Case Report

An Unusual Variant of Mid-penile Hypospadias with Intact Prepuce and Patulous Urethra : Addition of a Drop to the Sea of Hypospadias Variants

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Abstract

Hypospadias is one of the most common and extensively studied pathology of paediatric surgery. Although a plethora of research articles describing hypospadias and its variants exist, new variants are often encountered and they never cease to surprise us. We, in our case, are trying to add a drop to the sea of variants of hypospadias. A 2-year-old boy was brought with an abnormally large and patulous urethral opening at the mid-penile region with intact prepuce and chordee. In this case report, we will review the available literature on this rare variant and discuss its management.

Keywords: Intact prepuce, large mid-penile urethral opening and megameatus, mid-penile hypospadias, patulous urethra

Introduction

Hypospadias is a common developmental anomaly of the male external genitalia distinguished by aberrant growth of the ventral foreskin of the penis and the urethral fold, which results in improper positioning of the urethral opening proximal to its normal location at the tip of the penis.[1,2] Megameatus with intact prepuce (MIP) is an uncommon variant of distal hypospadias that has a widely splayed coronal or subcoronal meatus, a deep glanular groove, a normally conformed prepuce and no chordee.[3,4] Although its exact incidence is unknown, it occurs in between 3% and 6% of all hypospadias cases. Embryologically, as prepuce is the last structure to form, this epithelial layer originates from the coronal sulcus and encloses the glans circumferentially. Glans lamellae extend laterally and ventrally and become confluent with the margins of the urethral plate and dorsally with its opposite part.[5] Arrested development of the urethra may leave the meatus located anywhere along the ventral surface and leads to the arrest of the other structures like prepuce, which fails to form circumferentially. Twenty percent of all hypospadias have a urethral opening in the middle of the penile shaft with or without chordee.[6] To the best of our knowledge, no cases of intact prepuce along with mid-penile hypospadias have been yet reported as it defies the currently agreed on theories of embryological development. In this case report, we will review the available literature on this rare variant and present its management.

Case Report

A 2-year-old boy presented to our outpatient department (OPD) with complaints of passing urine from an opening on the undersurface of the penis with downward bending of the genitalia. On examination, intact prepuce (non-retractile) with a large urethral meatus was present in the mid-penile region. The urethra was patulous around the meatus with a chordee of approximately 30 degrees present and the glans was not

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visualised as it was covered with preputial skin. Testes were fully descended [Figure 1a-d].

After an initial pre-anaesthetic checkup and clearance, he was taken up for surgery under general anaesthesia. The intact preputial skin around the glans was dissected off till the corona. As there was intact prepuce, the frenulum was present and no glanular groove was noted [Figure 1d]. Glanular stitch was taken with a 5-0 round body polypropylene suture. To correct chordee, a circumferential incision around the dorsal aspect of the glans was taken and the penis was degloved. No chordee was present post-degloving, which was confirmed by Gitte's test [Figure 2a]. The splayed corpus spongiosum was dissected and freed from the corporal bodies. The patulous urethra noted at the urethral meatus in the mid-penile region was trimmed to prevent post-operative diverticulum formation. However, the urethral plate distal to the meatus and the glans groove were narrow. Glanular wings were raised and the 'Snodgrass incision' of the ventral urethral plate was made from the distal penile region to the glans. The urethral plate in the middle shaft was not incised as it was patulous and >1 cm width was present. Urethroplasty was done around 7Fr infant feeding tubes with 6–0 round body polyglactin suture [Figure 2b]. Spongioplasty was done over the neourethra till the corona as a second layer. The dorsal dartos flap was used as a watertight layer to cover the urethroplasty, and glansplasty was then done in two layers. Skin covering was done using Byar's skin flaps [Figure 2c], and compression dressing was done.

The post-operative period was uneventful. The dressing was changed on the post-operative day (POD) 5 and the urinary stent was removed on POD 13 as per our institutional protocol. Intravenous antibiotics (third-generation cephalosporin) were given till POD 5, after which the patient was shifted to oral antibiotics till the urinary stent was removed. After the removal of the urinary stent, the patient passed urine in the stream from the tip of the neourethra. The patient was reviewed in paediatric surgery OPD after 1 week and serial urethral calibration was done with 6/7/8 French (Fr) infant feeding tube, which went easily. At 1 year of follow-up after surgery, the patient is asymptomatic with neomeatus at the tip of the glans and passing urine in the stream from neourethra [Figure 3a and b] without any difficulty and has no fistula.

