites

  - Respiratory distress from pulmonary hypoplasia
    - Severity often does not correlate with degree of obstruction
    - Primary cause of death in newborns
• Infants
  - Urinary dribbling
  - Failure to thrive/ renal failure
  - Urosepsis

• Toddlers:
  - UTI
  - Voiding dysfunction

• School-age boy
  - Urinary incontinence
  - Eneuresis
Diagnosis-Investigations
Serum Creatinine

- Immediately after birth reflect maternal creatinine
- Therefore, serum values for creatinine and BUN should be obtained at least 24 hours after birth.
- Observing serial serum chemistries for several days to weeks is important to determine the true status of the newborn's renal function
Postnatal Ultrasound

- Initial investigation
- Done soon after birth when PUV is suspected
- To assess renal parenchyma
- Findings
  - upper tract dilatation (which is sometimes only unilateral)
  - perinephric urinoma (occasionally)
  - thickening of the bladder wall, with or without residual urine
  - if voiding views can be obtained, dilatation of the posterior urethra
Postnatal Ultrasound
MCUG

- **Mandatory** for all PUV evaluations
- Provides the definitive diagnosis – gold standard
- VUR is present in 40–60% of cases and is unilateral in approximately two-thirds of cases.

- **Findings**
  - dilated prostatic urethra, valve leaflets
  - detrusor hypertrophy, bladder diverticula, bladder neck hypertrophy, incomplete emptying
  - narrow penile urethra stream
Post natal VCUG

VCUG of PUV:
- trabeculated bladder
- dilated posterior urethra
- valve leaflets
Initial Management-Postnatal

- Resuscitate child
- Treat urinary infection often complicated by septicaemia
- Correct gross disturbances of electrolyte and acid-base balance, of which hyperkalaemic acidosis is the most serious form.
- Co-manage with paediatric nephrologist.
- Drain the bladder
Initial Management-Postnatal

Bladder drainage

- A neonate can be catheterized with a 3.5-5 F catheter/feeding tube. A VCUG is performed to see if the diagnosis is correct and whether the catheter is within the bladder and not in the posterior urethra.
- Position can also be checked with ultrasound to make sure tube is not coiled in dilated posterior urethra.
- An alternative option is to place a suprapubic catheter, perform a VCUG and leave the tube until the neonate is stable enough to perform an endoscopic incision or resection of the valve.
• Surgical treatment of the bladder outflow obstruction is best deferred until the general condition has been stabilised, usually after 2–7 days.

• **Vesicostomy and delayed valve ablation** if child is too small or **primary valve ablation** if endoscopes available/adequate urethra.

• Circumcise with valve ablation

• If vesicostomy is insufficient to drain the upper urinary tract, high urinary diversion should be considered. Diversion may be suitable if there are
  – recurrent infections of the upper tract,
  – no improvement in renal function and/or an increase in upper tract dilatation, despite adequate bladder drainage.
High diversions- Types of Ureterostomies

Valve ablation

Small paediatric cystoscopes (<8F) and resectoscopes are now available either to incise or to resect (loop, Bugbee, laser) the valve at the 4-5, 7-8 or 12 o’clock position, or at all three positions.

It is important to avoid extensive electrocoagulation as the most common complication of this procedure is stricture formation.

Figure 122-8 Cystoscopic photographs made from the distal urethra show the posterior urethral valves before ablation (A,) and after ablation (B,). The ureteral catheter has been passed through a perforation in the valve leaflet.
Fig. 4. An algorithm for the management of newborns with PUV. PUV: posterior urethral valve; MCUG: micturating cystourethrogram.
Long term follow up-What to look for?

• Growth
• Hypertension
• UTIs
• Renal function
• Bladder dysfunction.
• Upper tract dilatation
Functional assessment

Diuretic Radioisotope Scan

- DTPA OR MAG-3
- with urethral catheter in place
- Exclude obstruction and assess split renal function
LONG TERM MANAGEMENT-VUR

• VUR is very common in PUV patients (up to 72%) and it is described bilaterally in up to 32%.

• However, early removal of the renal unit seems to be unnecessary, as long as it causes no problems.

• Removal if persistent UTI.

• It may be necessary to augment the bladder and in this case the ureter may be used.
• Bladder dysfunction is not uncommon (70%)

• Delay in day- and night-time continence is a major problem.

• Poor bladder sensation and compliance, detrusor instability and polyuria (especially at night) and their combination are responsible for bladder dysfunction. This has been postulated as the reason for the late renal deterioration (80%)
LONG TERM MANAGEMENT-
Bladder Dysfunction-2

• Use of urodynamic testing to assess bladder compliance help identify patients at risk.

• Bladder dysfunction in boys with PUV changes during childhood and through adolescence

• The urodynamic pattern of hypercontractility generally found soon after valve ablation gradually changes to hypocontractility in many boys and this pattern seems to be the rule after puberty.
LONG TERM MANAGEMENT-DELAYED RENAL INSUFFICIENCY

• Approximately one third of patients progress to ESRD and the need for dialysis or transplantation.

• Progression of ESRD is accelerated at the time of puberty due to the increased metabolic workload placed on the kidneys.

• Renal transplantation in these patients can be performed safely and effectively. Deterioration of the graft function is mainly related to lower urinary tract dysfunction.
Newborn with possible PUV, UUT dilatation and renal insufficiency

- USG and VCUG
- Assessment of renal function and electrolyte disorders

Confirm diagnosis

Bladder drainage

Nephrological care if needed

No stabilisation

Valve ablation when baby is stable

Improvement in UT dilatation and RF

- Close follow-up
- Monitor urinary infection
- Monitor renal function
- Monitor bladder function and emptying

- Progressive loss of renal function
- Recurrent infections
- Poor emptying

Short term

Long term

Consider augmentation and Mitrofanoff

No improvement but stable

No improvement and ill

Consider diversion

Check residual PUV
- CIC if not emptying
- Consider overnight drainage
- Consider alpha-blockers
- Anticholinergics if OAB