Anomalies of the penis

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Abstract

Anomalies of the penis occur with regularity and cause much anxiety for parents of affected children. If left untreated, these problems may not only result in long-term physical sequelae, but also in poor psychosocial adjustment. This article describes the more commonly seen anomalies of the penis and their evaluation and treatment.

Introduction

Anomalies of the penis may be congenital or acquired. The physical findings may be obvious, but sometimes quite subtle. This can be challenging for the health care provider and it is important that they understand, diagnose, and treat these conditions.

The purpose of this article is to provide a concise overview of the more commonly seen penile anomalies in the pediatric population. The goal is to provide the primary care physician, urologist, and other specialists with the essential information to accurately evaluate and treat these conditions. The penile conditions discussed are organized into three sections: prepuce, meatus, and penile shaft.

Physical examination

The physical examination is essential in recognizing penile anomalies. One needs to know the normal anatomy in order to recognise the abnormal conditions. The normal meatus is vertical with a slit-like appearance at the tip of the glans. The normal penis should be straight without deviation of the glans or shaft. When defining the ventral and dorsal aspects of the penis, it is based on the erect penis. Therefore, the ventrum is the side closest to the scrotum and the dorsum is closest to the pubic region. Finally, the length of the penis is usually based on its stretched length. This is determined by gently pulling on the head of the penis while compressing the suprapubic fat pad. The distance from the pubis to the tip of the penis is the accurate determination of the stretched length. The range of normal penile length is based on the age of the child.

Prepuce

Balanoposthitis

Balanitis is an inflammation of the superficial layers of the glans penis while posthitis is an inflammation of the prepuce. However, the two conditions may coexist (balanoposthitis). Balanoposthitis results from infection or from irritation due to external irritants [1]. It occurs most frequently in children aged 2 to 5 years and is usually the result of inadequate hygiene. Symptoms may include redness, exudates, irritation, and swelling of the glans and inner prepuce. It is often mistaken for sexually-transmitted diseases, even in young children [2]. However, a sexually-transmitted chlamydial or gonococcal infection is associated with a urethral discharge in most children. The presence of a thin purulent exudate within the preputial–glanular sulcus in the absence of an urethral discharge may signify a streptococcal infection, which may occur even in the absence of a sore throat [3]. While balanoposthitis is a diagnosis based largely on history and physical examination, a rapid strep test and culture of secretions is helpful in determining a streptococcal origin. Balanoposthitis may improve with simple cleansing and application of either 0.5% hydrocortisone cream or antibiotic ointment 2 times per day. However, recalcitrant or known streptococcal disease should be treated with oral penicillin. For recurrent cases, circumcision should be considered.

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Phimosis

Although the majority of American boys are circumcised, 80% of the world does not practice routine circumcision [4]. Thus, phimosis, a non-retractile foreskin resulting from a tight distal preputial ring, is a commonly encountered problem. In neonates, there is a natural adhesion between the inner foreskin and the glans. The foreskin cannot be retracted without disruption of these natural adhesions; this physiologic phimosis is often confused with a true phimosis. As many as 96% of male newborns demonstrate physiologic phimosis, but up to 90% will have resolved by 3 years of age [5]. By this age, the formation of keratinized pearls of glanular secretions and epithelial debris along with intermittent erections causes the foreskin to detach from the glans [6]. Forceful disruption is not recommended due to associated pain and high rates of recurrence.

Balanitis xerotica obliterans (BXO) is a chronic dermatitis most often involving the prepuce, glans, meatus, and urethra. The exact etiology is unknown. The appearance on the foreskin is typically of a circumferential white plaque that may make it difficult to retract the foreskin. Corticosteroids have been shown to be effective for treating BXO, but surgical intervention may be necessary, including circumcision [7,8].

There are non-surgical and surgical options for treating phimosis. Several reports using various topical steroidal agents with variable regimens for treating phimosis have been reported. These have proven to be an effective alternative to surgery with reported success rates of between 67% and 95% [9–16]. Recently Palmer & Palmer directly compared the outcome of two different regimens of betamethasone using a grading system for phimosis with an over 80% success rate [17].

Surgically, there are several options available. Circumcision is the traditional surgical option, which is typically a safe, straightforward procedure. However, there are several associated complications that may occur, including infection, hemorrhage, amputation and urethro-cutaneous fistulas. The trapped penis is another potential complication that has traditionally been managed surgically with either incision of the cicatrix or formal surgical reconstruction. A trapped penis occurs when a tight phimotic ring (cicatricial scarring) forms after circumcision. Palmer et al. recently reported on successful management of this condition with topical betamethasone cream and manual traction [18].

Alternative surgical options are preputioplasty techniques to facilitate retraction of the prepuce. However, these are surgical procedures that require general anesthesia and suturing, thus associated with risks similar to those associated with circumcision. In addition, there are techniques described to lengthen residual skin with the intent of reforming the prepuce.

