NOTES
BLADDER & URETHRAL CONGENITAL DISORDERS

GENERALLY, WHAT ARE THEY?

PATHOLOGY & CAUSES

- Congenital abnormalities in bladder, urethra
- Benign/kidney failure/systemic involvement

CAUSES
- Interferences in fetal development

SIGNS & SYMPTOMS

- May be asymptomatic until complications develop

DIAGNOSIS

DIAGNOSTIC IMAGING

Prenatal ultrasound
- Diagnosis difficult

OTHER DIAGNOSTICS
- *Bladder exstrophy, hypospadias, epispadias:* visible at birth

TREATMENT

SURGERY
- See individual disorders

BLADDER EXSTROPHY

osms.it/bladder-exstrophy

PATHOLOGY & CAUSES

- Congenital disorder, inside-out bladder protruding out of abdomen
- Part of the exstrophy-epispadias complex (EEC) that includes epispadias and cloacal exstrophy
- Bladder fails to fully form anteriorly, pushed through front anterior abdomen wall

CAUSES
- *Occurs during embryological development:* overdevelopment of cloacal membrane disrupts development of the lower abdominal wall → prevents migration of the mesenchymal tissue towards midline → rupture of cloacal membrane → herniation of lower abdominal components through the lower abdominal wall surface

RISK FACTORS
- Genetic predisposition
- Biological males > biological females
- Firstborn > subsequent births
- Infants born to white parents

COMPLICATIONS
- Urinary and/or fecal incontinence, UTIs, abnormal gait, hip dysplasia, rectal prolapse; inguinal hernia, uterine prolapse

SIGNS & SYMPTOMS

- Observable changes in pelvis, pelvic floor, genitalia
  - Exposed bladder and urethra
  - Low-set umbilicus
  - Abnormalities of the pelvic bone, vertebral column, and spinal cord
  - Flattened puborectal sling, anus anteriorly displaced
  - *Biological males:* epispadias, absent dorsal foreskin, open prostate, shortened penis
  - *Biological females:* vagina wider, shorter, more vertically oriented; displaced, narrowed vaginal orifice; bifid clitoris; divergent labia
  - Epispadias

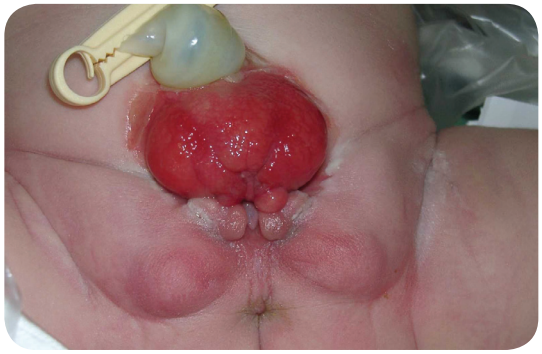

Figure 107.1 A newborn baby with a severe case of bladder exstrophy. The genitals are also grossly irregular.

DIAGNOSIS

DIAGNOSTIC IMAGING

CT scan

- CT scan detects skeletal abnormalities

Ultrasound and MRI

- Often made by prenatal ultrasound, can be confirmed by MRI

OTHER DIAGNOSTICS

- Clinically recognizable at time of delivery

TREATMENT

SURGERY

- Performed within first weeks of life
- Staged surgeries required over months/ years

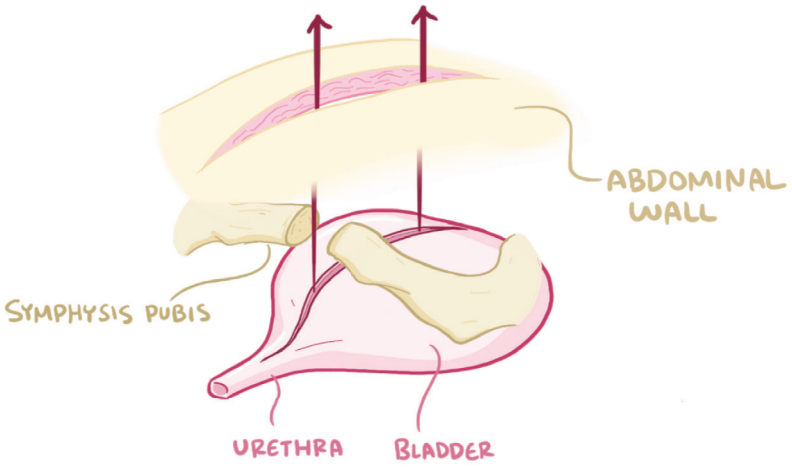

Figure 107.2 Illustration of the bladder pushing through the symphysis pubis and abdominal wall during bladder exstrophy.

