17 Management of Megmeatus Intact Prepuce/Concealed Hypospadias

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Abbreviations

GAP	Glanular approximation procedure
MAGPI	Meatal advancement and glanuloplasty
MIP	Megameatus intact prepuce
TUPU	Tubularized urethral plate urethroplasty
TIP	Tubularized incised plate urethroplasty

17.1 Introduction

Juskiewenski et al., in 1983, first time reported the description of the anomaly, a variant of hypospadias [1], and the terminology of Mega-meatus Intact Prepuce was first coined by Duckett and Keating [2]. This anomaly is also termed Concealed Hypospadias/Hidden Hypospadias. A deep glans groove characterizes MIP, a large meatus, and the glans is entirely covered by an intact prepuce. This unusual anterior hypospadias variant contributes approximately 3–6% of total hypospadias cases [2–5]. Concealed hypospadias is usually missed and presents late because it is covered by the prepuce. Many patients, who are un-noticed, may come to the clinician for any other medical problem because they usually do not have symptoms. Recognition at the time of circumcision in newborns and patients seeking advice for phimosis is likely to diagnose the entity. The diagnosis is often made during routine health checkups before the defence or any other services. The critical features identified in the diagnosis are (1) meatus located close to or just below the coronal margin; (2) A wide, splayed-out glans; (3) A deep glans cleft; (4) An abnormal urinary stream if witnessed. Duckett and Keating [2] recognized the distinct surgical challenges of the MIP variant first time in 1989. The controversy continues whether to operate on these patients or not [6]. But in modern-day hypospadiology, with more concern on the cosmesis, surgery is preferred. The techniques described are the "Pyramid procedure, Glans Approximation technique, Meatal Advancement and Glanuloplasty (MAGPI), Mathieu the peri-meatal-based flap technique, TIP and TIPU". The distinct anatomic features

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A. Bhat (ed.), *Hypospadiology*, https://doi.org/10.1007/978-981-16-8395-4_17

of the concealed hypospadias have led to the emergence of several techniques. Good cosmetic and functional results are preferred compared to those achievable with perimeatal-based flaps and MAGPI [7]. The glans approximation procedure (GAP) came into practice to overcome the challenges of a wide, deep glanular groove and a non-compliant fish mouth with the methods mentioned above [8]. Reconstruction of neourethra without dissection of glanular flaps in the GAP technique creates a neourethra with different diameters depending on the depth of the glanular cleft. The diameter of the neourethra may not match the original one, and the difference in caliber may cause pressure differentials that may predispose to fistula formation [3]. An intermediate tissue interposing layer is not used in the original GAP procedure; so, the two suture lines of glans overlie, increasing the fistula chances. The tubularized incised plate urethroplasty (TIPU) technique is the most commonly done procedure for distal hypospadias repair because of its better cosmetic and functional results. Now the same is being used for its MIP variant repair. As the urethral plate in MIP is very wide, so it does not require the incision in the urethral plate. Bhat et al. (2019) reported the excellent results of Tubularized Urethral Plate Urethroplasty (TUPU) in 13 cases of MIP in a retrospective study [9].

17.2 Embryology

The embryologic basis of the MIP variant of hypospadias is not clearly defined. The urethra develops by the fusion of the urethral plate from proximal to distal. The distal urethra is also a part of the urethral plate but grows from the tip of the glans to meet the proximal penile urethra at the coronal sulcus. A cuff of tissue lies at the margin of the sulcus from the prepuce. The arrest in the development and failure of fusion of the urethral plate results in hypospadias with a hooded prepuce. Duckett and Keating [2] postulated that a misdirected clefting of the glans proceeds down the already fused urethra creating the megameatus after normal folding of the proximal penile urethra and normal prepucial formation. The development of prepuce is independent of the glanular urethra, in their opinion. Nonomura et al. [10] proposed a deformation theory, i.e., ischemic or compressive change after completion of a normal urethra may result in a megameatus and a normally occurring prepuce. Megameatus Intact Prepuce is due to external compression of the penis in utero after completion of penis formation. The urethral plate is always wide and elastic, usually without chordee. Others feel it is embryologically related to the Megalourethra. But both of these theories fail to explain the embryology of Megmeatus Intact Prepuce. Bhat et al. [9] proposed the theory of both canalization and tubularization to develop the glanular urethra. Both canalizations of glans and closure of glanular plate are needed for the normal development of the glanular urethra, as opposed to the canalization alone. Thus, when dorsal canalization and the overlying prepucial tubularization are complete, but ventral urethral plate closure remains incomplete, that results in Megameatus Intact Prepuce (Figs. 17.1a, b and 17.2). But when canalization and tubularization do not meet to complete the normal urethra, then it may lead to partial duplication of the urethra with or without hypospadias (Fig. 17.1c).

