Andrology: Open Access
ISSN: 2167-0250
OPEN ACCESS Freely available online
Research Article

Hypospadias with Intact Prepuce: Evolving Morphology and Current Surgical Techniques

Ramaswamy Rajendran*

Department of Pediatric Urology and Pediatric Surgery, GG Hospital, Murinjapalam, Trivandrum, Kerala, India

ABSTRACT

A comprehensive research-report on the anatomy of hitherto designated Megameatus with Intact Prepuce (MIP) was recently published in early 2024. "Hypospadias with Intact Prepuce (HIP)" as the umbrella-term to contain cases with megameatus and all other cases of hypospadias with entire prepuce was proposed along with their current surgical techniques. The incidence of MIP is 3% to 6% among hypospadias. Presence of median raphe anomalies or long redundant prepuce may be pointer to the occult MIP. Cases may present post-circumcision in which case, iatrogenic hypospadias should be clinically excluded. Unlike popular belief, MIP can be associated with dorsal or ventral chordee. Glans-penis may have wide, spatulated or conical shape. MIP usually is distal hypospadias, but rarely midshaft hypospadias too. There is wide variation in the size of External Urethral Meatus (EUM) from normal to wide or enormously wide. Thus the conventional nomenclature "MIP" is inappropriate to some. On the other hand, HIP suits to all such cases irrespective of having megameatus or not. Urethral Plate (UP) may be wide and deeply clefted or not. Distal transverse septum in UP is rare. Associated distal urethral dilation is not omnipresent. Distal megalourethra associated with 8.33% of HIP had serious internal urinary anomalies. Thus a spectrum of anomalies of penile curvature, median raphe anomalies, long redundant prepuce, anomalous glans, EUM, UP and distal native urethra can be clubbed together under the umbrella-term "HIP". Duckett and Keating employed 'Pyramid Procedure' which involved wedge-excision of distal urethra and urethroplasty. Suture-lines overlapping and lack of necessity to excise distal urethra caused disfavour of this technique. Glanular HIP can be repaired by Glans Approximation Procedure (GAP), Tabularized Urethral Plate Urethroplasty (TUPU) or Meatal Advancement and Glanuloplasty Incorporated (MAGPI) depending upon the morphology. Coronal and subcoronal varieties can be corrected by MAGPI, Mathieu's, TUPU or Tubularised Incised Plate Urethroplasty (TIPU) where as penile shaft varieties can be repaired by TUPU or TIPU.

Keywords: Hypospadias with Intact Prepuce (HIP); Megameatus with Intact Prepuce (MIP); Megalourethra; Tubularised Incised Plate Urethroplasty (TIPU); Tubularised Urethral Plate Urethroplasty (TUPU)

INTRODUCTION

The first mention of mega meatusand intact prepuce was done by Juskiewenski in 1983. Duckett JW and Keating MA, are credited with the maiden description and designation of this peculiar category as Megameatus with Intact Prepuce (MIP) in 1989 [1]. They devised the most specific surgical technique for correction of this MIP as the "Pyramid Technique". Subsequently at different periods, authors internationally have published their observations on the anatomy of MIP and their success-stories from application of other corrective surgeries too. A comprehensive research-report on the anatomy of hitherto designated MIP was recently published in 2024 [2]. The nomenclature of "Hypospadias with Intact Prepuce (HIP)" as the umbrella-term to contain cases with mega meatus and all other cases of hypospadias with entire prepuce was proposed by these authors. The applicability of various remedial surgical techniques also was probed in this research.

