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Paediatric perineal skin pit: More than skin deep?

A 2.5-year-old boy was referred to our clinic for abnormal genitalia and a febrile urinary tract infection (*Escherichia coli* >10^5 colony-forming units/mL). At birth, his parents were informed that he had a "bilobed scrotum and a deep perineal skin pit" that may pose hygiene and cosmetic issues. They had never witnessed their son'surine stream as he was still in diapers. On examination,the boy was thriving, non syndromic, with a completely bifid scrotum, penoscrotal transposition, chordee, phimosis and a perineal skin pit close to the midline (Fig. 1). Bilateral testes were approximately 2mL in size and descended. The anus and sacrum were unremarkable.

Ultrasound scans of the kidneys and bladder were normal. The ultrasound scan was unable to trace the skin pit to its origin. Karyotype was normal with 46,XY chromosomes.

What is the suspected diagnosis of the skin pit?

A. Duplicated urethra
B. Perineal skin pit
C. Perineal fistula
D. Dysplastic hypospadiac urethra
E. Crohn's disease

Either a duplicated urethra or dysplastic hypospadiac urethra is likely. It is important to note that a perineal skin pit is only a description. The pit may extend deeper, and be a sign of a genitourinary malformation. A perineal fistula describes an abnormal connection between 2 mucosal lined tissues. While 1 end opens at the skin, it is necessary to delineate the anatomy where the second opening is. There were no signs of other sinuses, or fistulas to suggest Crohn's disease.

The boy underwent surgery with the aims to: (1) repair anomalies that cause urinary tract infection (UTI); (2) preserve urinary continence; and (3) ensure straight erection. He was found to have meatal stenosis that did not allow passage of an 8 Fr cystoscope. After meatoplasty, cystoscopy revealed a normal sized penile urethra and a separate opening in the posterior urethra distal to the verumontanum (Fig. 2). Methylene blue instilled via the cystoscope demonstrated urine egress from the perineal skin pit. Under cystoscopic visualisation, passage of a lacrimal probe through the skin pit and into the posterior urethra confirmed patency of the fistula and aided dissection. This tract was dissected close to its walls and excised. The junction between the penile urethra and the fistula was closed with continuous absorbable sutures, preserving the patency of the orthotopic urethra. The wound was closed in layers and the perineal body was reconstructed. A urethral stent was kept to decrease the risk of urine leak. The decision was made to repair the bifid scrotum in a separate sitting, to prevent a recurrent fistula from infecting the scrotal flaps. Histology confirmed the excised tract to be a complete urethra duplication with transitional epithelium surrounded by smooth muscle and scattered paraurethral glands.

The second operation took place 6 weeks later. The stent was removed, and cystoscopy with methylene blue instillation showed no fistula. The chordee was corrected by degloving the penis. The repair of the bifid scrotum involved preserving a 2cm penile dorsal skin bridge to improve vascular supply and lymphatic drainage. Subcutaneous scrotal flaps were rotated and transposed caudally to form a midline raphe. Circumcision was performed. Six months later, the child was continent and had a single, straight urine stream from the penile meatus (Fig. 3).

Urethral duplication (UD) is a rare congenital malformation, most often encountered in males.[1] It is a heterogeneous spectrum, classified by Effmann et al.[2] into 3 categories:

Type I: blind incomplete UD

- IA: distal end opens on the penile surface, but no communication with urethra or bladder;
- IB: proximal end opens from urethra and ends blindly in the periurethral tissue.

Type II: complete duplication with 2 separate urethras and 1 or 2 separate meatal openings

- IIA: 2 meatuses
  - IIA, 1: 2 non-communicating urethras arise from the bladder;
  - IIA, 2 (Y-type): second urethra arises from the first urethra and courses independently into a separate meatus;
- IIB: 1 meatus, but 2 urethras arise from the bladder or posterior urethra, uniting into a common channel distally.

Type III: as part of a partial or complete caudal duplication.

The complete, communicating UD with 2 meatuses (Y-type) occurs frequently with other abnormalities

Answer: A

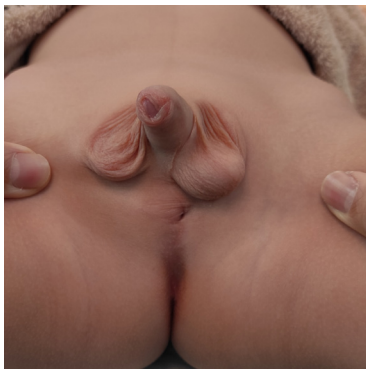

Fig. 1. Perineal skin pit close to the midsagittal plane.

