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

Paulina Magdalena, Gede Wirya Kusuma Duarsa

Urology Division, Department of General Surgery, Sanglah Hospital/ Faculty of Medicine, University of Udayana, Denpasar, Bali Indonesia

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, corpectomy 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. 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. 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.
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. 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 synchiae, hypoplasia, bifid scrotum, and high scrotum implantation.

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.

Most hypospadias occurs as an isolated condition, but associated anomalies include uni-bilateral cryptorchidism and microgenitalis. 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 microgenitalis 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 microgenitalis and warrant evaluation.

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.

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.

**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 uncircumcision 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.](image1)

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 corpectomy-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](image2)

During the first stage of the procedure, corpectomy-orthoplasty was successfully been performed without any complication during the procedures. Follow up 21 days after corpectomy-orthoplasty showing a straight penis. The patient can urinate with the stand-up position because the urine no longer flowing along the thighs.
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Testosterone therapy has generally been found effective in treating micropenis due to testosterone deficiency. Bin-Abbas et al., 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. 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.

### 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 synchia 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) Meato-plasty 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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### References


