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%)
Gastrointestinal - 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)

<table>
<thead>
<tr>
<th>Category</th>
<th>Characteristics</th>
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<tbody>
<tr>
<td>I</td>
<td>Renal dysplasia</td>
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<tr>
<td></td>
<td>Oligohydramnios</td>
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<td>Pulmonary hypoplasia</td>
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<td>Potter's features</td>
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<td></td>
<td>Urethral atresia</td>
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<td>II</td>
<td>Full triad features</td>
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<td></td>
<td>Minimal or unilateral renal dysplasia</td>
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<td></td>
<td>No pulmonary hypoplasia</td>
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<td></td>
<td>May progress to renal failure</td>
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<tr>
<td>III</td>
<td>Incomplete or mild triad features</td>
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<tr>
<td></td>
<td>Mild to moderate uropathy</td>
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<td></td>
<td>No renal dysplasia</td>
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<td></td>
<td>Stable renal function</td>
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<tr>
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<td>No pulmonary hypoplasia</td>
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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)

Prognosis
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