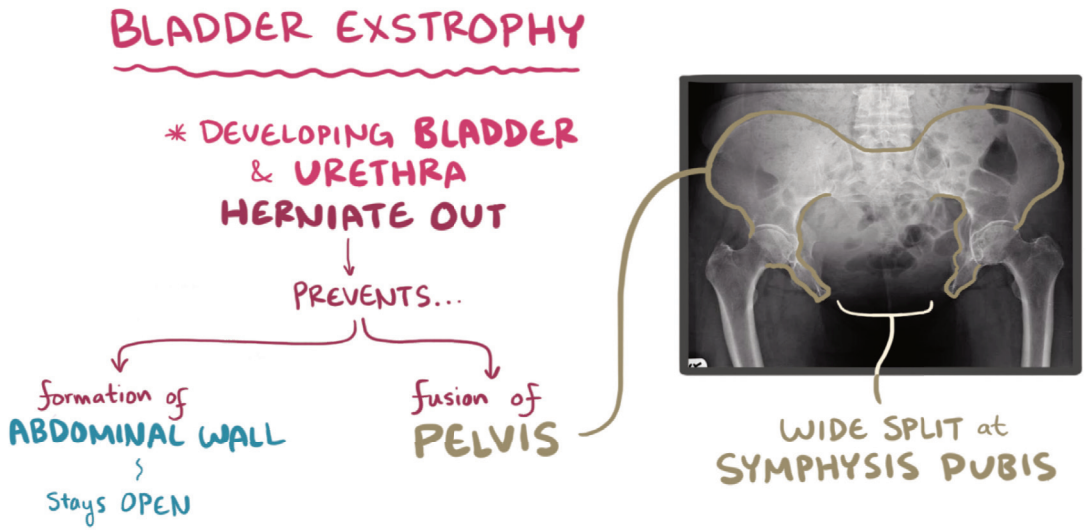

Figure 107.3 A plain pelvic radiograph demonstrating a wide symphysis pubis in a case of bladder exstrophy.

HYDRONEPHROSIS

osms.it/hydronephrosis

PATHOLOGY & CAUSES

- Dilation of renal pelvis, calyces associated with kidney atrophy
- Severe, long-standing hydronephrosis → kidney failure
- Urinary tract obstruction/compression → build up of urinary pressure → progressive dilation
- Dilation starts at blockage, continues up towards kidneys
  - *Hydroureter:* dilation of ureter
  - *Hydronephrosis/hydroureteronephrosis:* dilation of ureter, renal pelvis, calyces
- Grading
  - *0:* no dilation
  - *I:* dilation of renal pelvis
  - *II:* dilation of renal pelvis, calyces
  - *III:* moderate dilation of renal pelvis, calyces; mild cortical thinning, flattening of papillae
  - *IV:* severe renal dilation; cortical thinning

CAUSES

- *Fetus:* antenatal hydronephrosis
  - Often unknown, may disappear on own
  - *Congenital malformation:* ureteropelvic junction obstruction, vesicoureteral reflux
- *Children:*
  - *Congenital malformation:* ureterocele, posterior urethral valves
- *Adults:*
  - *Acquired disease:* kidney stones (most common cause), benign prostatic hyperplasia, blood clot, contiguous malignant diseases (prostate/bladder/cervix cancer, retroperitoneal lymphoma), contiguous inflammation (prostatitis, ureteritis, urethritis, retroperitoneal fibrosis), tissue scarring from injury/surgery, uterus enlargement during pregnancy

MNEMONIC: SIP BaN

Causes of acquired hydronephrosis

Stones
Inflammation
Prostate hypertrophy
Baby (pregnancy) / Blood clot
Neoplasm

SIGNS & SYMPTOMS

- Acute with sudden onset: intense pain in flank, called Dietl's crisis
- Nausea, vomiting