17.3 Classification [9]

The Megameatus Intact Prepuce is divided based on site of meatus into:

1. Glanular: The meatus is usually wide, and the frenulum is well-formed. A deep groove can be seen clearly. The stream is well-formed, and the appearance of the penis is almost normal (Figs. 17.1b and 17.3a)
2. Coronal: The meatus is located at the corona, two grooves are well seen, and the frenulum is attached proximally to the meatus. The urinary stream is well-formed (Fig. 17.3b)
3. Sub-coronal: Meatus just proximal to corona, frenulum not well-formed, and urinary stream is splayed (Fig. 17.3c)

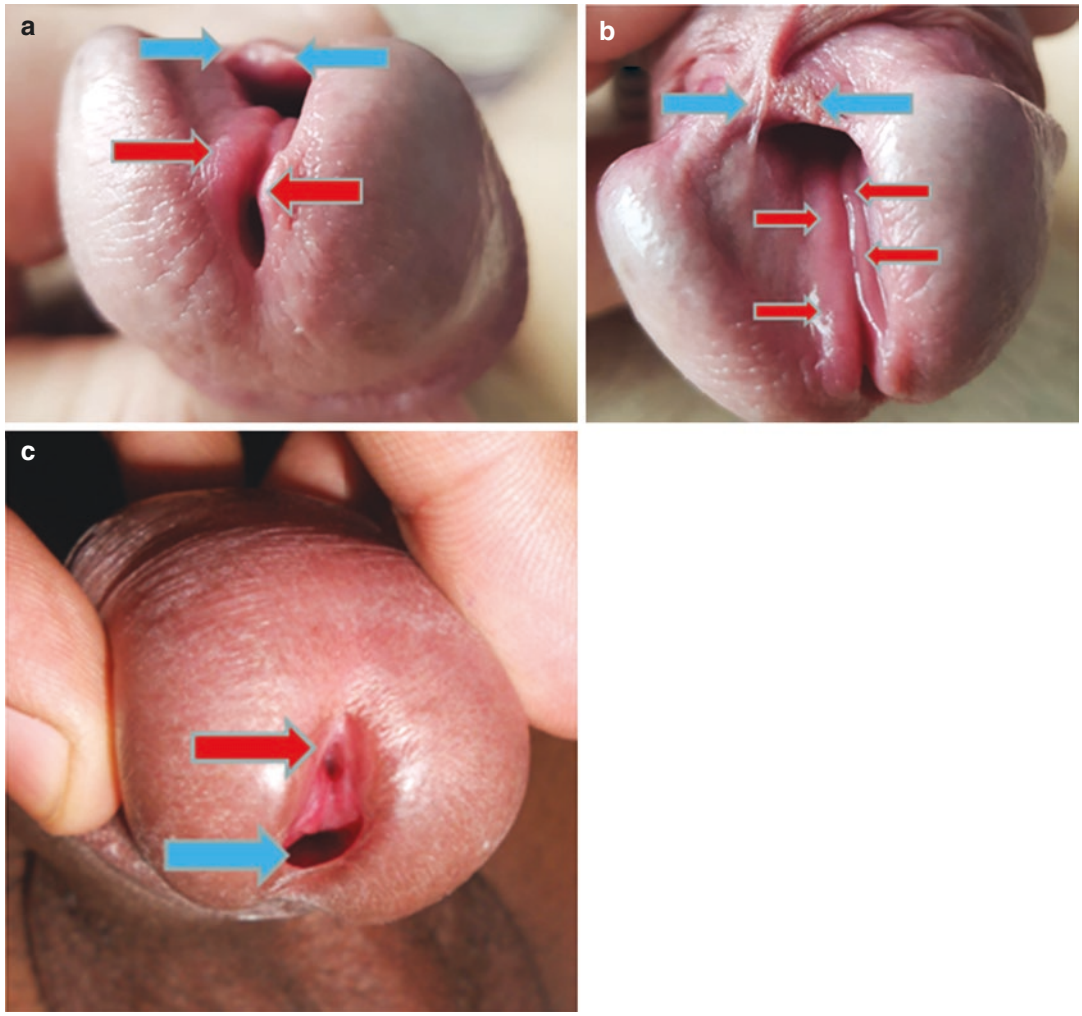

Fig. 17.1 Landmarks of incomplete development of the distal urethra. (**a**, **b**) Dorsal canalization is complete red arrows showing margins canalized urethra and ventral urethral plate closure remains incomplete tubularization as shown by the blue arrow. (**c**) Dorsal red arrow showing dorsal incomplete canalization leading to duplication of the urethra and a blue ventral arrow showing incomplete tubularization of Glanular hypospadias

4. Distal penile: This is the most severe form of MIP, meatus is located in the distal penile shaft, and the urinary stream is splayed (Fig. 17.3d)

17.4 Diagnosis

Hypospadias diagnosis is made at birth because of an incomplete prepuce, but this anomaly is frequently missed, as it is hidden due to the preputial covering. Many patients who are un-noticed may present for any other problem because they usually do not have symptoms. Recognition is common at the time of religious circumcision in newborns but may be diagnosed at adulthood, clinician's visit for another disease or during catheterization. Though it was reported that the anomaly is not associated with chordee, we found the anomaly is associated with dorsal curvature (Fig. 17.4a), sometimes may have ventral curvature (Fig. 17.4b), partial urethral duplication (Fig. 17.1c), and penile torsion (Fig. 17.5a, b).