METHODOLOGY

Apparently normal penis with entire prepuce masquerade the presence of hypospadias in HIP. Only if the prepuce is retracted or circumcised, HIP can be recognized. The incidence of HIP is 3%-6% among hypospadias. But incidence as high as 15% by Snodgrass, 118 cases over 2.5 years' period by Ben-David and as low as 1.26% by Bhat, et al., are published [3-5]. HIP is asymptomatic. Hence, it cannot be recognized unless prepuce is retracted or circumcised. In societies where practice of religious circumcision in newborn is prevalent, this anomaly is detected by the circumcising physician and circumcising such anomalous penis is aborted. The more severe variants of HIP may be associated with urinary spraying while micturition [1]. Presence of subtle external genital anomalies may be pointer to the occult HIP. Median raphe deviation, bifurcated raphe, raphe hyperpigmentation, chordee, penoscrotal web, undescended testis, or redundant long

Correspondence to: Ramaswamy Rajendran, Department of Pediatric Urology and Pediatric Surgery, GG Hospital, Murinjapalam, Trivandrum, Kerala, India, E-mail: pedsurgdrraj57@yahoo.in

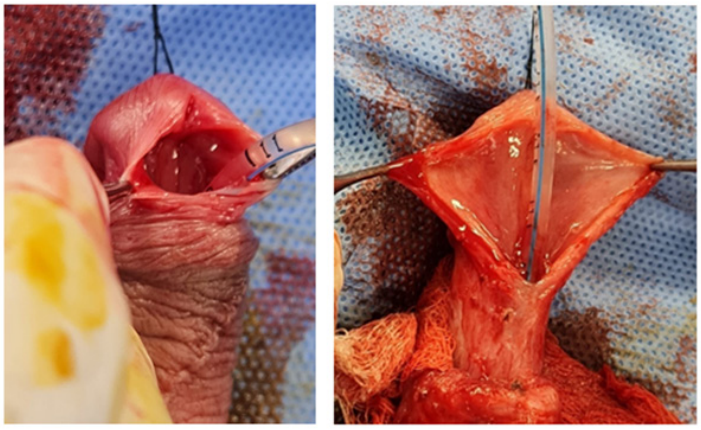
Received: 16-May-2024, Manuscript No. ANO-24-31469; **Editor assigned:** 20-May-2024, Pre QC No. ANO-24-31469 (PQ); **Reviewed:** 03-Jun-2024, QC No. ANO-24-31469; **Revised:** 10-Jun-2024, Manuscript No. ANO-24-31469 (R); **Published:** 17-Jun-2024, DOI: 10.35248/2167-0250.24.13.317

Citation: Rajendran R (2024) Hypospadias with Intact Prepuce: Evolving Morphology and Current Surgical Techniques. Andrology. 13:317.

prepuce should arouse suspicion of HIP [6,7]. Association of ureteropelvic junction obstruction, nocturnal enuresis, megalourethra, nonfunctioning kidney, vesicoureteric reflux also are observed [2,6]. Variable proportion of patients with HIP are already circumcised when they consult pediatric surgeon for remedy [1-3,8-10]. The circumcision physician may be dragged into litigation for this alleged iatrogenic hypospadias. But history of severe bleeding due to shearing of glans at circumcision and on examination scarring of glans along with stenotic meatus due to injury to the urethra differentiates iatrogenic hypospadias complicating circumcision from genuine HIP [9].