DIAGNOSIS

DIAGNOSTIC IMAGING

Abdominal ultrasound

- Dilation of renal calyces
- Increased anteroposterior diameter
- Dilated ureter, if obstruction is distal

Prenatal ultrasound

- Oligohydramnios if bilateral obstruction

Intravenous (IV) urography/pyelography

- Demonstrates distal obstruction

TREATMENT

SURGERY

- *Restore urine flow:* upper blockage
  - *Acute:* nephrostomy tube
  - *Chronic:* ureteric stent/pyeloplasty

OTHER INTERVENTIONS

- *Restore urine flow:* lower blockage
  - Urinary or suprapubic catheter

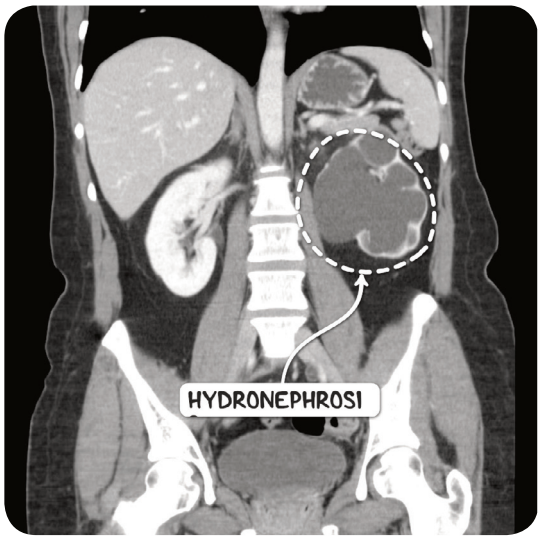

Figure 107.4 An abdominal CT scan in the coronal plane demonstrating severe hydronephrosis of the left kidney.

POSTERIOR URETHRAL VALVE

osms.it/posterior-urethral-valve

PATHOLOGY & CAUSES

- Congenital disorder, posterior urethra obstructed by membranous folds/tissue flap
- Most common cause of bladder outlet obstruction in infants who are biologically male
- Obstruction increases bladder pressure → bladder wall hypertrophy → decreases bladder compliance → repeats
- Obstruction increases bladder pressure → ureterovesical junction dysfunction → vesicoureteral reflux
- Urine retention by obstruction → urine backs up → bilateral hydronephrosis
- Severe obstructions *in utero* → oligohydramnios → Potter syndrome
  - Limb irregularities, facial anomalies, kidney failure, pulmonary hypoplasia

CAUSES

- *Unknown; theory:* abnormal integration of Wolffian duct → large plicae colliculi fuse anteriorly

SIGNS & SYMPTOMS

- Posterior urethra obstructed by membranous folds/tissue flap

DIAGNOSIS

DIAGNOSTIC IMAGING

Prenatal ultrasound

- Generally seen > 26 weeks gestation
- Noticeable distension and hypertrophy of bladder
- Possitlbe hydronephrosis and hydroureter
- *Keyhole sign:* distended proximal urethra and thick-walled bladder, resembles keyhole

Antenatal ultrasound

- Hydronephrosis (10% may be normal)
- Trabeculated and thick-walled bladder with elongation and dilation of posterior urethra
- Valve may be seen as echogenic line

Voiding cystourethrogram (VCUG)

- Dilation and elongation of posterior urethra
- Vesicoureteral reflux (in half of instances)
- Bladder trabeculation or diverticula
- Radiolucent linear band (representing valve)