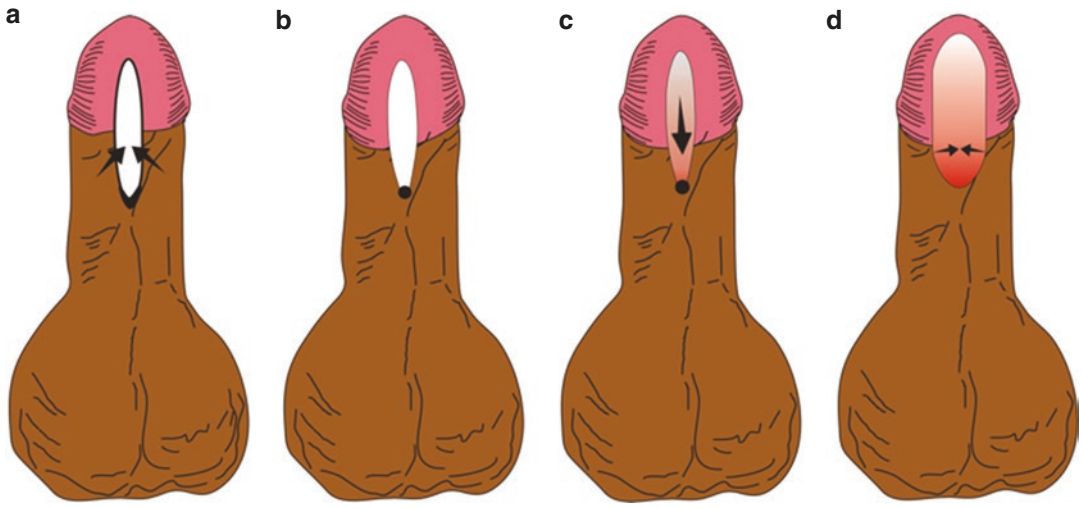

Fig. 17.2 Diagrammatic representation of the development of Megameatus Intact Prepuce. (**a**) Proximal to distal Closure of urethral fold. (**b**) The arrest of urethral fold closure and prepucial closure ventrally forms hypospadias (splayed glans and hooded prepuce). (**c**) Canalization of the glanular plate started and proceeded proximally. (**d**) Complete fusion of the urethral and prepuce fold. Canalization of glanular plate completed up to corona. However, there is no closure of glanular folds (incomplete tubularization)—Results in megameatus intact prepuce

