**Neuropathic bladder**

Rare
Girls > boys
Boys with more upper tract complications
CISC mainstay of treatment but only ~20% dry

**Aetiology**

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

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

Failed fusion of dorsal vertebrae (lamina)

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 myelomeningocele**
- 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 chorddee 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 chorddee (inverting caverno-cavernostomy)
Urethroplasty
Skin coverage (pedicled prepucial island flap)

Additional surgical procedures to improve continence in ~70% at 4-5 years

3. Cloacal extrophy
Rare (1:200,000)
Boys > girls
Combined bilateral hemibladder and central ileocaecal extrophy
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 extrophy
Male to female gender assignment (controversial)

Anorectal malformations
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)