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香港兒童醫院 Hong Kong Children's Hospital

Beyond Clinical Mimicry: Imaging Keys to Distinguishing Prune Belly Syndrome (PBS) from Posterior Urethral Valves (PUV)

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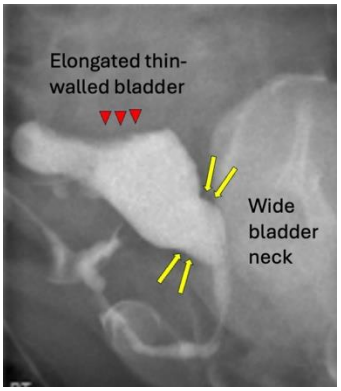
PBS and PUV are congenital urological anomalies primarily affecting male paediatric patients, both causing urinary tract obstruction but differing in pathology and imaging features.

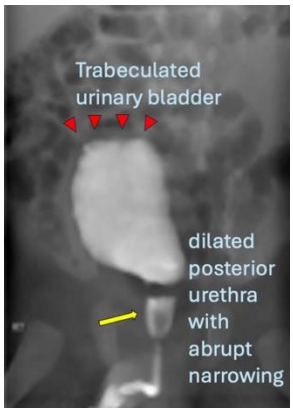
CASE 1

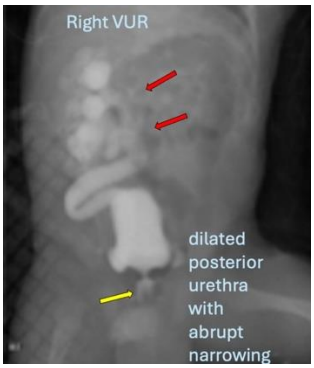
A 36-week male neonate with antenatal megacystis and lower urinary tract obstruction managed by fetal vesicoamniotic shunting. Postnatal findings included abdominal wall deficiency, bilateral intraabdominal testes, elongated bladder with trabeculation, and bilateral vesicoureteral reflux, consistent with PBS. The infant underwent emergency shunt removal and bladder repair, with multidisciplinary follow-up and initiation of clean intermittent catheterization.



CASE 2

A term male infant diagnosed antenatally with bilateral hydronephrosis and PUV. Postnatal imaging confirmed PUV and features of chronic bladder outlet obstruction with Grade 5 right vesicoureteral reflux (VUR). Endoscopic valve ablation was performed successfully. The patient experienced recurrent urinary tract infections managed with antibiotics prophylaxis and multidisciplinary monitoring.





PRUNE BELLY SYNDROME (PBS)

Pathophysiology & Core Defect:
A developmental malformation syndrome.
Primary issue is a **deficiency of the abdominal muscles**. Not caused by a physical obstruction.
RARE, 1 in 30,000-50,000 live male births[1].
Classic Clinical Triad[2]:
Deficient abdominal muscles ("prune-like" appearance).
Urinary tract dilation (megacystis, megaureter, hydronephrosis).
Bilateral cryptorchidism (undescended testicles).
Imaging Characteristics:
Bladder: Massively distended and **thin-walled**.
Upper Tracts: Severe hydroureteronephrosis.
VCUG (Voiding Cystourethrogram):
Shows a wide bladder neck and a normal-caliber urethra without narrowing[2].
Treatment:
Multidisciplinary repair, lifelong monitoring[3].

POSTERIOR URETHRAL VALVE (PUV)

Pathophysiology & Core Defect:
An obstructive uropathy. Caused by **abnormal development or persistence of embryonic membranes** in the posterior urethra.
These "valves" cause a mechanical obstruction to urine flow.
More common, 1 in 3800 live male births[4].
Key Features:
Obstruction **without** underlying muscle defects[5].
Cryptorchidism is **not** a feature.
Imaging Characteristics:
Bladder: Thick-walled, trabeculated.
Ultrasound: Classic **"keyhole sign"** (dilated posterior urethra with thick walled bladder).
VCUG: Dilated, irregular posterior urethra with an abrupt narrowing (at level of the valve)[6].
Treatment:
Early endoscopic valve ablation/ resection[7] and managing complications.

PUV and PBS are major causes of congenital urinary obstruction in boys. Distinct clinical and imaging features aid diagnosis; early multidisciplinary care improves outcomes.