Foreskin and penile problems in childhood

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Abstract

This article outlines the embryology, natural history and management of different conditions of the foreskin and penis in children. Although the classification of hypospadias is included, the management of this condition is not. Epispadias is not covered at all.

Keywords BXO; buried penis; circumcision; congenital mega-prepuce; hypospadias; Phimosis

Embryology and natural history of the penis and foreskin

The embryology of the external genitalia of the male is a complex developmental process. It starts at the 4th gestational week with genital tubercle. At the 8th week of gestation, the external genitalia is in the indifferent phase: a genital tubercle is present as well as paired labioscrotal folds and an endodermal urogenital sinus (Figure 1). The SRY gene on the Y-chromosome leads to differentiation of the gonad into a testis, with production of testosterone by the Leydig cells. This is converted into dihydrotestosterone by the action of 5-alpha reductase present in the external genitalia epithelium. Androgen stimulation causes elongation of the genital tubercle as well as fusion of the urethral folds enclosing the urethral groove moving proximally to distally creating the urethra. On the other hand, mesoderm within the urethral folds gives rise to the corpus spongiosum which fuses with the glans distally as well as the corpora cavernosa. Another peak of testosterone happens during the first 4 weeks after birth with subsequent enlargement of the penis that remains almost the same until further growth at puberty.

The prepuce forms between 13 and 18 weeks' gestation, overlapping with urethral development.[1] The preputial fold moves from the base of the glans distally until fusion with the glans and forms the midline raphae at the midline. Arrest of the final phase of urethral development will leads to 'hooded' foreskin that is deficient ventrally. Subsequent desquamation of the epithelial fusion allows foreskin separation and ultimately retraction. However, non-retractility occurs in 90% of boys at 6 months, 50% at 2 years, 10% at 5 years and 1% at 16 years.[2,3]

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Phimosis

Phimosis (Latin for 'muzzled') is narrowing of the preputial orifice that leads to inability to retract the foreskin. At birth, physiologic phimosis is present as adhesions between the prepuce and glans prevents retracting the foreskin. As the child grows, the two layers begin to separate as sloughed epithelial debris, or smegma, accumulates between them. However, physiological phimosis occurs in 90% of boys at 6 months, 50% at 2 years, 10% at 5 years and 1% at 16 years.[2,3] Clinically this can be recognized by the lack of scarring at the tip of the prepuce and by its mobility. Gentle retraction along the shaft will result in the introitus 'pouting'; if the introitus is gently held and the foreskin pulled distally then the skin will 'funnel' revealing a good sized opening (Figure 2a,b). 'Ballooning' of the foreskin is a normal occurrence during voiding in physiological phimosis. Recognition of normal physiological phimosis is important as the boy and family simply require reassurance and instructions for the boy to gently retract his own foreskin every time he passes urine and every night in the bath or shower, and time for his foreskin to become retractile. The boy needs to be the only person who retracts his foreskin, and generally needs to be over 5 years of age to perform this himself. Conservative treatment of physiological phimosis, particularly in the older child (delayed physiological phimosis), can be augmented by using topical low dose (0.1%) betnovate ointment applied once or twice a day for 3–6 months. Enthusiastic follow-up of this condition in the clinic is unnecessary. Failure to recognize normal physiological phimosis can lead to inappropriate circumcision, with a 2% risk of complications including bleeding, infection or meatal stenosis.[4,5]

Pathological phimosis results predominantly from balanitis xerotica obliterans (BXO), an idiopathic scarring process, or occasionally from forceful retraction of physiological phimosis, recurrent balanitis, or may be associated with incomplete/inadequate circumcision (Figure 2c).

Balanitis xerotica obliterans (BXO)

BXO is an idiopathic scarring condition of the prepuce. If left untreated, it can extend proximally to involve the glans, and distal urethra. Although rarely it can affect children less than 5 years of age, it is the most common cause of foreskin non-retractability in older boys, with an incidence of 1/100–1/200. This can be recognized clinically by a grey/white scar at the introitus of the foreskin, and by its immobility. This scar does not 'pout' or 'funnel' (Figure 2c). In adults this is a pre-malignant condition, but there are no case reports to date of development of penile malignancy complicating childhood BXO. Topical steroid application might be helpful in some cases but the mainstay of treatment is circumcision. There is a significant incidence of meatal stenosis (10%) complicating circumcision for BXO.