The gross size of the penis was normal-for-age in 83% patients and subnormal in the rest, as observed in the latest research [2]. It has been generally believed for many years that MIP is not associated with chordee, which is characteristic of majority of cases of hypospadias in general; if at al chordee is detected, it is dorsal chordee [11]. But Ekberli, et al., reported ventral chordee in 2 of their 31 MIP patients [6]. In 2021, a very high percentage of chordee was found by Ben-David, et al., dorsal chordee in 19% and ventral in 5% [4]. Dorsal chordee in 86% of cases had curvature ≥ 30° requiring ventral plication. The uniform characteristics of all 14 cases of Duckett JW and Keating MA [1], were absence of a ventral chordee, wide glans penis; wide mouthed "Blunderbuss" External Urinary Meatus (EUM) associated with wide distal urethra and wide, deeply grooved Urethral Plate (UP). The categorization of their cases as MIP is fully justified. In the subsequent years various authors reported diverse morphology in their individual case series of MIP, but still continued with the terminology of MIP until Ramaswamy justified the necessity of containing all such cases with diverse morphology of hypospadias having entire prepuce under the umbrella term HIP [2]. Shape of glans-penis varies widely. Duckett JW and Keating MA observed the glans in their cases of MIP "mirror that of balanic epispadias" [1]. Other authors observed wide and shovel like glans having deep navicular fossa, spatulated glans or presence of double grooves in the glans [5,9,12]. We observed variable shape of glans from wide shovel-like (in 75%) to conical shape (in 25%) in our series [2]. Most researchers found distal hypospadias in HIP but recently midshaft hypospadias also has been observed [1,2,4,5,10,12,13]. EUM was universally wide in the cases of Duckett JW and Keating MA, which was the basis of clustering them under the nomenclature MIP [1]. Large variation in the appearance of the EUM ranging from normal through mildly dilated to large fish mouth opening with 6 cm diameter has been reported afterwards [13-15]. We observed wide EUM in the vast majority of HIP but normal sized in 17% of our series as shown in Figure 1 [2]. There was no uniformity in the size of the wide meatus too. Thus, wide EUM is not universal hallmark of cases of hypospadias with entire prepuce. On the basis of wide variation in the size of EUM from normal to enormously wide in these cases of hypospadias, the conventional nomenclature of "MIP" is inappropriate to some. On the other hand, "HIP" suits to all such cases irrespective of having megameatus or not. Urethral Plate (UP) was wide and deeply clefted in cases of MIP of Duckett JW and Keating MA [1]. Wide (≥ 12 mm) or moderately wide (8 mm to 12 mm) UP was reported in MIP of other authors [5,12,14]. We observed variable UP characteristics. Majority (75%) had wide UP but only 50% looked deep. Even narrow (≤ 6 mm) and flat UP were owned by our HIP. Snodgrass WT and Khavari R, as well as we have seen distal transverse septum in UP causing deflection of urine stream, in a minority of HIP. This is corrected by dorsal UP incision [2,3]. Wide meatus was associated with widened distal urethra in the entire case-series of Duckett JW and Keating MA [1]. Distal urethra was wide and deficient of spongiosum in the cases of Faasse, et al., [16]. Attalla MF [13], specified that his case-series had no wide distal urethra. Other authors who contributed to the literature of MIP did not comment on the distal native urethra, from which inference can be drawn that no dilation of distal urethra accompanied MIP. Distal urethra was normal in all but one which was dilated (8.33%) [2]. This case was unique with enormously wide coronal meatus and adjacent urethra qualified to be termed as megalourethra shown in Figure 2. This megalourethra was tapering to normal at the mid-shaft level, was thin and not surrounded by corpus spongiosum but with normal corpora cavernosa. UP was very wide and deep but thin and dysplastic with wide shovel like glans penis. The presence of megalourethra provoked us to investigate him. This led to diagnose associated serious internal anomalies. Congenital heart disease (atrial septal defect), right side atrophic, nonfunctioning kidney (split renal function 1.62% as per isotope renogram), right side Vesico Ureteric Reflux (VUR) Grade IV-V, left side VUR Grade I, and developmental delay were the anomalies. In general there are no associated urological anomalies with MIP; so no radiological evaluation is needed in such patients [17]. However, Ekberli, et al., noticed genitourinary anomalies in 7/31 of his patients, as mentioned earlier [6].

Figure 1: HIP with different shapes of glans, different size of meatus and width of urethral plate; A) Wide (shovel-like) glans and wide, deep UP with distal septum; B) Conical glans and normal sized meatus in glanular HIP; C) Conical glans with narrow and shallow UP in thin long penis; D) Wide meatus and moderate width of UP; E) Normal meatus and UP of normal width but grooved; F) Dorsal vascular dartos flap for ventral transposition as superimposition layer used in TUPU and TIPU; G) Post-TIPU of subcoronal MIP shown in Figure (1D); H) 4 Months post-TIPU of the subcoronal MIP.



2(A) 2(B)

Figure 2: HIP with megameatus and megalourethra. A) Wide shelf-like UP with distal septum and megameatus; B) Megalourethra opened.