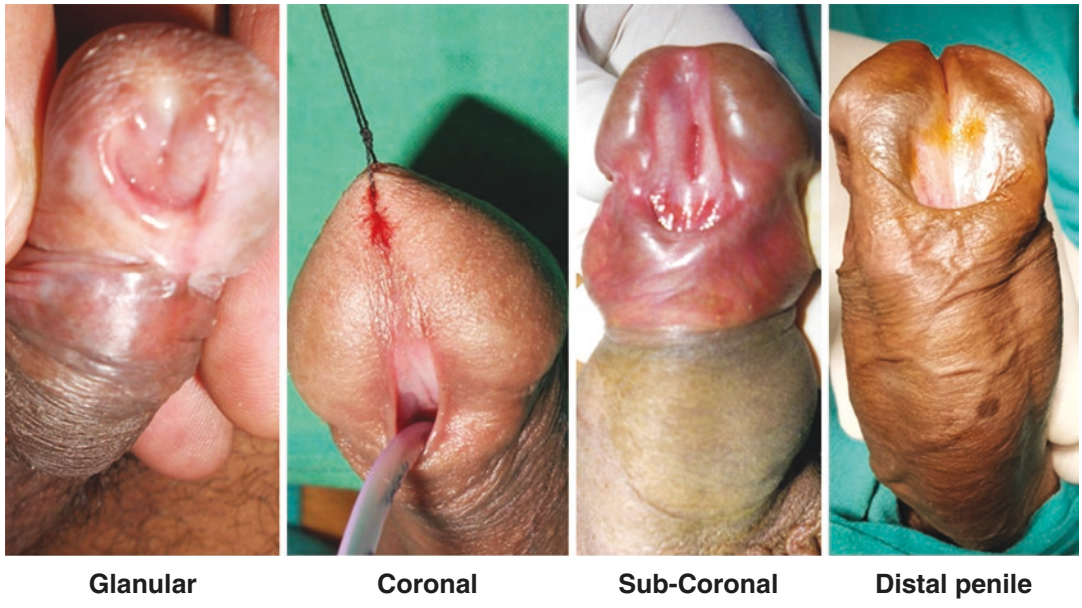

Fig. 17.3 Showing the classification of Megameatus Intact Prepuce

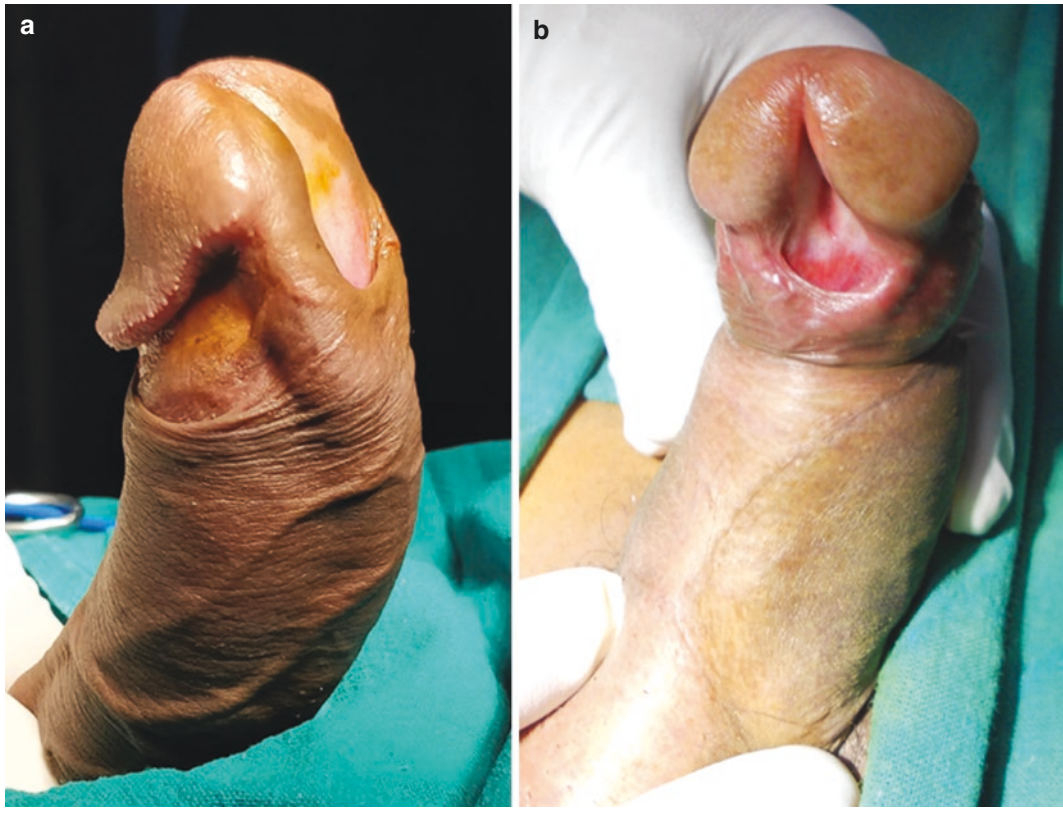

Fig. 17.4 Showing the associated Chordee with Megameatus Intact Prepuce. (**a**) Associated with dorsal curvature. (**b**) Associated with Ventral Curvature

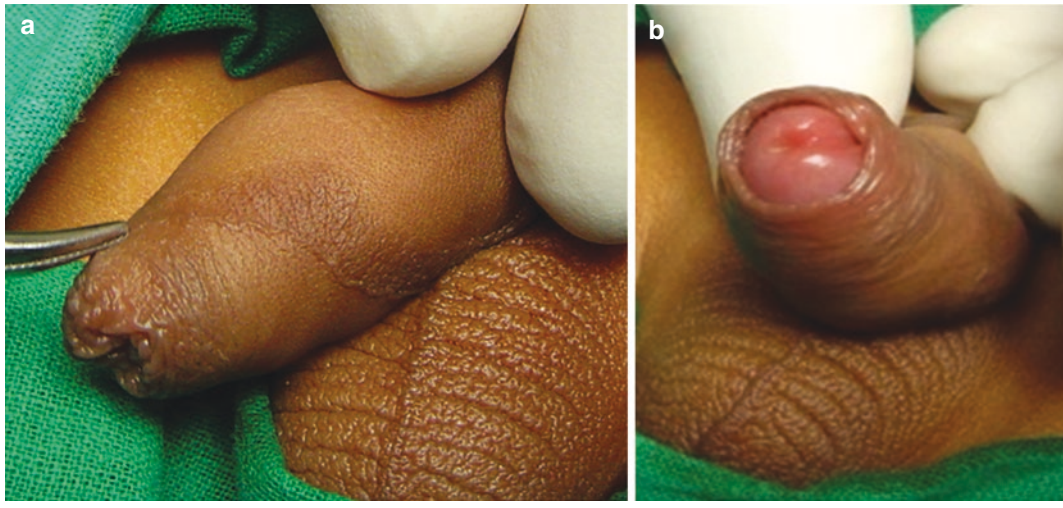

Fig. 17.5 Showing the associated penile torsion with Megameatus Intact Prepuce. (**a**) Median raphe going dorsally indicative of 180 degrees. (**b**) Showing partially retracted prepuce showing meatus dorsally

