(24)

Urology

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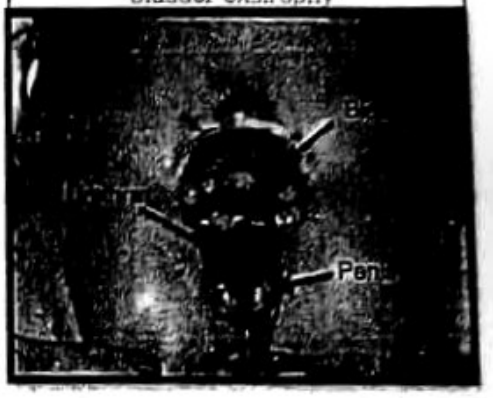
Congenital Anomalies of The Lower Urinary Tract

Bladder Exstrophy (Ectopia Vesicae):

This dreadful malformation is uncommon (1:50000). It is caused by failure of fusion of the pubic bones and anterior abdominal wall resulting in exposure of the bladder. The urinary bladder is open and exposed and the urethra is open. In males there is associated epispadias and in females there is bifid clitoris. There may also be bilateral inguinal hernia and VUR. The pubic bones are usually widely separated.

Sequelae: The exposed bladder is liable for recurrent severe infections. With time, irritation of the bladder mucosa will result in squamous metaplasia that may develop into malignancy.

Widely separated pubic bones in bladder exstrophy



The urinary sphincter is deficient leading to incontinence. The bony abnormality affects gait.

Treatment: The first step is to close the bladder. Next, there may be artificial sphincter and bladder augmentation. Bony defects may need osteotomy.

Hypospadias:

Overview: Hypospadias is one of the common urethral anomalies. Normally, the urethra opens in the tip of the glans penis (The external urethral meatus). In hypospadias, the meatus is situated more proximally on the ventral aspect of the penis.

Etiology: Hypospadias may develop due to incomplete fusion of the urethral plate during fetal life probably due to some maternal hormonal changes.

Epispadias:

Epispadias is a rare anomaly in which the urethral opening is located on the dorsum of the penis. It may be associated with upward penile curvature. It usually occurs in association with bladder exstrophy or other major anomalies. Treatment is surgical.

Epispadias

Posterior Urethral Valve:

Relatively uncommon anomaly occurs in males in which a membranous web is present in the urethra just distal to the veromontanum. This valve is a one-way valve preventing urine flow to the outside. With time it will cause back pressure on the kidney.

PUV is usually diagnosed antenatally with oligohydramnios and small-for-gestational age fetus. Antenatal ultrasound will show bilateral hydronephrosis and a distended bladder. If not diagnosed antenatally, it will present in early neonatal life with poor urinary stream, recurrent infection and may develop into uremia and renal failure.

Retrograde cystogram showin
prostatic urethra in a child wi

Diagnosis: A distended bladder is usually palpable per abdomen. Urinalysis reveals infection and renal function may be impaired. Ultrasound reveals overdistended bladder with possible bilateral hydronephrosis. Cystography reveals distended bladder and dilated urethra above the valve. Cystoscopy may show the valve.

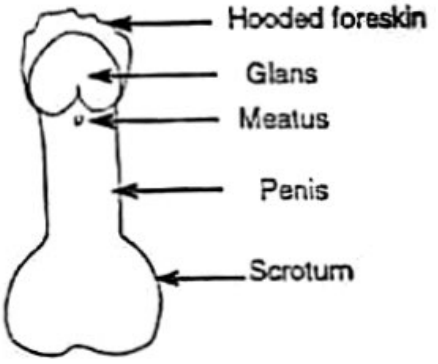
Treatment: Early treatment is by insertion of a urethral catheter to drain the urinary system an decrease pressure. Definitive management is by cystoscopic va ablation.

Clinical Presentation: As mentioned, the external urethral meatus is located more proximal on the ventral aspect of the penis or in the perineum in severe cases. Associated features are:

a. Poorly developed ventral part of the foreskin (Hooded prepuce).
b. Ventral curvature of the penis especially during erection due to poorly developed urethral plate. This curvature is called Chordee.

Hypospadias is classified according to the location of the meatus into:

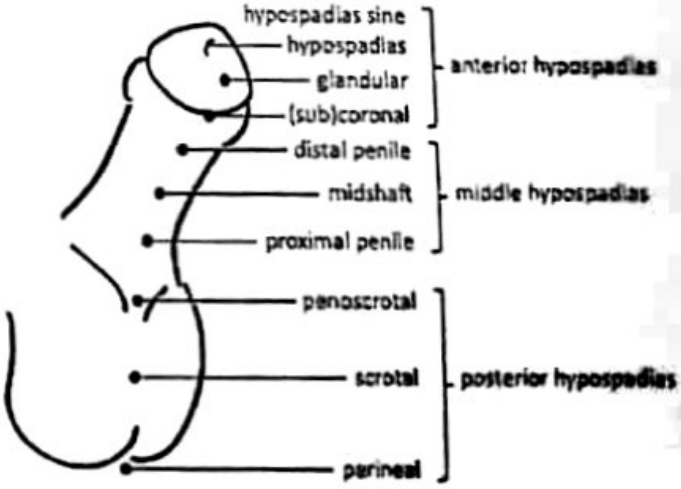
1. Glanular (Glandular).
2. Coronal.
3. Penile Hypospadias: the opening is on the penile shaft. This is further classified into:
    a. Distal shaft
    b. Mid-shaft
    c. Proximal shaft.
4. Penoscrotal
5. Scrotal
6. Perineal: This is the most severe form of the disease and associated with bifid scrotum and testicular maldescent.



The more proximal the opening, the more severe the disease is. In cases of perineal hypospadias, intersex conditions should be suspected.

Examination: In addition to position of the meatus, examination should include assessment of the caliber of the meatus as there may be associated meatal stenosis. One should also look for associated anomalies in the genitourinary system or elsewhere, especially testicular maldescent.

Treatment: The most important step in managing a child with hypospadias is to avoid circumcision as the foreskin may be utilized later for repair.



Surgery is indicated to improve sexual function, correct urinary stream and for cosmetic reasons. It is better performed prior to school age.