Smegma cysts

These result from the normal physiological process of separation of the prepuce from the glans (Figure 2d). The separation has occurred in an area of prepuce that has not connected with the outside yet, and results in a yellow painless lump visible through the foreskin/upper penile skin (Figure 2). With time this enlarges

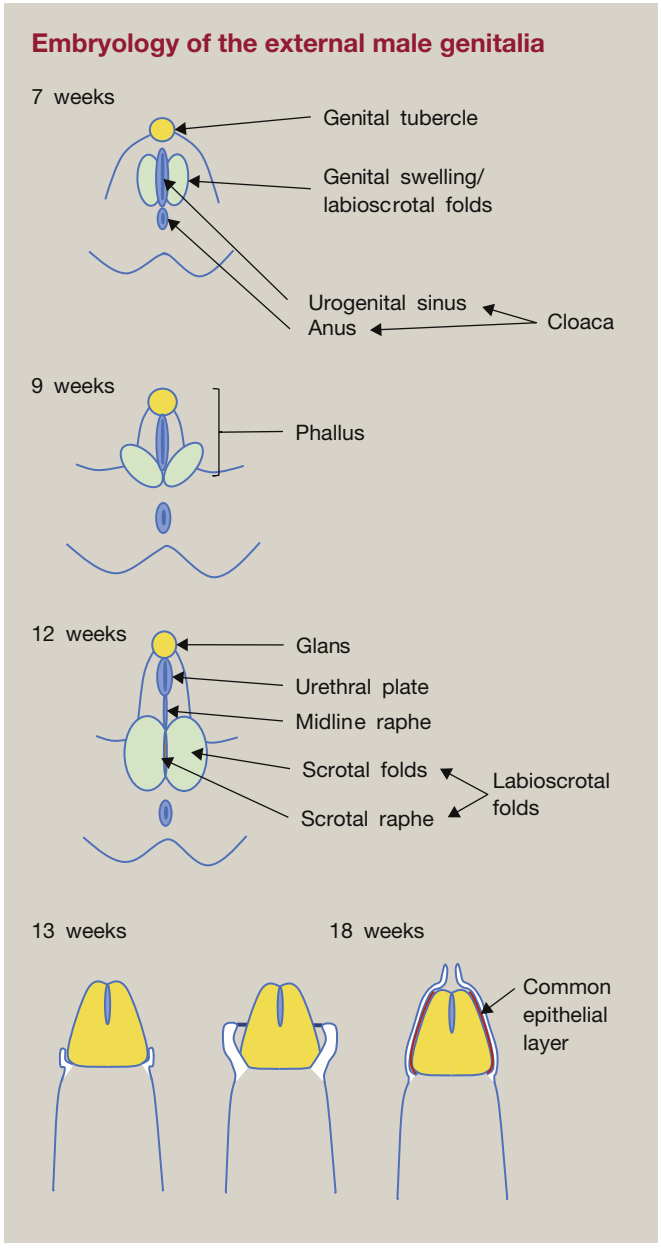

Figure 1

and the separation of preputial adhesions occurs resulting in cyst discharge of the smegma.

Paraphimosis

Paraphimosis occurs when a narrow foreskin has been forcibly retracted over the glans. The retracted foreskin forms a constriction ring around the shaft of the penis causing venous engorgement and painful swelling of the glans and distal penis. Retention of urine might occur as well as ulceration of the glans, and this represents an emergency condition. Reduction of the paraphimosis can often be achieved by gentle compression in a patient who is awake under local anaesthetic (ring block) but occasionally this requires a general anaesthetic. In case of reduction failure, a dorsal foreskin slit should be performed. Paraphimosis is not an absolute indication for circumcision.

Buried penis

Buried penis is a poorly defined term that covers a range of conditions. These include congenital megaprepuce, a concealed, subcutaneous penis associated with a thick suprapubic fat pad and penoscrotal web, it may be acquired as a result of excess weight gain in a prepubertal boy, or as a result of circumcision in a boy with a partially 'buried penis'.