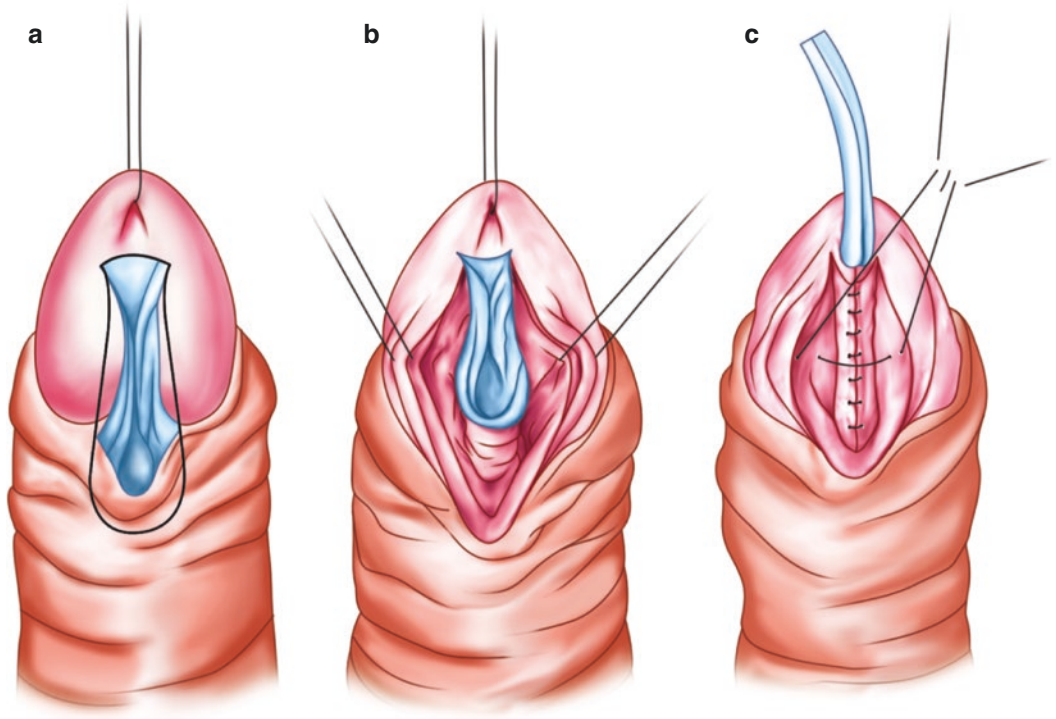

Fig. 17.6 Showing the Pyramid procedure in Megameatus Intact Prepuce. (**a**) U-shape incision around the urethral plate. (**b**) Raising the Glanular wings. (c) Tubularization of urethral plate and glansplasty started

17.5 Surgical Techniques

The surgical techniques for management of megameatus intact prepuce are:

17.5.1 Pyramid Procedure (Fig. 17.6)

1. A tennis racket shape incision is given around the margins of the urethral plate beside the groove of the glans down to the corona level (Fig. 17.6a).
2. The mobilization of the urethra is done up to the apex of the pyramid.
3. Then, the incision deepened proximally to create the glanular wings keeping the distal urethral plate intact dorsally (Fig. 17.6b).
4. Then, a small wedge is taken from the ventral tissue, and the distal urethra is sutured in continuity with the urethral plate (Fig. 17.6c).
5. In the next step urethra and the glanular wings are Tubularized over a stent to form the neourethra.
6. Then the glanuloplasty is done.

17.5.2 Modified Granular Approximation Procedure GAP (Fig. 17.7)

1. A U-Shaped incision is given around the Megameatus and the urethral plate.
2. Partial penile degloving is done.
3. Then, the glans wings are raised to assess the urethral width better (Fig. 17.1a).
4. The excess redundant urethral plate is trimmed to make the diameter of the urethra the same as that of the proximal urethra (Fig. 17.7b, c).
5. The urethral plate is Tubularized with 4–6/ Vicryl/PDS urethral tube according to the size of the urethra (Fig. 17.7d).

