Multi-stage repair surgery in combination with hormonal therapy for scrotal hypospadias with severe chordee, micropenis, and bifid scrotum

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Abstract

Proximal hypospadias includes proximal penile, penoscrotal, scrotal, and perineal types, which are a less frequent occurrence and correspond to 20% of total hypospadias. Surgical reconstruction is the only possible therapeutic option for hypospadias. The primary objectives of the reconstruction are to create a vertically slit orthotopic meatus, straighten the penis and establish good cosmetic results that include a conically shaped glans. Other essential aspects of the reconstruction are to avoid shortening the penis and optimal skin coverage. This study reports a case with abnormal spraying during urination. Severe forms of hypospadias are typically accompanied by an abnormal ventral curvature of the penis (chordee). Surgical correction of both chordee and hypospadias is recommended. When adequately corrected, it does not cause long-term problems, and a natural appearance of the penis is usually restored.

Keywords: *Scrotal hypospadias, severe chordee, micropenis, bifid scrotum, cordectomy orthoplasty, urethroplasty, scrotoplasty*

Introduction

Hypospadias is a congenital anomaly, which affects different structures of the penis and, eventually, the scrotum and perineum. A wide range of types of hypospadias can be observed according to the original site of the opening of the urethral meatus and to other associated deformities. They are found in varying configurations that range from an aspect of ambiguous genitalia to a well-formed penis with a superficial defect.

Some penile anomalies, often associated with hypospadias, are related to cosmetic and functional aspects of the penis. When in erection, it is considered a significant ventral curvature of the penis (chordee), a deflection angle of the shaft of the penis more significant than 20 degrees. In proximal hypospadias, the ventral curvature is often caused by fibrous chordee. Chordee is formed by residual fibrous tissue of the corpus spongiosum and is located on the ventral urethra, distal to the urethral meatus, in close contact with the tunica albuginea. Other significant causes of the ventral curvature began to be better defined with the introduction of the artificial erection technique.¹ The cutaneous chordee arising out of the asymmetric distribution of the skin around the penis can originate some degree of curvature both in distal and in proximal hypospadias. Also, there can be an asymmetry of the tunica albuginea, which accounts for the permanence of the curvature despite the excision of the entire fibrous chordee.² Another uncommon cause of penile curvature is the growth differential between the corpora cavernosa and the corpus spongiosum.

In proximal hypospadias, the prepuce is asymmetric, accumulating on the dorsum of the penis and being deficient in the ventral segment. The prepuce may be normal in distal hypospadias

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in up to 7% of cases. Urethral meatus stenosis is rarely found in proximal hypospadias, where it is observed in about 15% of cases. Axial kicking of the shaft of the penis occurs in 14% of patients and is not dependent on the degree of hypospadias.²

Proximal hypospadias is a less common occurrence and corresponds to 20% of total hypospadias. Proximal hypospadias is usually associated with scrotal malformations, such as penoscrotal synechia, hypoplasia, bifid scrotum, and high scrotum implantation.²

The most commonly used classification of hypospadias is Barcat's, and it is based on the location of the urethral meatus after the correction of the associated curvature of the penis. Proximal hypospadias includes proximal penile, penoscrotal, scrotal and perineal types in which the site of the urethral meatus is respectively the proximal third of the penis, the root of penis, scrotum or between the genital swellings and below the genital swellings.²

Most hypospadias occurs as an isolated condition, but associated anomalies include uni-bilateral cryptorchidism and micropenis.² Microphallus, or micropenis, is defined as a stretched penile length of less than 2.5 standard deviations (SDs) below the mean for age. Traditionally, the term micropenis refers to a penis that is otherwise typically formed, and the term microphallus has been used when associated hypospadias is present. The mean stretched penile length in a full-term newborn male is 3.5 cm. Measurements of less than 2-2.5 cm (2.5 SDs below the mean) in a full-term newborn male meet the definition of micropenis and warrant evaluation.³

Proximal hypospadias cause micturition problems besides limiting sexual intercourse and fertility, and require correction. Moreover, nontreated hypospadias put the patients at risk emotionally, regarding the acceptance of their body image, through the transference of parents' anxiety or the acknowledgment of the condition by friends or a sexual partner, which leads to embarrassing situations.⁴

Significant advances have been made in surgery for the correction of hypospadias. Proximal hypospadias remains the most significant challenge, but, despite many innovations and much progress, surgery can fail. Many authorities have introduced single-stage techniques, which have the purported advantage of correcting the defect with minimal hospitalization and family inconvenience. However, the more extensive published experience with these approaches would suggest that the ideal single-stage procedure has yet to be devised. A substantial number of children undergoing a single-stage procedure will have to undergo further surgery.⁵

Case Report

A 9 years old male presented with abnormal spraying during urination. A patient complained about a downward urinary spray since birth. Parents said that the opening of his urethra is located under the penis rather than at the tip. Every time he urinates, the urine flowing along the thighs, so the patient has to sit down to urinate.