Paraphimosis

Paraphimosis occurs when the foreskin is retracted proximal to the glans penis and not replaced back over the tip of the penis. The fibrous ring of foreskin may act like a tourniquet, resulting in venous congestion, edema of the glans and foreskin, and may possibly lead to ischemia and necrosis if left untreated. This condition is frequently mistaken for balanitis due to the pain and swelling. However, a thorough history and knowledge of the child's circumcision status should yield a correct diagnosis.

Correction of paraphimosis must be performed immediately by placing the foreskin into its normal anatomical position. This can be done at the bedside with or without a penile block or in the operating room with intravenous sedation or general anesthesia. Reduction is performed first by gentle, prolonged manual pressure around the distal penis including the glans and prepuce, which will temporarily relieve the penis of edema. Then the prepuce is grasped and pulled forward while simultaneously the glans is pushed back through the paraphimotic ring. This may be difficult and, rarely, a dorsal slit of the foreskin or formal circumcision is required to resolve paraphimosis. For recurrent cases, circumcision should be considered.

Meatus

Meatal stenosis

Meatal stenosis is defined as a narrowing of the urethral meatus. This is an acquired disorder affecting circumcised boys resulting either from meatal inflammation secondary to pro-

longed exposure of the urethral meatus to a moist diaper or other irritants, or from ischemic injury after division of the frenular artery at the time of circumcision [19]. Other causes include urethral instrumentation, trauma, or BXO. Meatal stenosis secondary to BXO demonstrates a characteristic whitish discoloration of the glans.

The diagnosis of meatal stenosis is typically made subjectively based on clinical experience. In hopes of providing a more objective measure, a study by Litvak et al. demonstrated that a boy aged less than 10 years should have a meatus of at least 8-French and boys of 11–12 years should accommodate at least 10-French [20]. Generally, a stenosed meatus does not obstruct urinary flow, but patients often demonstrate a fine caliber, narrow, forceful stream with a characteristic upward deviation. Standard treatment consists of meatotomy because simple urethral dilation results in high rates of recurrence.

Hypospadias

Hypospadias is an abnormal *ventral* opening of the urethral meatus and is frequently associated with ventral penile curvature and/or a hooded foreskin. The opening can be located anywhere from the glans to the perineum. It is a common condition with an incidence of approximately 1 in 300 newborn males. There is a familial pattern in that 6% to 8% of fathers of affected boys and 14% of male siblings demonstrate hypospadias [21]. It is usually an isolated phenomenon, but is frequently seen with cryptorchidism in 9% of patients and this rate increases with the severity of hypospadias [22,23]. Those children with hypospadias and a non-palpable testicle warrant genetic evaluation for they may genotypically be female. Also, the incidence of associated upper tract anomalies is no different than that of the general population and thus routine evaluation is unnecessary in those patients with isolated middle or distal hypospadias [22,24,25]. Screening with renal ultrasonography is typically done in cases of severe proximal hypospadias.

Once hypospadias is recognized or if one is unsure, circumcision should not be performed, but rather it should be evaluated by a surgical specialist. There are multiple corrective techniques depending on the severity of hypospadias and quality of tissue available. The surgical goal is to make a straight penis with a normal meatus. When considering anesthetic and psychosocial outcomes, an optimal time for repair is between 6 to 18 months of age. In long-term post-surgical follow-up, several studies have produced conflicting results regarding the quality of adult sexual functioning [26–28]. In a study by Mureau et al. the majority of adult patients who underwent childhood hypospadias repair tended to be more timid in initiating sexual contacts, but they experienced overall normal sexual functioning compared to the control group [29].

Epispadias

An epispadias is an abnormal urethral meatus located on the *dorsum* of the penis, anywhere from the glans to the penopubic junction. Unlike hypospadias, this relatively uncommon penile condition is associated with dorsal curvature, a ventral hood of foreskin, and splaying of the corporal bodies. It involves an absence of the dorsal aspect of the urethra and overlying skin. Epispadias may occur alone or in conjunction with bladder exstrophy. In proximal cases, the prostate and bladder neck may be inappropriately formed. Surgical goals of therapy are to return the urethra to a proper anatomical location, correct any curvature, and maintain urinary continence.

Penile shaft

Chordee

Chordee, or penile curvature, may be ventral (most common), dorsal, or lateral. The condition may occur independently or in association with various other congenital penile anomalies (hypospadias or epispadias). When associated with hypospadias, the cause of chordee is unclear and may be a part of early normal fetal development that has been arrested [5,21]. In the absence of hypospadias, chordee is postulated to result from either skin tethering, fibrotic superficial and deep penile fascia, or disproportion of the corpora [30]. When it occurs as a solitary anomaly, it is often unrecognized until curved erections are noticed in adolescence.

If a significant chordee is suspected, regardless of any associated anomalies, neonatal cir-

cumcision should be avoided until a trained surgeon is consulted. A simple degloving procedure under general anesthetic, whereby fibrotic tissue is excised, usually assists in resolving this condition. However, if the chordee persists, a corporeal plication procedure and/or grafting procedures may be necessary.