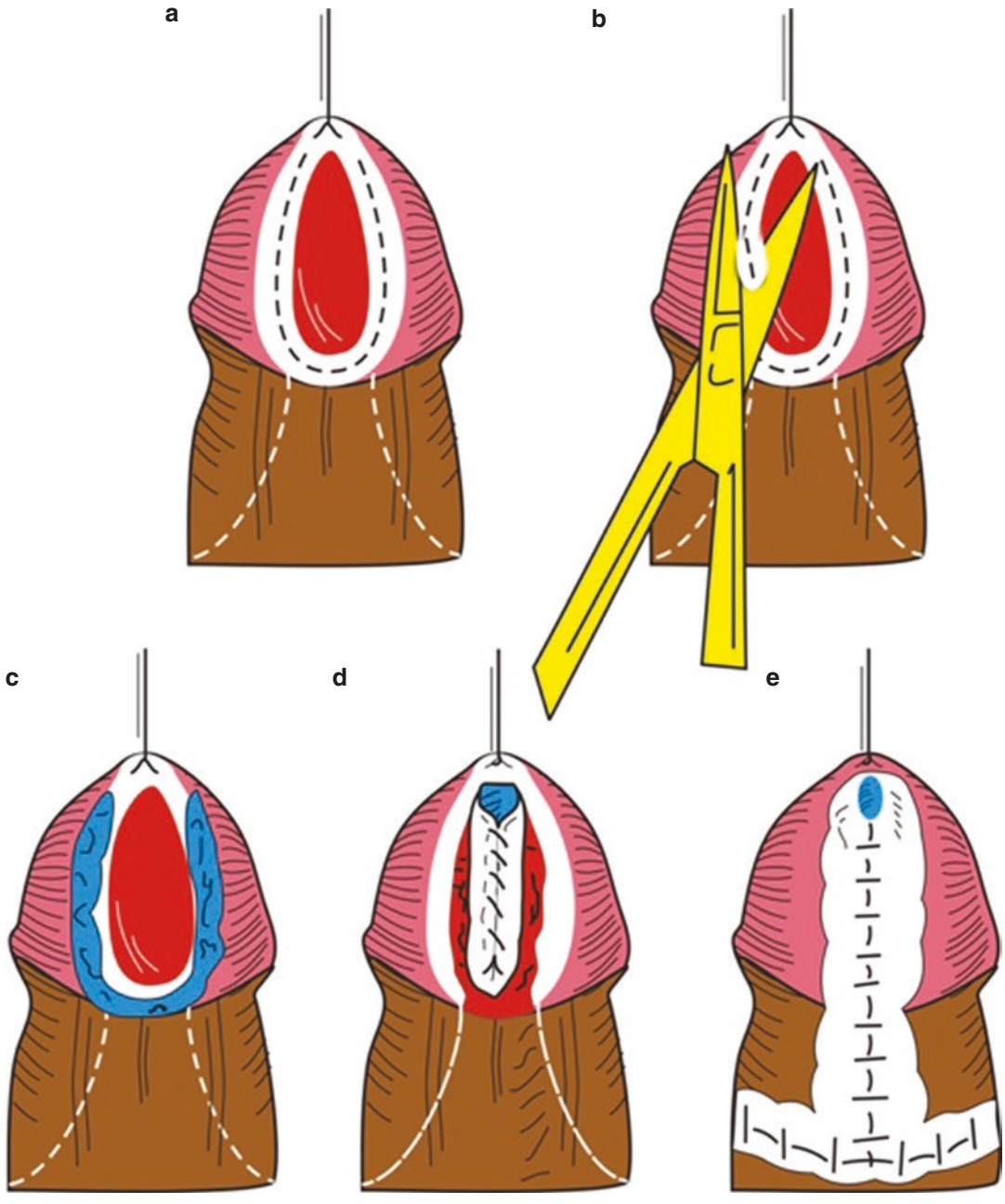

Fig. 17.7 Showing the GAP procedure in Megameatus Intact Prepuce. (**a**) The incision in the glans and distal penile shaft. (**b**) Excision of glanular margins. (**c**) Raising the glanular wing. (**d**) Closure of a urethral plate in the glans. (**e**) Closure of skin and Glans

6. Dorsal dartos or ventral dartos is harvested to cover the Neo-urethra as an intermediate layer (Fig. 17.7e).
7. Glanular wings are sutures over it to complete the urethroplasty and glansplasty followed by skin closure.

17.5.3 The Mathieu Technique

1. The vertical incisions are given on either side of the glans parallel to the urethral plate.
2. Same length and skin incision marked on the ventral surface of the shaft.
3. The glanular wings are dissected, ensuring the preservation of the urethral mucosa.
4. The excess mucosa is trimmed.
5. Parameatal based skin flap is raised.
6. The margins of the flap are sutured to the margins of the urethral plate after putting in the stent.
7. The glanular wings are sutured over the neo-urethra having an adequate meatus to complete the glanuloplasty followed by skin closure.

