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

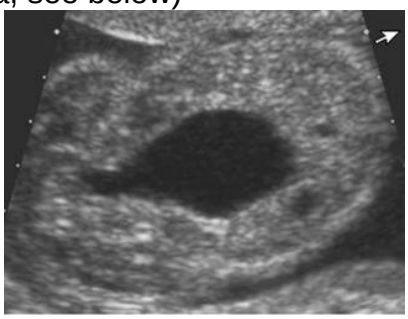
- Type 1 — 95% of valves
  - Bicuspid valves from veru, through membranous urethra (zone of EUS) to anterior urethral wall
- Type 2 — Now known not to be a/w posterior valves
  - Hypertrophied band of tissue in posterior urethra a/w dysfunctional voiding
- Type 3 — 5% of valves
  - Sheet membrane with 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

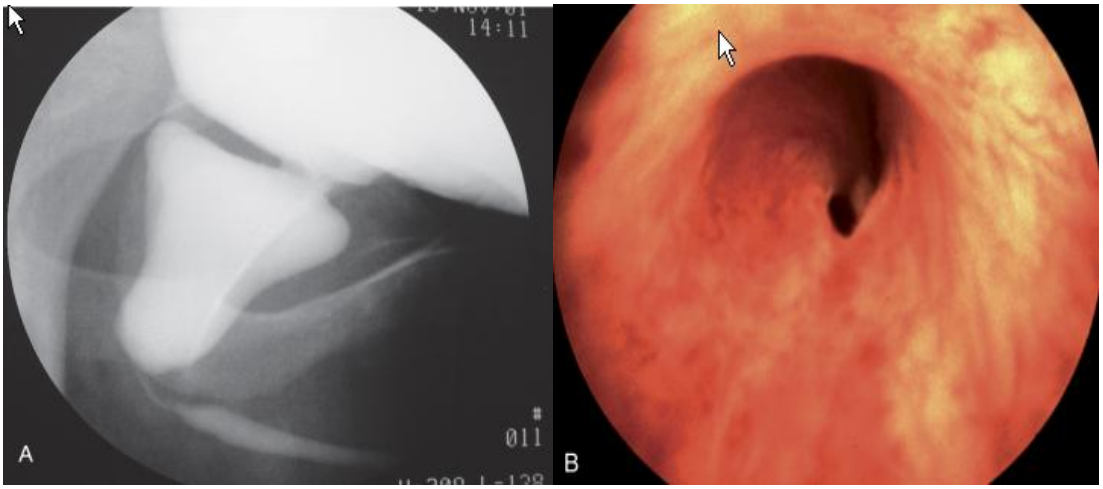
- Prenatally
  - Prenatal diagnosis in 80% (approximately 50% detected at routine second trimester scan; remainder later for obstetric indications)
  - Diagnosed on USS
    - Distended bladder and dilated upper tracts (DD = urethral atresia (fatal), prune belly and severe bilateral VUR)
    - Occasionally 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

<u>Other urethral abnormalities</u>

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 obststruction

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 MCUG

May be soft narrowing on cystoscopy

Not thought to be obstructive