Congenital megaprepuce

This occurs when there is a degree of penoscrotal transposition, a penoscrotal web, in association with a very tight preputial introitus. The dartos layer becomes massively hypertrophied. Obstruction to urine flow occurs, the foreskin becomes massively distended and acts as a 'second bladder' (Figure 3a,b).[6] Operative correction involves a ventral slit, allowing retraction of the prepuce, revealing the massively distended inner preputial layer to be excised following penile de-gloving. Closure of the shaft skin may benefit from ventral Z-plasty (Figure 3e). Reburying of the penis and meatal stenosis are possible complications of reconstruction.

Concealed, subcutaneous penis

This is secondary to thick suprapubic fat pad and penoscrotal web and is often also described as a buried penis (Figure 3f). However, the foreskin in this case does not act as a 'second bladder'. With penile growth, particularly during puberty, and development of a retractile foreskin, this condition is often self-correcting. Simple circumcision is contra-indicated in this condition as acquired buried penis can occur (Figure 3g).

Penoscrotal webbing

This occurs when the normal penoscrotal angle is obscured by a web passing onto the shaft of the penis. When severe, scrotalized tissue may connect to the distal ventral shaft and prepuce. In extreme cases this can give the impression of a buried, or concealed penis by itself.[7] Minor cases may require no treatment, but more severe webbing necessitates treatment, particularly if a circumcision is contemplated for pathological or cultural reasons. This may be performed by simple transverse incision and vertical closure, or by local flaps. The authors preferred method is by Z-plasty.[7,8]

Balanoposthitis 'balanitis'

Balanoposthitis is inflammation of the foreskin and penile shaft, and is almost always secondary to local infection under the foreskin. It may be associated with difficulty in passing urine or even with complete urinary retention, and can complicate physiological phimosis and BXO. Treatment involves culturing any pus present, the use of oral or systemic antibiotics, and simple analgesia. Topical antibiotic treatment can speed resolution. The patient can be discharged home if passing urine, but admitted if having difficulty. Giving the patient a warm bath can often result in spontaneous voiding without the need for urethral catheterization. A single episode of balanoposthitis is not an indication for circumcision in physiological phimosis, but repeated episodes may be.

'Balanitis' is a non-specific term that is often used to describe simple preputial irritation, clinically seen as redness of the