17.5.4 Tubularized Incised Plate Urethroplasty (TIP)

1. A circumscribing incision is given around the Mega meatus, and penile degloving is done.
2. Parallel longitudinal incisions are given approximately 6–8 mm apart to separate the glans from the lateral margins of the plate. The wings of the glans are developed and mobilized for subsequent tension-free glans closure.
3. After which, the urethral plate is deeply incised from the meatus to the end of the plate, just below the tip of the glans penis. Next, the incised urethral plate is tubularized over a catheter as a stent; the epithelium of the urethral plate is inverted toward the lumen to avoid fistula formation.
4. A vascularized dartos fascia flap covers the urethroplasty as a second layer; then, the prepuce is divided and rotated on both sides to cover the second layer.
5. Glanular wings are closed over neo-urethra to complete glansplasty, and skin closure is completed.

17.5.5 Tubularized Urethral Plate Urethroplasty (TUPU)

The prepuce is retracted, and a U-shape incision is given encircling the meatus, and only inner prepucial skin is incised (Fig. 17.8a, b, c). Dissection plane is created just proximal to the meatus at the level of tunica albuginea, and the urethral plate with spongiosum is mobilized up to mid-glans, and glanular flaps are raised (Fig. 17.8d). The margin of the urethral plate is trimmed in case of wider urethral plate than required. The tubularization of the urethral plate is done, keeping a wide urethral meatus (Fig. 17.8e). Spongiosum is sutured over the neourethra to complete the spongioplasty; glanular flaps are sutured for glansplasty (Fig. 17.8f, g). Inner prepucial skin closure reconstructs the frenulum creating normal meatus and intact prepuce (Fig. 17.8h, i). The following modifications in the classical Thiersch–Duplay technique are done for TUPU. The neourethra is covered with spongiosum in place of the dorsal dartos flap to preserve the prepuce, and frenuloplasty is done to create a normal-appearing penis and prepuce. Patients are followed at 1, 3, 6, 9, and 12 months. Postoperative results are excellent, with a wide slit-like meatus at the tip, well-formed prepuce, and frenulum.

17.5.6 TUPU in MIP with Chordee or Torque

Circum-coronal circumferential incision is given in cases with mild to moderate chordee or torsion, and complete penile de-gloving is done (Fig. 17.9a, b, c). The Gittes test is done to see the chordee correction (Fig. 17.9d). Then the urethral plate with spongiosum and proximal

Fig. 17.8 Showing the Tubularized Urethral Plate Urethroplasty in Megameatus Intact Prepuce. (**a**) Subcoronal Megameatus Intact prepuce. (**b**) Urethral stent inserted after the injection of 1:100,000 solution of adrenaline at the site of incision. (**c**) Inverted U-shaped incision given. (**d**) Urethral plate and spongiosum Mobilization. (**e**) Urethral plate tubularization started. (**f**) Spongioplasty. (**g**) Glanuloplasty. (**h** and **i**) Frenuloplasty and skin closure with intact prepuce

urethra is mobilized, and chordee/torque correction (Fig. 17.9e, f). If chordee/torque still persists, the urethral plate with spongiosum is mobilized into the glans. The Gittes test is repeated to confirm the chordee/torque correction. (Fig. 17.9g, h, i). Then the urethral plate is tubularized, and spongioplasty is done (Fig. 17.9j, k). The glanuloplasty is done to create a conical glans with the slit-like opening at the tip and Gittes test is done to confirm chordee/torsion correction (Fig. 17.9l, m, n). Then frenuloplasty & skin closure is done to create a normal frenulum and prepuce (Fig. 17.9l, m, n). A urethral catheter is put in and removed in 7–10 days and is followed up 1, 3, 6, 9, and 12 months. Patients/parents are asked to start retracting the prepuce after 4–6 weeks.

17.5.7 The Subcutaneous Frenulum Flap (Scuff)

1. A circum-meatal incision is given, and degloving of the penis is done.
2. Then glanular wings are raised.
3. The redundant mucosa trimmed and tubularization of the urethral plate is done over a stent.
4. Then, a well-vascularized inferior-based frenulum flap is developed.
5. The skin is de-epithelialized and advanced over the neourethra.
6. Then, the glanular wings are sutured to complete the glanuloplasty, and skin closure is done.

Fig. 17.9 Showing the Tubularized Urethral Plate Urethroplasty in Megameatus Intact Prepuce with ventral chordee. (**a** and **b**) Distal penile Megameatus Intact prepuce with moderate chordee. (**c**) De-gloving of the Penile skin. (**d**) Gittes test showing persistence of Chordee. (**e**) Urethral plate with spongiosum and proximal urethral mobilization and Lateral mobilization of Buck's Fascia. (**f**) Gittes test showing persistence of Chordee. (**g** and **h**) Urethral plate with spongiosum mobilization up to the mid-glans. (**i**) Gittes test showing correction of Chordee with glanular tilt. (**j**) Tubularization of the urethral plate. (**k**) Spongioplasty. (**l**) Glanuloplasty. (**m** and **n**) Gittes test showing correction of Chordee and glanular tilt corrected. (**o**) Frenuloplasty and skin closure