In physical examination, we found the opening of the urethra is in the scrotum, a downward curve of the penis. The penis was uncir-

cumcision and in a small size. The scrotum was bifid in the normal position, and bilateral testes were fully descended in the well-developed scrotum. No additional urogenital and anorectal malformations were detected by physical examination. The anus was normal in position and caliber.

Fig 1. The clinical picture of the penis and scrotum.

The patient was the product of a section caesarian delivery at 38 weeks of gestation with an uneventful prenatal period, and his birth weight was 3500 grams and length 50 cm (proportional for gestational age). He was the product of healthy non-consanguineous parents. Neither notable family history nor exposures to teratogens, alcohol, or drugs were noted. During pregnancy, his mother did not have routine follow-up examinations

Based on the history and clinical findings, the patients were diagnosed with scrotal hypospadias with severe chordee, micropenis, and bifid scrotum. The decision then made to perform multi-stage repair. The first stage of the procedure is cordectomy-orthoplasty for penile straightening. Six months later, we will do urethroplasty as the second stage of the procedure to relocate urethral meatus. For the third stage of the procedure, we will do scrotoplasty for cosmetic needs. Micropenis will be treated with hormonal therapy.

Fig 2. Post surgery presentation

During the first stage of the procedure, cordectomy-orthoplasty was successfully been performed without any complication during the procedures. Follow up 21 days after cordectomy-orthoplasty showing a straight penis. The patient can urinate with the stand-up position because the urine no longer flowing along the thighs.

Fig 3. The result after 21 days post operation

Discussion

With an incidence of 1:300, hypospadias is one of the most common genital anomalies in male newborns.⁶ Hypospadias is defined as an anomaly (hypo- or dysplasia) involving the ventral aspect of the penis.⁷ These malformations mainly comprise an abnormal ventral opening of the urethral meatus, an abnormal ventral curvature of the penis (chordee), and/or abnormal distribution of the foreskin. The extent of the malformation varies.

Such ectopic urethral openings (meatus) can be located at the tip of the glans penis (hypospadias sine hypospadias), glandular, coronal, subcoronal, along the penile shaft, penoscrotal, scrotal, or perineal. The form and extent of malformation of the urethral opening vary as well and is in some cases, widely gaping and resembling the mouth of a fish. Stenosis is rather rare.⁸

Generally, severe forms of hypospadias are typically accompanied by an abnormal ventral curvature of the penis (chordee). This is due to the difference in length between the ventral and the dorsal side of the penis (corpora cavernosal disproportion). Proximal hypospadias frequently has a penoscrotal transposition and/or bifid scrotum.⁸

Further abnormalities in hypospadias concern the prepuce. Typically, there is a dorsal hump with excessive skin on the dorsal and a scarcity of foreskin on the ventral aspect of the penis. In most cases, the frenulum is entirely missing. In the rare cases when the prepuce is normally developed, it must be preserved, and a circumcision avoided.⁸

Surgical reconstruction is the only possible therapeutic option for hypospadias.⁷ Notwithstanding the dispute on single-stage versus two-stage repairs, and agreement exists that procedure assignment in hypospadias repair is based on the patient's anomaly and the surgeon's experience and preference. However, the ultimate surgical goal is common and that is to reconstruct a healthy or near-normal appearance of penis mimicking that of post circumcision one, to have adequate caliber, water-proof neourethra extending to the apex of glans for upright voiding with regular urine stream, to create a straight penis adequate for sexual intercourse, and to prevent complications.⁹

The repair of proximal hypospadias involves the correction of several components of the hypospadias complex with the intent of optimizing long-term functional and cosmetic outcomes. These com-

ponents include ventral penile curvature, proximal location of the urethral meatus, ventral skin deficiency, abnormal glans morphology, abnormal division of the corpus spongiosum, penile torsion and penoscrotal transposition.¹⁰

There are five sequential steps for the successful repair of hypospadias: 1) Chordectomy orthoplasty or penile straightening, 2) Urethroplasty or reconstruction of the urethra, 3) Meatoplasty and Glanuloplasty, 4) Scrotoplasty, and 5) Skin Coverage.