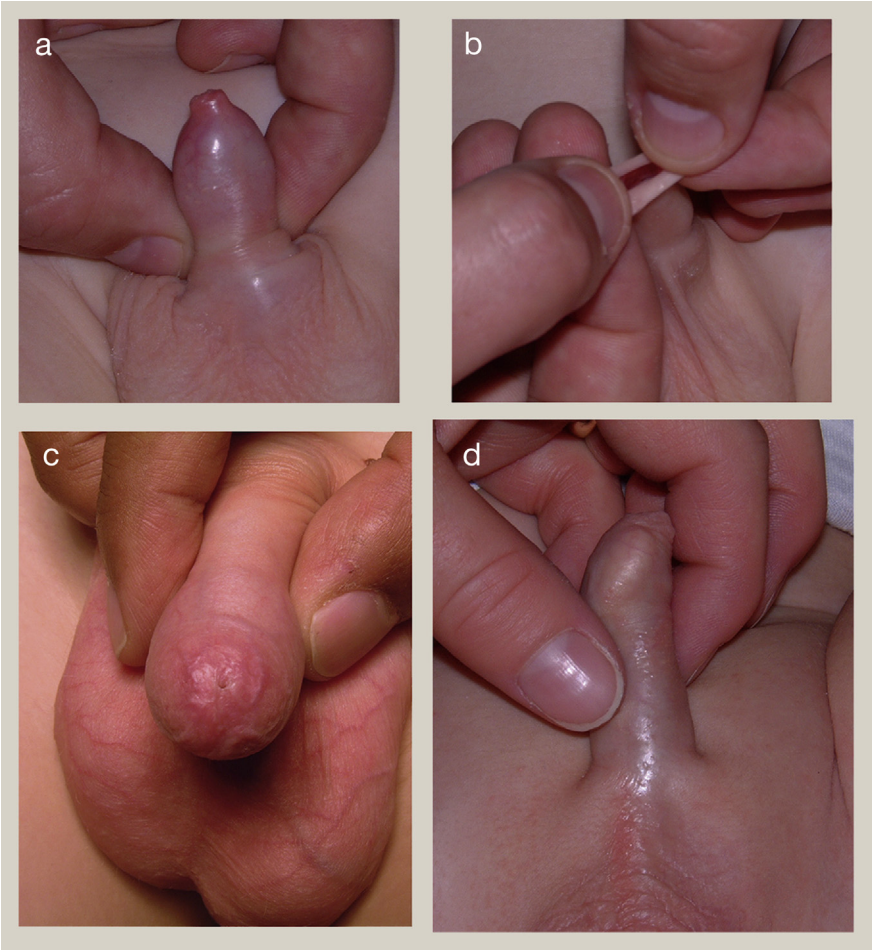

Figure 2 Retraction of normal physiological phimosis will result in pouting (a). whereas pulling the foreskin distally will show funnelling, (b); (**c**) balanitis xerotica obliterans; (**d**) smegma cyst.

foreskin only. This generally resolves with simple attention to hygiene, and resolution of physiological phimosis. Development of foreskin retractility can be encouraged by the boy's gentle retraction of his own foreskin when passing urine and at bath-time. This is not an indication for circumcision.

Circumcision

Circumcision is the operation to remove the foreskin (prepuce) in males. It is one of the most common surgical procedures performed, and one of the few procedures that can legally be performed in the UK without the need for medical qualification. The WHO estimates that globally 30% of males have been circumcised and that two-thirds of these are Muslim.[9] Circumcision can be performed in many different ways. Neonatal religious circumcision in the UK is often performed using a Plastibell. This involves a plastic cone being forced under the foreskin, separating the preputial adhesions all the way back to the coronal sulcus. A tight suture is tied around the foreskin external to this, constricting the blood supply. The distal skin becomes ischaemic and separates from the shaft skin. This technique is not widely used by paediatric urologists in the UK, who tend to perform a surgical circumcision. The foreskin is excised as a 'sleeve', haemostasis normally being achieved using bipolar diathermy. The wound may be closed with dissolvable sutures or tissue glue. Complications following circumcision are not uncommon and include bleeding, infection, meatal stenosis, excess removal of skin, inadequate removal of skin (risk of recurrent phimosis), and misalignment of shaft skin with glans (resulting in torsion or chordee (bending of penis)). Rarely the glans can be damaged, or even urethrocutaneous fistulae created.[10] The complication rate quoted varies widely, but is probably between 2% and 10%.[10]

Contraindications to circumcision in the community include presence of hypospadias (as the prepuce may be needed for surgical reconstruction of the defect) or presence of a buried penis (as the loss of the foreskin may lead to a permanently subcutaneous position of the penis).

Meatal stenosis is one of the most common complications after circumcision. It can also occur in patients with distal hypospadias who have undergone hypospadias repair or as a result of BXO. The pathogenesis is not well understood: it may be meatal ischaemia secondary to ligation of the fraenular artery during circumcision, or it may be ammoniacal dermatitis in the nappy acting on an exposed meatus, or a combination of these processes, or simply the scarring process in BXO. The principle symptoms include penile pain at micturition; a narrow urinary stream; and the need to sit or stand back from the bowl to