17.6 Choice of Technique

In a case report, a 41-year-old male was the only case managed conservatively, and authors advocated the patient participation in the surgical intervention decision [6]. But surgery is the treatment of choice in modern hypospadiology, giving excellent results in functionality and cosmesis in MIP patients. The good results of the Mathieu and the MAGPI are acceptable in distal hypospadias. Still, the techniques being not ideal for Mega meatus intact prepuce led to the development of techniques specifically to be used for the MIP. However, GAP is technically less demanding and achieves good results in glanular defects but has limitations. These are the unequal size of neo-urethra due to non-mobilization of glanular wings and superimposition of glanular suture lines without an interposing tissue layer which may lead to fistula. Dissatisfied with the results, people have modified the glanular approximation procedure by mobilizing glanular wings equal to the size of neo-urethra with that of the normal urethra for tension-free urethroplasty and added interposing a dorsal dartos layer. Though the cutaneous advancement techniques have also been used for megameatus intact prepuce, that may not be suitable for distal penile MIP, and also the glanular defect remains uncorrected.

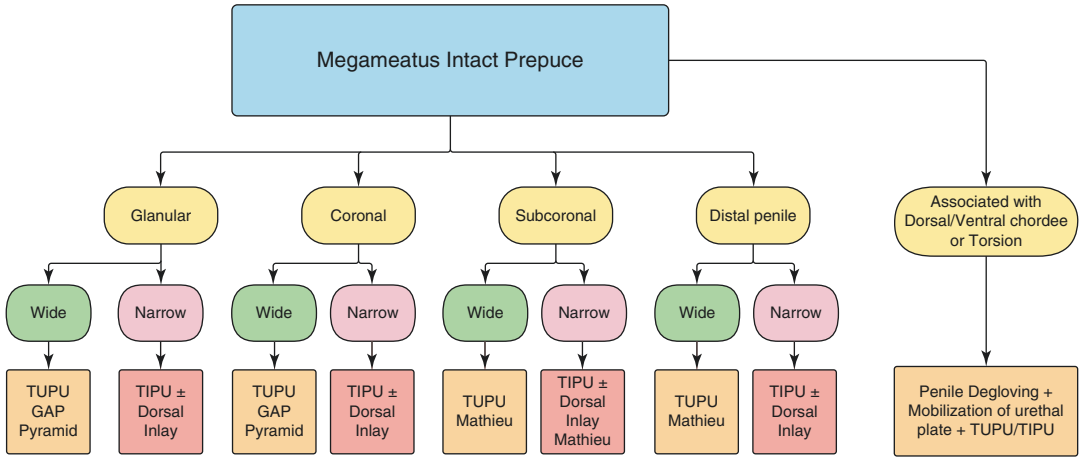

Fig. 17.10 Algorithm for management of Mega meatus Intact Prepuce

MIP is a surgical challenge in circumcised patients because the penile skin is usually thin and scarred and lacks the prepucial and dartos tissue. However, Snodgrass had good results in circumcised patients and narrated that circumcision does not affect the results of TIPU in MIP [11].

The anatomic characteristics of the MIP present a unique challenge to surgeons. The wide meatus and urethral plate dissection may create thin glanular wings more prone to dehiscence and urethral fistula formation. The TIP urethroplasty technique allows for better glanular wings and thicker glanular wings for glanular reconstruction. Because the lateral aspects of the urethral plate are not dissected as they are with the Pyramid and GAP. Nevertheless, the incision in the urethral plate is needed only in the narrow urethral plate, which is rare in MIP. So, we opted for tubularized urethral plate urethroplasty (TUPU) as it allows for more formal dissection of the glans wings on the expanse of the width of the urethral plate. Adding spongioplasty and frenuloplasty to tubularized urethral plate urethroplasty (TUPU) restores the normal urethral and penile anatomy. Based on the type of MIP and width of the urethral plate, an algorithm is proposed to manage MIP (Fig. 17.10). All kinds of MIP can be managed with TUPU or TIPU and GAP. Even minor to moderate curvature and torsion can also be corrected by mobilization urethral plate with spongiosum and urethra. The procedure is feasible with a good surgical outcome even after circumcision.

17.7 Conclusion

Megameatus Intact Prepuce is likely to be missed frequently due to the preputial covering. Patients being asymptomatic may present late. Examination of meatus by retracting the prepuce before circumcision is helpful in the early diagnosis and management of MIP. Surgical correction of MIP in the current era of increased cosmetic awareness is justified. It is a variant of anterior hypospadias, and the repair is feasible by the techniques used for distal hypospadias repair. Excellent functional and cosmetic outcome is seen with tubularized urethral plate urethroplasty (TUPU) with a few modifications like spongioplasty, prepuce preservation, and frenuloplasty. A wide urethral plate and well-developed spongiosum are available. Therefore, it should be the procedure of choice in all cases of MIP. Incised urethral plate urethroplasty is suitable for cases with a narrow urethral plate. Mathieu or onlay flap urethroplasty are rarely required for correction of MIP.

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