Neuropathic bladder

Rare

Girls > boys

Boys with more upper tract complications

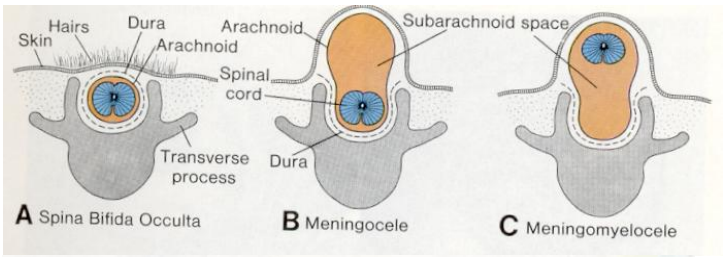
CISC mainstay of treatment but only ~20% dry

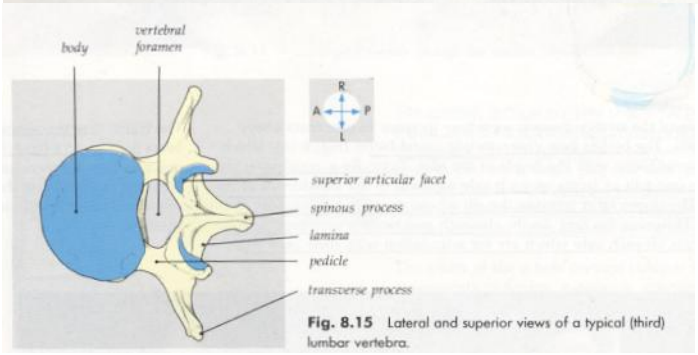
Aetiology

- Congenital
  - Myelomeningocoele
  - Spinal bifida occulta
  - Sacral agenesis
- Acquired
  - Trauma
  - Infarction
  - Tumour
  - Transverse myelitis

Myelomeningocoele (subtype of spinal bifida) represents commonest cause of congenital neuropathic bladder

- Failed fusion of dorsal vertebrae (lamina)
- Covered by skin - spina bifida occulta. More than one vertebrae causes bulging of meninges – meningocoele. Very large sacs contain cord – myelomeningocoele.
- Conus medullaris invariably involved – 95% have bladder/bowel dysfunction although 30% retain sacral cord reflexes





- Common associated findings
  - Hydrocephalus
    - Caudal displacement of medulla oblongata (Arnold Chiari malformation)
  - Kyphoscoliosis
  - Renal agenesis

Spina bifida occulta

Spinal dysraphism covered by skin, often with a hairy patch or atypical presacral dimple. Atypical presacral dimple defined as offcentre, > 2.5cm from anal verge and > 0.5cm deep. 40% of patients with atypical presacral dimple will have underlying spinal dysraphism. Combination of hairs/atypical dimple a/w highest risk of dysraphism. Diagnosis:

- If < 6 months old – spinal USS (ossification of spine at 6 months)
- If > 6 months old – spinal MRI

Classification of NLUTD in myelomeningocoele

- Suprasacral (discoordinated voiding)
  - Retained sacral conus reflexes (anocutaneous (S5); bulbocavernosus (S3,4))
  - Neurogenic detrusor overactivity
  - DSD
  - No sphincter paralysis
- Sacral (acontractile bladder)
  - Absent sacral conus reflexes
  - Often poorly compliant bladder
  - High pressure (retainers) or low pressure (wetters) dependent on intrinsic EUS tone (static sphincteric obstruction)
  - NB. Paralysis of EUS means that intrinsic tone may be overcome by external pressure – 'expressible bladder' pathognomonic for neuropathic bladder (Crede's manoeuvre)

Management neuropathic bladder

Principles

- #1 Preserve renal function
- #2 Provide continence (3 features)
  - (i) adequate storage
  - (ii) complete emptying*
  - (iii) voiding at will*

* CISC in all patients if possible (but only 20% continent)

Additional measures

- A. Detrusor hyperreflexia & detrusor non-compliance
  - Anticholinergics
  - Augmentation cystoplasty
    - Enterocystoplasty
    - Ureteocystoplasty (only grossly dilated ureters)
    - Detrusor myomectomy
    - Detrusor myomectomy + demucosalised enterocystoplasty
- B. Sphincteric incompetence
  - Medication (a-adrenergic agonists)
  - Periurethral bulking agents
  - Bladder neck suspension
  - Bladder neck slings
  - Pippe-Salle (aka Kropp onlay procedure – girls only)
  - AUS