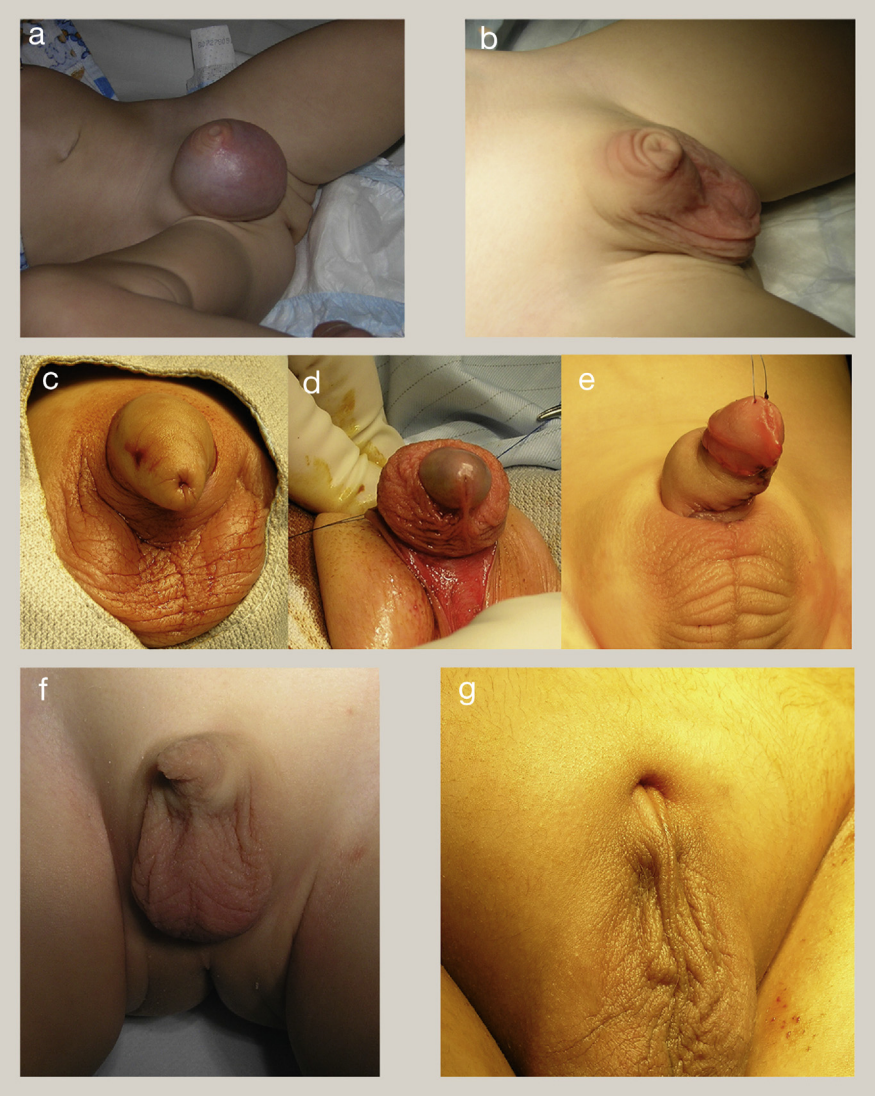

Figure 3 (**a**, **b**) Congenital megaprepuce; (c) decompressed megaprepuce at start of procedure; (d) ventral slit made, and massively extensive rugose inner preputial layer revealed; (e) following excision of excess inner preputial layer, shaft skin excised, and Z-plasty used to close skin ventrally; (f) peno-scrotal webbing, with peno-scrotal transposition, but no preputial obstruction, so no megaprepuce but partially buried penis; (g) circumcision performed in buried penis complicated by scarring trapping penis sub-cutaneously.

urinate, secondary to a deflected stream. Ventral meatotomy can be done under general anaesthesia.

Hypospadias

Hypospadias is defined by the abnormal ventral position of the meatus. Three other abnormalities coexist with the abnormal ventral opening, namely: incomplete foreskin, chordee (ventral bending of shaft), and meatal abnormality (may range from stenosis to megameatus). All four of these aspects can also occur in isolation. There are many classifications of hypospadias that all depend on a description of the position of the meatal opening (Figure 4a). All these preoperative classifications may be confounded by the presence of a hypoplastic urethra (Figure 4b) (which may only be obvious under general anaesthetic), or by chordee (release of this may reveal that the opening is much more proximal) (Figure 4b).

An underlying disorder of sexual differentiation (DSD) may be present – if one undescended testis (UDT) is present, then the risk of abnormal sexual differentiation is 15%, whereas if bilateral UDT is present this risk increases to 30%. Clinical classification should include position of the meatus, presence or absence of chordee, and description of the testes.

