MANAGEMENT OF THE BLADDER IN SPINA BIFIDA

Dan Wood
objectives

- Definition
- Epidemiology
- Risks
- Treatment options
objectives

- Definition
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- Risks
- Treatment options
definition

- abnormal development of the spinal canal and internecine spinal cord
objectives

- Definition
- Epidemiology
- Risks
- Treatment options
At least 25% of the clinical problems seen in paediatric urology are the result of neurologic lesions that affect lower urinary tract function.

90% of open spinal dysraphic lesions are myelomeningocele
incidence

- Marked decrease
- 1/1000 live births
- Reasons for fall
  - Antenatal diagnosis and TOP
  - Folic acid
objectives

- Definition
- Epidemiology
- Risks
- Treatment options
## Familial Risk

<table>
<thead>
<tr>
<th>Relationship</th>
<th>Incidence per 1000 LB</th>
</tr>
</thead>
<tbody>
<tr>
<td>General Population</td>
<td>0.7-1</td>
</tr>
<tr>
<td>Mother + 1 affected child</td>
<td>20-50</td>
</tr>
<tr>
<td>Mother + 2 affected children</td>
<td>100</td>
</tr>
<tr>
<td>Patient with myelodysplasia</td>
<td>40</td>
</tr>
<tr>
<td>Mother over 35 yrs</td>
<td>30</td>
</tr>
<tr>
<td>Sister of mother with affected child</td>
<td>10</td>
</tr>
<tr>
<td>Sister of father with affected child</td>
<td>3</td>
</tr>
<tr>
<td>Nephew who is affected</td>
<td>2</td>
</tr>
</tbody>
</table>
folic acid

- Affected family
  - 4mg OD for at least 2 months pre conception
  - General Pop’ n 400 µg OD

- Reduction by 50%
survival

Children green
Adults red
## Cause of death

Malakounides et al 2013

<table>
<thead>
<tr>
<th>Cause of death</th>
<th>Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sepsis (hindquarter amputation)</td>
<td>14</td>
</tr>
<tr>
<td>Sepsis (post ileal conduit)</td>
<td>17</td>
</tr>
<tr>
<td>Sepsis (pressure sores)</td>
<td>??</td>
</tr>
<tr>
<td>Sepsis (pressure sores)</td>
<td>??</td>
</tr>
<tr>
<td>Blocked shunt</td>
<td>11</td>
</tr>
<tr>
<td>Not given</td>
<td></td>
</tr>
<tr>
<td>Not given</td>
<td></td>
</tr>
</tbody>
</table>

Mean age of death 14 (0-25 years) 4.4 % NONE FROM RENAL FAILURE

Previous series upto 1/3 death from renal failure (Singhal et al 1999)
Functional results
Malakounides et al 2013

- 1/3 augmented (median age 10 years)
- 10.8% BN surgery (9 with AUS)
- 1/3 ACE (age 9 years)

Table 3  Urological surgeries.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Percentage</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Augmentations</td>
<td>26%</td>
<td>31</td>
</tr>
<tr>
<td>Mitrofanoff</td>
<td>23%</td>
<td>28</td>
</tr>
<tr>
<td>Artificial urinary sphincter</td>
<td>7.5%</td>
<td>9</td>
</tr>
<tr>
<td>Pippe Salle procedure</td>
<td>2.5%</td>
<td>3</td>
</tr>
<tr>
<td>Bladder neck sling</td>
<td>0.8%</td>
<td>1</td>
</tr>
<tr>
<td>STING anti-reflux</td>
<td>1.7%</td>
<td>2</td>
</tr>
<tr>
<td>Nephrectomy</td>
<td>1.7%</td>
<td>2</td>
</tr>
<tr>
<td>Intravesical botox injection</td>
<td>1.7%</td>
<td>2</td>
</tr>
<tr>
<td>Antegrade continence enema</td>
<td>44%</td>
<td>53</td>
</tr>
</tbody>
</table>
## Renal status

Malakounides et al 2013

<table>
<thead>
<tr>
<th>CKD stages</th>
<th>Description (corrected GFR in ml/min/1.73 m²)</th>
<th>%No. of patients (total 120)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>Normal GFR and imaging (≥90)</td>
<td>48% (58)</td>
</tr>
<tr>
<td>I</td>
<td>Kidney damage with normal GFR (≥90)</td>
<td>6% (7)</td>
</tr>
<tr>
<td>II</td>
<td>Kidney damage with mild reduction of GFR (60–89)</td>
<td>36% (43)</td>
</tr>
<tr>
<td>III</td>
<td>Moderate reduction of GFR (30–59)</td>
<td>8% (10)</td>
</tr>
<tr>
<td>IV</td>
<td>Severe reduction of GFR (15–29)</td>
<td>0.8% (1)</td>
</tr>
<tr>
<td>V</td>
<td>Kidney failure (&lt;15 or dialysis)</td>
<td>0.8% (1)</td>
</tr>
</tbody>
</table>