Bladder neck closure

c. Other surgical procedures
- Mitrofanoff
  - Appendix, fallopian tube, Monti
- Vesicostomy
- Urethral sphincterotomy
- Ileal conduit
- MACE

Exstrophy-epispadias complex

Failure of cloacal membrane to retract caudally prevents medial migration of mesenchyme = rotational abnormality of lower abdomen and pelvis
In order of severity cloacal exstrophy, bladder exstrophy and epispadias

1. Bladder exstrophy
    - Most common (1:50,000)
    - Boys > girls (3:1)
    - Increased risk 500x in offspring of adults with exstrophy
    - Usually detected by fetal ultrasonography
    - Features
        - Low-lying umbilicus
        - Exposed bladder plate
        - Pubic symphysis diastasis
        - Divarication of recti
        - Inguinal hernia (80% boys; 10% girls)
        - Anterior ectopia of vagina and anus
        - Epispadias always present
        - a/w VUR and inguinal hernia but not other congenital anomalies (cf. cloacal exstrophy)
    - Management
        - Traditionally 3-stage surgical repair:
        - (i) Primary bladder closure
            - neonatal period, typically with pelvic osteotomy
            - no attempt to close bladder neck – all incontinent
        - (ii) Epispadias repair (see below)
            - 1-2 years
        - (iii) Bladder neck closure
            - > 5 years
            - Bilateral ureteric reimplantation and tubularisation of trigone
            - Augmentation for those with low bladder capacity
            - Urethral sensation intact – ISC often problematic requiring mitrofanoff +/- BN closure
        - Recently a number of centres (Erlangen perhaps most famous) have advocated a single-stage repair at ~ 10 weeks
    - Outcome
        - 70% acceptable continence after staged repair (NB. Single most important factor predicting continence is bladder capacity)
        - 50% upper tract dilatation
        - Reasonable sexual function in males but impaired fertility
        - Normal fertility in females but uterine prolapse and fetal malpresentation

2. Epispadias
    - Less common cf. bladder exstrophy (1:120,000)
    - Boys > girls 5:1
    - May be glanular or penopubic
    - Features
        - Open urethral plate

- Dorsal chordee and lack of dorsal foreskin in males
- Bifid clitoris (manta ray) and patulous urethra in females
- Pubic symphysis diastasis
- Incontinence in all females and 70% males (only glanlular continent)

Management

- Surgical repair at 1-2 years e.g. Cantwell-Ransley
- Principles
  - Penile lengthening
  - Correction of chordee (inverting caverno-cavernostomy)
  - Urethroplasty
  - Skin coverage (pedicled prepucial island flap)
- Additional surgical procedures to improve continence in ~70% at 4-5 years

3. <u>Cloacal exstrophy</u>

- Rare (1:200,000)
- Boys > girls
- Combined bilateral hemibladder and central ileocaecal exstrophy
- Often vestigial penis and absent testes
- Frequent co-existent congenital anomalies
- Management
  - Termination of pregnancy
  - Immediate managment
    - Cover with saline packs and either clingfilm or a silo
    - Give prophylactic antibiotics (Trimethoprim 2mg/kg)
    - Refer Leeds or Great Ormond Street
  - Terminal colostomy and bladder repair as for classic exstrophy
  - Male to female gender assignment (controversial)

<u>Anorectal malformations</u>

1:5000
Males > females 3:2
May be high (above levator) or low (below levator)
High fistulas always associated with urinary fistula, low fistulas sometimes not
Two thirds of boys have high fistulas; two-thirds of girls low fistulas
Typically boys have worse functional outcome (except girls with cloacal malformation)
Commonly a/w other congenital abnormalities, often as a syndrome
Commonest syndrome VACTERL

- V – vertebral* (sacral agenesis)
- A – anorectal*
- C – cardiac
- TE – tracheo-oesophageal
- R – renal* (VUR)
- L – Limb (typically radial)
- * commonest

Investigation of newborn with anorectal abnormality

- V - screen by plain x-ray spine and spinal USS
- R – renal tract USS/MCUG/MAG3

Management of anorectal abnormalities (many surgical procedures)
e.g. Sigmoid colostomy + posterior sagittal anorectoplasty (Pena)
CHARGE syndrome (Coloboma, heart, choanal atresia, mental retardation, genital, ear)