

Common Pediatric Penile Conditions

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1. Introduction:

This chapter will introduce common penile anomalies that pediatric urologist will evaluate in their practice. The goal is to introduce the most common conditions seen in the outpatient setting as well as raise attention to common differential diagnosis for the acute scrotum.

Penile Anomalies:

Penile anomalies are common and can be congenital or acquired. For the most part, penile conditions can be categorized as involving the prepuce or the urethra. They can also include problems with penile shape or configuration or other congenital issues such as nevi or cysts. This review will cover penile conditions excluding **hypospadias** and **epispadias**.

2. Conditions Involving the Prepuce:

Most benign penile conditions will involve the prepuce in both circumcised and uncircumcised males.

2.1 Phimosis

Phimosis refers to a condition where the foreskin cannot be retracted to expose the glans and can be categorized as physiologic or pathologic phimosis. Physiologic phimosis occurs in all uncircumcised males and is present in all newborns and tends to resolve as the child matures (**Figure 1**).

Therefore, it is found in 100% of infants but only 5% of children approaching puberty.¹ The events leading to spontaneous resolution of physiologic phimosis are multifactorial. The preputial skin is very responsive to androgen and, therefore, during puberty the skin will soften. In all neonates, the preputial skin is fused to the glans but with time, epithelial cells between these two layers will slough and create a substance known as smegma. This collects between the layers and mechanically

separates these two layers. Lastly as the child grows the penis grows and will have intermittent erections, further adding to the mechanical separation of these layers.

Pathologic phimosis occurs when there is a ring of fibrotic scar tissue that prevents the foreskin from being retracted. This can result from recurrent irritation, repetitive forceful retraction or a condition known as balanitis xerotica obliterans (Bxo). In cases where Bxo is not present and patients are symptomatic, topical application of steroid ointment can be utilized. There have been two randomized controlled trials evaluating topical steroids versus placebo for the treatment of phimosis with reported success rates between 50 and 74% utilizing different strengths of betamethasone with and without hyaluronidase.^{2,3} Our clinical practice is to utilize 0.1% betamethasone ointment three times a day for up to 2 months. In cases where medical therapy is not effective, circumcision, if elected by the parents, is curative.

Another cause of pathologic phimosis as mentioned is Bxo. Bxo is a variant of lichen sclerosus and is an inflammatory condition which should be clinically suspected if there is a white hard ring in the distal preputial skin. Histologically it is characterized by hyperkeratosis and hyperplasia of the squamous mucosa along with inflammation, collagen deposition and loss of rete pegs.⁴ It is a progressive condition and therefore it can involve the meatus and urethra potentially causing meatal stenosis or urethral strictures.⁵ There has been one trial reporting that phimosis secondary to Bxo is responsive to topical steroid therapy. Kiss et al performed a randomized controlled trial comparing 0.05% mometasone furoate vs. placebo in 40 boys with Bxo. After once daily application for 5 weeks the authors reported “improvement” of phimosis in the treatment group.⁶ Another study reported that tacrolimus ointment (0.1%) applied twice daily for 3 weeks may provide improvement in Bxo secondary phimosis or meatal involvement.⁷ If medical therapy is unsuccessful or if the parents prefer, circumcision can be curative. Boys with Bxo where meatal or urethral involvement is suspected need clinical follow-up.



Figure 1: Physiologic phimosis

2.2 Secondary Phimosis or “Trapped Penis”

Secondary phimosis occurs when a child has been circumcised and the subcoronal incision creates a cicatrix that “traps” the penis. This condition has also been shown to be amenable to topical steroid therapy with a response in up to 64% of patients.⁸ Refractory cases can be treated surgically by circumcision revision.

2.3 Paraphimosis

Paraphimosis is defined by inflammation and edema of the distal penis and glans caused by constriction below the glans. This occurs when the prepuce is retracted to expose the glans but not replaced back over the glans (**Figure 2a**). Venous congestion creates more swelling. Paraphimosis is considered a medical emergency. In most cases the paraphimosis can be manually reduced (**Figure 2b**). Maneuvers to assist in this include a penile block for analgesia and oral or parenteral

pain medication. Compression of the edematous foreskin may also help in reduction. Other methods that have been described include wrapping the penis in a gauze soaked with hyper-osmolar solution and making holes in the edematous foreskin with a needle to allow the tissues to drain. If conservative maneuvers are not successful a dorsal slit or circumcision may be required. These patients usually present with significant pain or urinary retention. Although described, glans necrosis secondary to paraphimosis is rare.⁹



Figure 2a: Retraction of the phimotic band can cause constriction of the penis and cause distal edema.



