Posterior Urethral Valve & Bladder Dysfunction

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Definition

• Posterior urethral valve is an obstructing membrane in the posterior male urethral due to abnormal development in vitro.
• 1:8000 live birth.
• Can be various in degree of severity.
• The common cause of bladder outlet obstruction.
It was first described by Langenbeck in 1802, later popularized by Hugh Hampton Young in 1919.

The classification still used until today.

PUV is a misnomer because it is not a valve.

It carry no function and is abnormal development.
Classification (Young)

• Type I- ridge lying on the floor of the urethra, continuous with the verumontanum, which takes an anterior course and divides into two fork-like processes in the region of the bulbo-membranous junction. These processes are continued as thin membranous sheets, direct upward and forward which may be attached to the urethra throughout its entire circumference.

• Most commonest type.

• 95%.
• Type II- Bicuspid valve leaflet radiating from verumontanum to bladder neck. Possible due to hypertrophy of muscle.

• Type III- A disc of tissue distal to verumontanum with center opening. Can be found at difference level.
CLASSIFICATION

- Young classification 1919

Type I

Type II

Type III
Pathophysiology

• During the early stages of embryogenesis, the most caudal end of the Wolffian duct is absorbed into the primitive cloaca at the site of the future verumontanum in the posterior urethra. Failure of absorb lead to PUV.
Impact- Lower Urinary Tract

• Bladder dysfunction.
• Dilated post urethral.
• Due to long term obstruction, bladder become hypertrophy and abnormal large volume, lead to neurogenic bladder.
• Incontinent.
• Poor emptying → UTI.
Impact- Upper Urinary Tract

- Ureteral dilatation, reflux.
- Chronic renal insufficiency.
- ESRF.(50%)
- 10% end up in renal transplant.
Changes in Urinary tract

• The prostatic urethra is distended and the ejaculatory ducts may be dilated due to urinary reflux.
• The bladder neck is hypertrophied and rigid.
• The hypertrophied bladder occasionally has multiple diverticula.
• Nearly all valve patients have dilatation of both upper urinary tracts. This may be due to the valve itself and the high pressure in the bladder, or due to obstruction of the ureterovesical junction by the hypertrophied bladder.
• If there is secondary reflux, the affected kidney functions poorly in most cases.
Diagnosis

• Commonly diagnosed during antenatal check up- noted oligohydramnios, fetal hydronephrosis.
• Baby ix for UTI, renal impairment.
• Rarely this condition can manifested in adolescence age. Pt will present with obstructive symptom or post ejaculation dysuria.
Antenatal Scan

• Hydronephrosis, hydroureter.
• Dilated ejaculatory duct, distended bladder and urethral (Key hole sign).
• Thicken bladder wall.
• Oligohydramnios.
• Renal Dysplasia.
Voiding Cystourethrogram

- To assess the anatomy of bladder and urethral.
- To assess for any VUR.

DMSA/ MAG 3

- To look for renal scarring and differential the renal function.
Prenatal Management

- Regular ultrasound to monitor the oligohydramnios, progression of HU/HN.
- Refer to tertiary centre for maternity care, Paediatric as well as urology care.
- PUV causes oligohydramnios, amniotic fluid are necessary for lung develop, lack of amniotic fluid lead to pulmonary hypoplasia lead to respiratory failure.
Antenatal treatment

- Placement of vesicoamniotic shunt can resolved the issue of oligohydramnios, but it doesn’t improve the renal outcome of the baby.
- >20wk<32wk.
• Salam MA. Posterior urethral valve: Outcome of antenatal intervention. 2006

• 58 babies are divided into 2 gps,
• One gp had vesicoamniotic shunt and one gp had postnatal corrective surgery.
• 50% of the gp1 had normal function whereas the gp 2 60% had normal function.
Complication

- Dislodge shunt.
- Abortion.
- Intestine evisceration.
- Intrauterine death.
- Preterm delivery.
- Block/Infected Shunt
• Fetal Surgery for Posterior Urethral Valves: Long-Term Postnatal Outcomes. Nicholas Holmes; Michael R. Harrison; Laurence S. Baskin. 2001

• 1 fetal bladder marsupialization, 2 in utero ablation of valves and placement of vesicoamniotic catheter in 9 fetus.

• 8 out of 12 dies, 5 had chronic renal disease, 3 renal transplant.
Fetal cystoscopy

- It carried high abortion rate and low successful rate.
Antenatal ultrasound to predict postnatal renal function in congenital lower urinary tract obstruction: systematic review of test accuracy

RK Morris, a GL Malin, a KS Khan, a MD Kilby a,b

- Amniotic fluid index
- Renal cortical appearance.
- 54% sensitivity, 89% specificity.
• *Prenat Diagn* 2007; **27**: 900–911.

