Prune Belly Syndrome

Aka Eagle-Barrett syndrome or triad syndrome

- *Failed abdominal wall development*
- *Bilateral intra-abdominal testes*
- *Urinary tract anomalies*

Demographics

1:30,000 live births

Males >> females (95%)

Twins > singletons

Unknown aetiology – multiple theories

- Possibly temporary glanular urethral obstruction, leading to dilated urethra and bladder. Massively enlarged bladder proposed to inhibit normal development of abdo wall musculature

Associated with trisomy 18 and 21

Clinical features

- Failed abdominal wall development
  - Ranges from total absence to limited defects
  - Medial and inferior elements most commonly absent
- Bilateral intra-abdominal testes
  - Most commonly at level of CIA
  - Histology similar to non-PBS patients
  - Spontaneous paternity never reported
  - ICSI successful in a few case reports
- Urinary tract anomalies
  - Renal dysplasia
    - 50%, determines severity
  - Hydroureteronephrosis
    - lack of ureteric smooth muscle, distal > proximal
  - Massive bladder
    - pseudodiverticulum at dome
    - patent urachus in 25%
    - increased ratio of collagen to muscle
    - 50% void spontaneously with normal UDS and low PMRs
  - Posterior urethra
    - Dilated prostatic urethra
    - Hypoplastic prostate
    - Dilated, occasionally absent vas and seminal vesicles
    - Typically retrograde ejaculation
  - Anterior urethra
    - Megalourethra
    - Urethral atresia (fatal unless patent urachus)
  - Associated non-urological abnormalities
    - Overall seen in ~75%
    - Pulmonary – hypoplasia, pneumothorax (50%)
    - Orthopaedic - scoliosis, congenital hip dislocation (50%)
    - Gastointestinal - malrotation, atresia, stenosis, volv.(30%)
    - Cardiac - PDA, VSD, ASD, tetralogy (10%)

Diagnosis

Classic prenatal USS findings
- Hydroureteronephrosis
- Distended bladder
- Irregular abdominal wall

Typically not seen however. Difficult therefore to distinguish from causes of bladder outflow obstruction such as posterior valves

No correlation between the degree of hydroureteronephrosis and renal dysfunction

Classification (Described by Woodard 1985)

Category	Characteristics
I	Renal dysplasia
	Oligohydramnios
	Pulmonary hypoplasia
	Potter's features
	Urethral atresia
II	Full triad features
	Minimal or unilateral renal dysplasia
	No pulmonary hypoplasia
	May progress to renal failure
III	Incomplete or mild triad features
	Mild to moderate uropathy
	No renal dysplasia
	Stable renal function
	No pulmonary hypoplasia

Category 1 patients almost always either stillborn or die in perinatal period; exception is those patients with patent urachus.
Category 3 patients usually do well. Urologic intervention reserved for those with complications (i.e. UTI)
Category 2 patients intermediate risk. Intervention to prevent renal decline controversial

Management

Initial management in all patients
- MDT consisting of neonatology, nephrology, urology plus others
- U+E
- Renal tract USS
- VCUG with ABx cover if renal insufficiency or evidence of BOO.
- CXR for pneumothorax, pneumomediastinum, and pulmonary hypoplasia
- SPC for BOO
- ? Circumcision (recommended in Campbell's)

Orchidopexy

Complete transperitoneal mobilisation with division of vascular pedicles readily achievable if performed at ~6 months
Alternatively Fowler-Stephens (one or two-stage) or autotransplantation and microvascular anastomosis

Management of Category 2 patients (PBS and renal insufficiency)

Aim to prevent upper tract UTI
Reduction cystoplasty, ureteric shortening, tapering and reimplantation with or without abdominoplasty (eg. Monfort technique)
Recommended by some with good long-term preservation of renal function (Woodard)
Others advocate careful surveillance with intervention for repeated febrile UTI or renal deterioration (Woodhouse)

<u>Prognosis</u>

Nadir creatinine in infancy useful predictor of long-term function
If creatinine <60 umol/l long-term renal function usually stable
30% of patients with impaired renal function develop chronic renal failure before adulthood