Figure 2b: Treatment includes compression of the edema and readvancement of the foreskin over the glans.

2.4 Webbed penis

Penoscrotal webbing or a webbed penis occurs when there is poor or no separation of penile skin from scrotum. This creates a web of tissue and eliminates the appearance of a penoscrotal angle. This is most often congenital but can be iatrogenic during a circumcision if too much ventral skin is removed. The true incidence of webbed penis is not known but there are various approaches for surgical correction.¹⁰

2.5 Congenital Megaprepuce

Congenital megaprepuce occurs when there is a large surface area to the inner preputial lining. It is unclear if this occurs because of penile stretching during urination in a patient with phimosis or if this is congenital¹¹ (**Figure 3**). Penises with this condition often appear short and there is often significant pooling of urine in the inner foreskin. A surgical circumcision can be performed with possible need to excise the redundant megaprepuce along with repairing buried penis appearance.



Figure 3a: Concealed penis with phimosis.



Figure 3b: Retraction of foreskin reveals congenital megaprepuce.

2.6 Balanoposthitis

Balanoposthitis refers to inflammation of the glans (balanitis) and the foreskin (posthitis). Patients will usually present with an edematous, erythematous prepuce with or without discharge from the preputial opening. It is a relatively common condition, with a reported incidence of 6% in uncircumcised boys.¹² The etiology is often unclear but can include bacterial or candida infection, mechanical trauma, contact irritation, and contact allergies. Treatment is often with topical anti-inflammatory cream with topical antifungal therapy. Oral antibiotics are seldom necessary unless there is a concomitant urinary infection. Failure to resolve should prompt further investigation as this can be a sign of other underlying conditions such as sexually transmitted diseases and other etiologies.¹³

3. Structural and Anatomic Conditions:

3.1 Penile Agenesis

Aphallia (penile agenesis, absence of the penis) is an extremely rare and complex congenital penile malformation with a reported incidence of 1 in 10 million male newborns and only 100 cases reported globally in the literature.¹⁴ During early development, paired genital swellings on either side of the cloacal membrane merge to form a single tubercle which gives rise to the penis. Failure of this mesenchymal outgrowth results in this rare phenomenon.

The hallmark of this anomaly is a visually absent penis sometimes with a remnant midline appendage. The remnant tissue may contain the urethral meatus although meatal positioning may be presphincteric, postsphincteric or there may be urethral atresia. The location of the urethral meatus in relation to the anal sphincter has been used to classify the severity of the disease, a more proximal meatus correlating to more severe comorbid anomalies thus, higher mortality. A majority of patients with aphallia will have concurrent genitourinary anomalies such as cryptorchidism, renal agenesis, renal dysplasia, and imperforate anus, among others.

3.2 Penile Curvature: Torsion and Chordee

Curvature anomalies are relatively common and can be lateral, dorsal, ventral or rotational. Although most commonly associated with hypospadias, these curvature anomalies do occur in isolation and are estimated to affect 4-10% of all males.¹⁵

Penile torsion is a malformation of unclear etiology in which the corporal bodies are torsed or sometimes it is just the glans that is rotated. This rotation almost always occurs counterclockwise and can be associated with other abnormalities of the penis, scrotum or urethra. Isolated penile torsion is estimated to occur in 2–27% of males, with rotation of greater than 90 degrees reported in only 0.7%.¹⁶ Most children with this condition do not have symptoms and, therefore, surgical correction is mostly cosmetic although extreme torsion can interfere with micturition. The vast majority of adults with penile torsion do not report any sexual dysfunction.¹⁷

The term chordee is usually used to refer to ventral penile curvature which is most commonly seen

with hypospadias. There are multiple hypotheses as to why this occurs including a short urethra, corporal disproportion and abnormal ventral tissue.¹⁸ In a series of 87 patients with different types of curvature, Donnahoo in fact confirmed that these were the primary etiologies of curvature although a congenitally short urethra occurred in only 7% of patients. In another paper however, the authors found no evidence of fibrous bands or dysplastic tissue in the urethral plates of boys with hypospadias.¹⁹ There are various different techniques to correct chordee usually involving either plication of the longer side of the corporal bodies or expansion of the curved side with incisions, tissue inserts, or both.