Penile adhesions and skin bridges

Penile adhesions are a common complication of circumcision. They form as filmy attachments of the distal penile skin to the glans penis. Atalla & Taweela determined that these adhesions occur when the denuded glans is in close contact with raw areas of penile skin or remnant preputial tissue [31]. Glanular adhesions separate and resolve over time as epithelial debris accumulates beneath the adhesions and due to mechanical stretching from normal erections [32]. Less commonly, adhesions may further epithelialize and form a skin bridge that must be treated with excision, either with a local anesthetic in the consulting room or a general anesthetic in the operating room. Following either spontaneous breakdown or surgical excision, reattachment of the adhesion is prevented by strict adherence to a daily manual-retraction routine and/or by lubrication with an ointment for several weeks.

Penile cysts

Penile cysts may be congenital or acquired. The congenital form may occur at any point along the median raphe, extending from the urethral meatus to the anus [33]. These cysts are usually asymptomatic unless secondarily infected. If patients are bothered by the cosmetic appearance, surgical excision is the treatment of choice.

Acquired cysts are either due to the accumulation of smegma entrapped under unretracted prepuce (most common) or an epidermal inclusion cyst. The latter is a result of islands of epithelium left in the subcutaneous tissue as a result of surgery. They are usually asymptomatic. Surgical excision including the capsule surrounding the inclusion cyst should resolve this condition.

Concealed penis

A concealed penis (hidden penis, buried penis) is a normally developed penis hidden within the suprapubic fat. Patients usually present as neonates or as obese prepubertal males. Factors contributing to a concealed penis include excess suprapubic fat, inadequate attachment of the penile skin to the fascial layers at the base of the penis, scrotal webbing, abnormal tethering of the penis to the underlying dartos fascia, or a combination of these conditions. Secondary concealment, or a trapped penis, occurs after circumcision (see in *Phimosis*, above) or when too much penile shaft skin is removed during circumcision. In a child with concealment who subsequently undergoes neonatal circumcision, a concentric scar forms distal to the glans that will essentially trap the penis [34]. Many children with concealed penis undergo neonatal circumcision prior to repair, which makes corrective surgery more difficult [35,36]. Consequences of concealed penis include embarrassment among peers, balanitis, difficult hygiene, urinary tract infections, and inability to void with a directed stream, which may interfere with toilet training.

In the obese child, conservative therapy consists of weight loss through dietary restriction and vigorous exercise. However, early surgical correction is advocated for treating those patients with surgically correctable conditions. Several surgical procedures have been described, and the correct approach is based on the causative factor and severity of concealment [37]. Surgical intervention results in voiding with a directed stream and improved hygiene [38]. The majority of patients who undergo corrective surgery are satisfied with the results, however, the parents of infants and toddlers who undergo surgical repair report a higher degree of satisfaction than those with older children [39].

Microphallus

Microphallus (micropenis) is defined as an abnormally small, but otherwise anatomically normal penis [40]. This is distinguished from a concealed penis wherein the phallus is of normal size, but buried within the prepubic fat. Microphallus is defined as a stretched penile length less than 2.5 standard deviations below the mean, or less than 1.9 cm at birth [41]. Penile growth is dependent upon both testosterone and growth hormone, and abnormalities relating to either hormone may result in micropenis. Approximately half of these

patients have hypogonadotropic hypogonadism, 25% have hypergonadotropic hypogonadism, and 15% have end-organ androgen insensitivity [26,42]. Children with micropenis secondary to growth hormone deficiency have a normal-sized phallus at birth due to the presence of maternal growth hormone, however, micropenis and short stature will develop with age. In contrast, patients with growth hormone resistance demonstrate both microphallus and short stature at birth.

Treatment for microphallus is predicated by the cause. Thus, a thorough work-up is necessary, including a detailed history and physical examination with attention to abnormalities that raise the suspicion of a genetic syndrome or other co-existing urogenital anomalies such as cryptorchidism or hypospadias. A standard laboratory examination includes a genetic karyotype, gonadotropin level, and testosterone level. Other pituitary hormone levels also require investigation as deficiencies tend to occur together [40]. If the laboratory examination reveals an end-organ cause for microphallus and the testes are not palpable, diagnostic laparoscopy is indicated. In cases of hypogonadotropic hypogonadism, a renal ultrasound is indicated because of a known association with unilateral renal agenesis.

Treatment of microphallus involves replacement of the underlying deficiency. Most patients should be given a trial of testosterone intramuscular injection therapy at 4-week intervals until adequate penile length is achieved. Some regression of penile size should be anticipated in the years following treatment, and another course of treatment may be indicated later in childhood [43]. Although studies have yielded conflicting long-term results, treatment for patients with fetal androgen deficiency typically results in a normal-sized adult phallus with normal sexual function [44]. Furthermore, treatment with growth hormone yields a normal phallus size in the majority of patients with isolated growth hormone deficiency [42]. In cases of failed treatment, or in cases of androgen resistance, gender reassignment is often indicated.

Conclusion

Disorders of the penis are frequently encountered in the primary care setting. When recognized and treated properly, physical well-being is preserved and long-term psychosocial and sexual outcomes are improved significantly.

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