Penile straightening is achieved by removal of the fibrous remnants on the ventral aspect of the penis, and the urethra is created by tubularization of the urethral plate.¹¹ The procedure starts removing any fibrous tissue on the ventral aspect of the penis (chordee).

Correction of the location of the urethral meatus to a position in the glans with a cosmetically appealing appearance is the second component of proximal hypospadias repair. The decision on which urethroplasty technique to use depends on whether the urethral plate is preserved after or during the correction of ventral curvature (VC). In contemporary hypospadias surgery, there has been an increasing trend to preserve the urethral plate where possible.¹² This stems from a recognition that the urethral plate is not always comprised of fibrotic tissue that leads to VC, as once thought.¹³

The significance of scrotoplasty has been stressed not only for cosmetic reasons but also for a successful outcome of urethroplasty. The ventral aspect of the phallic urethra, where the skin is inherently deficient, is now covered by ventrally transposed scrotal skin. This procedure corrects bifid scrotum and at the same time, either supplies copious amount of skin for future urethroplasty when this is done as an intermediary procedure for an otherwise three-stage operation or protects the urethroplasty wound when it is combined with Cecil's procedure. Scrotoplasty also affords satisfactory functional outcome because it lengthens the penis and corrects the tendency for the erect penis to point into the scrotum. Despite these beneficial aspects of scrotoplasty, few reports deal specifically with correction of the bifid scrotum. By inclusion of scrotoplasty, however, the techniques described heretofore have the drawbacks either of totaling the whole number of procedures to three before completion or inclusion of the scrotal skin in the subsequent urethroplasty wound when scrotoplasty is combined with cordectomy.¹⁴

Less and early surgery have an influence on the psychological development of the child with genital deformity. Scrotal inlay in the urethroplasty is also condemned because of its hair-bearing tendency. With these in mind, we have revised the two-stage procedure, which eliminates these drawbacks. The initial procedure consists of cordectomy and concurrent scrotoplasty combined with urethral mobilization to obviate incorporation of the scrotal skin and also to shorten the wound in the future urethroplasty. The second and final stage procedure is urethroplasty from the perineum to the tip of the glans with meatoplasty.¹⁴

Testosterone therapy in the form of 3 monthly intramuscular (IM) injections has been used to increase penis size in infants and children.

Testosterone therapy has generally been found effective in treating micropenis due to testosterone deficiency. Bin-Abbas *et al.*,¹⁵ showed that 1 or 2 courses of 3 testosterone injections (25-50 mg) administered at 4-week intervals in infancy or childhood resulted in a sufficient increase in penis sizes to reach the reference range for age. With appropriate pubertal and adult replacement, patients achieved average adult penis size and reported sexual activity and appropriate gender identity.¹⁵ Thus, evidence suggests that treatment with testosterone during infancy or childhood primes the penis for later growth during puberty.

The incidence of complications of proximal hypospadias repair is far larger than with distal hypospadias. Generally, complication rates of penile, scrotal, and perineal hypospadias correction have been similar. Urethra-cutaneous fistula remains the most common complication encountered by all.¹

Conclusion

Proximal hypospadias is a less common occurrence but is the most severe manifestation of the hypospadias spectrum. Proximal hypospadias is typically accompanied by an abnormal ventral curvature of the penis (chordee) and usually associated with scrotal malformations, such as penoscrotal synechia hypoplasia, bifid scrotum, and high scrotum implantation.

Surgical reconstruction is the only possible therapeutic option for hypospadias. Surgical correction of both chordee and hypospadias is recommended. There are five sequential steps for the successful repair of hypospadias: 1) Chordectomy orthoplasty or penile straightening, 2) Urethroplasty, or reconstruction of the urethra, 3) Meatoplasty and Glanuloplasty, 4) Scrotoplasty, and 5) Skin Coverage. When adequately corrected, it does not cause long-term problems, and a natural appearance of the penis is usually restored.

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