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## A case of distal hypospadias masquerading as Megameatus intact prepuce

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### Abstract

Hypospadias is a common abnormality in males which the urethral opening is ectopically located on the ventrum of the penis as far down as in the scrotum or perineum. Classic hypospadias is characterized by the triad of ventral urethra defect, absence of dorsal prepuce and chordee. A megameatus with intact prepuce (MIP) is an unusual variant of an anterior hypospadias. It is characterized by a widely splayed coronal or subcoronal meatus, a deep glanular groove, a normally conformed prepuce, and no chordee. Here, we report a case of a 23-year-old male who was diagnosed as MIP clinically but found to be a hypospadias intraoperatively.

**Keywords:** Chordee, hypospadias, MIP, rare case

### Introduction

Hypospadias is a common congenital anomaly in males caused due to partial development of the urethral meatus hence it opens anywhere on the ventral side of the penis rather than the tip of it. It is seen in about 1 in 300 male live births<sup>[1]</sup>. Depending on the opening of the meatus, it can be classified as posterior, middle and anterior type, of which the anterior variety is the most commonly observed. The megameatus intact prepuce is a rare variant of hypospadias. It is usually not reported or diagnosed only while performing circumcision or retracting the foreskin. It accounts for only 3% of the hypospadias repair<sup>[2, 3]</sup>. In majority of the cases hypospadias has ventral meatal opening, absent dorsal prepuce and presence of chordee, while MIP has intact prepuce, absence of chordee and mild to large fish mouth meatal opening. Both the conditions can be clinically well differentiated. But the below reported case is of a rare finding with symptoms of MIP but patient had chordee which changed the diagnosis to distal hypospadias.

### Case Report

A 23-year-old male presented with a history of bending of penis since birth. Patient had an intact prepuce and on its retraction it was found to have a widely splayed subcoronal meatus and deep glanular groove (Figure 1A). All these phenotypical features mimicked the diagnosis of MIP. However, the patient also had ventral chordee, which usually is not an associated feature of MIP. Patient also had a tight frenulum which could be the cause of chordee (Figure 1B). Intraoperatively after penile degloving a Y shaped distal 2.5 cm of corpus spongiosum and thin distal urethra of length 2.5 cm were found (Figure 2A). Even after the release of frenulum, chordee persisted. On further dissection of the underlying buck's fascia about 80% of chordee correction was achieved. Both the wings of the Y shaped spongiosum were released and sutured over the thin urethra in a Y to I manner. Finally, glanuloplasty was done and a neomeatus was created at the tip of glans. Figure 2B shows improvement on postoperative day 1.

### Discussion

The term hypospadias was first considered by Galen in second century AD. It is a congenital anomaly seen in males in which the urethral opening is dystopic and located at any position on the ventral side of the penis. It maybe abnormally located anywhere in the glans, shaft of penis, perineum or scrotum. It is also associated with atretic corpus spongiosum, ventrally deficient foreskin and a ventral curvature of the penis (chordee).

Depending on the location of opening of the meatus, hypospadias can be classified into various types. One of the rare types of hypospadias is megameatus intact prepuce variant. In this type the patient has a mildly dilated or a large fish mouth meatal opening. But it is usually detected later in life or at the time of circumcision. Clinically if the patient has a mild meatal dilatation, he shall have a normal urine stream while if the opening is wide he may present with a splayed urine stream. As the name suggest the prepuce is intact and normal and also there is no curvature of the penis.

Embryologically in males the genital tubercle enlarges and further develops from the fifth week of gestation. During this process the urethral groove deepens and the urethral folds on its either side enlarge and fuse towards the glans. But in hypospadias there is an incomplete fusion of these urethral folds which results in an incomplete urethra with hooded foreskin.

About 50% to 70% of hypospadias cases are the anterior or distal types (1,4). The surgical repair of distal hypospadias has been successful in majority of the cases with comparatively lower chances of complications [5, 6]. Several surgical techniques have been advocated for repairing anterior hypospadias. Some of these techniques are MAGPI, Mathieu, Arap, Snodgrass, Mustard and Barcat, among which Mathieu and Snodgrass are the most commonly used techniques [1, 4].

Some hypospadias variants present with a normal foreskin concealing a glanular to distal shaft meatus. These generally have a deeply grooved urethral plate, which sometimes extends laterally under the skin edge creating a phenotype known as the

megameatus with intact prepuce. The diagnosis of these variants is made during circumcision or when the foreskin becomes retractable. Embryological origins are unclear, but it appears to be related to megalourethra [7].

One has to be aware of the variant in order to avoid confusion. The features of MIP variant include a spatulated glans with a distal, wide patulous meatus at the deep subcoronal groove with an intact foreskin, a thin corpus spongiosum and importantly no ventral chordee. If curvature is present, it undoubtedly suggests dorsal hypospadias [8]. MIP has no effect on the flow of urination or sexual functioning. It's only the appearance which might disturb a child's psychology and hence cosmetic remodelling is usually indicated.

Several technical approaches are used for MIP reconstruction, including the glanular approximation procedure (GAP), the pyramid procedure, and tabularised incised plate (TIP) urethroplasty [3, 8, 9].

Our above case had a rare presentation with features of MIP but presence of chordee which is a feature of distal hypospadias but again in hypospadias the prepuce is not intact. Thus the patient neither fits into a classical hypospadias because there was an intact prepuce nor into a MIP as there was chordee associated with other findings. A prompt clinical examination is the key for identification of MIP. The foreskin should be fully retracted and the glans and the urethral meatus should be examined to confirm the diagnosis before planning any surgical intervention. The preputial and ventral urethral meatal development is independent of each other.



**Fig 1:** (A) Widely splayed subcoronal meatus with deep granular groove seen after retraction of prepuce and (B) associated chordee



**Fig 2:** (A) Y shaped distal spongiosum and thin distal urethra and (B) postoperative day 1

### Conclusion

Prompt diagnosis of MIP is a challenge as deciding the future management depends on it. Both MIP and distal hypospadias can have a psychological and functional impact on the patients. Hence diagnosis and management should be considered on both cosmetic and functional grounds.

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