TREATMENT

SURGERY

- Surgical ablation of membrane
- Prenatal surgery

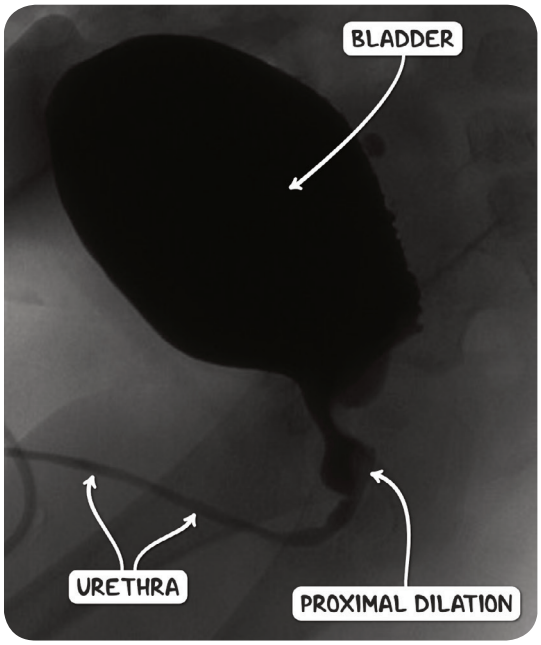

Figure 107.5 A lateral view of a micturating cystourethrogram demonstrating a proximally dilated urethra in case of posterior urethral valve.

VESICOURETERAL REFLUX

osms.it/vesicoureteral-reflux

PATHOLOGY & CAUSES

- Retrograde flow of urine from the bladder into the ureters and kidneys
- Grading
  - *Grade I:* urine goes into ureters
  - *Grade II:* urine fills entire ureter, renal pelvis
  - *Grade III:* urine fills, stretches ureter, renal pelvis
  - *Grade IV:* ureter swollen, curvy; renal pelvis, calyces swollen, distorted
  - *Grade V:* urine fills ureter, pelvis, calyces; swell completely
- *Primary vesicoureteral reflux (most common type):* due to congenital defect at ureterovesical junction (congenital absence/shortening of intravesical portion of ureter)
  - Inadequate closure of the ureterovesical junction → urine builds up in bladder → ureter fails to act as valve → urine returns to ureters
- *Secondary vesicoureteral reflux:* due to failure of the ureterovesical junction to close during bladder contraction; often due to a blockage in urinary tract
  - Pressure increases in urinary tract → urine follows path of least resistance, back into ureters

RISK FACTORS

- Genetic predisposition
- *Neonates:* prenatal hydronephrosis
- *Children:* febrile UTIs
- Individuals of white, Northern European descent

COMPLICATIONS

- Recurrent UTIs, pyelonephritis, renal scarring/fibrosis, hypertension, kidney failure
- *Infants:* asymptomatic, fever, lethargy, poor appetite
- *Children:* discomfort with urination; bowel and bladder dysfunction

SIGNS & SYMPTOMS

- *Infants:* asymptomatic, fever, lethargy, poor apetite
- *Children:* discomfort with urination

DIAGNOSIS

DIAGNOSTIC IMAGING

Abdominal ultrasound

- Assesses renal parenchyma for scarring or anatomical abnormalities
- Presence of hydronephrosis

VCUG

- Should be performed on first UTI in child < six years old
- Used for grading
- Presence of other anatomical abnormalities

TREATMENT

SURGERY

- Primary vesicoureteral reflux
  - Surgery to repair valve at ureterovesical junction
  - *Infants, children:* no intervention; child grows → ureters lengthens → valve function improves
- Secondary vesicoureteral reflux
  - Surgery to remove blockage

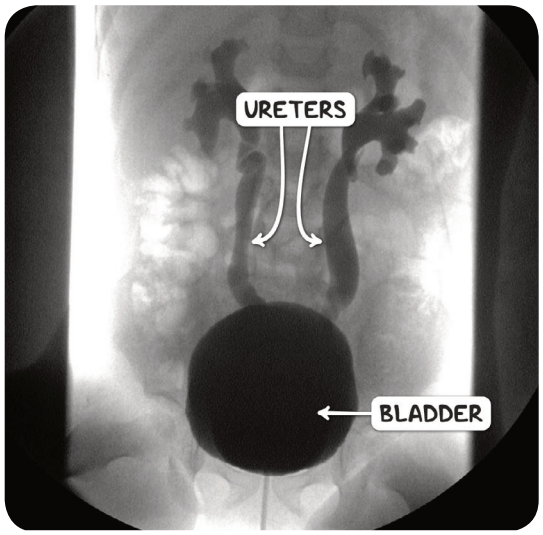

Figure 107.6 A voiding cystourethrogram demonstrating bilateral vesicoureteric reflux.