• Sodium >100mmol/dl
• Chloride >95mmol/L
• Osmolality >200mOsm/L
• Total protein >20mg/dL
• Calcium >8mg/dL
• Beta 2 macroglobulin >3g/mL
• All are poor indicator.
Biomarker

- Still at developmental status.
- Endothelial-1
- Epidermal growth factor
- Transforming growth factor beta 1
When to intervene

• Aim are to preserved the renal function and provided adequate environment for lung maturation.
• B/L HU/HN with oligohydramnios.
• Renal function using urine analysis.
• Renal dysplasia.
Post natal treatment

• Bladder drainage- baby born with suspected PUV, an immediate VCUG is necessary if possible. It can done by placing the urinary catheter or 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.
• Valve ablation- once the creatinine level decreased or normalized, valve ablation using peds resectoscope to resect 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.

• Three months following initial treatment, a control VCUG or a re-look cystoscopy should demonstrate the effectiveness of the treatment, depending on the clinical course.
• Vesicostomy- If the child is too small and/or too ill to undergo endoscopic surgery, a vesicostomy is used to drain the bladder temporarily. If initially a suprapubic tube has been inserted, this can be left in place for 6-12 weeks. Otherwise, a cutaneous vesicostomy provides an improvement or stabilisation of upper urinary tracts in over 90% of cases.
• High diversion- If bladder drainage 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. The choice of urinary diversion depends on the surgeon’s preference for high loop ureterostomy, ring ureterostomy, end ureterostomy or pyelostomy.

• Reconstructive surgery should be delayed until the UUT has improved as much as can be expected.
Figure 8: An algorithm providing information on assessment, treatment and follow up of newborns with possible PUV

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

No improvement but stable

No improvement and ill

Consider diversion

Short term

Long term

Consider augmentation and Mitrofanoff

CIC = clean intermittent catheterisation; OAB = overactive bladder; PUV = posterior urethral valve; RF = renal function; USG = urinary specific gravity; UT = urinary tract; UUT = upper urinary tract; VCUG = voiding cystourethrogram.
### Conclusions and recommendations posterior urethral valves

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<th>PUV</th>
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| **Diagnosis** | An ultrasound can indicate a PUV, but a VCUG is required to confirm the diagnosis.  
  - Split renal function is to be assessed by DMSA scan or MAG III clearance.  
  - Serum creatinine is the prognostic marker. | 3 | B |
| **Treatment antenatal** | A vesico-amniotic shunt is effective in reversing oligohydramnios, but it has a relatively high complication rate. There is no difference in the renal outcome and long-term results. | 1b | A |
| **Treatment postnatal** | After bladder drainage and stabilisation of the child, endoscopic valve ablation should be performed.  
  - In case the child is too small, a vesicostomy is an option for bladder drainage.  
  - If bladder drainage is insufficient to drain the UUT, and the patient remains unstable, high urinary diversion should be considered (see Fig. 8). | 3 | B |
| **Follow-up** | Life-long monitoring is mandatory (bladder dysfunction; end-stage renal failure) in all patients.  
  - Those with serum creatinine nadir above 80 μmol/L have a poor prognosis. Despite optimal treatment 10-47% of cases develop end-stage renal failure.  
  - High creatinine nadir and severe bladder dysfunction are risk factors for renal replacement therapy.  
  - Renal transplantation can safely be performed if bladder function is stable. | 3 | B |

*DMSA = dimercaptosuccinic acid scan; VCUG = voiding cystourethrogram.*
Bladder dysfunction

• Post urethral valve causes various degree of urinary organ damage, and the organ mainly involved is bladder.

• Interference of upper tract drainage and incontinent are the two long term problem.
Interference with urinary drainage

• The bladder causing upper tract obstruction mainly in newborn and late childhood.

• PUV causing bladder thicken, trabeculated with saccules and diverticuli. It causing strong bladder contraction and low compliance → obstruction.

• Late childhood, bladder less contractile and low compliance, poor sensation → neurogenic obstruction.
Incontinence

- 80% of child had delay continence upto 5yr old.
- 53% of the pt dry only by 12 yr old.
- Majority improved by age of 20.
- Incontinence mainly cause by poor sensation, polyuria, poor compliance, detrusor instability.
Urodynamic study

• UDS although are difficult to perform in children but is a good tool to diagnosed bladder dysfunction in children.
• Mainly three type of feature- myogenic failure, detrusor hyperreflexia, poor compliance.
• Myogenic failure lead to overflow incontinence and incomplete voiding.
• Detrusor hyperreflexia lead to frequency and urge incontinence.
• Poor compliance means small bladder volume $\rightarrow$ inadequate storage $\rightarrow$ obstruction to upper tract drainage.
Treatment

• Myogenic failure- double voiding, timed voiding, alpha blocker, CISC.
• Detrusor hyperreflexia- anticholinergic.
• Poor compliance- augmentation cystoplasty.
• Treatment are usually multiple procedure.
• Baby with high bladder neck and sensate urethral, it often made CISC difficult, the other option are appendicovesicostomy and follow by CIC.