Congenital anomalies

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Posterior urethral valves The incidence of posterior urethral valves is around 1 in 8000 live male births. The valves are membranes that have a small posterior slit within them. They typically lie just distal to the verumontanum and cause obstruction to the urethra. They function as flap valves; although they are obstructive to antegrade urinary flow , a urethral catheter can be passed retrogradely without any di ffi culty . Posterior urethral valves need to be detected and treated as early as possible to minimise the degree of renal failure. Diagnosis Antenatal ultrasound shows a distended bladder, dilated prostatic urethra and hydroureteronephrosis. The presentation varies according to the severity of the obstruction. The more severe the obstruction, the earlier the presentation. If the diag - nosis is not made antenatally , babies typically are presented by parents because of voiding complaints and urinary tract infection (UTI). Rarely the valves are incomplete, and the patient may present in adolescence or adulthood. Impaired renal function is assessed by ultrasound to check r enal cortical thickness and by nuclear renography to check - for di ff erential r enal function. Investigations include a voiding cystourethrogram (VCUG), which shows a dilated posterior (prostatic) urethra (Figure 85.1). The bladder is hypertrophied and often shows diverticula. Typically , there may be vesicouret - eral reflux.

The diagnosis and treatment of phimosis • The diagnosis and treatment of erectile dysfunction • The common diseases of the penis and urethra and the • principles of their surgical management Figure 85.1 A voiding cystourethrogram showing a dilated bladder with a dilated prostatic urethra above an obstruction at the level of the posterior urethral valves (courtesy of Dr Shashank Shrotriya, Pune, India).

Initial treatment is by catheterisation to drain the urine and decompress the bladder and upper urinary tracts. The valves themselves can be di ffi cult to see on urethroscopy because the flow of irrigant sweeps them into the open position. Definitive treatment is by endoscopic ablation of the valves. Long-term follow-up is required in view of the associated vesi coureteral reflux, bladder dysfunction and renal impairment. Summary box 85.1 Posterior urethral valves /uni25CF /uni25CF /uni25CF /uni25CF The incidence of hypospadias is around 1 in 300 male live births. It is the most common congenital abnormality of the urethra. Diagnosis is made on physical examination. There are three characteristic features, including an ectopic ventrally located urethral meatus; usually a ventral penile curvature - (chordee); and an incomplete dorsal hood prepuce. Hypospadias is classified according to the position of the meatus (Figure 85.2a–d). /uni25CF Glanular hypospadias: the ectopic meatus is placed on the glans penis, but proximal to the normal site of the external meatus, which is marked by a blind pit. /uni25CF Coronal hypospadias: the meatus is placed at the level of the coronal sulcus. /uni25CF Penile hypospadias: the meatus is on the underside of the penile shaft. /uni25CF Penoscrotal hypospadias: the meatus is at the level of the penoscrotal junction. /uni25CF Perineal hypospadias: this is a rare and severe abnormal - ity . The scrotum is bifid, and the urethra opens between

Posterior urethral valves are congenital membranes that cause obstruction to the urinary tract in the male Antenatal ultrasound typically shows a distended bladder, dilated prostatic urethra and hydroureteronephrosis Treatment is by endoscopic valve ablation Patients need long-term follow-up in view of recurrent UTI, bladder dysfunction and renal impairment (a) Hooded foreskin Glanular Coronal Penoscrotal Perineal (c) (d) Figure 85.2 (a) Hypospadias classi /f_i cation. (b) Coronal hypospadias. tion in which

the scrotum is placed superior and anterior to the penis. (b) (e) (c) Midpenile hypospadias. (d) Hypospadias with penoscrotal transposi

(e) Urethrocutaneous /f_i stula seen in multiple failed hypospadias surgeries.

important to consider disorders of sexual development, which are usually associated with undescended testes, her nia and micropenis. Treatment Surgery for distal hypospadias is often for cosmetic reasons. This is usually treated by a tubularised incised plate urethro plasty . Proximal hypospadias with chordee needs surgical correction and may involve a two-stage repair. The first stage corrects the penile curvature and the second stage r epairs the urethra. Circumcision should be avoided as preputial skin may be required for future repairs or revisions. Surgery for hypospa dias is best performed by experts in hypospadias surgery and is typically undertaken before the age of 18 months. Failed hypospadias repair can present as urethrocutaneous fistula (Figure 85.2e). Epispadias Epispadias is very rare. In penile epispadias, the urethral opening is on the dorsum of the penis and is associated with an upward curvature of the erect penis (Figure 85.3). Epis padias often coexists with bladder exstrophy and other severe developmental defects. Summary box 85.2 Hypospadias /uni25CF /uni25CF /uni25CF /uni25CF Urethral diverticulum Congenital urethral diverticulum is rare. It is commonly seen post urethroplasty where genital skin is used for augmentation (Figure 85.4). Typically , patients present with postmicturition dribble. Diagnosis is made by urethrography and the divertic ulum is repaired by surgery .

Hypospadias is diagnosed clinically by a ventrally placed urethral meatus, a hooded foreskin and penile curvature In severe cases with coexisting testicular maldescent and micropenis, consider disorders of sexual development as a diagnosis Avoid circumcision as the prepuce may be used in procedures to correct the abnormality Surgical treatment should be undertaken by experts

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