POSTERIOR URETHRAL VALVES

THE JAMAICAN EXPERIENCE
A SINGLE CENTRE STUDY

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3rd Paediatric Nephrology Conference October 21st 2018
Posterior urethral valves (PUV) Definition

- This is a congenital abnormality that causes varying degrees of urethral obstruction due to embryological anomalies.
- It is the most common cause of lower urinary tract obstruction in male infants.
PUV - Incidence

- **Jamaica**
  - Gomes et al (2016) – 1.4% MCUG for UTI
  - 1:2750 live births over review period

- **Internationally**
  - 1: 5000- 8000 live births

PUV 41% chronic renal failure in Jamaican children < age 12 years (2007-2012)
Embryology

- Excessive development of normally placed urethral folds
- Remnant of the urogenital membrane
- Anomalous junction between the ejaculatory (Wolffian) duct and the prostatic utricle (Mullerian duct)
- Fusion of seminal coliculus with the roof of the urethra
Primary and Secondary Pathology

- Bladder and bladder neck hypertrophy
- Detrusor muscle hypertrophy
- Vesicoureteric reflux
- Pyelonephritis and scarring of kidneys with associated sequelae
Presentation

- Antenatal evidence of hydronephrosis, mainly bilateral
- Acute urinary retention at birth with palpable bladder
- Poor urinary stream
- Urinary tract infection
- Failure to thrive
- Progressive renal failure
Investigations

- Plain abdominal x-ray
- Ultrasonography
- Micturating cystourethrogram
- Renal scan
- Urethrocystoscopy
ANTENATAL ULTRASOUND
Normal MCUG
Dilated posterior urethra
Residual Urine Post Micturition
VALVE LEAFLETS

BUGBEE ELECTRODE

VERUMONTANUM
Management

- Preliminary resuscitation
  - Correct infection
  - Correct azotemia
  - Correct dehydration
  - Correct electrolyte imbalance
  - Correct acid / base imbalance
Management

- Bladder drainage (urethral catheterization)
- Monitor post-obstructive diuresis
- Endoscopic valve ablation
- Whittaker hook electrode
- Urinary diversion
Management

- Preliminary resuscitation
  - Correct infection
  - Correct azotemia
  - Correct dehydration
  - Correct electrolyte imbalance
  - Correct acid / base imbalance
Management

- Bladder drainage (urethral catheterization)
- Monitor post-obstructive diuresis
- Endoscopic valve ablation
- Whittaker hook electrode
- Urinary diversion
Urinary Diversion

- Cutaneous vesicostomy
- Cutaneous ureterostomy
- Cutaneous pyelostomy
Figure 1 - Blocksom vesicostomy. A) The skin, rectus fascia, and muscle are divided. The urachus is identified and pulled into the wound. B) and C) The urachus is excised, the posterior and anterior serosa of the bladder are approximated to the rectus fascia, and the bladder mucosa is approximated to the skin. (From Hinman F Jr: Atlas of Pediatric Urology. Philadelphia, W.B. Saunders Co., 1994, with permission).
VESICOSTOMY
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[Abel C, Stanley A, Miller M, Reid M]
79 Patients (1 Antigua)
Age at diagnosis

- < 1mo
- 1-5 mo
- 6-11mo
- 1-5 yrs
- 6-10 yrs
- >10yrs

Number
Reason for investigation

- Urinary tract infection 93%
- Voiding disorder 19%
- Abnormal antenatal ultrasound 8.3%
  [73/79 studied]
- Abdominal mass 19%
Ultrasound abnormal (74%)

- Bilateral hydronephrosis 64.4%
- Thick walled bladder 59.7%
- Small kidney 2.6%
Micturating cystourethrogram (MCUG)

- 78/79 studies found
- Diagnostic in 100%
- Vesico-ureteric reflux (VUR) 34.6%
- High grade VUR 69.9%
Renal glucoheptonate nuclear scan

- Done in 29.5%
- Abnormal in 52.2%
Age at diagnosis and surgery
PROGNOSIS
Definitions

- GFR calculations by Schwartz formula
  - not possible (no heights)
Definitions

- **Renal impairment**
  - serum Creatinine > 2SD for age

- **Significant renal impairment**
  - Serum creatinine ≥ 100umol/l

- **CKD**
  - renal impairment at 1 year or later post op]

- **Significant CKD – CKD ≥ 3 (chronic renal failure)**
  - Assumed if serum Creatinine > 500umol/l

- **End stage renal disease – CKD 5**
  - Assumed if serum Creatinine > 1000umol/l
Follow up duration

- Mean 2.8 years (range 4 days – 9.9 years)
Renal function

<table>
<thead>
<tr>
<th>Serum creatinine levels</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>High creatinine at diagnosis</td>
<td>53.2</td>
</tr>
<tr>
<td>High creatinine 1 month post</td>
<td>27.8</td>
</tr>
<tr>
<td>High creatinine $\geq$ 1 year post (CKD)</td>
<td>34.7</td>
</tr>
</tbody>
</table>

- Significantly elevated creatinine (range 110-1665umol/l) 9.7%
- End stage renal disease (creatinine 1665 umol/l) 1.4%
Age at diagnosis vs CKD

- > 10 yrs
- 6-10 yrs
- 1-5 yrs
- 6-11mo
- 1-5 mo
- < 1mo

CKD% of group vs number tested
Age at first surgery vs CKD

- CKD (%) of group
- Number tested

>10 yrs

6-10 yrs

1-5 yrs

6-11mo

1-5 mo

< 1mo

0 20 40 60 80
Observations predictive of CKD

**Predictive**
- All CKD had abnormal ultrasound
- High peak Creatinine
- VUR
- Referral from WRHA
  - (p-value 0.03)
  [statistical analysis incomplete]

**Not predictive**
- Age at diagnosis
- Age at first surgery
COMPARISONS
Presentations - UTI

<table>
<thead>
<tr>
<th>COUNTRY</th>
<th>%</th>
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<tbody>
<tr>
<td>Jamaica</td>
<td>96.3</td>
</tr>
<tr>
<td>Nigeria</td>
<td>90.5</td>
</tr>
<tr>
<td>South Africa</td>
<td>71.4</td>
</tr>
<tr>
<td>USA</td>
<td>43.3</td>
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</table>
### Hydronephrosis

<table>
<thead>
<tr>
<th>COUNTRY</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jamaica</td>
<td>64.4</td>
</tr>
<tr>
<td>African studies</td>
<td>83-89</td>
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</tbody>
</table>
Comparisons

- Thick walled bladder – more common in our study than internationally
- Fewer renal scans – cost factor
<table>
<thead>
<tr>
<th>COUNTRY</th>
<th>CKD %</th>
<th>ESRD</th>
<th>DEATH</th>
<th>LOST TO FOLLOW UP OR DEAD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jamaica</td>
<td>34.6</td>
<td>1.4</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>India</td>
<td>42.7</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nigeria</td>
<td></td>
<td>31%</td>
<td>69%</td>
<td></td>
</tr>
<tr>
<td>Egypt</td>
<td>36.5</td>
<td>18</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>USA</td>
<td>20</td>
<td>8.6</td>
<td></td>
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</tr>
</tbody>
</table>

Definitions of CKD vary or are unstated in some series.
Summary

- **PUV**
  - more common in our population than internationally
  - most diagnosed after UTI
  - few diagnosed antenatally (91.8% false negative)
  - 1.4% ESRD 34% CKD
  - Progression may occur
  - Need for long term follow up

- PUV 41% Chronic renal failure in Jamaica children < age 12 yrs
THE END