3.3 Buried penis

The term “buried penis” is misleading because it could include secondary phimosis, congenital megaprepucce, concern about excess foreskin after circumcision, and other congenital conditions (**Figure 3**). In most cases, however, the terms hidden, buried or concealed penis are used to refer to the covering of the penis with a large suprapubic fat pad. This condition resolves when infants grow and the fat pad disappears, so in these cases there is no indication for surgical repair.

3.4 Penile Duplication

Penile duplication or diphallia is an extremely rare condition occurring in approximately one in every 5.5 million live births.²⁰ It is usually subdivided into true diphallia and a bifid phallus and then further sub-categorized into partial or complete duplication.²¹ True complete diphallia is defined by complete penile duplication, each with two corpora cavernosa and one corpus spongiosum. If there is only one corpus cavernosum in each penis, the term bifid phallus is used. True diphallia is the less common and usually presents with a wider range of associated malformations such as bladder and urethral duplication, exstrophy vesica, renal anomalies, bifid scrotum, anorectal malformations, bowel duplication, and vertebral anomalies. Bifid phallus is usually associated with less severe malformations.²² There is proposal for a new classification system based on the clinical and surgical implications for each group:²³ true penile duplication (each with 2 corpora and spongiosum), hemiphalluses (2 penises with 1 corpora each), pseudoduplication (normal penis with accessory non-functional penis) and partial duplication (duplication involving only the distal penis in the sagittal or coronal plane; glans duplication; **Figure 4a** and **Figure 4b**).



Figure 4a: Glans duplication, coronal plane.



Figure 4b: Glans duplication, coronal plane.

3.5 Penile Lymphedema

Penile lymphedema can be congenital or acquired and may be self-limited or permanent (**Figure 5**). Treatment of this disease is dependent on its etiology since the cause of lymphedema usually determines the natural history of the disorder. A good classification and review of the etiologies is provided by Dr. McDougal in his 2003 review of the topic.²⁴ In children, lymphedema is usually transient and secondary to irritation. Insect bites or external trauma are the most common etiologies for penile lymphedema in children and these will spontaneously resolve. Care must be taken, however, not to miss rarer causes, including Crohn's disease and trauma.



Figure 5: Idiopathic penile edema.

4. Postcircumcision conditions:

4.1 Penile adhesions

Attachments of the foreskin back to the glans after circumcision.

Penile skin bridges are dense scar adhesions that cannot be easily separated and require incision to divide.

Physiologic adhesions: The prepuce has adhered down to the glans after circumcision. These adhesions are physiologic and the natural history is resolution over time similar to physiologic phimosis.

4.2 Epidermal inclusion cysts

Epidermal inclusion cysts usually occur under two circumstances. First would be congenital rests of

skin buried during development most commonly along the median raphe. More commonly they occur when epithelial tissue is trapped during a circumcision or other surgical procedure. They can occur anywhere along the raphe and subcoronal margin and presents as a small, enlarging white lesions which grow subcutaneously. Depending on the size, number, and symptoms, these can be treated with topical steroids to thin the epithelial layer and allow for the lower trapped epithelium to extrude. It could also be drained with a small incision after topical anesthetic is applied or a surgical excision can be performed.

4.3 Meatal stenosis

Meatal stenosis is a common condition almost exclusively occurring in circumcised males. The preputial skin normally covers the meatus. In circumcised children, the meatus is exposed to urine and feces and may also rub on the diaper and clothing. There have been 2 posited hypotheses for why meatal stenosis occurs. The 1st involves repetitive irritation of the meatus secondary to the factors mentioned above. The 2nd is the possibility that a circumcision, by cutting the frenular artery may cause ischemia to the meatus. The secondary hypothesis has been recently questioned given that the frenular artery usually bleeds away from the meatus not towards the meatus. Although the etiology is not completely clear, the fact remains that meatal stenosis is the most common long term complication after circumcision.²⁵ Symptoms of meatal stenosis usually involve a narrow stream, having to strain to void, prolonged voiding and, perhaps most commonly, an upward deviation of the urinary stream.²⁶ In the vast majority of children a meatotomy can be performed as an office procedure after topical anesthetic application. The meatal web is simply crushed with a hemostat and then divided. A meatoplasty with excision of ventral webbing and securing urethral mucosa to glans mucosa with small absorbable sutures can be in the operating room as well.

5. Conditions Involving the urethra

There are multiple conditions that can involve the urethra. These range from urethral agenesis to urethral diverticuli, congenital anterior urethro cutaneous fistula, megalourethra and others.²⁷ Management depends on the condition, the symptoms and the presence or absence of urinary obstruction.

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