Posterior urethral valves
Originally described by Hugh Hampton Young in 1919
Males only
Incidence 1:5000
Possibly higher incidence in Down’s syndrome; No a/w VACTERL
Young originally described 3 types, I II and III
- Type 1 95% of valves
  Bicuspid valves from veru, through membranous urethra (zone of EUS) to anterior urethral wall
- Type 2 Now known not to be a/w posterior valves
  Hypertrophied band of tissue in posterior urethra a/w dysfunctional voiding
- Type 3 5% of valves
  Sheet membrane with central aperture. Probably a variation of type 1
Now believed that all patients have a type 3 iris-shaped deformity, termed a congenitally obstructing posterior urethral membrane (COPUM), which is converted to a ‘type 1’ deformity by he passage of a catheter after birth. Typically arise at 7 weeks gestation due to abnormal interaction of mesonephric duct confluence and urogenital sinus. Early severe obstruction causes bladder outflow obstruction (valve bladder), upper tract dilatation and renal dysplasia. Later mild obstruction a/w bladder outflow obstruction without dysplasia.
Presentation
  Prenatally
  Prenatal diagnosis in 80% (approximately 50% detected at routine second trimester scan; remainder later for obstetric indications)
  Diagnosed on USS
    Distended bladder and dilated upper tracts (DD = urethral atresia (fatal), prune belly and severe bilateral VUR)
    Occasionally keyhole sign (dilated bladder and proximal urethra; see below)

  Oligohydramnios, pulmonary hypoplasia, moulding defects (Potter’s facies, talipes etc.)
  Potter’s facies (4)
    Flattened nose
    Recessed chin (hypognathism)
    Prominent epicanthal folds
    Low-set ears
  Predictors of poor functional outcome and early renal failure:
Detection before 24 weeks
Thickened bladder wall
Echobright kidneys (dysplastic)
Oligohydramnios

Postnatally
AUR, palpable abdominal mass, listlessness in neonates
UTI, growth retardation in older children

Diagnosis
USS Dilated upper tracts, thick-walled bladder, occasionally dilatation of proximal urethra
MCU Diagnostic
Dilated proximal urethra with ‘sail in the wind’ appearance of valve; dilated trabeculated bladder; VUR in ~ 50% cases
Perineal USS – research tool only

Management
Prenatally
Fetal vesico-amniotic shunting
Theoretical benefit from animal studies in preventing decline in renal function and correcting oligohydramnios
RCT never been performed and long-term follow-up difficult, therefore ? beneficial in humans
Results not encouraging – mortality 40-60% (Fetal Surgery Register, Elder 1987) but performed on patients with very poor prognosis
May have limited role
Elective pre-term delivery
Theoretical benefit – earlier treatment in late onset cases but no evidence base
Late termination of pregnancy
Often for oligohydramnios with marked dilatation

Postnatally
Correction of electrolyte abnormalities first followed by early incision of valves
Correction of electrolyte abnormality
Urethral or suprapubic catheter drainage 2-7 days
5-8F infant feeding tube (Foley balloon can block ureteric orifices)
Vesicostomy for severe renal impairment or when neonatal endoscopic instruments unavailable
Endoscopic valve ablation
4 and 8 o’clock positions
With increasing diameter of urethra:
   a. Whittaker hook
   b. Neonatal cystoscope and bugbee electrode
   c. Paediatric resectoscope and loop

Prognosis
Poor prognostic features in COPUM
Antenatally
   Oligohydramnios
   Early pre-natal detection < 24 weeks
   Thickened bladder wall
   Echobright kidneys
   High B2 microglobulin in urine
Postnatally
   Early presentation < 1 month
   Bilateral VUR
   Impaired renal function (see below)
   Proteinuria
   Daytime incontinence (after 5 years old)
   UDS showing poor compliance or detrusor failure
Renal failure
30% impaired renal function [primary dysplasia, BOO, VUR and UTI and persistent bladder dysfunction 2’ to valve bladder]
Predicting renal failure on follow-up:
   Oligohydramnios
   Early prenatal detection
   Clinical presentation within 6 months
   Proteinuria
   Bilateral VUR**
   Impaired continence > 5 yrs
   ** recognised good prognosis features (‘pop-off’ mechanisms) include unilateral VUR with dysplasia (aka VURD: massive reflux into non-functioning kidney preserves function in other kidney), ascites, perinephric urinoma and bladder diverticulum

Valve bladder
Features
   Poor sensation
   Hypercontractility
   Poor compliance
   Bladder neck hypertrophy
   Concentrating defect leading to high urine output (which increases cycling - therefore more upper tract
dysfunction. To avoid overnight cycling Koff popularised concept of overnight catheterisation)
Typically high resting intravesical pressures, poor emptying and incontinence, leading to eventual bladder atonicity
UDS findings
10% detrusor overactivity (young)
65% poorly compliant high pressure bladder
25% myogenic failure (old)
Managed by double void and ISC, but compliance poor due to preserved sensation. May require augmentation + mitrofanoff

Other urethral abnormalities
Anterior urethral valves
Incidence 1:40,000 (8x less common than posterior valves)
40% bulbar; 30% penoscrotal; 30% penile
No upper tract pathology
Diagnosis with MCUG
Mx = endoscopic incision

Urethral diverticulum
Usually wide-mouthed at penoscrotal junction
Distal lip may give obstruction
Usually UTI or obstruction
Conservative or excision/grafting

Megalourethra
Dilatation without obstruction
a/w absence of corpus spongiosum
may be part of prune belly syndrome

Cowper’s gland cysts
Arise level with urogenital diaphragm (EUS)
Dilatation can cause urethral obstruction

Urethral duplication (Shotgun urethra)
Sagittal (over and under) common
Typically orthotopic urethra with accessory dorsal – usually excised
Collateral (side by side) uncommon

Posterior urethral polyps
Fibroepithelial and pedunculated – excise

Cobbs collar
Controversial
Slight narrowing of urethra distal to urethral sphincter on MCU
May be soft narrowing on cystoscopy
Not thought to be obstructive