RESULTS AND DISCUSSION

Thus the previously popular "MIP" had no chordee, had no hinting external penile anomalies, was a rare variant of distal hypospadias, had wide glans-penis with megameatus-dilated distal urethra complex, had wide, deeply clefted UP and had no associated urinary anomalies. But today we have ample data to challenge the above uniform morphology in cases of hypospadias having entire prepuce. A spectrum of anomalies can be clubbed together under the umbrella-term "HIP". Such cases may have ventral/dorsal chordee, median penile raphe anomalies or long redundant prepuce to give clue to covert HIP, may have distal or midpenile EUM which may or may not be wide, glans-penis and UP have variable features, distal native urethra is undilated in general but rarely dilated as shown in Table 1. If there is association of megalourethra, such case can be associated with serious internal urinary anomalies. MIP is a large subgroup under HIP.

In the modern era with the variable morphology of HIP, treatment may be cosmetic remodeling of ventrally placed, cosmetically unappealing urethral meatus, or functional correction of dorsal/ventral chordee and of midpenile or distal hypospadias. Occassionally, surgery is adopted due to spraying of urine during micturition or to avoid child's or parent's psychological disturbance. Duckett JW and Keating MA originally devised "Pyramid Procedure" as panacea for MIP [1]. The megameatus, deep glanular UP, and widened distal urethra, universal to their cases, are dissected as a unit, a wedge of distal native ventral urethra is excised, urethra and UP are tabularized, and glans is repaired. Excellent results with no complications in their hands could not be reproduced by others. Overlapping of two suture lines risk fistula formation as a complication. In the absence of distal urethral dilation in most cases of HIP, tailoring of distal urethra is unnecessary. Moreover only minimal dissection around distal urethra is necessary, extensive mobilization of UP is not warranted. The pyramid procedure has not gained popularity. Snodgrass WT and Khavari R [3], implemented the procedure to reconstruct virgin cases and circumcised cases of HIP by Tubularised Incised Plate Urethroplasty (TIPU). Most patients already circumcised underwent a ventral "Y" incision along the glans wings converging below the meatus and continuing down the median raphe to the penoscrotal junction. Dorsal midline UP incision was performed routinely followed by UP tabularization. A barrier layer of dartos obtained from either the dorsal or ventral aspect of the penis covered the neourethra in all cases before glanuloplasty [3]. Faasse, et al., excised ventral part of distal native urethra in their cases. Tubularisation with or without midline dorsal UP incision along with superimposition layer of dorsal prepucial vascular dartos flap completed the repair [16]. Glanular HIP can be repaired by Glans Approximation Procedure (GAP) or Tubularised Urethral Plate Urethroplasty (TUPU) if UP and meatus are wide; in presence of conical glans and normal-sized meatus, MAGPI is suitable [10]. Coronal or subcoronal HIP has many choices for repair. MAGPI in presence of conical glans and normal-sized meatus, Mathieu's if UP not wide and the formed urethra is short, TUPU for cases with wide UP and wide meatus, and TIPU for cases where UP is not wide enough [3,6,12]. Penile shaft varieties can be repaired by TUPU or TIPU according to the width of UP as shown in Figure 3. Does circumcised status of HIP unfavorably affect reconstruction? Prior circumcision need not adversely affect reconstruction, even though the presence of prepuce is beneficial [3]. Hypospadias was an absolute contraindication to circumcision, since the foreskin might be needed for surgical correction. Recently comparison of results of 70 post-circumcision MIP hypospadias repair with 69 post-circumcised non-MIP classic hypospadias repair arrived at the conclusion that reoperation rates in MIP hypospadias are high compared the to the results of children who underwent non-MIP hypospadias repair of following neonatal circumcision. This suggests strong evidence of the possible role of previous circumcision in the surgical challenge of reconstructing MIP hypospadias [18].