Discussion

Although hypospadias is one of the most common anatomical congenital malformations of the urethra in males, its exact etiology is not known and is believed to be an amalgamation of genetic, endocrine, and environmental factors. Despite a spectrum of physical features described in the literature, hypospadias with a few aberrant physical features still occur



Figure 1: (a) Normal-looking genitalia with prepuce, (b) Lateral view showing downward bending of the penis with intact prepuce, (c) Intact prepuce on the ventral side (marked by *) with large urethral meatus at the middle of the shaft (marked by arrow), Glans not visualised, (d) View after dissecting off the intact prepuce from the glans

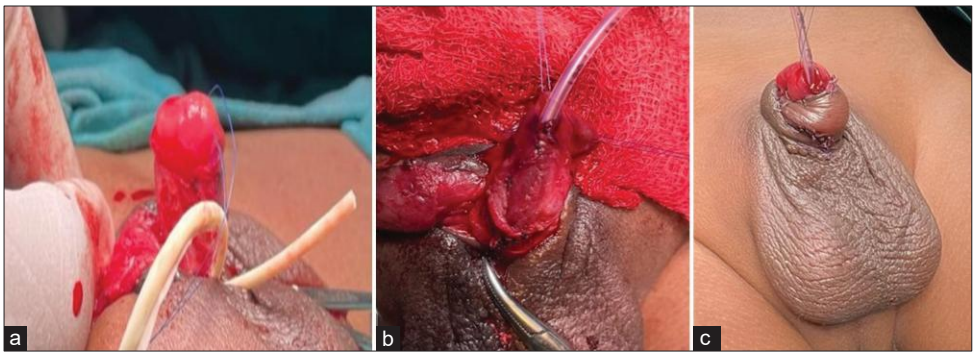


Figure 2: (a) Lateral view showing penis after degloving, note absence of chordee on Gittestest, (b) After Snodgrass incision over distal penile urethral plate till the glans, Urethroplasty done over 7 fr. Infant feeding tube, (c) Immediate Post-operative image. Neomeatus seen at the glans with no chordee or torsion

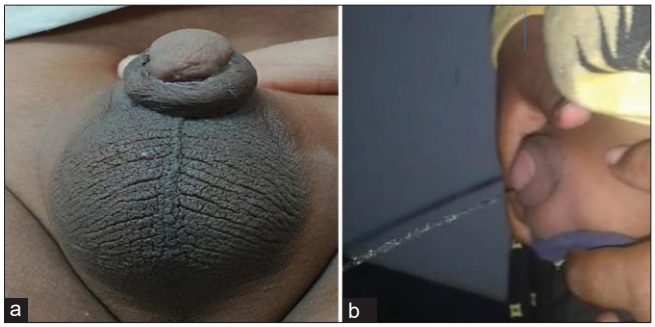


Figure 3: At 1-year follow-up (a) Meatus present at the tip of glans, (b) Child passing urine in good stream from neomeatus

and tend to surprise physicians as they sometimes defy the proposed embryology. Our case is one such example as the presence of intact prepuce ventrally despite a hypospadiac urethral meatus at the mid-penile region defies the proposed current embryologic theory, As prepuce is the last structure to form, arrest in the development of urethra leads to arrest in development of prepuce ventrally leading to the physical appearance of a ventrally deficient prepuce in a classical case of hypospadias.[7]

The variant of hypospadias described in the literature that most closely resembles our case is the MIP variant. However, in the MIP variant, the formation of prepuce is normal with the distal urethra open and it has been theorized that over canalisation or persistent splitting of the distal, ventral urethra may have caused the disunion of the glans and an abnormally large meatus with a deep glans cleft.[4,6,8] Our case is different from MIP because of the following reasons; First, the location of the urethral meatus is distal and ventral in the MIP variant, whereas in our case location of abnormal urethral meatus was in the midpenile region ventrally. Second, the MIP variant typically has a deep glans groove, whereas in our case, the glans groove was shallow. Third, the MIP variant has no associated chordee, whereas in our case chordee was present, which required penile degloving for correction. Therefore, to the best of our knowledge, our case is the first such case described in the literature.

As we encountered a new variant anatomy, the surgical procedure had to be tailored as per the anatomy. Therefore, after the chordee was corrected post-penile degloving, we gave a vertical incision over the distal urethral plate and glans, just as we do in Snodgrass urethroplasty with excision of the proximal patulous urethra to avoid the formation of any diverticulum in future. Therefore, the technique used by us is a combination of tubularized incised plate urethroplasty in the distal part and Thiersch-Duplay urethroplasty in the proximal part of the repair. Both of these techniques are well established in the literature.[8,9] Due to the difference in anatomy between our case and MIP variant, procedures such as the modified glans approximation procedure or modified pyramid procedure described in the literature for the management of the MIP variant[4] cannot be used for the management of our case.

In conclusion, despite a plethora of variants and spectrum described in the literature, unusual variants like our case can be encountered rarely. The management of such variants requires tailoring the surgical management to the variant anatomy so that the best possible surgical results can be obtained and complications such as urethrocutaneous fistula, meatal stenosis and urethral diverticulum can be avoided.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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