Ref. [19].
Neurogenic Causes
Congenital

- Myelomeningocele
  - Most at level lumbar vertebrae then sacral, thoracic, and cervical areas, in decreasing order of frequency
- Spina bifida occulta
- Syringomyelia
- Lipoma of cord
- Cord tethering
- Sacral agenesis
Myelomeningocele

Renal failure commonest cause of death - rare with sensory level below L4, common above T10

Incidence 48/100,000 live births, 1990 - decreased with influence of folic acid and terminations

McGuire - no patient has developed renal failure at Ann Abour since 1983
Associated Problems

- **Hydrocephalus**
  - 85% of children with myelomeningocele have Arnold-Chiari malformation
  - may affect dexterity and intellect

- **Mobility**
  - neurological level > L3 (quads): wheelchair

- **Spinal deformity**
  - unable to self catheterise

- **Upper tracts**
  - kidney damage with high pressure bladder
  - DLPP >40cmH$_2$O (Maguire)
Aim of Management

Achieve ‘normal’ bladder function

- Healthy kidneys
- Bladder empties completely
- Dry
- No UTI
Examination

- Inspect back - USS Spine
- Palpable bladder?
- Lower limb reflexes ± sensation/ambulation
- Anocutaneous reflex – conus intact?
Upper Tract Damage

- 20% affected by 2 yrs, 50% of boys at risk
- UTI due to bladder dysfunction
- VUR due to bladder dysfunction
  - Treat bladder
- Outflow obstruction – DSD or static
- Poor compliance

- Gross SUI + empty bladder = safe
Urodynamics

- Measure bladder pressures during filling
  - Slow fill to expected capacity – 10% of expected capacity/min (Nijman)
  - Capacity = (age in years x 30) + 30 ml

- Catheter (eg 6F double lumen) – to fill bladder
  - SPC – neurologically intact
  - Urethral - CIC

- Pressure transducers
  - Overactive - spontaneous contractions
  - Compliance – pressure < 20cm H₂O within capacity

- EMG – perineal electrode, ?DSD
Urodymanics

- **Timing – controversy**
  - Maguire advocates UDS in first few months with early CIC if potential for upper tract damage ie ↑DLPP
  - Others, upper tract imaging first, UDS when abnormality demonstrated
  - Repeat fill and void sequences

- **Bladder pressures do not correlate with level** (Blavias 1977)
Remember Neuropathic Bowel

- Assess at same time as bladder
- May need ACE at same time – Malone
objectives

- Definition
- Epidemiology
- Risks
- Treatment options
early evaluation

- Spinal closure
- USS & PVR
- Early intervention
  - CIC vs Crede manouvre
- 15-20% will have abnormal UT at Δ
- Antibiotic prophylaxis
Aggressive management early

- CIC
  - Start early
  - Kochakarn et al
    - 6.88 months vs 45 months
    - Normal Creatinine 66% vs 39%
    - HN 27% vs 58%
    - Cystoplasty needed in 14% vs 32%
  - Burden for parents
  - Bladder may worsen to 2 years
Continence

- Beware improving continence
- >40 cm H₂O
- Watch upper tract
  - AUS – 77% with revision
  - 13% erosion
  - May only require a cuff
  - BN is better
treatment options

- CIC
- Anticholinergics
- Botox
- Cystoplasty

SAFE BLADDER

Remember spinal level may not match clinical syndrome
Summary of Treatment options

- Overactivity + poor compliance
  - Anticholinergics + Cl
  - Botox
  - Bladder augmentation ± Mitrofanoff / Monti

- Sphincter weakness
  - α agonist – Ephedrine, ????Duloxetine
  - Suspension/sling/closure/periurethral injection/AUS
    - Brantley-Scott AUS 77% success rate (with revisions) mean follow-up 5yrs, erosion rate 13%

- Sphincter spasm
  - Botox
  - ?Diversion
SURGERY – WHEN IT’S NEEDED

Clam Cystoplasty and Mitrofanoff Channel
Neobladder and Mitrofanoff Channel
Ileal Conduit
Replacement of the Lower Urinary Tract

<table>
<thead>
<tr>
<th>Reservoir</th>
<th>Conduit</th>
<th>Continence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bladder</td>
<td>Urethra</td>
<td>Sphincters</td>
</tr>
<tr>
<td>Stomach</td>
<td>Appendix</td>
<td>Mitrofanoff</td>
</tr>
<tr>
<td>Ileum</td>
<td>Ureter</td>
<td>Ileo-caecal valve</td>
</tr>
<tr>
<td>Colon</td>
<td>Ileum</td>
<td>Kock Nipple</td>
</tr>
<tr>
<td>Rectum</td>
<td>Anus</td>
<td>Anal Sphincter</td>
</tr>
<tr>
<td></td>
<td>Fallopian</td>
<td>AUS</td>
</tr>
</tbody>
</table>
Two Key Urological Objectives