In many cases with very wide UP but glans wings are thin, there needs deep dissection of UP and excision of its thin margins eventually require TIPU. Bhat did three modifications in the classical Thiersch-Duplay technique of TUPU in their cases. First, the neourethra was covered with spongiosum in place of dorsal dartos flap, the second, prepuce was preserved, and the third, frenuloplasty was done to create a normal appearing penis [5]. Tubularization of UP followed by Mathieu modification of superimposition with de epithelialized inframeatal vascular flap has been practiced by Duan, et al., and Cendron M [12,14]. Cendron M, concluded that after adopting the inframeatal flap dissection as the first step, and using traction on this inframeatal flap, UP injury was averted, which is an added benefit of this technique [14]. We used TUPU technique (Thiersch Duplay) to construct neourethra in glanular and coronal types of HIP. All sub-coronal and some coronal types were repaired by TIPU. This tubularisation involved

epithelium and spongiosum together as a single layer. A protective superimposition layer was often used. In the case of megalourethra, the glans wings were thin and shelving, and lateral parts of very wide UP were excessively thin, unhealthy, and traumatized on dissection. Post-excision of severely damaged, vascularity-compromised peripheral portions of UP and ventral part of megalouretha, narrow UP resulted; consequently TIPU was unavoidable for repair [2]. A hinging midline incision of UP relieves suture line tension on neourethra. To prevent suture line overlapping and to add vascular layer in TIPU, buttressing by ventral or dorsal vascular dartos flap should be preferred [3]. Distal urethral injury and UP injury in a minority of cases can be effectively solved [2]. Excellent postoperative results of us and other authors are testimony to continue with the above techniques of repair.

Table 1: Findings of various researchers in Megameatus Intact Prepuce (MIP).

Author	Incidence	Chordee	Glans	E.U.M	U.P	Distal lip in U.P	Distal urethra	Associated anomaly	Operation
Duckett JW and Keating MA, [1]	(14 cases)	-	-	Wide	Wide, deep	-	Wide		Pyramid procedure
Attalla MF, [13]	(14 cases)	-	-	Not wide. Subcoronal	-	-	Not wide		Pyramid procedure
Bar-Yosef, et al., [10]				Coronal/ Glanular/ distal shaft					TIPU, GAP, MAGPI
Snodgrass WT and Khavari R, [3]	15% (63 cases)								TIPU
Bhat, et al., [5]	1.26% (13cases), Spectrum	-	Double groove+	Glanular, coronal, subcoronal., distal penile	Wide	+			TUPU, frenuloplasty GAP
Cendron M, [14]	5% (25cases)	-	Wide, splayed	Large variation in size and shape	Irreglar, wide	-			Modified Mathieu
Duan, et al., [12]	5% (25cases)	-	Wide, shovel-like	Coronal, distal penile, glanular	0.8 cm-1.2cm	-			TIPU, GAP, Duplay, Mathieu, MAGPI
Ekberli, et al., [6]		2/31						Medianraphe anomalies 10/31, Others 7	TUPU, TIPU, meatoplasty
Ben-David, et al., [4]	5% (25cases)	Dorsal19%, Ventral5%		Glanular, coronal, subcoronal, distal penile, midpenile					
Ramaswamy, et al., [2]	4.72% (12cases)	nil	Broad 9, conical 3	Wide 10, Normal 2	Variable, width	2	Megalourethra=1	Serious urinary anomaly in 1	TUPU, TIPU

Note: M.I.P=Megameatus Intact Prepuce; EUM=External Urethral Meatus; UP=Urethral Plate; TIPU=Tubularised Incised Plate Urethroplasty; TUPU=Tubularised Urethral Plate Urethroplasty; GAP=Glans Approximation Procedure; MAGPI=Meatal Advancement and Glanuloplasty Incorporated.