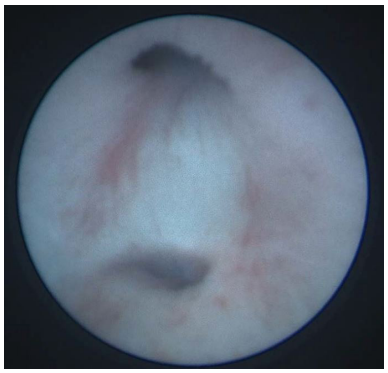

Fig. 2. Cystoscopy revealed a separate opening (at the 6 o'clock position) of the posterior urethra distal to the verumontanum. The orthotopic urethra leading to the bladder neck is 12 o'clock position.

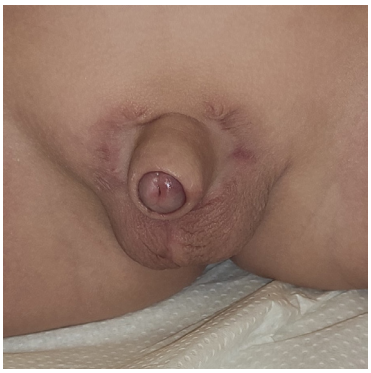

Fig. 3. After surgical repair of the urethral duplication, meatal stenosis, penoscrotal transposition, bifid scrotum, chordee and phimosis, acceptable functional and cosmetic outcome was obtained.

e.g. anorectal malformations;[3] vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities—collectively known as VACTERL association; and cloacal exostrophy.[4] It has 2 urethral channels—1 penile (usually stenotic), and 1 posterior that ends in the perineum or anorectum. These channels result in 2 separate urine streams, which become obstructed if 1 or both of the openings are stenosed. UTI occurs due to stasis and proximity of the ectopic urethral opening to the anus. With repeated infections, there is a risk of metaplasia and squamous cell carcinoma.[5]

Surgical management of a Y-type UD is individualised according to the symptoms, associated abnormalities and anatomical findings.[6] When the bladder outflow is via both urethras and the orthotopic urethra is patent but stenotic, meatal or urethral stenosis is treated with meatoplasty or dilatation respectively. Complete excision of the ectopic urethra removes the risk of urine stasis and infection. When the bladder outflow is mainly via the ectopic urethra and the orthotopic urethra is severely strictured, treatment would be excision of the diseased urethra, and urethroplasty (from the ectopic urethra to the glans) by tubularising flaps or grafts.[7] Urethroplasty is performed commonly with preputial skin in a single procedure or 2 stages depending on length of defect and surgeon's preference. When the ectopic urethra is the main outflow tract and rectally implanted, this is mobilised via the anterior or posterior sagittal transrectal approach, before the urethroplasty.[8] When both urethras are steno-atretic with no bladder outflow, an emergency cystostomy at birth is performed, with subsequent bladder augmentation and urethroplasty.[9]

In our patient, meatal stenosis, phimosis and duplicated urethra (Fig. 4) led to UTI. If the perineal skin pit had merely been excised and the scrotum reconstructed, the duplicated urethra would have been missed. Recurrent UTI and fistula would make late excision, through scarred planes, more difficult. Our case illustrates the importance of recognising the perineal skin pit as sign of a malformation and providing accurate advice to parents and patients.

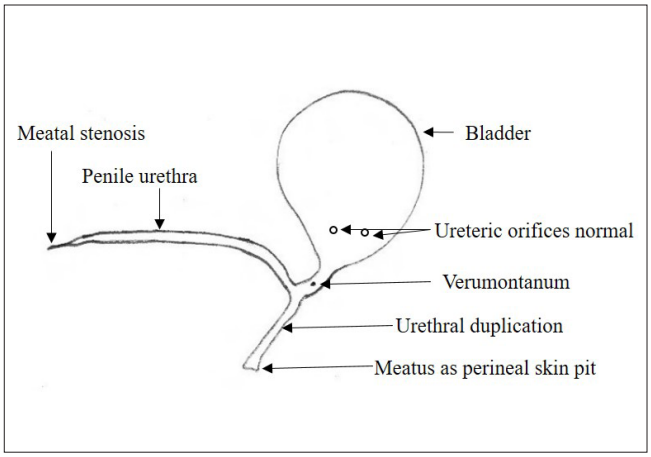

Fig. 4. Diagram of patient's complete urethral duplication with bifurcation distal to the verumontanum and 2 meatuses. The orthotopic penile urethral meatus was stenosed and the ectopic urethra ended as a perineal skin pit.

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