- Preservation of upper tracts
- Continence/dry
Bladder Reconstruction Options

- Ileal Conduit
- Enterocystoplasty
  - Clam (Augmentation)
  - Substitution (Supratrigonal or subtrigonal)
- Neobladder
  - Orthotopic
  - Heterotopic
Ileal Conduit

- Complications:
  - Redundant loop: 0-13%
  - Incisional hernia: 3-5%
  - Stones: 0-9%
  - Ureteroileal obstruction: 0-14%
  - Metabolic: 0-1%
  - UTI: 0-23%

Stomal Complications

Parastomal hernia 4.5-31%

Stomal retraction 0-31%

Stomal stenosis 0-15%

Stomal bleeding 0-8%
Clam Cystoplasty

Described in 1888 in Italian dogs

Popularised for IDO by Bramble in 1980s

85% cure
Ileocystoplasty

With thanks – Tony Mundy
Cystoplasty - Early Complications

- Death: 0-3.2%
- Adhesive SB obstruction: 3-6.6%
- Wound infection: 1.5-9%
- Reoperation for bleeding: 0-3%
- VP shunt infection: 0-20%

Greenwell TJ, Venn SN, Mundy AR. Augmentation cystoplasty. BJU Int 2001; 88: 511-525
Cystoplasty Complications

- Infection
  - Bacteriuria in 75%
  - Septic episodes in 5-20% per year
  - Bowel in symbiosis with bacterial flora
  - Bacterial translocation on distension
  - Decreased bacteriostatic action of urine
Cystoplasty Complications

• Perforation
  • Neuropaths
    – 23-25% mortality
    – difficult to diagnose
      – Post emptying contrast CT cystogram
    – Explore if any doubt

Mitrofanoff Harvest

- 1st described in 1980
- Continent supravesical catheterisable channel
- Appendix on vascular pedicle
Contra-Indications

- Insufficient dexterity
- Can be used in quadriplegia

Mitrofanoff complications

- catheterisation difficulties
  - 27% of Mitrofanoff and 60% of Yang/Monti
- 10-40% stomal stenosis
- 2% stomal prolapse
- 28% sacculation
- 16-50+% revision rate

Sahedevan K et al. Is continent diversion using the Mitrofanoff principle a long term viable option for adults requiring bladder replacement. BJU Int 2008; 102: 236-240
Mitrofanoff complications

- 40% UTI
- 40-100% stones (higher rate both Mitrofanoff and cystoplasty)
- high patient acceptance
- patients with both urethra and Mitrofanoff prefer Mitrofanoff

Fishwick JE, Gough DC, O’ Flynn KJ. BJU Int 2002; 85 (4): 496-497
AND IF YOU IGNORE THEM....
Stone formation  Hamid et al 2008

- Cystoplasty patients
- 2 Groups
  - 1 had formed stones
  - 2 No stone Hx
- No significant differences with metabolic screen
- Group 2 41% 24hr uo, 173% urinary citrate lower pH (p<0.005)
- Group 1 Higher urinary Ca$^{2+}$
- Urethral Drainage – 8%
- Mitrofanoff - 21 to 100%  Kroner 1998, Woodhouse 1998
## Cancer - Risk Factors

**Husmann JPU 2008 n=153**
**Median FU – 27 years**

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>% Malignancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asymptomatic Bacteriuria</td>
<td>No difference</td>
</tr>
<tr>
<td>Symptomatic UTI</td>
<td>No difference</td>
</tr>
<tr>
<td>Colonic segment</td>
<td>5 % (same as Ca colon risk)</td>
</tr>
<tr>
<td>Ileal segment</td>
<td>4.5 % (vs 0.1 population risk)</td>
</tr>
<tr>
<td>Neurogenic</td>
<td>2 %</td>
</tr>
<tr>
<td>PUV</td>
<td>12 % (NB ESRD)</td>
</tr>
<tr>
<td>Renal transplant</td>
<td>20 %</td>
</tr>
<tr>
<td>BEX</td>
<td>8 %</td>
</tr>
</tbody>
</table>

NB Gastric segments –
- Short lag time to Ca
- Assoc of gastric to SB anastomosis with Ca
- Atrophic Gastritis
What we do....

- Review at 3-6 months with USS and bloods
- 1 year USS, GFR, MAG-3, Bloods
- Then annual bloods and USS
- GFR every 5 years

- Bloods – FBC, U&E, Chloride, Bicarbonate and B12.
thank you.........any questions?