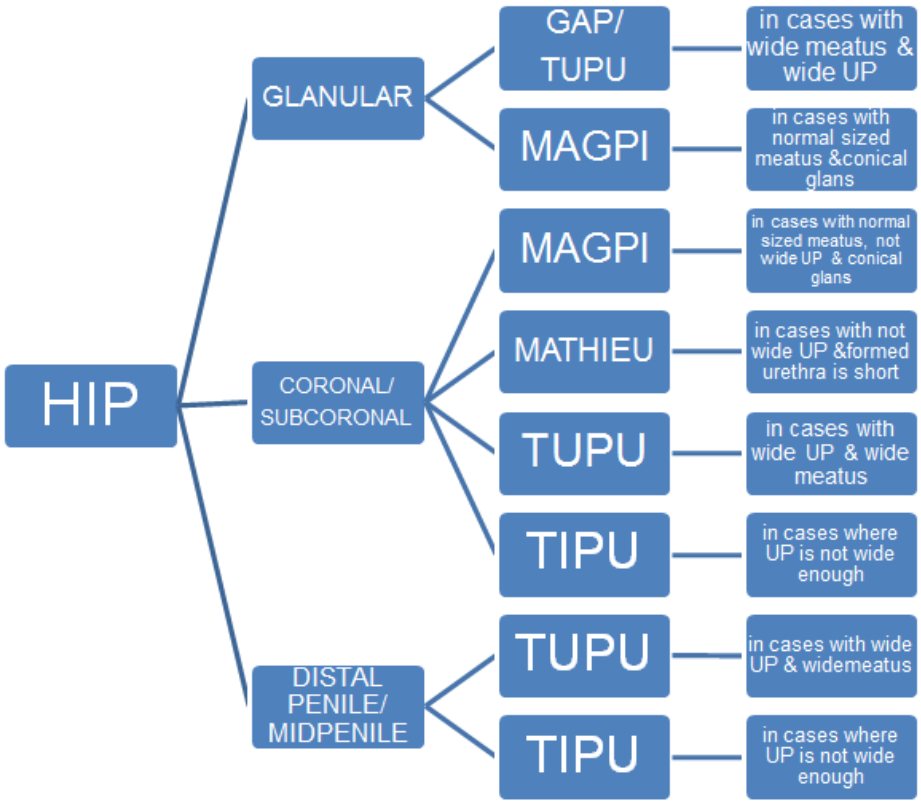


Figure 3: Current operations used for repair of HIP; Note: HIP: Hypospadias with Intact Prepuce; GAP: Glans Approximation Procedure; TUPU: Tubularised Urethral Plate Urethroplasty; MAGPI: Meatal Advancement and GlanuloPlasty Incorporated; TIPU: Tubularised Incised Plate Urethroplasty; UP: Urethral Plate.

CONCLUSION

Hypospadias with intact prepuce is a rare variant of hypospadias. Some such cases have no megameatus and hence cannot be termed MIP, contrary to convention. Furthermore, MIP may or may not have wide and deep UP/wide glans penis/wide distal urethra which were described as universal. All cases of hypospadias having intact prepuce can be covered by the umbrella term "HIP"; MIP is a large subgroup under HIP. HIP may have penile median-raphe anomalies or long redundant prepuce. HIP has a spectrum of anomalies of penile curvature, glans penis, EUM, and UP. Significant dilatation of the distal urethra (megalourethra) is an occasional association with HIP. Reconstruction of penis in HIP by tubularization of UP without or with midline incision and superimposition layer or with MAGPI in selected cases gives excellent results.

ACKNOWLEDGMENTS

I am deeply indebted to my previous colleagues Dr. Samir Morsi Hegab (Consultant & Head of Department), Dr.Ghazi Mukattash (Consultant), Dr.Hany Fawsy (Specialist), Dr.Saif Seed Ghalib (Specialist), Dr. Mohamed Shawky (Specialist), Dr. Rayan Ahmed Baz, (Specialist), Dr. Mohamed Eid (Specialist) and Dr. Motaz Ahmed Mohamed (Resident), in Department of Pediatric Surgery, Maternity & Children's Hospital, Najran, Government of Saudi Arabia, for their contribution in this research and involvement in management of cases. I also gratefully Acknowledge the administration of Maternity & Children's Hospital, Najran, Government of Saudi Arabia, for permitting to publish clinical data of research conducted in that hospital.