The aim of surgery is to create a straight (enough) penis, with a meatus at the tip. This will allow the boy to stand up and pass urine forwards, and when an adult achieve straight erections, be able to ejaculate forwards and so achieve successful coitus. Surgery is generally performed at 1–2 years of age under general anaesthetic. Glanular hypospadias may require no treatment. Distal hypospadias may be reconstructed by parameatal flap (Mathieu hypospadias repair) or by the tubularized incised plate procedure (Snodgrass procedure). More proximal hypospadias, often associated with chordee may be reconstructed by two-stage

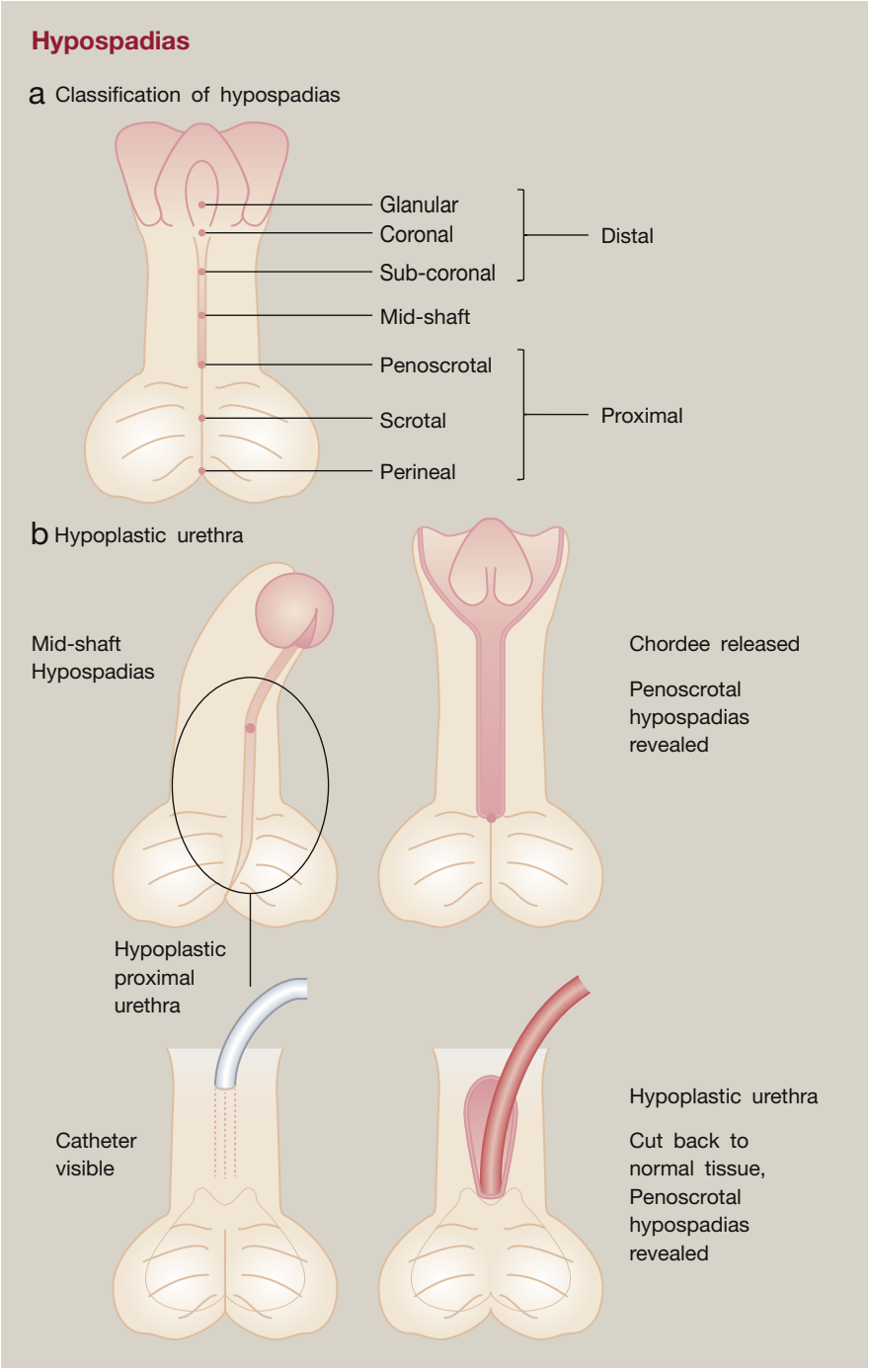

Figure 4

Bracka hypospadias repair. Short-term complications include urethrocutaneous fistula, stricture, wound breakdown. Long-term complications are poorly described, but can include: recurrence of chordee, stricture, resulting in impaired urinary flow, meatal retraction, altered sensation, and chronic pain. ◆

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