FUNDING

Nil.

COMPETING INTERESTS

None.

REFERENCES

1. Duckett JW, Keating MA. Technical challenge of the megameatus intact prepuce hypospadias variant: The pyramid procedure. J Urol. 1989;141(6):1407-1409.
2. Ramaswamy R, Hegab SM, Fawsy H, Ghalib SS, Shawky M, Mukattash G. Hypospadias with intact prepuce: A spectrum of anomalies and their reconstruction. J Indian Assoc Pediatr Surg. 2024;29(2):129-136.
3. Snodgrass WT, Khavari R. Prior circumcision does not complicate repair of hypospadias with an intact prepuce. J Urol. 2006;176(1):296-298.
4. Ben-David R, Kupershmidt A, Dekalo S, Herzberg H, Mano R, Dubi-Sobol A, et al. Dorsal penile curvature and megameatus intact prepuce hypospadias: A common association in a rare variant of hypospadias. J Pediatr Urol. 2021;17(4):517-e1.
5. Bhat A, Bhat M, Bhat A, Singh V. Results of tubularized urethral plate urethroplasty in Megameatus Intact Prepuce. Indian J Urol. 2017;33(4):315-318.
6. Ekberli G, Ate U, Sözduyar S, Gurbanov A, Göllü G, Kolo lu M, et al. Megameatus intact prepuce and associated anomalies. J Contemp Med. 2020;10(4):542-545.
7. Fahmy MA, El Shenawy AA, Altramsy A, Samahy OA, Yehya A, Othman D. Penile median raphe anomalies as an indicator of megameatus intact prepuce anomaly in children undergoing routine circumcision. Urology. 2018;121:164-7.

8. Belloli G. The cutaneous advancement procedure for megameatus and intact prepuce repair. Pediatr Surg Int. 1994;9:153-155.

9. Peretz D, Westreich M. Pseudo-iatrogenic hypospadias: The megameatus intact-prepuce hypospadias variant. Plast Reconstr Surg. 2003;111(3):1182-1185.

10. Bar-Yosef Y, Binyamini J, Mullerad M, Matzkin H, Ben-Chaim J. Megameatus intact prepuce hypospadias variant: Application of tubularized incised plate urethroplasty. Urology. 2005;66(4):861-864.

11. Duan SX, Li J, Jiang X, Zhang X, Ou W, Fu M, et al. Diagnosis and treatment of hypospadias with megameatus intact prepuce. Front Pediatr. 2020;8:128.

12. Duan S, Jiang X, Zhang X, Ou W, Fu M, Chen K, et al. Megameatus intact prepuce treated with urethral plate-preserving surgery: A retrospective study of an unusual hypospadias variant. Transl Androl Urol. 2019;8(6):583.

13. Attalla MF. Subcoronal hypospadias with complete prepuce: A distinct entity and new procedure for repair. Br J Plast Surg. 1991;44(2):122-125.

14. Cendron M. The megameatus, intact prepuce variant of hypospadias: Use of the inframeatal vascularized flap for surgical correction. Front Pediatr. 2018;6:55.

15. Muhammad UM, Abdulkadir A, Sharfuddeen AM, Bashir Y. Megameatus Intact Prepuce in a two years old boy in Katsina Northwestern Nigeria. Casereports ReDelve: RD-CRP. 2019;10015.

16. Faasse MA, Dray EV, Cheng EY. Repair of megameatus: A modified approach. J Pediatr Urol. 2015;11(2):100-101.

17. Erikci VS. Glanular approximation procedure in hypospadias variant of Megameatus Intact Prepuce: A mini review. EC Paediatr. 2020;9:01-3.

18. Herzberg H, Ben-David R, Mendelson T, Dubi-Sobol A, Bashi T, Savin Z, et al. Megameatus intact prepuce variant reconstruction: Long-term outcomes and comparison to post-circumcision hypospadias repair. J Pediatr Urol. 2024;20(1):38-e1.